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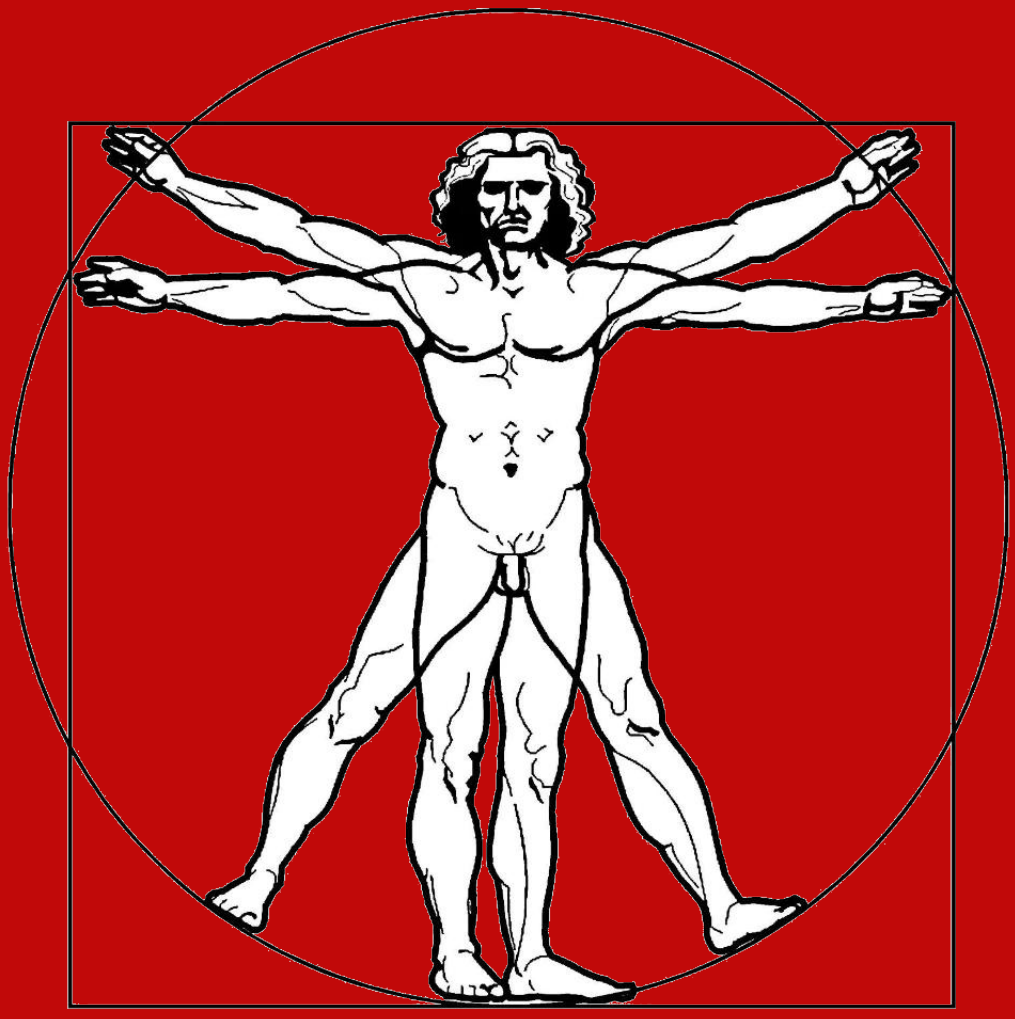

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Skopje, June 2022

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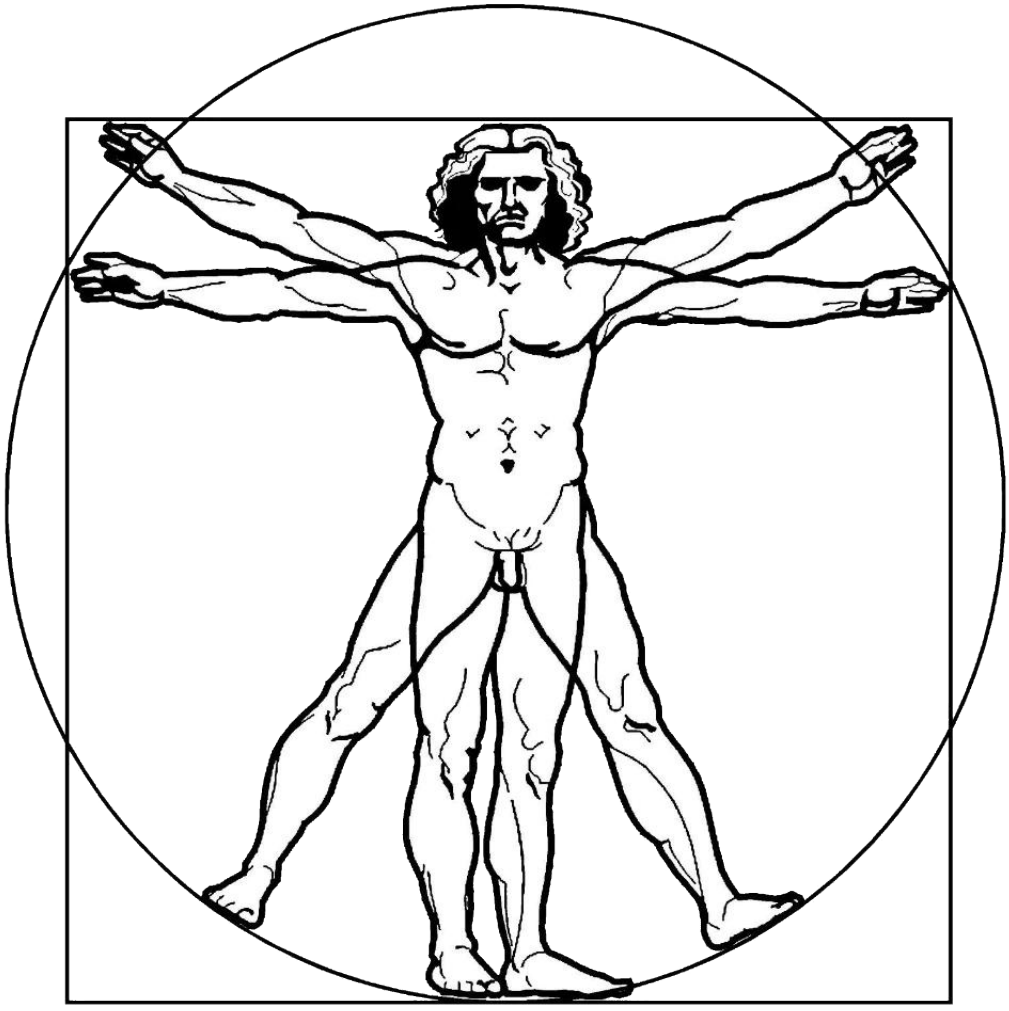

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Skopje, June 2022

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НАМЕСТО ВОВЕД

Драги колеги, пријатели и соработници,

Имам посебна чест и задоволство да го објавам новиот број на нашиот Македонски ортопедско-трауматолошки гласник. За жал, нашиот план за објавување на нови броеви во изминатиот период не можеше да се оствари заради специфичните околности во врска со пандемијата со Covid 19. Во овој момент, пред се, ви посакуваме на сите да бидете здрави и безбедни.

Како и претходно, весникот ќе биде печатен на англиски јазик со апстракти на македонски и англиски. Содржината на овој број се состои од стручни и научни трудови од полето на мускуло-скелетната онкологија комбинирани со трудови кои опфаќаат некои аспекти на ендопротетската, спиналната хирургија и дистракционата остеогенеза. Посебна чест и задоволство за сите нас е можноста да објавиме труд на светски познатиот ортопедски хирург Проф. Рајнер Коц и сор. од Виена. Нашата цел е секое наредно издание да се состои од публикации кои ќе го прошират полето на научен и стручен интерес во областа на ортопедијата и трауматологијата.

Во име на сите членови на редакцискиот одбор, би сакал да се заблагодарам на сите автори, коавтори и соработници за нивниот активен придонес и помагање во нашите заложби за објавување на трудовите. Во оваа прилика, се извинуваме и за неизбежните технички грешки во ова издание, како и во наредните броеви на весникот.

Рецензентите и другите членови на редакцискиот одбор ќе продолжат со своите добри намери за вклучување на сите заинтересирани за објавување трудови од различни медицински области, првенствено од областа на ортопедијата и трауматологијата, во нашиот Македонски ортопедско-трауматолошки гласник. Во таа смисла, ќе бидеме среќни да ја продолжиме традицијата на документирање и анализа на резултатите од нашата професионална работа и споделување на тие искуства во земјава и во странство.

Претседател на редакцискиот одбор,
Научен соработник д-р Ненад Атанасов

INSTEAD OF INTRODUCTION

Dear colleagues, fellow doctors, friends and supporters,

It is my special honor and pleasure to announce a new number of our journal “Acta Orthopedica et Traumatologica Macedonica”. Unfortunately, our plan to publish the journal in the past time period could not be achieved due to the specific circumstances regarding the situation with Covid 19 pandemic. For the moment, above all, we wish you all to stay healthy and safe.

Like in the previous time period, our journal will be again published in English language with supporting abstracts in Macedonian. The content in this number is consisted of articles focused on musculoskeletal oncology as a topic combined with some articles and case reports in the field of endoprostethic surgery, spine surgery and distraction osteogenesis. It is a special honor and pleasure for all of us to be able to publish a paper from the world famous orthopedic surgeon Prof. Rainer Kotz et al. from Vienna. Our goal is every forthgoing issue to be published with articles covering broader field of interest of orthopedic surgery and traumatology.

On behalf of all members of the Editorial Board, I would like to thank to all the authors, co-authors and other collaborators for their active contribution and help in our publishing efforts. In addition, we apologize for the inevitable technical mistakes in this and other issues of the Journal.

The members of the Board and our reviewers will proceed their intentions and efforts to involve everybody who has interest for publishing in field of medicine and especially in orthopedic surgery and traumatology in our “Acta Orthopedica et Traumatologica Macedonica”. Thereby, we will be happy to continue the tradition of documenting and analysis of the results of our professional work and sharing our experiences in our country and abroad.

President of the Editorial Board,
Ass. Prof. Dr. Nenad Atanasov

THE ESTABLISHMENT OF ORTHOPEDIC TUMOR SURGERY IN A PRIVATE HOSPITAL "A single surgeons experience"

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ABSTRACT

Experience with 154 tumor cases in a private institution with a single surgeon showed excellent results in respect to margins with 45 wide resection out of 49 primary tumors including 8 pelvis locations.

Key words: bone tumor, limb salvage, single institution experience

The Tumor Center for orthopedic oncology at the Univ. Clinic for Orthopedics at the Medical University of Vienna developed under my direction from 1984 – 2009 with new innovative methods and gained international recognition. After that period I established a tumor center from 2010 to date at the Wiener Privatklinik, a private hospital with 145 beds. Thanks to the personal experience and the support of a radiology centerx)with all common modern examination options with immediate availability, as well as the expertise of an internationally recognized pathologist in the sector of bone and soft tissue tumorsxx), 141 patients with bone and soft tissue tumors were able to be identified in the period from 2010 to 2020. Most of the patients came from neighboring countries and were mostly paid for by the state, as the cases involved were difficult and could not be successfully treated in their home country.

A total of 154 operations were performed by the tumor team. These are 49 primary malignant bone tumors (Tab.1); 21 soft tissue sarcomas, 15 metastases of carcinomas, 39 benign bone lesions and 30 revision surgeries.

The surgical treatment of these patients by the senior surgeon (R.K.) together with his experienced team prevented complications to a large extent - it was never necessary to transfer a patient for intensive medical supervision outside the home hospital. It should be noted that there were included 19 pelvic operations, of which two external and seven internal hemipelvectomies - with endoprosthetic replacement - were performed. One focus was put on the salvage of the extremities after tumor resections with tumor prostheses. It concerned 14 distal femur, 4 proximal tibiae, 3 prox. Femur. 14 humerus and scapula and 2 total knee prostheses. The GMRS system from Stryker (Lit. 1) was used primarily on the lower extremity, the HHMRS system from Stryker on the upper extremity and later the humerus system from Zimmer. A special focus was the biological reconstruction, especially on the lower extremity through the fibula per tibia transplantation. Resection-Replantations were also performed, the so-called rotation plasty on the lower extremity (Lit. 2) and the Tikhoff Linberg resection (Lit. 3) on the upper extremity (Fig. 1a,b). A total of 5 growing prostheses (Fig.2a,b) were implanted in children (Lit.4).

The high number of revisions (n=30) in the whole serie and 15 of that in our own cases resulted from the early surgical intervention in cases of woundhealing disturbences and/or hematomas, what was decisive for good results.

In difficult cases, cooperation with vascular surgeons and plastic and reconstructive surgeons – all present in the Wiener Privatklinik - was also necessary. An abdominal surgeon and/or urologist was always involved in the operations on the sacrum, and in all cases embolic complications could be prevented in the pelvic operations by implanting aninferior vena cava filterxxx). The necessary pre- and postoperative chemotherapy was carried out almost entirely in the home country and the patient was referred to the Wiener Privatklinik, i.e. the tumor center, for an operation if there was no longer any possibility of limb preservation in their own country.

x) P.PELOSCHEK: Radiology Center, Wiener Privatklinik
xx)S.LANG: Senior Pathologist, MedUniWien
xxx) M.FUNOVICS, Radiology Center, Wiener Privatklinik

Table 1: Diagnoses of 49 primary malignant bone tumors

Osteosarcoma	24
Ewing´s Sarcoma	05
Chondrosarcoma	07
Malignant fibrous histiocytoma and dediff. Sarcoma, Chordoma	13
TOTAL	49

Table 2: Location and surgical margins in 49 primary malignant bone tumors

	wide	***marginal***	***intralesional***
dist. Femur	14		
prox. Tibia	04		
prox. Femur	03		
Total Knee	02		
Humerus und Scapula	14		
Pelvis	04	02	02
Radius, Tibia, Fibula, Calcaneus	04		
TOTAL	45	02	02

92,1% adequate margins, histological proven by S.L.

Illustrations

1. 12-year-old child – Osteosarcoma of the proximal humerus

Figure 1a: intraoperative situs with tumor resection

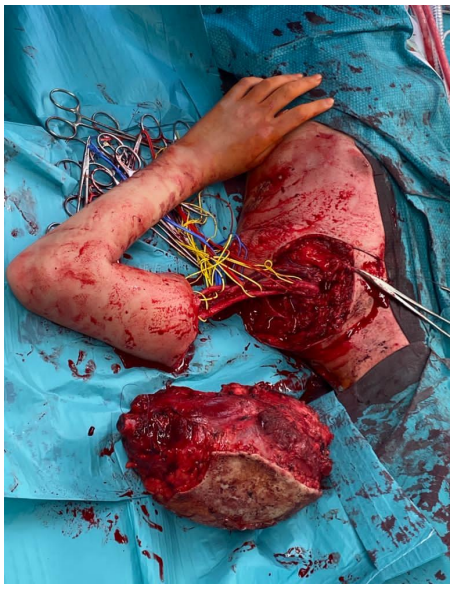
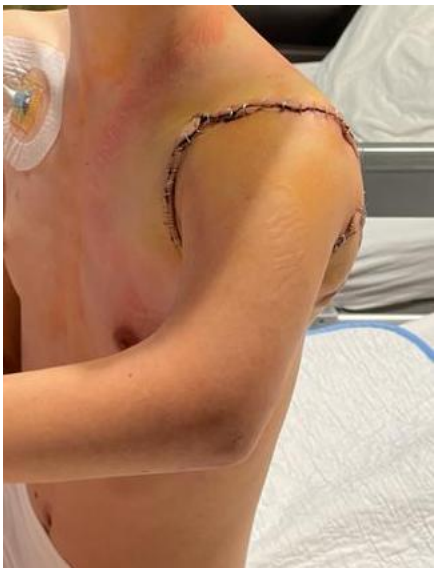


Figure 1b: postoperative replantation



2. 4-year-old child – Ewing´s sarcoma of the tibia

Figure 2a: Situs after resection and interim spacer made of bone cement

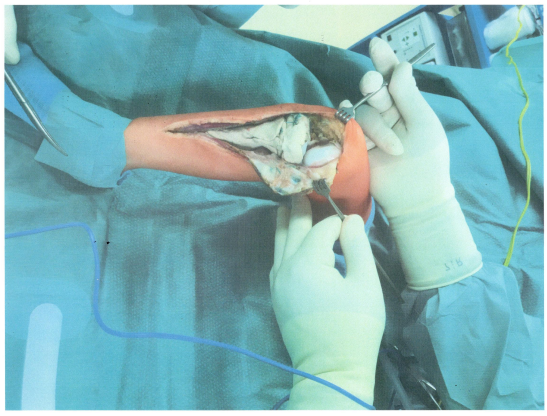
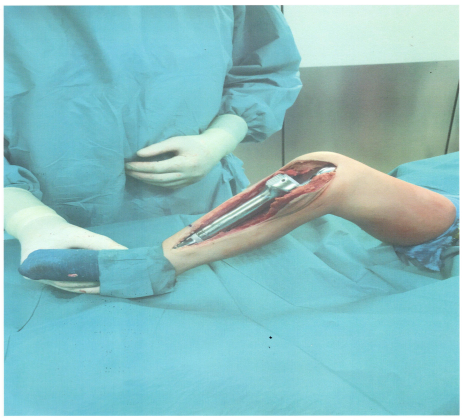


Figure 2b: Implantation of a tibia Stanmore children growing prosthesis



3. 50-year-old male patient - chondrosarcoma of the right pelvis with further internal hemipelvectomy, extracorporeal radiation with 100 Gray and replantation

Figure 3a: Condition of the replant after two years

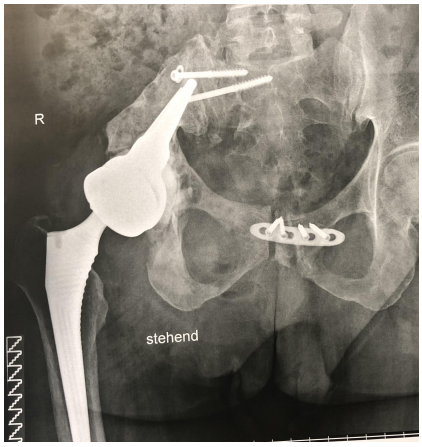


Figure 3b: Clinical performance of the patient with free standing on the operated leg

Requirements for establishing a tumor center in a private hospital

- An experienced trained surgeon for tumor orthopedics with a trained team (assistants and anesthesiologist)
- All possibility of imaging examinations on site and immediately
- Disposal of expertise in pathological examination of sarcomas

Checklist:
- Exact planning of the surgical steps
- A realistic estimation of the blood consumption
- Provision of the appropriate endoprosthetic device
- Post-operative care of the patient over one night in a recovery area with two intensive care beds
- Availability of an oncological team with regular tumor board meetings
- Experienced physiotherapy department
- A support team for foreign patients through an appropriate foreign office with interpreters
- Continuous support for inquiries by a foreign office and lively internet traffic with the transmission of external imaging materials and the resulting digital advice
- Communication with the local insurance companies and other government agencies that provide for the approval of an operation abroad
- Carry out the medical visits - in individual cases - on site the patients country, to be able to make reliable and competent predictions for the surgery

Conclusion:

Thanks to 35 years of experience from 1974 – 2009 (Lit.5) and expansion of the operating modalities in the general hospital, the experienced senior surgeon(R.K.) with an educated team of assistants (F.A., D.T.) was able to achieve excellent results also in a private hospital xxxxx). It has also been shown that when the fate of the patient is in one hand and the preparation and performance of the operation is carried out responsibly, permanent cures and good functional results are achieved even in largest pelvic surgeries.

xxxxx) The Wiener Privatklinik received the Certificate "World's Best Hospitals 2021" by Newsweek

Literature:

1. Kotz R (2010) Megaprostheses: KMFTR-GMRS. Orthopedist 39: 922-30
2. Kotz R, Salzer M (1982) Rotationsplasty for childhood osteosarcoma of the distal part of the femur. JBJSurg A 64: 959-69
3. Windhager R, Millesi H, Kotz R (1995) Resection-replantation for primary tumors of the arm. An alternative to fore-quarter amputation. JBJSurg B 77: 176-84
4. Kotz R, Windhager R, Dominkus M et al (2000) A self-extending pediatric leg implant. Nature 406: 143-4
5. Kotz R (2014), Progress in musculoskeletal oncology from 1922-2012. Int.
6. Orthopaedics (SICOT) 38:1113-1122

USE OF RADIOGRAPHIC CLASSIFICATION SYSTEM IN DISTRACTION OSTEOGENESIS

ПРИМЕНА НА РАДИОГРАФСКА КЛАСИФИКАЦИЈА ВО ДИСТРАКЦИОНАТА ОСТЕОГЕНЕЗА

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АПСТРАКТ

Цел на трудот: Евалуација на клиничкиот исход кај пациентите лекувани со дистракциона остеогенеза со различни класифицирани радиографски карактеристики на новото коскено ткиво, преку вредностите на индексот на коскено зараснување (ИКЗ)

Материјал и методи: Студијата опфаќа 27 пациенти со елонгација на екстремитетите по методот на Илизаров лекувани на Клиниката за ортопедски болести во Скопје од 2013 до 2020. Анализирани се 27 сегменти користејќи го класификациониот систем на Ru Li, Michael Saleh, Lang Yang, Les Coulton. Класификацијата е базирана на обликот на калусот и на карактеристиките на дистракционата остеогенеза. Резултатите се класифицирани во 2 групи: група со резултати за индексот на коскено зараснување (ИКЗ) под 50 дена/цм постигната елонгација и група над 50 дена/цм. Резултатите се корелираат со обликот на калусот и со типот на дистракциона остеогенеза според користената класификација.

Резултати: 8 од 10 пациенти со тип 1 на облик на калусот покажаа порапидно коскено зараснување од остатокот на пациенти во оваа група. (80%) 5 од 8 пациенти со тип 2 презентираа вредности за ИКЗ под 50дена/цм. Кај останатите пациенти со тип 3,4 и 5, ИКЗ беше поголем од 50 кај 66,66%. Само еден пациент презентираше тип 5 облик на калусот со ИКЗ од 69,45. Според типот на дистракциона остеогенеза, пациентите со тип 2,6 и 9 покажаа ИКЗ под 50 дена/цм постигната елонгација. Кај тип 8 и 10, мораше да се смени режимот на дистракција, а кај пациентот со тип 1, неопходна беше модификација на конструкцијата.

Заклучок: Користење на радиографскиот класификационен систем при елонгации на екстремитетите, може не само да ги презентира, туку и да ги предвиди карактеристиките на новото коскено ткиво, а со тоа да укаже на можните корекции во елонгациониот процес и подобрување на финалниот исход во лекувањето.

Клучни зборови: дистракциона остеогенеза, радиографска класификација, индекс на коскеното зараснување (ИКЗ)

ABSTRACT

Purpose: To evaluate the clinical outcome in patients with different radiographic features of the regenerate within the radiographic classification system judged by the bone healing index (BHI)

Material and methods: The study has included 27 patients with limb lengthening using the Ilizarov method at the Clinic for Orthopaedic Surgery in Skopje from 2013 to 2020.27 segments have been analyzed using the classification system of Ru Li, Michael Saleh, Lang Yang, Les Coulton. The classification is based on the shape of the callus and on the type of feature of distraction osteogenesis. The results were classified in two groups: Results with BHI under 50 days/cm and over 50 days/cm. The results for BHI were correlated with the shape of the callus, and with the type of feature at the distraction site.

Results: In 8 of 10 cases grouped in the first of five callus shape modalities, there was a more rapid bone healing than in the rest of the cases (80%).In 5 of 8 patients classified in the second type (67,5.%), the BHI was under 50 days/cm. In the other patients representing type 3,4 and 5, BHI was over 50 days/cm in more than 66,66%, Only one patient presented type 5, with BHI of 69,45. In patients with 2,6 and 9 type of feature, BHI was under 50 days/cm. In type 8 and 10, we had to change the rhythm of distraction.In one patient with type 1 of feature, a final modification of the frame was necessary.

Conclusion: Use of this classification system in limb lengthening procedures can not only detect, but also predict the healing pattern of the bone and thereby allow therapeutic corrections to be made at the appropriate time in order to improve the final outcome of the treatment.

Key words: distraction osteogenesis, radiographic classification, bone healing index (BHI)

INTRODUCTION

Monitoring of bone changes is mandatory in limb lengthening procedures.(1) Serial radiographic examinations of the newly formed bone at the distraction gap formed by gradual pulling apart the bone fragments in distraction osteogenesis using a circular external frame, is undoubtedly one of the most important factors for successful outcome of the treatment.(2) The detection of initial radiographic indicators of new bone formationon native radiographs, as well as trabecular bone and a clearly outlined cortical margin in latter stages of bone formation, leads to an adequate planning of artificial gradual segmental lengthening.(3) Bone healing is a long process, which involves remodeling of the regenerate, and its completion is often difficult to define, which makes the removal of the frame one of the most challenging decisions in this kind of surgical treatment. Unfortunately, there are very few systems of radiographic classification in various age groups, which accurately present the progression of bone healing in different stages of limb lengthening. The majority of studies presenting the efforts of the authors to create an adequate classification system, have the disadvantage of relatively small sample size and a large number of influencing factors which limits the interpretation of their findings.(4-7) The studies mostly analyze the results of radiographic classification systems in children and teenagers. The classification of Ru Li, Michael Saleh, Lang Yang, Les Coulton presented in a study which refers to radiographs and medical data in patients from various age groups who underwent limb lengthening or lengthening with some minor deformity correction, is used in our study.(8) The aim of our study is to correlate the clinical outcome in patients with different radiographic features of the regenerate within the above mentioned classification system judged by the bone healing index (BHI).

MATERIAL AND METHODS

The study has included 27 patients who have underwent a limb lengthening procedure using the Ilizarov method at the Clinic for Orthopaedic Surgery in Skopje from 2013 to 2020. The whole number of patients involved in the study was initially larger, but those patients who had inadequate follow up were excluded. No patients were excluded between the time of surgery and the time of frame removal. During the radiographic examination in our patients, special attention has been paid on the following preconditions:

1. Proper positioning of extremities necessary for receiving standard and correct results
2. Standard and constant conditions for each examined segment i.e. bone
3. Maintaining constant distance between the X-ray source and the examined segment in each stage of bone formation
4. The X-ray beam directed at the centre of the distraction/correction site
5. X-ray examination performed in antero-posterior and medio-lateral projection
6. High degree of visibility and clear presentation of the radiographic parameters. (Only obvious parameters with a high degree of clarity were taken in consideration during the process of analyzing the results.)

The monitoring of new bone formation was simultaneously performed with ultrasonography with 7.5 MHz linear transducer at the distraction site in longitudinal and transversal plane at the same day. We considered the consolidation and remodeling stage accomplished once a non-invaginated continuous cortical margin appeared on sonograms in longitudinal plane and a continuous hyper-reflecting solid line in transversal plane, with no cystic formations larger than 2 cm in diameter and presence of three of four cortical margins and a trabecular bone structure without a radiolucent zone or any cystic changes on the A-P and lateral radiographs.

The analysed results were received in three different stages of new bone formation

1. Stage of initial bone formation
2. Stage of strain of tension and advanced new bone formation
3. Stage of mineralization, bone consolidation and remodeling

Within the clinical material, 27 segments have been radiographically analayzed using the classification system of Ru Li, Michael Saleh, Lang Yang, Les Coulton presented in a study from 2005. The classification is based on the shape of the callus, as well as on the type of feature of distraction osteogenesis.

The five callus shapes of distraction osteogenesis are:

Shape 1. The regenerate is wider than the original bone

Shape 2. The regenerate is the same width as the original bone

Shape 3. The regenerate is narrower than the original bone with attenuated midportion.

Shape 4. The regenerate is mainly at the one side of the distraction gap. It has been observed that the anterior on the lateral view and the medial on the AP view at the tibia lengthening show a callus defect; this is in contrast with sound callus in posterior and lateral side.

Shape 5. The regenerate is a thin pillar in the central portion and occurs more in poor conditions and complex deformity correction.

The 10 types of feature of distraction osteogenesis are :

Type 1 is sparse low density callus typically first seen in the distraction gap approximately 30 days after the osteotomy.

Type 2 is low density uniform stripes, seen 2 months after osteotomy. The newly formed bone appears to consist of longitudinal striped columns homogeneously bridging the two ends of the osteotomy.

Type 3 shows some newly formed bone as dense speckled regenerate 2 months after osteotomy; it is heterogeneous and bridges the two ends of the osteotomy.

Type 4 has new bone adjacent to the sides of the osteotomy with a central radiolucent zone 3 months after osteotomy.

Type 5 occurs 4 months after osteotomy. The callus is ovoid and sclerotic around the bone ends with sparse sclerotic regenerate in the mid-portion of distraction gap; the regenerate is of poor quality.

Type 6 has uniform intermediate density occurring 4–5 months after osteotomy. The new bone bridges the gap with well distributed and dense ossification. The callus region appears in radiographs as a relatively sclerotic zone. This area of remodeling appears as a zone of relative ossification behind the sclerotic zone. The bone becomes structurally continuous.

Type 7 has an intermediate density with an irregular and heterogeneous appearance occurring 5 months after osteotomy. The bone formed in the gap is amorphous with heterogeneous ossification and includes multiple cysts.

Type 8 occurs 6–7 months after osteotomy. Some of the distraction gap is filled with a dense ossification but shows a radiolucent saw-tooth line across the mid-zone. Types 5–8 have an intermediate radiographic density and are typically seen during early consolidation.

Type 9 shows normal density bone with homogeneous ossification of the regenerate densely corticalized at 9 months after the osteotomy. The two ends of the osteotomy are bridged by homogeneous new bone with two continuous cortices.

Type 10 has cyst defects and a heterogeneous bone density. In 10 months after the osteotomy, the fragments are completely fused with a dense and irregular ossification of the regenerate that appears solid on the radiograph with independent small defect areas. The two ends of the osteotomy are bridged by a continuous segment of bone, but it presents an irregular appearance or a discontinuity of at least one of its cortices.

From the point of view of clinical outcome, the results were classified in two groups: Results in a group of patients with BHI under 50 days/cm and in a group over 50 days/cm. The results for BHI were correlated with the shape of the callus, as well as with the dominant type of feature of the distraction osteogenesis radiographically detected at the distraction site throughout the process of new bone formation.

RESULTS

In the whole group of 27 patients in this study, 10 patients were 5 to 18 years of age, 9 of them between 19 and 29 years, whereas 8 over 30 years of age. In three of the patients the lengthening procedure was combined with simultaneous gradual correction of angulatory deformities of minor extent. In the majority of patients, the whole frame was fixed on the bones with tensioned wires only, except in those 5 cases where hybrid use of wires and half pins was necessary. Tensioning of the wires was achieved using a circular wire tensioner. The corticotomy was performed either using an osteotome to crack the bone after a drilling procedure with multiple drilling holes, or using a Gigli saw in 11 patients. (figure 1)

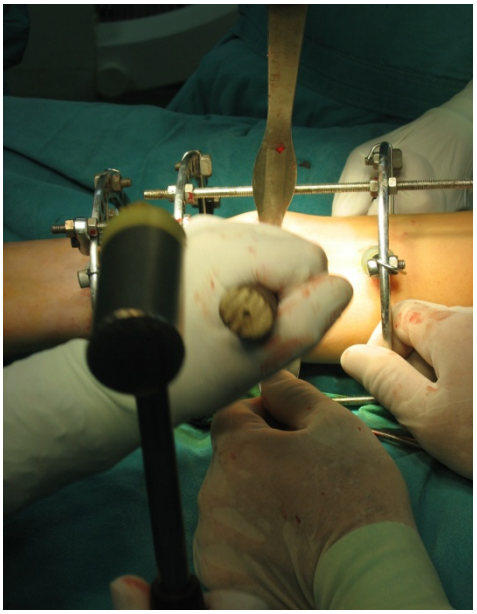

Figure 1: Corticotomy of fibula using an osteotome in surgical treatment of lower leg shortening.

Intraoperatively, the wires were inserted using a power drill at a slow speed with a removal of the drill once the wire exited and use of a mallet to finish the contact between the wire and the ring on the opposite side. At the end of the operative procedure, gauzes with betadine in form of squares were placed on each pin site and an occlusive dressing was used around. 48-72 hours postoperatively, the whole region around the pin sites was left uncovered. An additional use of gauzes was necessary only in cases of exudates on the pin sites. The condition of each pin site was recorded daily, and a crust removal was performed if necessary. Pin site care was accomplished by local use of bacitracin-neomycine antibiotic spray which meant not merely a local prevention of pin site infection but also helped in

maintaining dry pin sites. A single preoperative parenteral dose of Ceftriaxon, which was continued post-operatively for 72 hours usually was sufficient in the general antibiotic treatment of the patients. The additional use of antibiotics was needed only in cases of osteomyelitis or pin site infections after the patients were discharged. Since no skin tension was detected on our clinical material, there was no need of release of skin or subcutaneous tissue.

A latency period prior to the beginning of gradual distraction was 10-14 days. The daily rate of segmental lengthening was 1 mm, divided in 4 increments of 0.25 mm. All the patients were encouraged of early weight bearing by means of crutches, and a full weight bearing as soon as possible. At the end of the treatment, the external frame was removed in the operating room under sedation. Once the construct was removed, patients have been wearing a cast for 4-6 weeks with an increase in the extent of weightbearing. The average lengthening was 67.8 mm, from 34 mm to 108 mm. The average EFT (external fixation time) was 245.34, whereas the average BHI (bone healing index) was 47.68 days/cm, ranging from 36.76 to 76,88.

The results of two groups of patients with different values of BHI in correlation with the radiographic shape and features of the regenerate are presented in table 1 and 2.

Table 1

Shape of callus	Analyzed segments	Patients with BHI < 50	Patients with BHI > 50
1	10	8 (80%)	2 (20%)
2	8	5 (62,5%)	2 (37,5%)
3	3	1 (33,33%)	2 (66,66%)
4	4	1 (25%)	3 (75%)
5	2		2 (100%)

Table 2

Feature of distraction osteogenesis	Analyzed segments	Patients with BHI < 50	Patients with BHI > 50
1	1		1 (100%)
2	3	3 (100%)	
3	2	1 (50%)	1 (50%)
4	2		2 (100%)
5	3	1 (33,33%)	2 (66,66%)
6	5	5 (100%)	
7	4	2 (50%)	2 (50%)
8	2		2 (100%)
9	4	4 (100%)	
10	1		1 (100%)

Types 3,4 and 5 in the classification of callus shapes were followed by increased values of BHI in our patients, which means a prolonged bone consolidation and remodeling. Figure 2 and 3.

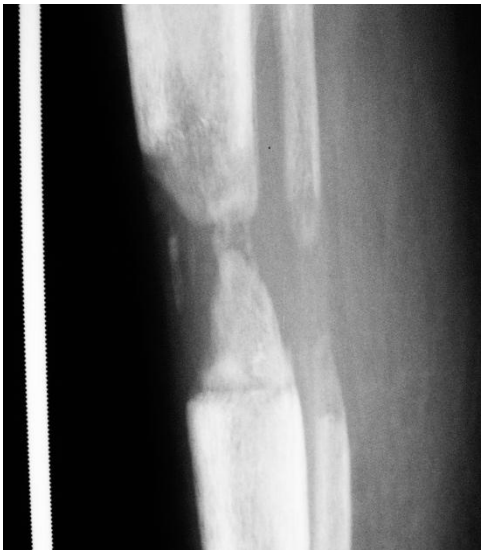

Figure 2: X-ray of distraction gap, of lower leg in stage of lengthening. The regenerate is narrow compared to the surrounding bone with visible attenuation of mid-portion. (type 3 of callus shape)

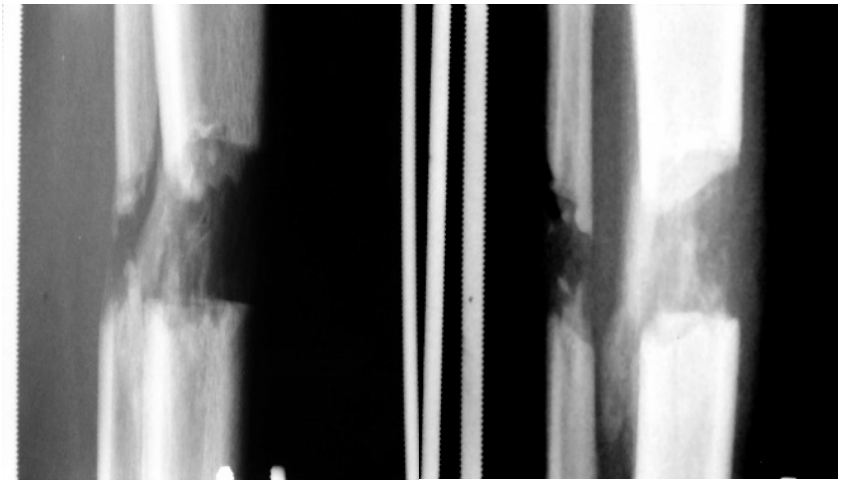

Figure 3: X-ray of lower leg presents a defect in the callus formation in the anterior portion on the lateral view and in the medial of the A-P view (type 4 in the classification of callus shape)

In 8 of 10 cases in our study classified in the first of five callus shape modalities according to the used classification system, there was a sufficient and more rapid bone healing than in the rest of the cases in this group (80%). The judging parameter for bone healing was the value of BHI under 50 days/cm, the minimal value for BHI was36.76 days/cm in these group. In 5 of 8 patients classified in the second type of shape of the callus at the distraction site (67,5.%), the BHI was under the limit of 50 days/cm, too. In the other patients representing callus shape 3,4 and 5 at the distraction gap, BHI was over the value of 50 days/cm in more than 66,66%, with maximal value of 76,88 which was indicative of some form of delayed bone healing. Only one patient presented type 5 callus shape, with BHI value of 69,45.(table 1) Generally, types 3,4 and 5 in the classification of callus shape were followed by increased values of BHI in our patients, which was indicative for a prolonged bone consolidation and remodeling. (Figure 2 and 3). Nevertheless, none of the patients which represented these types of callus shape needed some form of modification in the lengthening procedure in order to stimulate bone healing.

The types of feature of distraction osteogenesis were also related to the clinical outcome in our patients, judged by the BHI. In patients with 2,6 and 9 type of feature, all the patients showed a value for BHI under 50 days/cm gained lengthening. 3 of them were with type 2, five with type 6, and four of them with type 9.In type 7, two of the patients presented BHI value under 50, whereas the other two patients over 50, both not over 55 days/cm. In type 8 and 10, we had to change the rate and the rhythm of distraction due to a poor bone response in the segment, (figure 4) whereas in one patient with type 1 of feature of distraction osteogenesis, not only the distraction regimen had to be changed but a modification of the frame was necessary in order to achieve an enhancement in new bone

formation at the stage of bone consolidation and remodeling. (Figure 4)

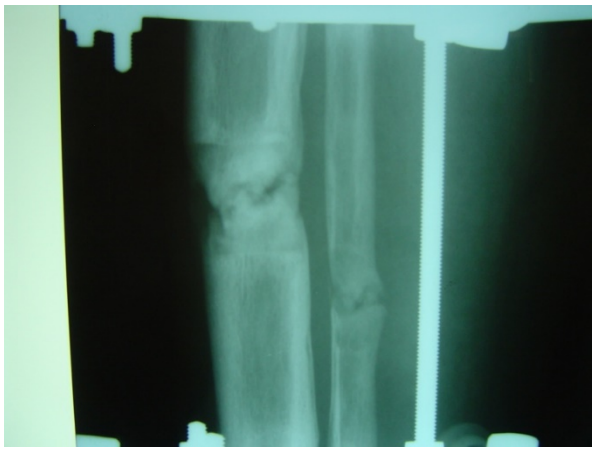

Figure 4: X-ray of lower leg presenting feature type 8 with its radiolucent saw-tooth line across the mid-zone and sclerotic bone formation. Progress towards delayed union.

DISCUSSION

Segmental limb lengthening which starts with a positioning, surgical fixation and tensioning of the external frame completed with corticotomy of long bone(s) in the segment and continues with gradual distraction of bone fragments and soft tissues using the method of "strain of tension" is a process laden with numerous complications.(9,10) Apart from bone infections, neurological disorders, joint contractures etc., the greatest threats are either the poor bone consolidation of the newly generated bone, or its premature healing, both due to inadequate planning of rate, frequency or rhythm of surgical distraction. This leads to fractures, distortion of the axis, or lack of osseous consolidation of the newly formed bone.(11) In order to reduce the number of such complications, a permanent monitoring of the entire process is necessary, from the initial moment of treatment, until the distraction apparatus is removed. In that manner, plain radiography is considered one of the most useful imaging modalities for evaluation of the bony alignment and the extent of osseous production especially in the latter stages of new bone formation. Native radiographs, at beginning of lengthening allow visualization of the primordial hazy densities within the distraction site. The progression of bone healing from the central zone of collagenous growth to the more peripheral columns of mineralized bone results in a distinctive radiographic appearance in lengthening procedures. The superiority of the diagnostic radiography over other diagnostic methods of evaluation of newly formed bone at the distraction or compression site is most evident in the stages of bone mineralization and remodelling, due to the possibilities of the radiographic method for visualization of the trabecular bone structure, as well as to the advantage of the observation of the whole bone and especially the zone of new bone formation in many different planes and projections. Therefore, using the native radiography as a method of choice in the monitoring of the whole new bone formation process, enables not only an optimal dynamics of surgical correction, but leads to a proper decision when to remove the external stabilizer and to allow weight bearing of the lengthened limb.

Radiographic classification of the patterns of newly formed bone at the distraction site is of great importance in predicting the final outcome of the treatment. Unfortunately, there are very few systems of radiographic classification which accurately present the progression of bone healing in different stages of limb lengthening. Most of studies, have the disadvantage of relatively small sample size and a relatively large number of influencing factors which limits the interpretation of their findings. The studies mostly analyze the results of radiographic classification systems in children and teenagers. The classification of Ru Li, Michael Saleh, Lang Yang, Les Coulton as a proved classification system in various age groups of patients treated with limb lengthening or lengthening with some minor deformity correction, is used in our study.(8) The most useful benefit of this classification is the ability to relate radiographic features at various stages to healing progress and possible problems that could lead to a poor outcome.

In our study the correlation of the clinical outcome in patients with different radiographic features of the regenerate within the above mentioned classification system was judged by the bone healing index (BHI). With some disadvantages in the study such as an obviously small sample size, our results present a large pleomorphism in the radiological presentations of the regenerate especially from the point of view of the feature of distraction osteogenesis. All the modalities from the used classification system had at least one representative case in our clinical material. The success in the clinical outcome judged by the values for BHI, was mostly dependent on the callus shape and on the feature of distraction osteogenesis, and in a big extent match the results in the literature.(5,7,11) Generally, a homogeneous pattern is more likely to lead to a good outcome, whereas a heterogeneous pattern is more likely to progress to a poor outcome. In vast majority of cases, homogeneous callus was generally seen in adolescents, whereas, in adults, it was the heterogeneous pattern of the newly formed bone at the distraction site.

CONCLUSION

Radiographic classification of the regenerate is of great importance in determining the features of bone at the distraction site throughout the whole process of new bone formation. The classification system based on callus shapes and types of radiographic features offers valuable data received from monitoring and recording of the healing process.Use of this classification system in distraction osteogenesis during limb lengthening can not only detect, but also predict the healing pattern of the bone and thereby allow therapeutic corrections to be made at the appropriate time in order to improve the final

outcome. This improvement in the treatment would enable earlier frame removal and full weight bearing of the limb.

REFERENCES

1. Babatunde O.M, Fragomen A.T, Rozbruch S.R.Noninvasive quantitative assessment of bone healing after distraction osteogenesis. HSS J2010;6(1):71–8
2. Kumar TS, Mahapatra S. Assessment of Regenerate in Limbs by Ilizarov External Fixation. Int J Sci Stud2016;4(4):97-101
3. Starr KA, Fillman R, Raney EM. Reliability of radiographic assessment of distraction osteogenesis site. J PediatrOrthop. 2004;24:26–29.
4. Catagni M. The radiographic classification of bone regenerate during distraction. Williams & Wilkins London 1991; 53–57.
5. Donnan LT, Saleh M, Rigby AS, et al. Radiographic assessment of bone formation in tibia.J PediatrOrthop 2002; 22:645–651.
6. Hamanishi C, Yasuwaki Y, Kikuchi H, et al. Classification of the callus in limb lengthening. Radiographic study of 35 limbs. ActaOrthop Scand. 1992; 63:430–433.
7. Minty I, Maffulli N, Hughes TH, et al. 1994.Radiographic features of limb lengthening in children. ActaRadiol.1994; 35:555–559.
8. Li R, Saleh M, Yang L, Coulton L. Radiographic classification of osteogenesis during bone distraction. Journal of Orthopaedic Research. 2006; 24(3):339-47.
9. Valentin Antoci, MD, PhD, Craig M. Ono, MD, Valentin Antoci, Jr., PhD, and Ellen M. Raney, MD. Pin-Tract Infection During Limb Lengthening Using External Fixation. Am J Orthop. 2008; 37(9):E150-E154.
10. Ilizarov GA: The tension-stress effect on the genesis and growth of tissues:The influence of stability of fixation and soft tissues preservation. Clin.Orthop. 1989; 238: 249-281
11. Speigelberg B, Parratt T, Dheerendra SK, Khan WS, Jennings R, Marsh DR. Ilizarov principles of deformity correction.Annals of the Royal College of Surgeons in England.2010;92(2):101–105

PSEUDOTUMOR FOLLOWING METAL-ON-POLYETHYLENE TOTAL HIP ARTHROPLASTY: A REPORT OF THREE CASES

ПСЕВДОТУМОР ПОСЛЕ МЕТАЛ НА ПОЛИЕТИЛЕН ТОТАЛНА АРТРОПЛАСТИКА НА КОЛК: ПРИКАЗ НА ТРИ СЛУЧАИ

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АПСТРАКТ

Вовед: Една од ретките компликации кај тоталната артропластика на колкот се псевдотуморите. Тие се дефинираат како грануломатозни или цистични лезии кои немаат инфективно или неопластично потекло. Обично постои латентен период од 2 до 15 години од имплантирањето на ендопротезата до клиничка манифестација на лезијата. Најчести симптоми се болка, оток и нелагодност, но исто така разлабавување на компонентите на ендопротезата или компресивна симптоматологија може да се манифестира. Потенцијалните механизми за настанување на псевдотуморите се реакција на страно тело, хиперсензитивна реакција и реакција кон партиклите од прекумерно трошење на компонентите.

Приказ на случаи: Прикажуваме три случаи на псевдотумори кај метал-на-полиетилен тотална артропластика на колк. Првиот случај е 76-годишна жена со болка и ограничени движења во колкот, 18 години после татлна артропластика на колкот. Вториот случај е 53-годишен маж кој презентира болка и оток во десната трохантерна регија, 21 година после тотална артропластика на колкот. Третиот случај е 55-годишна жена која презентира безболна, голема промена во проксималната надколеница, 13 години после тотална артропластика на колкот. Тројцата пациенти беа третирани оперативно со екстрипација на псевдотуморот.

Дискусија: Пациентите со болка во колкот или препоните, оток или колекција на течност после тотална артропластика на колкот треба внимателно да се евалуираат, посебно за присуство на инфекција. Незапознаеноста на клиничарите и радиолозите со псевдотуморите може да ги наведе на погрешно толкување на овие промени. Третманот на псевдотуморите е контраверзен бидејќи не постои јасен консензус за оптимален третман и надзор на оваа компликација.

Заклучок: Препознавањето на псевдотуморите е многу важно со оглед на зголемениот број на хируршки интервенции и последователно постоперативни компликации. Псевдотуморите се ретка, но важна компликација која се појавува кај сите типови на импланти кои се користат во артропластиката на колкот.

Клучни зборови: Псевдотумор, тотална артропластика на колк, метал-на-полиетилен

ABSTRACT

Introduction: One of the rare complications of the total hip arthroplasty are pseudotumors. They are defined as granulomatous or destructive cystic lesions with nonneoplastic and noninfective origin. Typically there is latent period of 2 to 15 years from the implantation of the endoprosthesis to the clinical manifestation of the lesion. Most common symptoms are pain, swelling and discomfort but also, loosening of the components of the endoprosthesis or compressive symptomatology can occur. Potential mechanisms of development of psudotumors are foreign-body reaction, hypersensitivity reaction and excessive wear debris reaction.

Case reports: We report three cases of pseudotumors following metal-on-polyethylene total hip arthroplasty. The first case is 76-year-old women presented with pain and limited motion of the right hip, 18 years after the hip arthroplasty. The second case is 53-year-old man presented with pain and swelling in the right trochanteric region, 21 years after the hip arthroplasty. The third case is 55-year-old women presented with painless, large mass in the left proximal thigh, 13 years after the hip arthroplasty. All three patients were treated operatively with extirpation of the pseudotumor.

Discussion: Patients with hip or groin pain, a mass, or a fluid collection following total hip arthroplasty should be carefully evaluated, especially for the presence of infection. Unfamiliarity with pseudotumors may lead clinicians and radiologists to misinterpret these masses as worrisome for malignancy. The management of pseudotumors is controversial since there is no clear consensus for optimal treatment or surveillance.

Conclusion: The recognition of pseudotumor is very important considering the increasing number of surgical procedures and consequential postoperative complications. Pseudotumors are a rare but important complication occurring in hip replacement surgery using all different types of implants.

Key words: Pseudotumor, total hip arthroplasty, metal-on-polyethylene

INTRODUCTION

Total hip arthroplasty revolutionized the treatment of hip osteoarthritis. The advancement in technology of bioengineering, component materials, fixation of both cemented and uncemented endoprosthesis and minimal invasive surgery led to very good long-term results. Not accidentally total hip arthroplasty is considered for the operation of the century.[1] The number of total hip arthroplasties is increasing. Only in USA, 270 000 primary total hip arthroplasties are performed each year.[2,3] With the increasing number of performed interventions the number of postoperative complications is on the rise. One of the rare late postoperative complications is development of pseudotumor around the hip joint. Pseudotumors are defined as granulomatous or destructive cystic lesions with nonneoplastic and noninfective origin, presenting near the components of the endoprosthesis and resemble tumor. They can present as small or large, solid or fluid filled masses with or without communication with hip joint.[4] Usually there is a latent period of 2 to 15 years following the initial total joint arthroplasty before the pseudotumor becomes clinically or radiologically apparent.[5] Most common symptoms caused by pseudotumors, regardless if it is metal-on-metal or metal-on-polyethylene are pain, swelling and discomfort but also loosening of the components, compressive symptomatology as neuropathy, venous compression, thrombosis or compression of other vital structures can occur.[4,6] Potential mechanisms of development of psudotumors are foreign-body reaction, hypersensitivity reaction and excessive wear debris reaction. The pathogenesis of pseudotumors developed in total hip arthroplasties with metal-on-polyethylene bearing is considered to be result of the reaction of the macrophages. Polyethylene debris is taken up by macrophage giant cells that release prostaglandin E2, which resorbs bone, causing the implant to loosen and leading to a vicious cycle of wear and loosening.[5,7,8] The pathogenesis of the pseudotumors developed in total hip arthroplasties with metal-on-metal bearing are explained with the delayed hypersensitivity reaction induced by metal,suggesting that a type-IV immune response plays a role in

Ipseudotumor pathogenesis.[9] The exact incidence and prevalence are not known Tallroth et al. obtained a lesion incidence rate of 4.6% in revision metal-on-polyethylene total hip arthroplasty with a predominance in males.[7] Some of the lesions are asymptomatic.

CASE REPORTS

Case 1

A 76-year-old woman was admitted to our clinic on February 2022 complaining of pain and loss of motion in the right hip. The pain started 2 years ago. 18 years ago she had undergone cemented total hip replacement because of fracture of the right hip using 28 mm metal-on-polyethylene femoral head. On physical examination pain during motion of the hip and shortening relative to the other leg was present. There was no suggestion of any sign of infection on physical examination. Laboratory analysis showed normal red cell counts, electrolytes and c-reactive protein. Plain radiographs showed signs of migration and loosening of the acetabular component of the endoprosthesis (figure 1). Osteolysis was also present. Computed tomographic (CT) scan revealed homogeneous, lobulated, mass surrounding the right acetabulum (figure 2). At the time of revision surgery, loosening and migration of the acetabular component was present and granulomatous tissue around the right acetabulum was encountered. Extripation of the mass was performed and reconstruction of the acetabulum with reconstruction cage was performed. New cemented polyethylene liner was implanted (figure 3). The patient had an uncomplicated postoperative course.

Figure 1: Preoperative plain radiography of the right hip

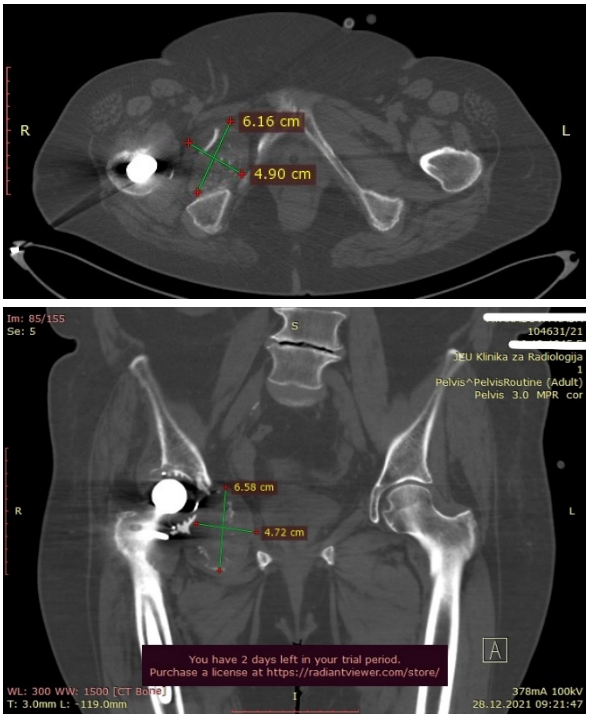


Figure 2: Computed tomography at the level of the lesion

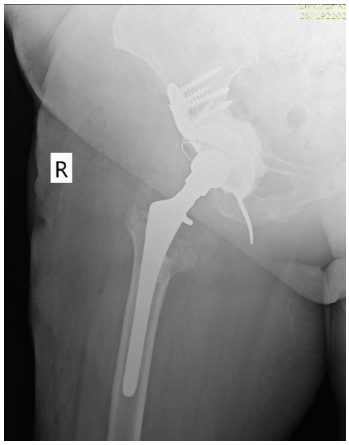


Figure 3: Postoperative plain radiograph

Case 2

A 53-year-old man was admitted to our clinic on May 2021 complaining of pain and swelling of the left hip. He described his pain mild to moderate. 21 years ago he had undergone uncemented left total hip replacement because of osteoarthritis using 28 mm metal-on-polyethylene femoral head. The used liner was ultra high molecular weight polyethylene (UHMWPE). On physical examination, an obvious and tender soft tissue mass surrounding the left hip, the left trochanteric region and extending slightly in the left buttock was present (figure 4). There was no suggestion of any sign of infection on physical examination. The patient had history of kidney transplantation. Laboratory analysis showed normal red cell counts and electrolytes. C-reactive protein was

elevated at 61.04 mg/L (normal value up to 6 mg/L). Serum urea was 10.39 mmol/L (normal 2.7-7.8mmol/L), serum creatinine was 144.74 mmol/L (normal 45-109 mmol/L) and serum ucric acid was 591.03 mmol/L (normal 150-450mmol/L). D-dimer was 669.9 ngr/mL (normal 0-500 ngr/mL). Plain radiographs showed no signs of migration or loosening of the components of the endoprosthesis (figure5). Computed tomographic (CT) scan revealed homogeneous, lobulated, fluid filled mass surrounding the left trochanter (figure 6). The measured size of the mass was 14 cm x 9 cm x 7 cm. At the time of revision surgery, thick granulomatous tissue around the left trochanter was encountered with no communication with the hip joint (figure 7). Extripation of the mass was performed and the tissue was sent to histologic examination. Histologic results confirmed the diagnosis of inflammatory pseudotumor. The patient had an uncomplicated postoperative course.

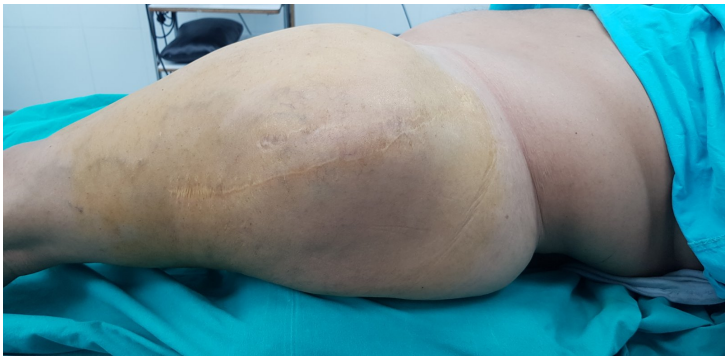


Figure 4: Preoperative site of the lesion

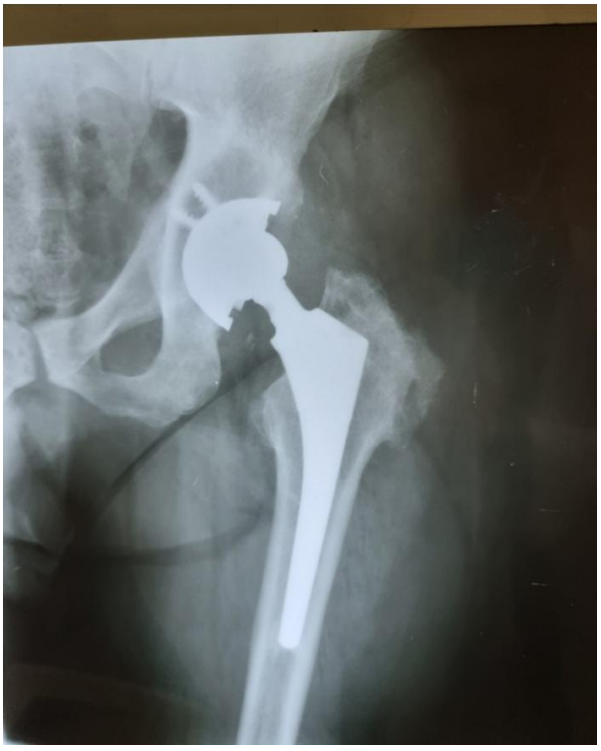
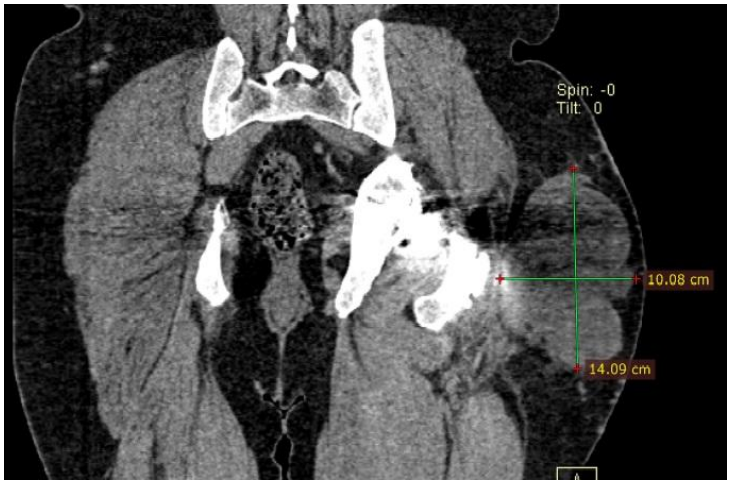


Figure 5: Preoperative plain radiography of the left hip



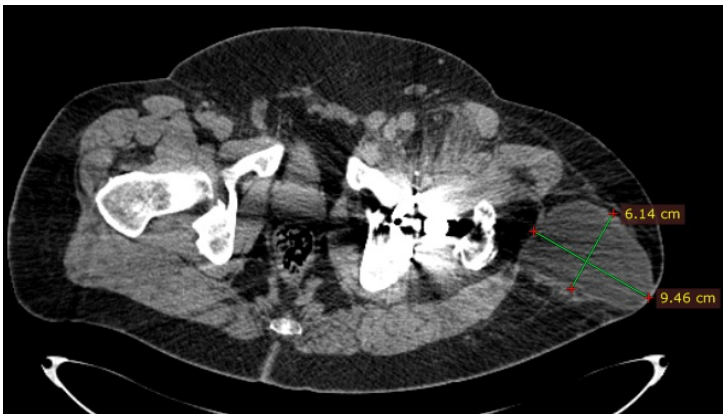


Figure 6: Computed tomography at the level of the lesion

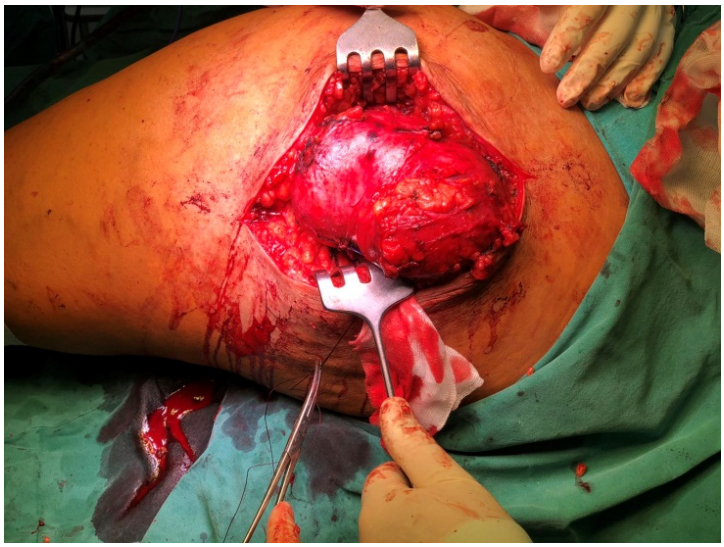


Figure 7: Intraoperative image before extripation of the lesion

Case 3

A 55-year-old women presented in 2019 in our clinic with a painless, large mass expanding the anterior and lateral aspect of the left proximal thigh. 13 years ago she had undergone left total hip arthroplasty because of osteoarthritis using 28 mm metal-on-polyethylene femoral head. The used liner was standard high density polyethylene. She had no history of wound problems, local warmth, fever or other symptoms possibly suggesting infection. Past medical history was essentially unremarkable. The physical examination at the time of admission showed painless, firm tissue mass filling the anterior and lateral aspect of the proximal thigh. Plain radiographs showed no signs of migration or loosening of the components of the endoprosthesis. The computed tomography revealed massive homogeneous lobulated soft tissue mass expanding around the anterior and lateral region of the proximal femur and hip joint. At the time of revision surgery, the thick granulomatous tissue connected with the hip joint was encountered (figure 8). Extripation of the mass was performed and the tissue was sent to histologic examination. Histologic results confirmed the diagnosis of pseudotumor. The patient had an uncomplicated postoperative course.

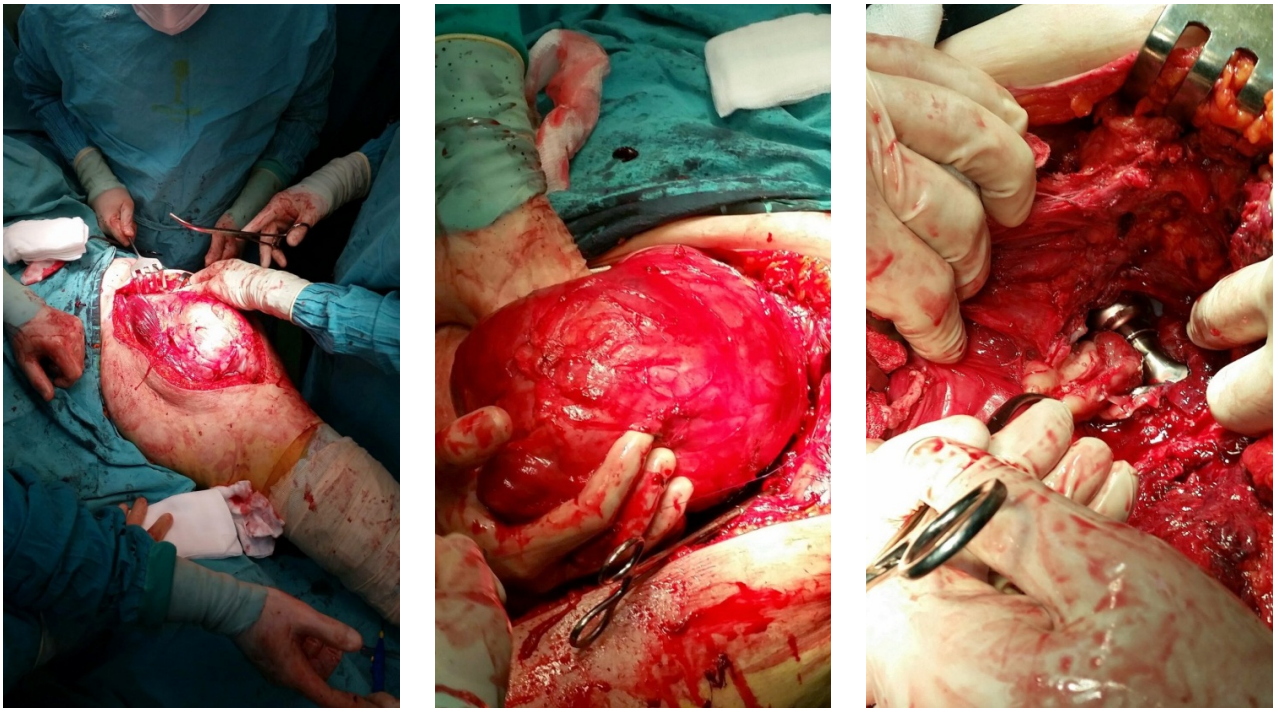


Figure 8" Intraoperative images before and after the extripation of the lesion

DISCUSSION

Pseudotumors are relatively rare complication following total hip arthroplasty. The clinical appearance may vary from asymptomatic to massive soft tissue masses accompanied with osteolysis and aseptic loosening of the endoprothesis which requires revison.[10] Patients with constant hip or groin pain, a mass, or a fluid collection following total hip arthroplasty should be carefully evaluated, especially for the presence of infection. The histological characteristics of an infection are distinctly different from those of an adverse immune reaction or an inflammatory foreign-body response. Up to now, there is no reliable blood or other screening test that offers a high predictive value for subsequent pseudotumor development.[11] Radiographic investigations are significant in visualizing pseudotumor size, location, communication with the joint and assessment of the stability of the implant. Plain radiographs have low sensitivity in detection of the pseudotumors compared to magnetic resonance imaging (MRI) which provides excellent evaluation of the periarticular soft tissues.[12] Anatomical classification system has been developed based on MRI findings on pseudotumors, consisted of three groups. Type I are thin-walled cystic masses (cyst wall <3 mm), Type II are thick-walled cystic masses (cyst wall >3 mm, but less than the diameter of the cystic component), and Type III are predominantly solid masses. [13] Computed tomography (CT) has advantages in visualizing bones, implants, bone cement as well as presence of heterotopic ossification, osteolysis, periprosthetic fracture and metalosis.[14] All types of

total hip arthroplasty (metal-on-metal, metal-on-polyethylene, ceramic-on-polyethylene) are associated with development of pseudotumors.[15,16,17,18] Pseudotumor formation has been even reported after unipolar hemiarthroplasty [19] The etiopathogenesis of the pseudotumors is not confirmed entirely but there are factors that are associated with their development. Those are excessive wear debris, foreign-body reaction andmetal hypersensitivity.[4] Wear debris is significant cause for development of pseudotumors. Wear debris is generated by mechanical wear, surface corrosion or combination of both and consists of both particulate and soluble form. Metal-on-metal articulations generate approximately 6.7 x 1012 to 2.5 x 1014 particles every year, which is 13,500 times the number of polyethylene particles produced from a typical metal-on-polyethylene bearing. Despite this finding, the actual volumetric wear of a metal-on-metal articulation is lower because of the very small size of the particles (generally <50 nm) compared with polyethylene particles, which are rarely <0.1 μm.[20] Howie et al. explored bursal masses and identified excessive wear of the polyethylene component as the cause of the masses. [21] Santavirta et al. and Austin and Stoney recognized excessive polyethylene debris as the primary factor in the causation of granulomatous reactions.[8,22,23] Willert et al. Also found that polyethylene wear products alone can cause massive osteolysis by triggering foreign-body granuloma formation at the bone-cement interface.[24] Pseudotumors may be caused by a foreign-body reaction to methylmethacrylate, polyethylene, or metal adjacent to a total joint implant. Polyethylene debris is taken up by macrophage giant cells that release prostaglandin E2, which resorbs bone, causing the implant to loosen and leading to a vicious cycle of wear and loosening.[4,5] Metal hypersensitivity reaction has been identified as one potential cause of pseudotumor development. There is an ongoing debate on whether the immune-mediated response is an adaptive response to excessive debris from high wear and is dose-dependent, or whether it is an innate hypersensitivity response, which is independent of dosage and is initiated at low levels of wear.[4] Histological analysis of the tissue specimens in the study by Willert et al. revealed an active cellular reaction with diffuse and perivascular infiltrates of lymphocytes and plasma cells, increased endothelial venules, fibrin exudation, accumulation of macrophages with drop-like inclusions, and infiltrates of eosinophilic granulocytes and necrosis. These histological findings were described asaseptic lymphocyte-dominated vasculitis associated lesion(ALVAL).[25] The absence of a metal-on-metal bearing does not preclude the formation of a pseudotumor in the hip Additional sites of metal-on-metal contact for hip prostheses are the head-neck taper junction and neck-stem junction of the femoral component of hip prostheses.[26] Psuedotumors may represent differential diagnostic dilemma. They must be differentiated form malign, benign and tumor-like lesions of bones and soft tissues. Despite the fact that periprosthetic primary malignant tumors are rare in the setting of hip arthroplasty, with an estimated incidence of 1.43/100,000, unfamiliarity with pseudotumors may lead radiologists and clinicians to misinterpret these masses as worrisome for malignancy. This situation not uncommonly leads clinicians to request biopsy. However, biopsy is not without risk, since pathologists unfamiliar with pseudotumors may be confused by the histology or may misinterpret tissue specimens as suspicious for malignancy.[27,28] Seroma and hematoma are differentiated from a pseudotumor by their development in the immediate postoperative period and subsequent resolution over time.[29] Soft tissue abscess is an additional periarticular mass which must be distinguished from pseudotumor. Local or systemic symptoms and signs related to pain, erythema, fever, malaise, and palpable mass prompt a clinical work up to rule out this diagnosis.[30] The management of pseudotumors is controversial since there is no clear consensus for optimal treatment or surveillance.[12] Treatment for pseudotumors in patients with metal-on-polyethylene implants varies. Pseudotumors in patients with a cemented total hip replacement can be satisfactorily treated with a cementless revision prosthesis combined with cancellous bone-grafting.[31,32] Surgeons can proceed to revision arthroplasty using techniques appropriate for aseptic loosening.[21] If there is no loosening of the components only extirpation of the lesion is performed.

CONCLUSION

The recognition of pseudotumor is very important considering the increasing number of surgical procedures and consequential postoperative complications. Pseudotumors are a rare but important complication occurring with all types of hip replacements. Potential causes of pseudotumors may include foreign-body reaction, hypersensitivity, and wear debris. There is no

clear consensus for optimal treatment, it may depend on the extent of lesion or the loosening of the endoprosthesis.

REFERENCES

1. Learmonth, I. D., Young, C., &Rorabeck, C. (2007). The operation of the century: total hip replacement. The Lancet, 370(9597), 1508-1519. doi:10.1016/s0140-6736(07)60457-7
2. Kurtz S, Ong K, Lau E, Mowat F, Halpern M. Projections of primary and revision hip and knee arthroplasty in the United States from 2005 to 2030. J Bone Joint Surg Am 2007;89:780-5
3. Davis, D. L., & Morrison, J. J. (2016). Hip Arthroplasty Pseudotumors: Pathogenesis, Imaging, and Clinical Decision Making. Journal of clinical imaging science, 6, 17. https://doi.org/10.4103/2156-7514.181493
4. Daniel J, Holland J, Quigley L, Sprague S, Bhandari M. Pseudotumors associated with total hip arthroplasty. J Bone Joint Surg Am. 2012 Jan 4;94(1):86-93. doi: 10.2106/JBJS.J.01612. PMID: 22218386.
5. Harry J. Griffiths; James Burke; Thomas A. Bonfiglio (1987). Granulomatous pseudotumors in total joint replacement. , 16(2), 146–152.doi:10.1007/bf00367764
6. Rabin DN, Smith C, Kubicka RA, Rabin S, Ali A, Charters JR, Rabin H. Problemprostheses: theradiologic evaluation of total joint replacement. Radiographics. 1987;7:1107-27.
7. Santavirta, S., Hoikka, V., Eskola, A., Konttinen, Y., Paavilainen, T., &Tallroth, K. (1990). Aggressive granulomatous lesions in cementless total hip arthroplasty. The Journal of Bone and Joint Surgery. British Volume, 72-B(6), 980–984. doi:10.1302/0301-620x.72b6.2246301
8. Austin, R. T., & Stoney, P. J. (1982). Granulomatosis of bone from high density polyethylene. Injury, 13(5), 414–418. doi:10.1016/0020-1383(82)90097-3
9. Grote, Caleb W et al. "Pseudotumor from Metal-on-Metal Total Hip Arthroplasty Causing Unilateral Leg Edema: Case Presentation and Literature Review." BioResearch open access vol. 7,1 33-38. 1 Mar. 2018, doi:10.1089/biores.2017.0035
10. Jeanrot, C., Ouaknine, M., Anract, P., Forest, M., &Tomeno, B. (1999). Massive pelvic and femoral pseudotumoral osteolysis secondary to an uncemented total hip arthroplasty. International Orthopaedics, 23(1), 37–40. doi:10.1007/s002640050300
11. Mont MA, Schmalzried TP. Modern metal-on-metal hip resurfacing: importantobservations from the first ten years. J Bone Joint Surg Am. 2008;90 Suppl 3:3-11
12. Van der Weegen W, Sijbesma T, Hoekstra HJ, Brakel K, Pilot P, Nelissen RG. Treatment of pseudotumors after metal-on-metal hip resurfacing based on magnetic resonance imaging, metal ion levels and symptoms. J0 Arthroplasty 2014;29:416-21
13. Hauptfleisch J, Pandit H, Grammatopoulos G, Gill HS, Murray DW, Ostlere S. A MRI classification of periprosthetic soft tissue masses (pseudotumours) associated with metal-on-metal resurfacing hip arthroplasty. Skeletal Radiol2012;41:149-55.
14. Cahir JG, Toms AP, Marshall TJ, Wimhurst J, Nolan J. CT and MRI of hip arthroplasty. Clin Radiol 2007;62:1163-71
15. Sculley WF, Teeny SM. Pseudotumor associated with metal-on-polyethylene total hip arthroplasty. Orthopedics 2013;36:e666-70.
16. Hsu AR, Gross CE, Levine BR. Pseudotumor from modular neck corrosion after ceramic-on-polyethylene total hip arthroplasty. Am J Orthop (Belle Mead NJ) 2012;41:422-6.
17. Mao X, Tay GH, Godbolt DB, Crawford RW. Pseudotumor in a well-fixed metal-on-polyethylene uncemented hip arthroplasty. J Arthroplasty 2012; 27:493.e13-7
18. Smith AJ, Dieppe P, Vernon K, Porter M, Blom AW; National Joint Registry of England and Wales. Failure rates of stemmed metal-on-metal hip replacements: Analysis of data from the National Joint Registry of England and Wales. Lancet 2012; 379: 1199-204
19. Whitehouse MR, Endo M, Masri BA. Adverse local tissue reaction associated with a modular hip hemiarthroplasty. Clin OrthopRelat Res 2013; 471:4082-6.
20. Keegan GM, Learmonth ID, Case CP. Orthopaedic metals and their potential toxicity in the arthroplasty patient: a review of current knowledge and future strategies. J Bone Joint Surg Br. 2007;89:567-73.
21. Howie DW, Cain CM, Cornish BL. Pseudo-abscess of the psoas bursa in failed double-cup arthroplasty of the hip. J Bone Joint Surg Br. 1991;73:29-32
22. Santavirta S, Konttinen YT, Hoikka V, Eskola A. Immunopathological response to loose cementless acetabular components. J Bone Joint Surg Br. 1991; 73:38-42.
23. Santavirta S, Hoikka V, Eskola A, Konttinen YT, Paavilainen T, Tallroth K. Aggressive granulomatous lesions in cementless total hip Joint Surg Br. 1990;

72:980-4.
24. Willert HG, Bertram H, Buchhorn GH. Osteolysis in alloarthroplasty of the hip. The role of ultra-high molecular weight polyethylene wear particles. Clin OrthopRelat Res. 1990;25.
25. Willert HG, Buchhorn GH, G¨obel D, K¨oster G, Schaffner S, Schenk R, Semlitsch M. Wear behavior and histopathology of classic cemented metal on metal hip endoprostheses. Clin OrthopRelat Res. 1996;329 Suppl:S160-86.
26. Erve, R. H. G. P. van. (2013). The absence of a metal-on-metal bearing does not preclude the formation of a destructive pseudotumor in the hip—a case report. Acta Orthopaedica, 84(6), 598– 599. doi:10. 3109/17453674.2013.862461
27. Visuri T, Pulkkinen P, Paavolainen P. Malignant tumors at the site of total hip prosthesis. Analytic review of 46 cases. J Arthroplasty 2006;21:311-23.
28. Singh C, Kaplan A, Pambuccian SE. Necrotic granulomatous pseudotumor following metal-on-metal hip arthroplasty: A potential mimic of sarcoma on fine needle aspiration cytology. DiagnCytopathol 2012;40 Suppl 2:E104-8.
29. Wu JS, Hochman MG. Soft-tissue tumors and tumorlike lesions: A systematic imaging approach. Radiology 2009;253:297-316.
30. Turecki MB, Taljanovic MS, Stubbs AY, Graham AR, Holden DA, Hunter TB, et al. Imaging of musculoskeletal soft tissue infections. Skeletal Radiol2010;39:957-71.
31. Eskola A, Santavirta S, Konttinen YT, Hoikka V, Tallroth K, Lindholm TS. Cementless revision of aggressive granulomatous lesions in hip replacements. J Bone Joint Surg Br. 1990;72:212-6
32. Tallroth K, Eskola A, Santavirta S, Konttinen YT, Lindholm TS. Aggressive granulomatous lesions after hip arthroplasty. J Bone Joint Surg Br. 1989;71:571-5.

GIANT CORPORAL LIPOMA TREATED WITH LIPOSUCTION

ГИГАНТСКИ ЛИПОМ НА ТЕЛОТО ТРЕТИРАН СО ЛИПОСУКЦИЈА

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АПСТРАКТ

Липомите се најчестите бенигни мезенхимални неоплазми со потекло од адипозното ткиво. Најчесто со јавуваат поткожно, заради што ја деформираат контурата на телесната површина и го менуваат природниот изглед. Заради тоа, најчеста индикација за нивно отстранување е козметското нагрдување. Традиционално, класичната отворена хируршка аблација е метода на избор за нивно лекување. Со цел да се отстрани липомот без лузни и да се смали постоперативниот морбидитет, во последните децении се развија нови техники за третман. Од нив, липосукцијата е доста популарна со постојан пораст на употреба заради лесната применливост, безбедноста, високата комплијанса на пациентите, но пред се заради малата оперативна инцизија/лузна и радикалност. Глобално, поткожните липоми се најчестата индикација за не-козметска апликација на липосукцијата.

Во трудов презентираме случај на гигантски поткожен липом на телото кој што успешно е отстранет со липосукција. Прикажани се постоперативниот тек, при што постигната е радикалност како и отсуство на рецидив во период од 36-месечното следење. Соодветна предоперативна дијагностика и евалуација е круцијална при селекција на липомите кои може да се третираат на овој начин.

Клучни збоорови: липом, липосукција, липектомија

ABSTRACT

Lipomas are the most frequent benign mesenchymal neoplasms that origin from adipose tissue. Usually arising from the subdermal fat, they deform body contour and affect natural appearance. Therefore, main indication for their removal is of cosmetic reason. Traditionally, open classical surgical ablation is a mainstay of their treatment. In order to achieve scar- less removal and decreased postoperative morbidity, many new techniques have been innovated in the last decades. Out of them, liposuction has gained popularity with increasing rate of utilization due to simple applicability, safety, high patient compliance, but mostly because of small incision/scar and eventual total removal. World widely, subcutaneous lipomas are the most common indication for non-cosmetic application of liposuction.

In this paper, we present a case of giant lipoma of the body that has been removed successfully with liposuction. Total tumor removal is achieved as shown in the postoperative period without a recurrence in a period of 36 months follow-up. Appropriate preoperative diagnostic and evaluation is crucial when selecting lipomas that can be treated in this way.

Key words: lipoma; liposuction; lipectomy

INTRODUCTION

With an incidence of 1-2.1/1000, lipomas are the most common neoplasms in human body, being a benign tumors of mesenchymal origin from the adipose tissue.[1] Mainly emerging from the subcutaneous fat, usually located on the trunk, extremities and nuchal area, they are easy to be palpated and inspected. These most frequent forms have classic appearance of soft, circumscript, painless and mobile solitary lesions, that concerns patients typically in 4th and 5th decade of their life, after a period of long lasting slow growth.[1,2] However, lipomas can develop at any age, at any region, at different organs and can have deeper and atypical localizations. An intraoseal, intramuscular, intradural and even lipoma in the kidney and spleen, have been descriped in the literature. [3] They can vary in dimensions; if larger then 10cm in diameter they are referred as giant.[2]

According to WHO classification of soft tissue tumors, histologically, there are many forms of lipoma, depending on the tissue that is co-present beside adipose cells. These comprise conventional lipoma, angiolipoma, lipomatosis of the nerve, lipoblastoma, hybernoma, mielolipoma, chondroidlipoma, spindle cell and pleomorphic lipoma [4]. Conventional lipoma, simple lipoma or common lipoma is the same term that reflects the clinical entity of the most common form.[5] When clinicians discuss about lipoma, they generally refer it to conventional type. It is composed of mature fat cells – adipocites without atypia that differ them morphologically from normal fat cells. The cells are arranged in lobules divided with fibro-vascular septa or trabecules. Fine capsule that envelopes the tumor is usually present. [4,6,7]

Anamnesis (history, pain growth), physical examination (mobility, bordering, tenderness) and tumor's features (location, consistency, smoothness, margins) are usually sufficient for getting the diagnosis done. However, basic work-up consisted of ultrasound investigation with linear probe and fine needle aspiration biopsy can be used to conclude it.[8,9] In superficially located cases, it is often enough to get the data needed. Further investigations can be used in doubtful cases, as their rarer malign counterparts, especially atypical lipoma and low grade liposarcoma, can present clinically quite similar.[10] Moreover, lipoma can transform into malignancy what is a rare scenario. MRI is a imaging technique of choice in this case, although a CT scan can be useful also. [11]

Patients seek operation mainly because of cosmetic concerns, body disfigurement, pain (when nerve is compressed), increased growth or due to cancer phobia [5-12] Treatment traditionally comprises open surgical ablationoften requiring larger incisions, longer postoperative morbidity and eventual postoperative complications[1,5] Based on the idea of less scarring and less postoperative morbidity,new modalities have emerged lately.[13-15] Among them, liposuction raised popularity since its introduction in mid-1970s as a minimal invasive approach with high patients' compliance and high satisfaction rate, being a safe and effective method as well. There have been many reports for liposuction assisted lipectomy in cases of small (< 4cm),[16] medium (4-10cm)[15,17-19] and giant lipomas (>10cm).[20-23] Safety standards of its application are well documented. [24] However, main concern is about the probable higher recurrence rate noted in some studies[20], but not in others.[19]

This paper presents a case of successful removal of conventional lipoma with liposuction and postoperative follow up of 36 months.

CASE REPORTS

We present a case of 61-years old male patient, with a corporal conventional lipoma treated with liposuction.

Patient noticed the condition twenty years ago, as a small lump with slow painless growth over the years. No other symptomswere reported except for the disfiguration of the body. Location is lateral aspect of thoraco-abdominal wall, with a portion on the back. He wanted the removal because of fear of its growth, which has been accelerated in the last years. He noted impairment with garments and aesthetic mutilation as well. Clinically, it is a mobile, subcutaneous, well – bordered tumor,soft and painless on pressurewhich is26x14cm gross.(Figure 1).Ultrasound examination with linear probe showed well – encapsulated soft tissue tumor suprafascially located over the muscles. Fine needle biopsy showed Ist classification group. These additional data were in consistency with the clinical evaluation and assumed benign tumor of lipomatous origin. Preoperatively, usual blood count was done and operation was planned in local anesthesia with mild sedation. Single shot wide spectrum antibiotic was given intravenously 30 minutes before the operation.

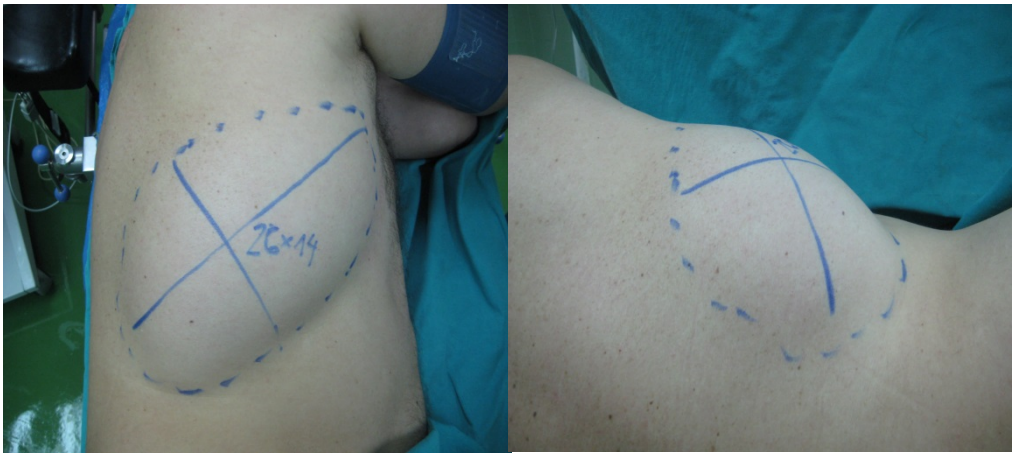

Figure.1: Preoperative photography and markings

Infiltration of 2ml 1% Lidocaine and 0.01% adrenaline in the planned 0.5cm incision was done. Few moments after, incision was made and infiltrating cannula was incorporated into the tumorous tissue. Tumescent infiltrating technique was used: about 200 ml of Klein's solution (0.1% lidocaine and 1:1 milion adrenalin in 1000ml 0.9% saline solution) was infiltrated under pressure in the lipomawith a blunt 3mm cannula until skingettingthe aspect of orange peel. (Figure 2a) After a waiting period of 10-15 minutes while gentle massaging, liposuction was conducted using 30cm long, blunt Mercedes 3mm cannula (Byron®) and manually made negative pressure with 60 cc syringe that fits the cannula. Manually made vacuum with syringeis enough for the liposuctionof a lipoma (Figure2b); no expensive equipment likepowered suction device or ultrasound isrequired. Liposuction is done until reaching the smoothness of the overlying skin and when bloody aspirate predominates in syringe as a typical sign of end point. Aspirate was filtrated and the hard part sent for pathohystological examination. Incision was closed with one resorptive subcutaneous suture and compressive dressing was applied. Patient was discharged same day from the hospital. Checkup was scheduled in 3 days with advice to wear compressive dressing and to continue usual obligations meanwhile. Wearing compressive garment (shirt with elastin) for 3 weeks was advised. Analgesics were prescribed in case of necessity.

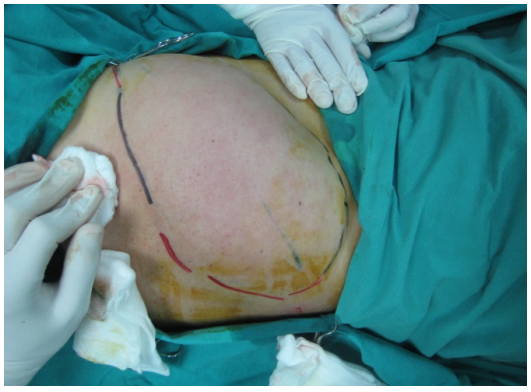

Figure 2a: Infiltration of the lipoma until orange peel aspect of the overlying skin.

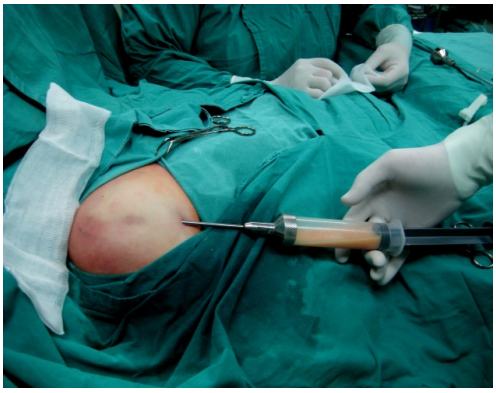

Figure 2b: Liposuction with syringe made vacuum

Patient generally felt well first critical days. He complained about mild pain that was increasing when changing posture, but not as strong as to take painkillers. On the first checkup, there was bruising and swelling over the plane that diminished in 2 weeks.(Figure3) Lipoma was removed totally. Over the period of one and three months checkups, there were no signs of recurrence, pain or skin irregularities. Incisional scar was almost invisible. Smoothness of the skin contour was achieved and patient was satisfied. After a year, excellent results can be seen (Figure 4): no signs of recurrence, superior skin alignment and almost invisible incisional scar. The same findings were noted on 36th month of follow up. Pathohystology results were in favor of lipomatous benign lesion - lipoma.

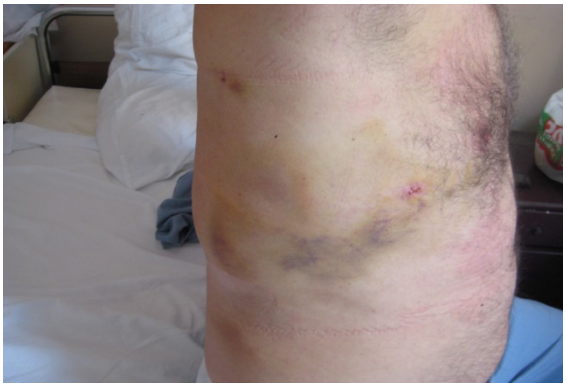
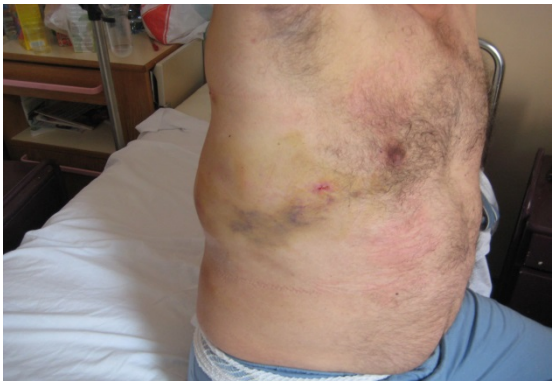

Figure 3: Postoperative bruising and swelling on 3th postoperative day

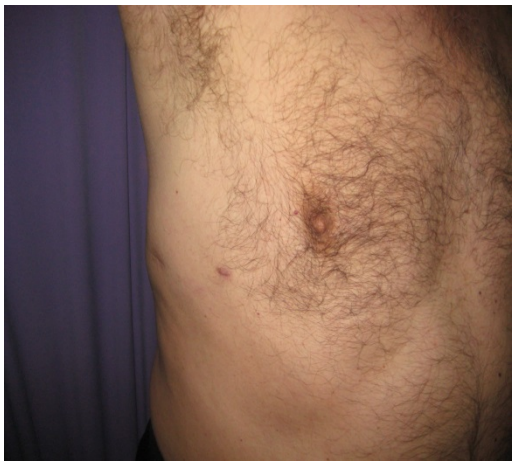
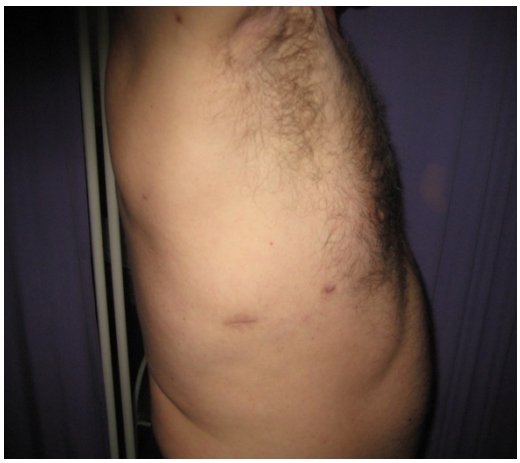

Figure 4: Appearance and result one year after op.

DISCUSSION

Lipoma are usually solitaire lesions, but can be multiple or diffuse as lipomatosis, or associated with other conditions as syndroms and congenital appearences (e.g. Madelung's, Dercum's disease etc.).[3] Male to female ratio is 1:2 [5] They appear usually after puberty, when fat accumulation is abundant, grow slowly and painless. [1,2] For the most common, conventional lipomas, etiology and pathogenesis is unknown though are more often in obese patients. Conventional lipoma, however, have typical chromosomic abberations without familial appearance and genetic transmissions[4]. Our case is a patient with conventional type of solitaire lipoma with negative familial anamnesis of the condition.

To diagnose lipoma usually is not difficult. Clinical presentation is of biggest importance prior diagnosis. Diagnosis "lipoma" emerges only if the mass is clinically present. [7] Encapsulated and well-defined, as benign lesions they are painless and slow growing. Still when the tumor is fast growing, painful and larger than 10 cm and/ or has atypical or deeper location, one have to bear in mind the possibility of sarcoma, most of which are atypical lipomas (or well -defined liposarcomas).[25] Imaging techniques such ultrasonography, CT and MRI as well as fine needle/core biopsy are useful applications. [8-12] MRI is highly sensitive in the detection of well-differentiated liposarcomas and highly specific in the diagnosis of simple lipomas.[10] Accurate diagnosis before any attempt for liposuction - assisted lipectomy is imperative. If accuracy is doubtful, tissue samples prior liposuction can be taken in manner of open biopsy.[12] However, fine needle aspiration biopsy is a high sensitive method for differentiating benign and malign soft tissue tumors.[9] Aspirate from the liposucted lipoma should be sent for pathologic examination. Studies have demonstrated that cell integrity in lipoaspirate is not damaged thus adequate pathohistology can be done accordingly.[26] Hereby, misdiagnosing liposarcoma of any type can be annulled successfully. Diagnostic pathway of our patient comprised anamnesis, clinical examination, ultrasonography, fine needle biopsy and microscopic examinations of the lipoaspirate.

In 1985, Rubenstein et al. were first to treat lipoma with liposuction[27], and so far, lipoma is the most frequent entity of its non – cosmetic application.[28] Advantages

are smaller scar, less pain, good cost – effectiveness, shorter operative time, better final surface contour, high patient compliance, ability to remove more lipomas through fewer incisions, small complication rate, ability to remove a tumor from distant operative site aesthetically acceptable.[28] Liposuction is indicated in suprafascial/subcutaneous, lipomatous masses, uni- or multilateral with size greater than 5 cm where diagnosis is well established. Smaller lipomas in areas where scar is to be avoided also can be removed.[16] Some authors opposed difficulties when removing giant lipomas[20] Our lipoma is giant and we did not have any problem; we had cannula long enough as to reach the very end border of the lesion. Difficulties might arise when a shorter cannula is used and in this case an additional counterincision is reasonable solution.

Minor sequels such are bruising, hematoma, immediate dimpling, light pain and swellings are concomitant to liposuction. They are mainly self-resolving. Infection is unusual. Liposuction is safe procedure when following guidelines. [24] On the other hand, open surgery includes larger scars, risk of hematoma, infections, seroma formation, dehiscence, hypertrophic scar, skin invaginations. The main concern about liposuction is its questionable ability to achieve radical removal of the lipomatous tissue and higher recurrence rate. Rubistein noted that it's difficult to remove the fibrous capsule with liposuction.[27] Raemdonck et all, in the only comparative study in the literature, showed higher recurrence risk in giant lipomas treated with liposuction compared to open surgery. This study consists of 30 cases. [20] Wilhelmi et al. treated small facial lipomas, with no recurrence in a follow up period from 11 months up to 10 years. However, this is non -comparative, prospective study consisting of only 5 cases. [16] Case reports of giant lipomas treated with liposuction showed no recurrences in 2 years follow up period.[21,22] Al-basty proposed capsule extirpation with forceps through liposuction incision or additional counter incision in larger lipomas as to prevent recurrences. In 6 years follow-up period of 16 patients, there were no recurrences.[19] Using the proposed technique in a study of 21 patients with 31 lipomas, Choi et all. found lipomatous remnants in three cases, no recurrences in 12 patients in a follow up period of 2 or more years.[17] According to their opinion, the three postoperative remnants occurred due to the hardness of tissue as location was dorsal region in all cases. One can notice, that Al-Bastyused dry technique of liposustion where end point lipectomy can be better visible, whereas Choiused tumescent technique, where lipoma is intentionally preoperatively infiltrated, thus end point lipectomy is less obvious. Both studies showed no late recurrences in long follow - up period. The proposed modification has also been applied in the largest andmost recent published study that reports no recurrence in 44 treated lipomas for mean period of 6 years follow-up.[29] In our case, there are no signs of recurrence in a period of three years follow up. In non- fibroticlipoma, as in our case, it's possible to destroy capsule mechanically and then liposucted. Otherwise forceps usage seems reasonable.

As comparative studies that compare open surgery versus liposuction - assisted lipectomy are lacking, one has to inform the patient about the probable higher risk of recurrence. The recurrence risk of open surgery is about 2%; there are not many cases in the literature about liposuction - assisted lipectomy that can estimated that risk of 2% or higher.[29] Therefore, these statements about higher recurrence rate are based on small series or are observational in nature. Nevertheless, in order the scientific truth to be concluded finally, larger randomized prospective studies are needed.

REFERENCES

1. Hodl S.Regionale und spezielle Erkrankungen des Fettgewebes.In: Braun-Falco O, Plewig G, Wolff HH, Burgdorf WHC,Landthaler M. Dermatologie und Venerologie.5. Auflage. SpringerVerlag,Heidelberg 2005; 1021-23
2. Koh HK, Bhawan J. Tumors of the skin. In: Moschella SL,Hurley HJ, eds. Dermatology. 3rd ed. Philadelphia: Saunders, 1992; 972-999
3. Shiffman AM. Lipomatous Disorders: Classification and Review of the Literature J CosmSurg Med2005; 5(3): 26-36
4. Fletcher CDM, Unni KK, Mertens F. Pathology and Genetics of Tumours of Soft Tissue and Bone, World Health Organization Classification of Tumours 2002
5. Bancroft LW, Kransdorf MJ, Peterson JJ et al. Benign fatty tumors: classification, clinical course, imaging appearance, and treatment.Skeletal Radiol 2006; 35:719-733
6. Kempson RL, Fletcher CDM, Evans HL, Henrickson MR, Sibley RS. Tumors of the Soft Tissues, Atlas of Tumor Pathology, AFIP Third Series, Fascicle 30, 2001

7. Shankar V.Soft Tissue Tumors, Adipose tissue, Lipoma.Available from www.pathologyoutlines.com, last updated 4 November 2012, accessed on 20th December 2017
8. RahmaniG.,McCarthyP.,BerginD.The diagnostic accuracy of ultrasonography for soft tisselipomas:a systematic review. ActaRadiologica Open 2017;6(6): 1-6
9. Iyer VK. Cytology of soft tissue tumors: Benign soft tissue tumors including reactive, nonneoplastic lesions. J Cytol 2008;25:81-6
10. Gaskin CM, Helms CA. Lipomas, Lipoma Variants, and Well-Differentiated Liposarcomas(Atypical Lipomas): Results ofMRI Evaluations of 126 ConsecutiveFatty Masses. AJR. 2004;182:733–739.
11. Burt AM, Huang BK. Imaging review of lipomatousmusuloskeletallesions.SICOT J 2017; 3, 34
12. Dalal KM, Antonescu CR, Singer S. Diagnosis and management of lipomatous tumors. J SurgOncol 2008; 97:298-313
13. Rotunda AM, Ablon G, Kolodney MS. Lipomas treated withsubcutaneous deoxycholate injections.J Am Acad Dermatol2005;53: 973–978.
14. Rittes PG The Use of Phosphatidylcholine for Correction of Localized Fat DepositsAesth. Plast. Surg 2003; 27:315–318
15. Pinski KS, Roenigk HH JrLiposuction of lipomas.DermatolClin.1990;8(3):483-92
16. Wilhelmi BJ, Blackwell SJ, Mancoll JS, Phillips LG.Another indication for liposuction: small facial lipomas.Plast Reconstr Surg.1999;103(7):1864-7
17. Choi CW, Kim BJ, MoonSE et al. Treatment of lipomas assisted with tumescent liposuction. JEADV2007;21: 243–246
18. Apesos J , Chami R.Functional applications of suction-assisted lipectomy: A new treatment for old disorders.Aest Plast Surg, 1991; 15 (1): 73-79
19. Al-basti HA, El-Khatib HA.The use of suction-assisted surgical extraction of moderate and large lipomas: long-term follow-up. 2002; 26(2):114-7
20. Raemdonck D, De Mey A, Goldsehmidt D: The treatmentof giant lipomas. Acta Chir Belg 1992; 92:213-217
21. Ilhan H, Tokar B. Liposuction of a pediatric giant superficial lipoma.J Pediatr Surg 2002;37(5):796-8
22. Nichter LS, Gupta BR. Liposuction of giant lipoma. Ann PlastSurg 1990; 24:362
23. Medina CR, Schneider S, Mitra BS, Spears J, MitraA.Giant submental lipoma: Case report and review of the literature. Can J Plast Surg 2007 ;15(4):219-222
24. Housman TS, Lawrence N, Mellen BGet al.The safety ofliposuction: results of a national survey. Dermatol Surg2002; 28: 971–978
25. Johnson CJD, Pynsent PB, Grimer RJ. Clinical features of soft tissue sarcomas. Annals of the Royal College of Surgeons of England 2001; 83(3):203–205
26. Shiffman MA, Mirrafati S. Fat transfer techniques: the effect of harvest and transfermethods on adipocyte viability andreview of the literature.Dermatol Surg 2001; 27(9):819–826
27. Rubenstein, R., Roenigk, H., Garden, J.M., Goldberg, N.S.,Pinski, J.B.: Liposuction for lipomas. J DermatolSurgOncol1985; 11(11):1070–1074
28. Coleman WP 3rd. Noncosmetic applications of liposuction. J Dermatol Surg Oncol 1998; 14(10):1085 1090
29. Copeland-Halperin LR, Pimpinella V, Copeland M. Combined liposuction and excision of lipomas:long-term evaluation of a large sample of patients. PlastSurgInt2015; Available at http://dx.doi.org/10.1155/2015/625396 Accessed 03.08.2017

TREATMENT OF NEUROMUSCULAR SCOLIOSIS A REVIEW

ТРЕТМАН НА НЕУРОМУСКУЛНА СКОЛИОЗА ПРЕГЛЕД НА ЛИТЕРАТУРАТА

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АПСТРАКТ

Кај децата со невромускулни заболувања често се развива сколиоза, која прогредира и по достигнување на скелетна зрелост. Невромускулната сколиоза влијае на квалитетот на животот, ја отежнува секојдневната нега на овие пациенти и може да предизвика кардиопулмонални компликации. Конзервативниот третман може да го успори напредувањето на деформитетот, но не се очекува да ги постигне целите на дефинитивен третман. Со напредокот на медицинските и хируршките техники во последните години, со повеќе пациенти со невромускулна сколиоза, со бројни коморбидитети и со со покомплексни деформитети се лекуваат оперативно. Во оваа прегледна студија ја разгледуваме достапната литература која го обработува хируршкиот третман на невромускулната сколиоза, со фокус на воспоставените принципи за индикациите и постапките на корекција на `рбетните деформитети кај невромускулните сколиози.

Клучни зборови: невромускулна сколиоза, спинална фузија, церебрална парализа

ABSTRACT

Children with neuromuscular disorders frequently develop scoliosis, which tends to progress even after skeletal maturity, significantly impairs their quality of life and daily care and may cause cardiopulmonary complications. Conservative treatment may slow curve progression, but usually cannot satisfy the needs as definitive treatment. With the recent advances in medical and surgical techniques, more neuromuscular scoliosis (NMS) patients with comorbidities and more complex deformities are being treated surgically. With this article we aim to review the available literature addressing surgical treatment of NMS, with focus on established principles of indications and procedures for deformity correction in NMS and review of different surgical techniques.

Key words: neuromuscular scoliosis, spinal fusion, surgery, cerebral palsy

INTRODUCTION

Neuromuscular scoliosis (NMS) occurs in patients with neurological or muscular disorders. In these disorders, due to poor muscular control of trunk and pelvis, weakness and spasticity, there is inadequate vertebral column support that leads to scoliosis development. Cardiac, gastrointestinal and respiratory compromise, as well as other musculoskeletal and neurologic conditionsare frequent in these patients and cause increased risk of perioperative complications. Therefore, management of the scoliosis requires careful planning and multidisciplinary approach.

The most prevalent cause of NMS is cerebral palsy (CP), followed by Duchene muscular dystrophy (DMD) and other central, peripheral neurologic or muscular diseases. 1Therefore, most scientific data on NMS comes from

studies of cerebral palsy patients and is generalized for all patients. However different diseases have specifics which should be considered when planning treatment of the scoliosis.

Rationale for operative treatment of NMS

Scoliosis incidence and progression

Overall incidence of NMS in patients with CP is reported to be 15%-25%.2NMS occurrence and progression is associated with the degree of neuromuscular involvement of the patient. The Gross Motor Function Classification System (GMFCS), a five-level system estimating functional capacity, where level I represents the highest level of function and level V the lowest is most commonly used to estimate this involvement.3 (Table 1) In a population-based study of 962 individuals, Hagglund et al. found 15% incidence of NMS. The incidence increased with age. They found that the overall frequency and severity of scoliosis increased with GMFCS level, from 0–1% in GMFCS level I to 42–55% in GMFCS level V based on clinical or radiographic examination. NMS appeared at younger age in patients with higher GMFCS level.4

Table 1. GMFCS classification

GMFCS level	Description
I	Walks without limitations. Limitations in more advanced motor skills
II	Walks without assistive devices. Limitations walking outdoors and in the community
III	Walks with assistive mobility devices. Limitations walking outdoors and in the community
IV	Self-mobility with limitations. Children are transported or use power mobility outdoors or in the community
V	Self-mobility severely limited even with the use of assistive technology. Limitation in maintaining antigravity head and trunk postures.

In other neuromuscular diseases, there is different incidence according to type of neuromuscular involvement and ambulatory status. For example, all children with spinal muscular atrophy (SMA) type 2 develop scoliosis at approximately 3 years of age and by the age of 10, average curve is more than 540, making all SMA type 2 patients candidates for scoliosis surgery at approximately that age. The incidence of spinal deformity in patients with DMD varies between studies. Corticosteroid treatment has been shown to improve outcomes and reduce scoliosis incidence and progression. In a study by Alman et al, 76% of untreated patientsand 17% of patients treated with corticosteroids developed a scoliosis with a curve > 20°.5

The rate of progression of the scoliosis curve depends on several factors. In adult, non-ambulatory patients with CP, it has been found to vary between 3.00 and 4.40 per year.6 Multiple studies show that curves progress most rapidly in non-ambulatory, quadriplegic patients (GMFCS IV and V).7,8 In a more recent study of spinal deformity progression assessing several radiographic parameters, in the GMFCS level IV–V group, the scoliosis Cobb angle and thoracic kyphosis angle increased by 3.4°and 2.2° per year, respectively and apical vertebral translation was increased by 5.4 mm per year. There was no significant increase in GMFCS levels I-III.8 Unlike idiopathic scoliosis (IS), where curve progression is unusual after skeletal maturity, in patients with NMS curves tend to progress beyond skeletal maturity.

The size of the curve is another factor that influences progression. (Figure 1) Gu et al. reviewed 110 paediatric patients with spastic quadriplegic CP. The authors determined that those patients with a deformity > 40° by the age of 12 were more likely to progress than those with a curve of ≤ 40° by the same age and consider this value as an indication for surgery.9 Another study showed thatrate of progression varies according to size of curve; larger curves (>50o) progress almost twice as fast than smaller curves (<50o).7 Thoracolumbar curves are more likely to progress compared to other localizations. 10

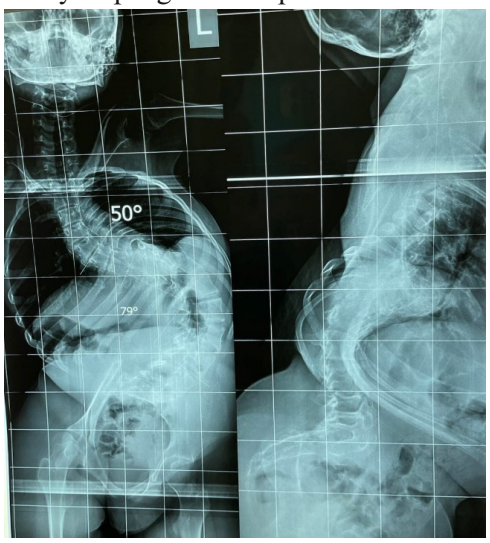

Figure 1. Large collapsing thoracolumbar curve in a 13-year-old female patient with unidentified type of muscular dystrophy.

Factors like height, weight, limb length inequality or hip dislocation, although considered important, have not been proven to influence scoliosis incidence or progression.

Ambulatory patients usually develop double thoracic and lumbar curves without pelvic obliquity (group I curves). (Figure 2) The typical scoliotic curve of NMS is a long, C – shaped collapsing curve affecting the thoracic and lumbar spine and causing pelvic obliquity (group II curve).

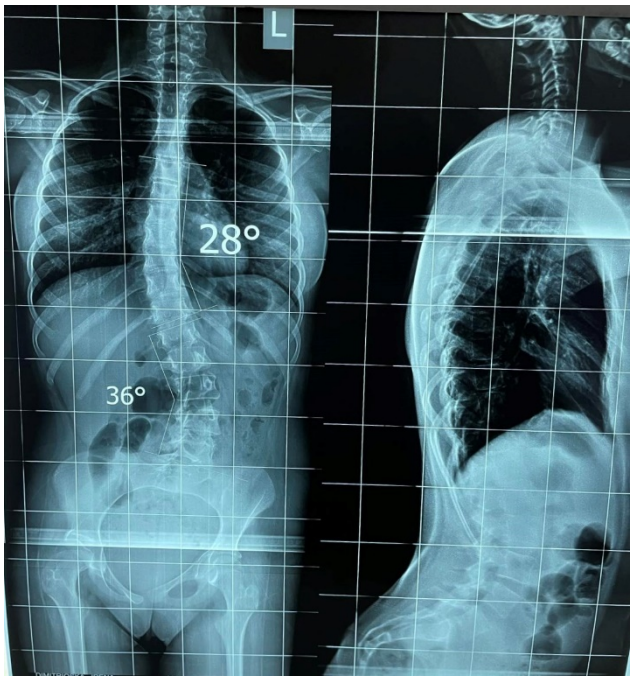

Figure 2. Ambulatory 16-year-old female with SMA type 3.

Neuromuscular scoliosis functional and clinical implications

The tendency of scoliosis progression, especially in children with impaired ambulatory function, leads to progressive loss of function and increasing difficulty with daily care. In larger curves, walking and sitting may become difficult and painful. Pelvic obliquity impairs seating, causes asymmetrical loading and may cause pressure sores in the buttock areas. In severe curves, ribs rest on the iliac crest, causing ulcerations in these areas. These changes cause sitting intolerance in wheelchair – bound patients, confining them to bed.

Severe rotation and shortening of the trunk, in addition to restricted thoracic compliance causes deterioration of cardiopulmonary function, which may add on to already impaired function caused by inherent muscle weakness due to primary disease.11 Feeding may also become more difficult, worsening preexisting poor nutritional status. Increased pain may exacerbate seizures. Associatedmedical co-morbidities such as gastro esophageal reflux, swallowing disorders, repeated aspiration may also get aggravated with progression of the spinal deformity.

Conservative treatment

Goals of treatment in NMS are curve progression control, improvement of posture and sitting, facilitation of daily care and alleviation of pain. Conservative treatment in NMS is directed at postponing operative treatment, but has been shown to have very limited effect in slowing down curve progression.

In a retrospective study of spinal orthosis in 86 patients with spastic quadriplegic cerebral palsy, there was curve progression at rate of 4.2O per year despite brace treatment. According to this study, curve progression slowed down in older patients and in patients with significant initial correction with the orthosis.[12] Previous studies also show that the effect of spinal orthoses on curve control is ambiguous.[13]

Bracing as a method for scoliosis control in NMS has been poorly tolerated by patients. Rigid orthoses cause pain and pressure ulcers. Therefore, some authors propose use of flexible braces or special brace manufacturing techniques, which differ from the concepts for brace production of IS.[14,15] Brace wearing in NMS should be restricted to time when patient is upright, and is not recommended during sleeping or lying down, in order to prevent further muscle atrophy or pressure sores.

The most important goal of bracing is attainment of trunk control, improved sitting posture and head and upper extremity control. This leads to improvement of overall activities of daily living in NMS patients, since arms are no longer used to support the body in sitting position. A study analyzed data from the Swedish National Cerebral Palsy Surveillance Program and Registry, which included 2800 children with CP. Of them 9% used spinal orthoses, which represents only 30% of the children with scoliosis. Almost all children at GMFCS levels III to V used spinal orthoses to improve function with one or more of the following goals: improve stability/ positioning; improve arm/hand function; or improve

head control and this goal was achieved in 90% of them. Only a third of the children wore the brace to prevent deformity. Deformity prevention was inversely proportional to scoliosis severity, with cases of mild scoliosis achieving greatest prevention.[16]

Findings of this study and several other studies suggest that bracing for deformity prevention should be focused on early prevention of mild deformities rather than prevention of collapse of large curves in later stages. [13,16,17]

In contrast, in patients with DMD, bracing is usually contraindicated and early surgery is favored, because it rarely if ever stops the progression of the curve. In cases where it slows the progression of the curve, surgery becomes indicated later in life, when pulmonary function has declined and the risk for cardiopulmonary complications is significantly increased.[18]

Adapted seating in specifically designed wheelchairs or "sitting shells" is another option for obtaining sitting position, head and upper extremity control in patients with NMS. Wheelchair adaptations include shoulder/waist harnesses, lateral chest support or modular seating systems and head support. These devices do not provide deformity correction and may be used in adjunction to bracing. On the other hand, for patients with severe neuromuscular involvement, they may be better tolerated than braces, are simple to use for carers, and do not restrict respiratory movements, which is particularly important for children at high risk for chest infections.[19]

Medicaments have been shown to prevent scoliosis development and progression in certain types of NMS, by controlling the underlying disease. Prolonged steroid treatment significantly increased survival rate and decreased incidence of scoliosis in patients with DMD in several studies.[5,20] Recent studies show significant improvement of motor function in SMA patients treated with intrathecal antisense oligonucleotide medication – nusinersen, but control of scoliosis progression has not been proven. [21](Figure 3) Intrathecal application of baclofen via a pumps used to control spasticity in CP patients and other neuromuscular disorders and has been shown to positively affect function, however it has been associated with scoliosis development or progression. Studies suggest that close monitoring for NMS is needed in patients with intrathecal baclofen pumps. [22]

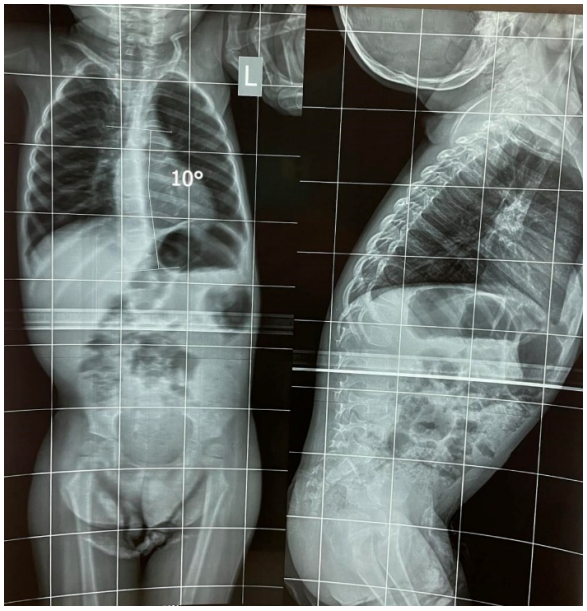

Figure 3. A 3-year-old boy with SMA type 1, treated with nusinersen (Spinraza). There is yet insufficient data to predict development of the deformity in treated SMA type 1 with prolonged life expectancy.

Optimal timing for NMS scoliosis surgery

Scoliosis surgery in patients with NMS depends on the underlying condition, the functional capacity and needs of the patient and the long-term prognosis. In ambulatory patients with NMS, surgery aims to obtain and maintain normal spine balance and preserve ambulatory function. In non-ambulatory patients, progressive deformity with truncal shift or pelvic obliquity that causes sitting difficulties, pressure sores or diminishing pulmonary and gastrointestinal function are reasons for which surgical treatment is indicated. (Figure 4)

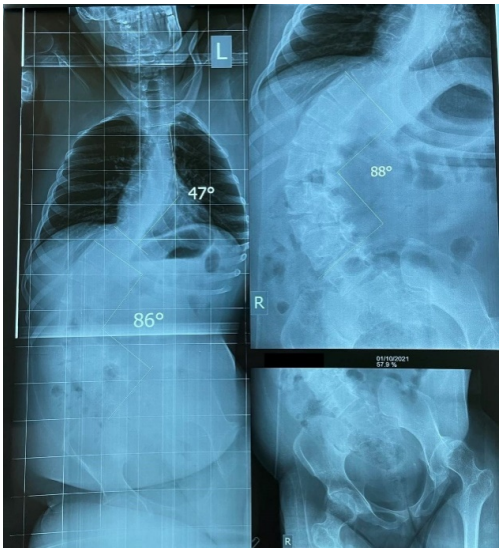

Figure 4. A 22-year-old male with SMA type 2, with large thoracolumbar curve and pronounced pelvic tilt impeding seating positioning.

In CP, curve progression is maximal during puberty and usually this is the period when surgical treatment is obtained. Surgical intervention is considered for curves larger than 40o and significant deterioration of function. Curves larger than 50o are considered unstable and expected to collapse even after skeletal maturity and should be fixed. Early onset curves should be treated expectantly and with bracing until sufficient skeletal maturity (i.e., closure of the triradiate cartilage) has been obtained. Early spinal fusion should be delayed in early onset curves as much as reasonably possible, in order to allow for Th1-Th12 growth and maximization of the size of the ribcage. If the scoliotic curve in CP remains flexible, larger curves may be permitted to develop in order to allow for sufficient growth. Flexible curves of 60-90 degrees are relatively easy to reduce in CP. When the curve becomes stiff, this is indication for surgery.

The relationship between growth and respiratory function was studied extensively in several papers, that showed a direct correlation between the results of respiratory function tests and the height ofthe T1-T12 segment measured on skeletally mature patients who had been operated on as children for congenital scoliosis.[23,24] These studies showed that spinal fusion over more than 60% of thoracic spine before 8 years of age reduced vital capacity for more than 50%.

In addition, patients with dystrophies have reduced thoracic compliance, making their respiratory tolerance to deformity lower than in patients with IS with the same Cobb angle. In patients with NMS, respiratory problems are not always proportional to the Cobb angle and to trunk collapse, as evidenced by the Th1-Th12 distance.[25] However, it is certain that when Cobb angle exceeds 100o, FVC is reduced by 30%; above 120o, there is a risk of respiratory failure and chronic pulmonary heart disease.

In patients with DMD, Roberts & Tsirikos suggest surgery should be considered in patients with progressive spinal curvatures greater than 25 degrees in order to prevent respiratory compromise produced by the deformity.[1] Other authors also state thatIf spinal fusion is the desired treatment for scoliosis in the large majority of DMD patients not on steroid treatment, the optimal time for correction is when the deformity is between 20° and 30°.[26] With time, the curve deteriorates, as do respiratory and cardiac function, making surgical intervention more difficult.

Additionally to performing a pulmonary function test, regular monitoring of the Th1-Th12 height has been suggested as an indicator for the seriousness of the situation and the effects of treatment. This distance should be greater than 20 cm at skeletalmaturity to avoid severe restrictive lung disease.[24] These concepts of restrictive lung disease and scoliosis have recently been named “thoracic insufficiency syndrome” by Campbell and Smith.[27] This condition occurs when the thorax no longer allows normal breathing or harmonious lung growth.

Studies show that reduced lung function may happen due to a less compliant thoracic cage because of the stiffening caused by the thoracic instrumentation. Thus reducing the Cobb angle does not always result in better respiratory function.[28]

What is the optimal surgical approach for NMS scoliosis surgery?

Spinal fusion

Surgical treatment of NMS aims to obtain solid bony union of the spine to prevent deformity progression, to correct the deformity of spine and pelvis and to restore standing or seating balance. In ambulatory patients with CP, curve characteristics and biomechanics are similar to IS, and in these patients the principles of treatment of IS can be used. However, majority of NMS surgery is performed in non-ambulatory patients, in which the ultimate goal is to maintain upright posture and sitting balance. In these patients, technical considerations are different with regards to proximal and distal extent of instrumentation and pelvic fixation, as well as the surgical approach used. Also, in these patients, preoperative planning should include careful evaluation of cardiopulmonary function and effects of anesthesia.

The surgical approach for treatment of NMS has changed significantly over the past few decades. Although combined anterior – posterior approaches (ASF/PSF) were used traditionally,[29] with the evolution of posterior fixation systems, most cases now can be treated with posterior – only surgery – posterior spinal fixation (PSF). Many recent studies have shown that use of posterior-only pedicle screw constructs offers excellent curve correction with a minimal complication rate when compared with the anterior–posterior approaches.[30,31]

The choice of low-profile polyaxial or monoaxial transpedicular screws alone or in combination with sublaminar hooks and wires and cobalt-chromium rods allow for increased correction forces needed to resist the lever arms of long curves in the setting of poor muscle control. Reduction screws only can be used, while keeping in mind that correction maneuvers should be performed with force distribution over few segments, in order to avoid screw loosening or dislocation in the osteoporotic spine of these patients. With these screws, both rotational and translation deformity can be corrected. Correcting lumbar lordosis is a very important step in NMS deformity correction, since it is important for balancing walking pattern in ambulatory patients and seating balance in those who are wheelchair bound. Different studies show good results with diverse fixation techniques. Lowenstein et al. observed a non-significant trend toward better correction of the main thoracic curve in all - screw versus hybrid hook-screw instrumentation. [32] According to a study by La Rossa et al, hybrid constructs (lumbar screws, thoracic acrylic clamps, thoracic hooks at the upper end of the curve) appear safe and effective and their mechanical performance is comparable to all-level screws construct. In addition, they offer shorter operative time and low vascular and neurologic risks combined with a satisfying stability.[33]

Due to upper thoracic kyphosis, poor head control and poor muscular control, the rate of junctional kyphosis and loss of fixation at the cephalad end of instrumentation in NMS surgery ranges between 30%-62%.[34] Therefore instrumentation in NMS should end proximally at least at Th2 level. For these uppermost levels, hybrid constructs with pedicle screws and hooks can be used. Also, in this area, care must be taken to preserve the soft tissues as much as possible and to not fully correct the local cervico – thoracic kyphosis.[35]

In ambulatory patients with no pelvic tilt and in a selected subset of non-ambulatory patients with pelvic tilt below 15o, instrumentation can stop at the L4-L5 level. This will allow for mobility of the lumbosacral junction, which is necessary for proper gait and will provide easier sitting positioning. In addition, stopping the instrumentation at this level shortens surgery time, decreases bloodloss, nonunion rates and infection, as well as hardware prominence. However, in non-ambulatory children with significant spasticity, even with mild pelvic tilt, stopping at L4-L5 is a risk due to the recurrence and progression of pelvic obliquity.[36] Fusion to the pelvis should be performed in children in which the apex of the curve is at L3 or lower, those with lumbosacral junction abnormalities and those with pelvic tilt above 15°. [37] S1 bicortical screws with iliac screws or double iliac screws can be used. Also S2 alar iliac screws can be used instead of traditional iliac screws, eliminating the need for offset connectors, which have been proven to be a weak spot where these implants frequently fail.38 These fixation points should not be used for reduction maneuvers and supplemental iliac screws with intraoperative distraction rods or halo - femoral traction should be considered.[39] Halo-femoral traction in cases of severe scoliosis above 100° provides good deformity correction, but it also elongates the thoracic cavity improving the compromised pulmonary function.[40]

In the past, ASF was used in order to achieve correction goals in rigid curves. However, recently, the use of advanced instrumentation techniques and different posterior osteotomies, including Ponte osteotomies, pedicle subtraction osteotomies and Posterior Vertebral Column Resection (PVCR) have largely diminished the role of anterior surgery. In extremely severe and stiff curves, PVCR has been proven to give good results but with a high risk of major complications, as was shown by Sponseller et al. in hisseries of 23 children with neuromuscular scoliosis.[41]Aydinli et al. treated 20 patients (4 with NMS) with severe 3-dimensional deformity with flexibility of less than 20%, who underwent PVCR by the modified Suk technique. They achieved 60% scoliotic curve correction and 62% kyphotic curve correction, with average blood loss of 1072mL and no neurologic complications. They conclude that modified Suk technique of PVCR reduces the potential for neurologic injury and 60% of blood loss happens during and after posterior bone resection.[42]

The addition of an anterior approach, particularly with 3-column pedicle screw fixation, is reserved for patients with very stiff or rigid spinal curves, young patients at risk for crankshaft phenomenon, patients with dysplastic posterior elements, and large spinal curves requiring anterior release or shortening of the anterior column to achieve the deformity correction goals. Menga et al. analyzed 5191 adult CP patients who underwent spinal fusion between 2001 and 2013 in the USA. The majority underwent PSF (86.5%), followed by the ASF/PSF approach (9.3%) and the rest had anterior surgery only. They concluded that only the rate of PSF surgery in the USA increased significantly during the studied period,

whereas ASF and ASF/PSF numbers remained the same. Combined ASF/PSF techniquewas associated with the longest hospital stays,highest hospital charges, and increased complications.[43]

The Dutch guideline for the treatment of scoliosis in neuromuscular disorders recommends that the indication for surgical correction of scoliosis should be made early in DMD and SMA type 2 patients (Cobb's angle 20°), such that surgery is less complicated, shorter, and safer and it is more likely that the pelvis can be left out of the fusion trajectory. In DMD, PSF should be performed from high thoracic (Th2-3) at least to L4 or L5. Pelvic fixation gives better possibilities to correct severe curves, but increases time of surgery and blood loss. In SMA type 2 PSF should be performed. The technique varies with age and should consider possible remaining growth. Instrumentation should be segmental. Allograft bone can be used supplemented with locally harvested autologous bone or bone substitutes.[44]

Implant density has been studied mostly for IS, with studies showing conflicting results of high and low implant density constructs, with better correction and maintenance of correction considered as advantages of high density constructs, as opposed to shorter surgery times, less blood loss, less screw insertion complications and decreased costs for the low density constructs.[45] Wolfram et al. investigated implant density in 26 patients, of which 10 had NMS. Average implant density was 88% and they found no differences in correction rates, pre and postoperative kyphosis, surgery time, blood loss, ICU stay or complications between the high and low – density group. They conclude that implant density of over 70% can be suggested as adequate operative treatment for idiopathic and neuromuscular scoliosis.[46]

Fusionless techniques

In young patients with large but flexible curves, growth preservation techniques can be considered. Most frequently used systems are growing rods, as a distraction – based technique. Here, they are applied in a setting of muscular imbalance and frequently poor soft tissue coverage in a malnourished child. Therefore, certain guidelines that refer specifically to NMS patients have to be respected in order to avoid mechanical and other complications. Spinal fixation should use multiple solid anchors. Fixation should not be onto the ribs, which are fragile, but rather onto the vertebrae, with supra- or sub-laminar hooks, which resist detachment better than screws. Proximal anchorage should extend up to the first thoracic or even last cervical vertebrae, to avoid the risk of proximal junctional kyphosis. In non-ambulatory patients, distal fixation should be supported by the pelvis, with solid, stable, small anchorage. Pelvic fixation by ilio - sacral screws meets these requirements. Spinal assembly should be fairly symmetrical, to neutralize balance disorder in the trunk and pelvis and distribute stress. This means it must be bilateral, supported by the pelvis. To reduce operative risk in fusion, surgery should be as non-invasive as possible, restricting the approach to two short incisions over the anchor points.[28] (Figure 5)

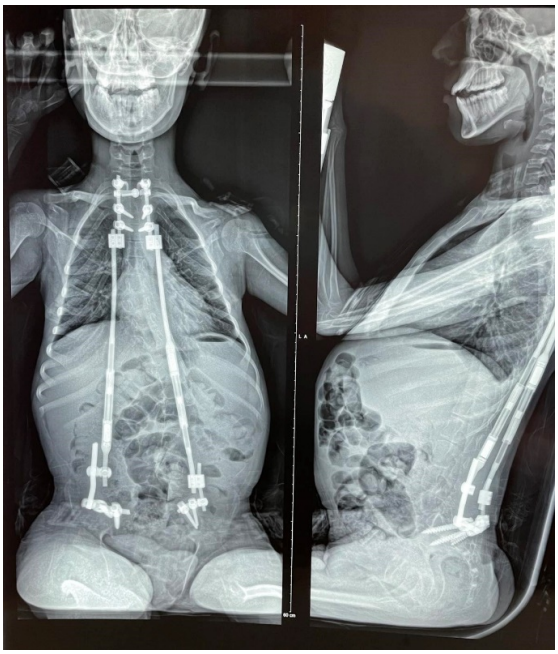

Figure 5. Double growing rod technique used in a 10-year-old patient with SMA type 2.

In a study of 100 NMS patient treated with fusionless surgery from T1 to the pelvis (bilateral double rod sliding construct anchored proximally by four hooks claws and distally to the pelvis by iliosacral screws through a minimally invasive approach), Miladi et al obtained significant correction of spinal deformity and pelvic obliquity during minimum 2 years of radiographic follow up. 26% of the patients experienced complications, which included but were not limited to 12 implant related complications and 16 infections, 2 of them necessitating implant removal.[28] In another study of 59 SMA patients that underwent minimally invasive fusionless surgery with bilateral sliding rod construct from T1 to the sacrum, anchored proximally by double-hook claws and distally by iliosacral screws, Gaume et al. showed significant correction of spinal deformity and pelvic obliquity, and

reduced rate of complications. The correction of spinal deformity was maintained at long term, not requiring definitive fusion at the end of growth. [47]

Enercan et al. have proposed a modified growing rod technique (while using some of the principles of the SHILLA technique of guided growth), where apical and intermediate anchor screws are added with muscle – sparing technique and correction is achieved with intraoperative halo – femoral traction. The most proximal and most distal screws were fixed and the rest of the screws were left with nonlocked set screws to allow vertical growth. The lengthening reoperations were performed every 6 months. They postulate that these screws will stop apical rotation and provide better sagittal and coronal curve control, while decreasing implant – related complications.4[8]

Another distraction - based technique is the Vertical expandable prosthetic titanium rib (VEPTR) which has been used to treat early onset NMS in recent years. In VEPTR, which can expand the ribs and thoracic cage and promote the growth and development of the ribs, a titanium rib distractor is placed on the concave side of the spinal curve, indirectly correcting thoracic scoliosis in children. Because scoliosis in children is often associated with thoracic insufficiency syndrome, VEPTR can be used to correct the deformity of spine and chest while preserving the capacity for growth of the spine and lungs. Studies show satisfactory deformity correction with complication rates that are comparable to other distraction – based techniques.[49,50]

Guided growth systems can also be used in carefully selected subsets of NMS patients. Modified Luque trolley technique with segmental sublaminar spinal instrumentation without fusion has been shown to be safe and effective in treating NMS in low – tone NMS patients, without need for multiple distraction – based surgeries or auto fusion.[51]

Outcomes and complications

Complications following NMS surgery are more frequent compared to IS surgery, due to characteristics of primary illness and variety of comorbidities these children have. In a large retrospective study comparing children with NMS and IS after spinal surgery, those with NMS had longer lengths of hospital stay, higher total charges, higher complication rates, higher number of secondary procedures, and higher rates of disposition to other facilities or to home health care services.[52]

The risk of pulmonary complications in neuromuscular patients following spinal surgery is well known and includes respiratory failure, aspiration, pneumonia, pneumothorax, or pleural effusion. Scoliosis surgery itself causes transient decrease of vital capacity. A meta-analysis of 15,218 patients with NMS showed that pulmonary complications are the most prevalent (22.71%), followed by implant-related complications (12.51%), infections (10.91%), neurological complications (3.01%), and pseudoarthrosis (1.88%).[53] Patients with NMS have 10-fold increased risk for infectious complications compared to their IS counterparts, as shown in a study of 1347 procedures performed in 946 patients with different scoliosis etiologies. In this study, non-idiopathic scoliosis and extension of instrumentation to the pelvis were identified as a risk factors.[54]

NMS patients bleed more during surgery than IS patients. Studies show that these patients are at 7-fold bigger risk for losing more than 50% of their blood volume which in itself increases risk for other types of complications.55 Anti-fibrinolytics agents, in particular tranexamic acid, significantly reduce the intraoperative estimated blood loss associated with posterior spinal fusion with no adverse effects.

Perioperative mortality is high in patients with NMS. Numbers vary greatly, depending on study, follow-up period and underling condition. Mortality from 1% up to 28% for CP have been reported in the literature.[55–57]

Assessing outcomes can be difficult for NMS patients, who very frequently can not respond appropriately to patient reported outcome tools. Also, even though complication rates of surgery are high, studies show that there is high level of satisfaction with surgery. A systematic review showed that for NMS patients with spastic quadriplegia, parent or caregiver satisfaction with surgery ranged from 85 to 91.7%. In one of the analyzed studies, 91.7% of the parents or caregivers reportedthat they would repeat the procedure under thesame conditions and another study reported that 95.8% of parents reported that the benefits of thesurgery offset the risks.[58] In the general population of CP patients, satisfaction of parents and caregivers goes up to 99%, although studies cannot clearly demonstrate improvement of function, school attendance or decrease of

comorbidities.5[9]

Quality of life is perhaps the most important outcome measure in any postoperative NMS patient. In a systematic review, the evidence suggests an improvement in postoperative quality of life in CP and muscular dystrophy patients who underwent scoliosis surgery. Nevertheless, the authors commented there are conflicting reports and the literature is currently lacking well-controlled, prospective studies. [60]

CONCLUSION

The advances of understanding neuromuscular disorders and advances in implants and surgical techniques caused increase in numbers of NMS surgery in the past decades and have provided options for complex reconstructive procedures while lowering the risk for complications and mortality in these fragile patients. However, since complication rates are still high, careful individual assessment of each patient's physiological status and needs should be performed and parents and 'caregivers' wishes and expectations should be taken into consideration when deciding for scoliosis surgery. Timing of surgery should depend not only of deformity magnitude, but also on possibilities for optimization of physiological and nutritional parameters, while having in mind the natural course of the underling condition that may lead to further physiological deterioration. Proper assessment of patient physiology and biological, mechanical and 3-dimensional characteristics of the spinal deformity should be undertaken in order to choose appropriate correction and fixation technique for each case respectively.

REFERENCES

1. Roberts SB, Tsirikos AI. Factors influencing the evaluation and management of neuromuscular scoliosis: A review of the literature. J Back Musculoskelet Rehabil. 2016;29(4):613-623. doi: 10.3233/BMR-160675
2. Persson-Bunke M, Hägglund G, Lauge-Pedersen H, Ma PW, Westbom L. Scoliosis in a total population of children with cerebral palsy. Spine (Phila Pa 1976). 2012;37(12). doi:10.1097/BRS.0b013e318246a962
3. Palisano RJ, Rosenbaum P, Bartlett D, Livingston MH. Content validity of the expanded and revised Gross Motor Function Classification System. Dev Med Child Neurol. 2008;50(10):744-750. doi: 10.1111/ j.1469-8749.2008.03089.x
4. Hägglund G, Pettersson K, Czuba T, Persson-Bunke M, Rodby-Bousquet E. Incidence of scoliosis in cerebral palsy: A population-based study of 962 young individuals. Acta Orthop. 2018;89(4):443-447. doi:10.1080/17453674.2018.1450091
5. Alman BA, Raza SN, Biggar WD. Steroid Treatment and the Development of Scoliosis in Males with Duchenne Muscular Dystrophy. J Bone Jt Surg - Ser A. 2004;86(3):519-524. doi:10.2106/00004623 - 200403000-00009
6. ME M, DS M, RT H. Natural history of scoliosis in the institutionalized adult cerebral palsy population. Spine (Phila Pa 1976). 1997;22(13):1461-1466. doi:10.1097/00007632-199707010-00007
7. JG T, SR S. Progression of scoliosis after skeletal maturity in institutionalized adults who have cerebral palsy. J Bone Joint Surg Am. 1988;70(9):1290-1296. https://pubmed.ncbi.nlm.nih.gov/3182881/. Accessed August 16, 2021.
8. Yeol S, Chin L•, Chung Y, et al. Annual changes in radiographic indices of the spine in cerebral palsy patients. doi:10.1007/s00586-014-3746-4
9. Y G, JE S, JM K, et al. Natural history of scoliosis in nonambulatory spastic tetraplegic cerebral palsy. PM R. 2011;3(1):27-32. doi:10.1016/J.PMRJ.2010.09.015
10. N S, S E, K O, H K, K T. Natural history of scoliosis in spastic cerebral palsy. Lancet (London, England). 1998;351(9117):1687-1692.doi:10.1016/S0140-6736(98)01302-6
11. Inal-Ince D, Savci S, Arikan H, et al. Effects of scoliosis on respiratory muscle strength in patients with neuromuscular disorders. Spine J. 2009;9(12):981-986. doi:10.1016/J.SPINEE.2009.08.451
12. Terjesen T, Lange JE, Steen H. Treatment of scoliosis with spinal bracing in quadriplegic cerebral palsy. Dev Med Child Neurol. 2000;42(7):448-454. doi:10.1017/S0012162200000840
13. Olafsson Y, Saraste H, Al-Dabbagh Z. Brace treatment in neuromuscular spine deformity. J Pediatr Orthop. 1999;19(3):376-379. doi:10.3233/978-1-60750-903-5-332
14. Jang J, Park Y, Lee S, et al. The effect of a flexible thoracolumbar brace on neuromuscular scoliosis: A prospective observational study. Medicine (Baltimore). 2021; 100 (32):e26822 .doi : 10. 1097/ MD. 0000000000026822
15. Brunner R. Development and conservative treatment of spinal deformities in cerebral palsy. J Child Orthop. 2020;14(1):9-16. doi:10.1302/1863-2548.14.190127

16. Pettersson K, Rodby-Bousquet E. Prevalence and goal attainment with spinal orthoses for children with cerebral palsy. J Pediatr Rehabil Med. 2019;12(2):197-203. doi:10.3233/PRM-180596
17. Nakamura N, Uesugi M, Inaba Y, Machida J, Okuzumi S, Saito T. Use of dynamic spinal brace in the management of neuromuscular scoliosis: a preliminary report. J Pediatr Orthop B. 2014; 23 (3) :291-298. doi: 10.1097/ BPB. 0000000000000034
18. Karol LA. Scoliosis in patients with Duchenne muscular dystrophy. J Bone Joint Surg Am. 2007;89 Suppl 1(suppl_1):155-162. doi:10.2106/JBJS.F.00506
19. Kotwicki T, Jozwiak M. Conservative management of neuromuscular scoliosis: personal experience and review of literature. Disabil Rehabil. 2008;30(10):792-798. doi:10.1080/09638280801889584
20. Lebel DE, Corston JA, McAdam LC, Biggar WD, Alman BA. Glucocorticoid treatment for the prevention of scoliosis in children with Duchenne muscular dystrophy: long-term follow-up. J Bone Joint Surg Am. 2013;95(12):1057-1061. doi:10.2106/ JBJS.L.01577
21. Coratti G, Cutrona C, Pera MC, et al. Motor function in type 2 and 3 SMA patients treated with Nusinersen: a critical review and meta-analysis. Orphanet J Rare Dis. 2021;16(1). doi:10.1186/S13023-021-02065-Z
22. Walker KR, Novotny SA, Krach LE. Does Intrathecal Baclofen Therapy Increase Prevalence and/or Progression of Neuromuscular Scoliosis? Spine Deform. 2017;5(6):424-429. doi:10.1016/ J.JSPD. 2017.03.006
23. Dimeglio A, Canavese F. The growing spine: how spinal deformities influence normal spine and thoracic cage growth. Eur Spine J 2011 211. 2011;21(1):64-70. doi:10.1007/S00586-011-1983-3
24. Karol LA, Johnston C, Mladenov K, Schochet P, Walters P, Browne RH. Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. J Bone Joint Surg Am. 2008;90(6):1272-1281. doi 10.2106/JBJS.G.00184
25. Cunin V. Early-onset scoliosis: current treatment. Orthop Traumatol Surg Res. 2015;101(1 Suppl):S109-S118. doi:10.1016/J.OTSR.2014.06.032
26. Shapiro F, Zurakowski D, Bui T, Darras BT. Progression of spinal deformity in wheelchair-dependent patients with Duchenne muscular dystrophy who are not treated with steroids: Coronal plane (scoliosis) and sagittal plane (kyphosis, lordosis) deformity. Bone Jt J. 2014;96 B(1):100-105. doi:10.1302/0301-620X.96B1.32117/ASSET/ IMAGES/ LARGE/32117-GALLEYFIG5.JPEG
27. Campbell RM, Smith MD. Thoracic Insufficiency Syndrome and Exotic Scoliosis. J Bone Jt Surg. 2007;89(suppl_1):108-122. doi:10.2106/JBJS.F.00270
28. Odent T, Ilharreborde B, Miladi L, et al. Fusionless surgery in early-onset scoliosis. Orthop Traumatol Surg Res. 2015;101(6 Suppl):S281-S288. doi:10. 1016/J.OTSR.2015.07.004
29. Drummond DS. Neuromuscular scoliosis: recent concepts. J Pediatr Orthop. 1996;16(3):281-283. doi:10.1097/00004694-199605000-00001
30. Modi HN, Suh SW, Hong JY, Yang JH. Posterior multilevel vertebral osteotomy for severe and rigid idiopathic and nonidiopathic kyphoscoliosis: a further experience with minimum two-year follow-up. Spine (Phila Pa 1976). 2011;36(14):1146-1153. doi:10.1097/ BRS.0B013E3181F39D9B
31. Jackson TJ, Yaszay B, Pahys JM, et al. Intraoperative Traction May Be a Viable Alternative to Anterior Surgery in Cerebral Palsy Scoliosis ≥100 Degrees. J Pediatr Orthop. 2018;38(5):e278-e284. doi:10.1097/ BPO.0000000000001151
32. Lowenstein JE, Matsumoto H, Vitale MG, et al. Coronal and sagittal plane correction in adolescent idiopathic scoliosis: a comparison between all pedicle screw versus hybrid thoracic hook lumbar screw constructs. Spine (Phila Pa 1976). 2007;32(4):448-452. doi:10.1097/01.BRS.0000255030.78293.FD
33. La Rosa G, Giglio G, Oggiano L. Surgical treatment of neurological scoliosis using hybrid construct (lumbar transpedicular screws plus thoracic sublaminar acrylic loops). Eur Spine J. 2011;20 Suppl 1(Suppl 1). doi:10.1007/S00586-011-1751-4
34. Sink EL, Newton PO, Mubarak SJ, Wenger DR. Maintenance of sagittal plane alignment after surgical correction of spinal deformity in patients with cerebral palsy. Spine (Phila Pa 1976). 2003;28(13):1396-1403. doi:10.1097/01.BRS.0000067088.99346.73
35. Mineiro J, Yazici M. Technical aspects of surgical correction of spinal deformities in cerebral palsy. J Child Orthop. 2020;14(1):30-40. doi:10.1302/1863-2548.14.190167
36. Jones-Quaidoo SM, Yang S, Arlet V. Surgical management of spinal deformities in cerebral palsy. A review. J Neurosurg Spine. 2010;13(6):672-685. doi:10.3171/2010.5.SPINE09669
37. Hasler C, Brunner R, Grundshtein A, Ovadia D. Spine deformities in patients with cerebral palsy; the role of the pelvis. J Child Orthop. 2020;14(1):9-16.

doi:10.1302/1863-2548.14.190141
38. Wu Z, Schwend RM, Anderson JT, Marasigan JAM, Price NJ. Iliac screw instrumentation to the pelvis in children with neuromuscular and syndromic scoliosis. No lateral connectors and respect sagittal balance. Spine Deform. 2021;9(4):1115-1123. doi:10.1007/S43390-021-00287-6
39. Vialle R, Delecourt C, Morin C. Surgical treatment of scoliosis with pelvic obliquity in cerebral palsy: the influence of intraoperative traction. Spine (Phila Pa 1976). 2006; 31(13): 1461-1466. doi: 10.1097/01.BRS. 0000219874.46680.87
40. Hamzaoglu A, Ozturk C, Aydogan M, Tezer M, Aksu N, Bruno MB. Posterior only pedicle screw instrumentation with intraoperative halo-femoral traction in the surgical treatment of severe scoliosis (>100 degrees). Spine (Phila Pa 1976). 2008;33(9):979-983. doi:10.1097/BRS.0B013E31816C8B17
41. Sponseller PD, Jain A, Lenke LG, et al. Vertebral column resection in children with neuromuscular spine deformity. Spine (Phila Pa 1976). 2012;37(11). doi:10.1097/BRS.0B013E318244460D4
42. Aydinli U, Kara K, Mutlu M, Yaray O. Results of Posterior Vertebral Column Resection: Surgical Modification of Suk Technique. Glob spine J. 2018;8(5):478-482. doi:10.1177/2192568217739854
43. Menga EN, Bernstein DN, Thirukumaran C, McCormick SK, Rubery PT, Mesfin A. Evaluating Trends and Outcomes of Spinal Deformity Surgery in Cerebral Palsy Patients. Int J spine Surg. 2020;14(3):382-390. doi:10.14444/7050
44. Mullender MG, Blom NA, De Kleuver M, et al. A Dutch guideline for the treatment of scoliosis in neuromuscular disorders. Scoliosis. 2008;3(1). doi:10.1186/1748-7161-3-14
45. Luo M, Wang W, Shen M, Luo X, Xia L. Does higher screw density improve radiographic and clinical outcomes in adolescent idiopathic scoliosis? A systematic review and pooled analysis. J Neurosurg Pediatr. 2017;19(4):448-457. doi:10.3171/2016.11. PEDS16334
46. Wolfram JM, Kristen VA, Cip J, Bach CM. Influence of implant density and flexibility index on curve correction after scoliosis surgery. Musculoskelet Surg. 2021. doi:10.1007/S12306-021-00707-W
47. Gaume M, Saudeau E, Gomez-Garcia De La Banda M, et al. Minimally Invasive Fusionless Surgery for Scoliosis in Spinal Muscular Atrophy: Long-term Follow-up Results in a Series of 59 Patients. J Pediatr Orthop. 2021;41(9):549-558. doi:10.1097/BPO.0000000000001897
48. Enercan M, Kahraman S, Erturer E, Ozturk C, Hamzaoglu A. Apical and intermediate anchors without fusion improve Cobb angle and thoracic kyphosis in early-onset scoliosis. Clin Orthop Relat Res. 2014;472(12):3902-3908. doi:10.1007/S11999-014-3815-3
49. Abol Oyoun N, Stuecker R. Bilateral rib-to-pelvis Eiffel Tower VEPTR construct for children with neuromuscular scoliosis: a preliminary report. Spine J. 2014;14(7): 1183-1191. doi: 10.1016/J.SPI NEE. 2013.07.484
50. Almajali A, Obeidat M, Bashmaf O, Wagokh R, Harahsheh B, Alzaben R. Early Childhood Scoliosis Management by Vertical Expandable Prosthetic Titanium Rib (VEPTR): Experience of Royal Medical Services (RMS). Med Arch (Sarajevo, Bosnia Herzegovina). 2020;74(6):433-438. doi:10.5455/MEDARH.2020.74.433-438
51. Rosenfeld S, Schlechter J, Smith B. Achievement of Guided Growth in Children With Low-Tone Neuromuscular Early-Onset Scoliosis Using a Segmental Sublaminar Instrumentation Technique. Spine Deform. 2018;6(5):607-613. doi:10. 1016 / J.JSPD.2018.02.012
52. Murphy NA, Firth S, Jorgensen T, Young PC. Spinal surgery in children with idiopathic and neuromuscular scoliosis. What's the difference? J Pediatr Orthop. 2006; 26(2): 216-220. doi: 10.1097/01. BPO. 0000206516.61706.6E
53. Sharma S, Wu C, Andersen T, Wang Y, Hansen ES, Bünger CE. Prevalence of complications in neuromuscular scoliosis surgery: a literature meta-analysis from the past 15 years. Eur Spine J. 2013;22(6):1230-1249. doi:10.1007/S00586-012-2542-2
54. Mackenzie WGS, Matsumoto H, Williams BA, et al. Surgical site infection following spinal instrumentation for scoliosis: a multicenter analysis of rates, risk factors, and pathogens. J Bone Joint Surg Am. 2013;95(9):800-806. doi:10.2106/JBJS.L.00010
55. Modi HN, Suh SW, Yang JH, et al. Surgical complications in neuromuscular scoliosis operated with posterior- only approach using pedicle screw fixation. Scoliosis. 2009;4. doi:10.1186/1748-7161-4-11
56. Tsirikos AI, Lipton G, Chang WN, Dabney KW, Miller F. Surgical correction of scoliosis in pediatric patients with cerebral palsy using the unit rod instrumentation. Spine (Phila Pa 1976). 2008;33(10): 1133-1140. doi: 10.1097/ BRS. 0B013E31816F63CF

57. Sitoula P, Holmes L, Sees J, Rogers K, Dabney K, Miller F. The Long-term Outcome of Early Spine Fusion for Scoliosis in Children With Cerebral Palsy. Clin spine Surg. 2016;29(8):E406-E412. doi:10.1097/ BSD.0000000000000184
58. Legg J, Davies E, Raich A, Dettori J, Sherry N. Surgical correction of scoliosis in children with spastic quadriplegia: benefits, adverse effects, and patient selection. Evid Based Spine Care J. 2014;5(1):038-051. doi:10.1055/S-0034-1370898
59. Cloake T, Gardner A. The management of scoliosis in children with cerebral palsy: a review. J Spine Surg. 2016;2(4):299-309. doi:10.21037/jss.2016.09.05
60. Mercado E, Alman B, Wright JG. Does spinal fusion influence quality of life in neuromuscular scoliosis? Spine (Phila Pa 1976). 2007;32(19 Suppl). doi:10.1097/ BRS.0B013E318134EABE

THE IMPORTANCE OF FOLLOWING THE ORIGINAL PONSETI METHOD IN TREATMENT OF CLUBFOOT: THE IOWA EXPERIENCE

СЛЕДЕЊЕ НА ОРИГИНАЛНИОТ ПОНСЕТИ МЕТОД ПРИ ТРЕТМАНОТ НА ВРОДЕНО КРИВО СТАПАЛО: ИСКУСТВО ОД АЈОВА

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АПСТРАКТ

Др. Игнацио Вивес Понсети е ортопедски хирург со шпанско потекло, роден на 03.06.1914 и починал на 18ти Октомври 2009 во Ајова, САД, каде што живеел и работел. Тој бил брилијантен педијатриски ортопедски хирург, прочуен по неговиот метод на конзервативен третман на вродената кривата стапала, што и ден денес претставува златен стандард. Неговиот труд од 1963та година е еден од ретките во ортопедијата кој радикално го променил дотогашниот начин на третман. Методот по Понсети лесно се учи, но за жал и подлегнува на многу модификации, што негативно влијае на исходот од третманот. Тоа става дополнителен акцент на потребата од учење на точните чекори опишани од д-р Понсети. Првиот автор на овој труд беше на студиски престој во Детската болница на фамилијата Стед во Ајова, каде што го учеше методот на местото на неговиот изум, под водство на др. Хозе Моркуенде, наследникот на др. Понсети, кој на извонреден начин го продолжува наследството на д-р Понсети. Започнавме да го користиме оригиналниот метод во октомври 2019. Од тогаш, 16 пациенти, 7 со билатерална, 9 со унилатерална зафатеност (23 стапала вкупно) се третирани и евалуирани. Се трудевме во огромна мерка верно да го пратиме точниот протокол на Понсети, опишани во неговиот труд. Обично, за корекција на првите три компоненти од деформитетот потребни се четири до десет гипсени имобилизации, изведувани на неделно ниво. Еквинусот се коригира последен. Со цел да се избегне пролонгирано поставување на гипсови и последователна појава на “rocker bottom” деформитет, корекцијата на еквинусот се постигнува со едноставна перкутана ахилотенотомија во локална анестезија. Кај шеесет и пет проценти од пациентите имаше добри резултати, триесет и еден процент беа со просечен исход, додека кај еден пациент резултатот беше лош. Во споредба со оригиналниот труд на Понсети од 1963 година, не постојат сигнификантни разлики во резултатите, освен поголем процент на лош резултат кој вклучува само еден пациент во нашата серија.

ABSTRACT

Dr. Ignacio Vives Ponseti, an American orthopaedic surgeon with Spanish origin, was born on June the 3rd, 1914 and died at the age 95 in Iowa, USA, on October 18th 2009. He was a brilliant pediatric orhopaedic surgeon, best known for his method of nonoperative treatment of clubfoot, that has become a gold standard of clubfoot treatment. His iconic paper on clubfoot management (1963) is one of the few manuscripts in orthopaedic literature which has radically changed the practice as we know it now. The Ponseti method is easy to learn but, unfortunately easy to modify because modification negatively affects the treatment outcome. That emphasizes the need to learn and follow the exact steps explained by Dr. Ponseti. The first author of this article had the opportunity to learn this method in the place where it all started, guided by Dr. Jose Morcuende, the successor of Dr. Ponseti who continues his legacy in the brightest way possible. After return from the stay at Iowa Stead Family Children's Hospital 16 patients and 23 feet were treated with best possible adherence to the Ponseti Method.

We began using the original Ponseti method in October 2019. Since then, 16 patients, 7 bilateral, 9 unilateral (23 feet in total) have been treated and evaluated. We strove as much as possible to reproduce Ponseti's strict casting protocol faithfully, as explained in the paper. Usually, for correction of the first three components 4 to 10 casts are necessary, changed on a weekly basis. Equinus is the last deformity to be corrected. In order to avoid prolonged casting and concomitant appearance of rocker bottom deformity, the correction of equinus is facilitated by a simple operative procedure in local anesthesia. Sixty five percent of the patients had good results, 31% had acceptable results, in one patient there was poor result. Compared to the original Ponseti paper from 1963, there is not any significant difference in the results, except the bigger percent of poor results that involves only one patient in our series.

INTRODUCTION

Dr. Ignacio Vives Ponseti, an American orthopaedic surgeon with Spanish origin, was born on June the 3rd, 1914 and died at the age 95 in Iowa, USA, on October 18th 2009.[1] He was a brilliant pediatric orhopaedic surgeon, best known for his method of nonoperative treatment of clubfoot, that has become a gold standard of clubfoot treatment. His iconic paper on clubfoot management (1963) is one of the few manuscripts in orthopaedic literature which has radically changed the practice as we know it now.[2]

But, until the 1990s there wasn't any interest about his technique for treating clubfoot. Given that the results of surgical treatment can beso discouraging, parents looked for alternative treatments. That's what caused Dr. Ponseti to come out of retirement which launched a new era in the clubfoot treatment. The importance and execution of this method was sealed in 1996 when he published his book "Congenital Clubfoot: Fundamentals of Treatment".[3] This and the emerge of internet brought a new sight of this forgotten method.[4]

It has always been clear to most surgeons that the initial treatment of idiopathic congenital clubfoot should be serial gentle manipulations to stretch the contractures, with serial casting and splinting to maintain the correction obtained by stretching.[5, 6, 7] But, with the previous conservative methods, that were deficient with understanding of the nature of the deformity and the normal functional anatomy of the foot, the success rate was 10-50%, as 50-90% of the patients were referred to extensive surgery.[8, 9, 10,11] In those patients, the main concern is functional outcome that often abruptly deteriorates in their adult life.[12]

The Ponseti method is easy to learn but, unfortunately easy to modify because modification negatively affects the treatment outcome. That emphasizes the need to learn and follow the exact steps explained by Dr. Ponseti.Because many parents were now seeking out physicians trained in the Ponseti Method, orthopaedic surgeons began traveling to Iowa City from around the world to train with Dr. Ponseti and become proficient in his method. Now, more than two decades later, the Ponseti Method is the standard of treatment worldwide. The first author of this article had the opportunity to learn this method in the place where it all started, guided by Dr. Jose Morcuende, the successor of Dr. Ponseti who continues his legacy in the brightest way possible.

After return from the stay at Iowa Stead Family Children's Hospital 16 patients and 23 feet were treated with best possible adherence to the Ponseti Method. Besides demographic data we evaluated the number of

casts used, that indicated the severity of the clubfeet, the need of tenotomy, as well as appearance of recurrence of the deformity.

Material and methods

We began using the original Ponseti method in October 2019. Since then, 16 patients, 7 bilateral, 9 unilateral (23 feet in total) have been treated and evaluated. We strove as much as possible to reproduce Ponseti's strict casting protocol faithfully, as explained below.

The guidelines for the treatment developed by Ponseti and followed at our institution are as follows:[2, 3]The casting is performed by a treating surgeon and a nurse/casting technician. The molding should be done exclusively by the surgeon. Simultaneous correction of all the components of the deformity (cavus, adductus, varus), except for the equinus that is corrected last (figure 1).

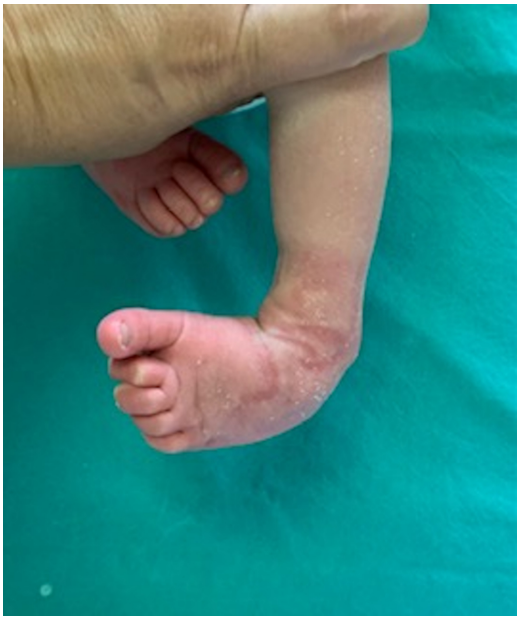

Figure 1. Not treated clubfoot in 10 days old newborn. It has all the components of the deformity-cavus, adductus, varus and equinus

The cavus should be corrected with the first cast using supination of the forefoot regarding the hindfoot by applying counter pressure on the superiolateral aspect on the head of the talus. The padding is thin and in one layer only followed by the application of plaster cast in two sections, the first below the knee, in order to obtain enough time to mold the cast before it becomes stiff, by contouring the lateral malleolus and emphasizing the posterior crease of the calcaneus (figures 2 and 3).

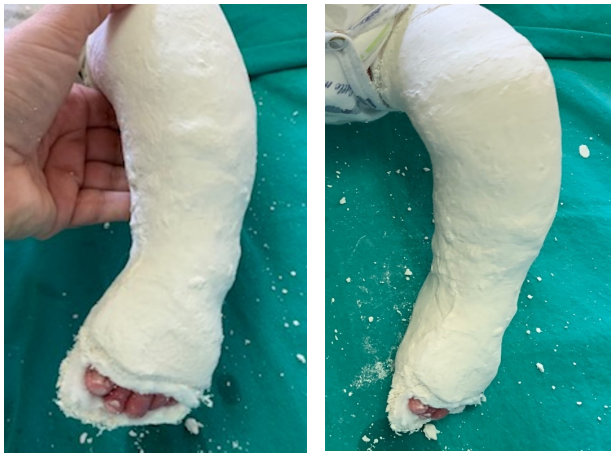

Figure 2 and 3. After the first manipulation, correction of the cavus with supination of the first metatarsal bone regarding the hindfoot is achieved.

Then the other section of the cast is added that covers the thigh with the knee in 90 degrees of flexion. The long cast is essential to avoid cast slippage and to correct tibial torsion. Distally, the toes are opened from the dorsal side, leaving enough cast on the plantar side, to avoid crowding and flexing of the toes. Once the longitudinal arch of the foot is corrected with supination, the entire foot is abducted under the talus. When the foot is abducted fully under the talus, the calcaneal varus will correct without touching the heel (figures 4, 5, 6 and 7).

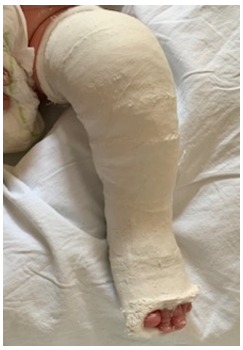

Figure 4

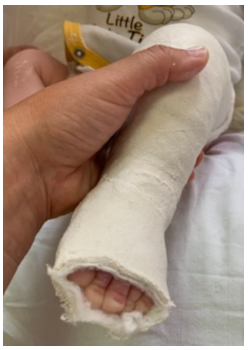

Figure 5

Figure 6

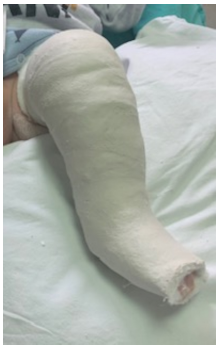

Figure 7

Figures 4, 5, 6 and 7. Serial casting with weekly change of the casts until abduction of 70 degrees is achieved.

Equinus is the last deformity to be corrected. In order to avoid prolonged casting and concomitant appearance of rocker bottom deformity, the correction of equinus is facilitated by a simple operative procedure in local anesthesia which is administrated half an hour before the operation. The intervention is performed in the operating room, older babies (more than six months) get an additional sedation. The patient is placed supine on the operating room, while an assistant holds the baby's foot and knee in flexion, parallel to the operating table. We use knife number 15, which is placed from medial to lateral, parallel to the tendon, with the blade facing proximally. When the tendon is probed by gentle anterior placement of the knife, the blade is rotated for 45 degrees posteriorly and the tendon is severed completely, followed by a increase of the dorsiflexion of about 15 degrees. The surgeon thumb from the opposite hand is placed over the skin in order not to damage it. No sutures are necessary. Afterwards, pressure is applied for several minutes to stop the bleeding and sterile thin gauze is placed over the heel. The last cast is applied in the same manner as all the previous casts with extreme correction of equinus (dorsiflexion of 15 degrees) and adductus (abduction of 70 degrees). These extremes are necessary to avoid relapse (figure 8).

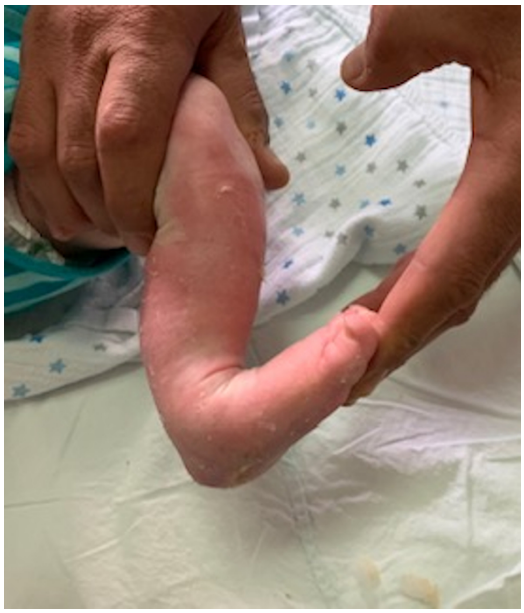

Figure 8. Achieved correction after serial casting and tenotomy in the same patient as in figure 1. Photography taken after five cast changes and tenotomy, at three and half months of age. On this visit boots and bar were placed

The last cast is worn for three weeks and afterwards an abduction brace is applied to the baby's feet. Abduction of the feet is set to 60-70° on the affected side and 40° on the normal side. In bilateral cases abduction is set to 60-70° in both feet.The length of the bar should be identical to the shoulder width of the baby. This brace is worn constantly in the first three months, for 23 hours daily and afterwards it's worn for 14 hours a day, in the next three to five years (figure 9).

Figure 9: Abduction boots and bar brace that needs to be worn 23 hours daily for the first three months after the tenotomy and 14 hours daily for the next 4-5 years.

Usually, for correction of the first three components 4 to 10 casts are necessary, changed on a weekly basis.

If any of the components of the clubfeet reoccurs (cavus, varus, adductus and/or equinus), it complies to appearance of a recurrence. Depending on the severity of the recurrence, it can be addressed by increase of the time of the wearing of the brace, additional manipulation with or without tenotomy (complete tenotomy, not lengthening) of the Achilles tendon.Criteria for a complete tenotomy were: feeling of a sudden pop as the tendon is severed, correction of equinus deformity by achieving bigger dorsiflexion and a palpable gap in the substance of the tendon.The tendon fully regenerates following percutaneous tenotomy.[13]

Of out sixteen patients, eight were female and eight were male. Two of them were twins, with unilateral clubfeet. The other twin didn't have any anomalies. From the unilaterally affected patients, six feet were right and three were left. Our patients underwent 3-9 cast changes (5 casts in average). The patients presented from ten days to six months in age, average 7 weeks. Seven patients, twelve feet were previously treated elsewhere. Percutaneous Achilles tenotomy was performed in eleven patients, sixteen feet.

RESULTS

In most of our patients, the deformity was corrected with five casts in average. In eleven patients more than six casts were used which indicates bigger severity of the deformity. Dimeglio and Pirani clinical scoring systems before and after the treatment are insufficient to predict the course and results of treatment by the Ponseti method.14 We also have found that significant correlation in predictive factors (i. e. complexity, number of casts, need for tenotomy, probability of recurrence) is exposed by clinical evaluation before the initiation of Ponseti treatment and after the removal of the second cast the accuracy is higher. We clinically evaluated the persistence of any of the components of the clubfeet in all our patients after the initial treatment following the Ponseti method. The clinical components analyzed were ankle dorsiflexion, heel varus, adduction of the forefoot as well as tibial torsion(table 1).

Ankle dorsiflexion (degrees)	**Heel varus (degrees)**	**Heel varus (degrees)**	**Tibial torsion (degrees)**	**Results**
>10	0	0-10	0	Good (n:15, 65%)
0-10	0-10	10-20	Moderate	Acceptable (n:7, 31%)
0	Over 10	Over 20	Severe	Poor (n: 1, 4%)

Table 1. Results

The cavus deformity, once corrected properly by the first and maybe the second cast rarely recurs. In one patient there was recurrence of this deformity, but it was corrected by recasting and up lifting the first metatarsal.

In four patients hindfoot varus and forefoot adductus reappeared. In all of them noncompliance to the brace wearing was reported by the parents. If the deformity persist after two and a half years, anterior tibial tendon transfer to the cuboid bone will be suggested.

Tibial torsion was completely corrected in all patients, except in one calf in a patient with bilateral deformity.

Dorsiflexion of the ankle lacked in four patients. In two of them the tendon was sectioned at the initial treatment. In one of them repeated percutaneous Achillotenotomy was performed.

The results are collected in table 1. Sixty five percent of the patients had good results, 31% had acceptable results, in one patient there was poor result.

DISCUSSION

In our case series, as well as in many works before, it has been proven, that following the original Ponseti method gives fruitful results.[2, 15, 16, 17]

We compared our results to those of the original Ponseti paper from 1961.[2] It involves evaluation of sixty seven patients with total of ninety-four clubfeet. The results in 71 per cent of the feet were good; in 28 per cent a slight residual deformity persisted; and in one foot a poor result was obtained, as compared to ours, 65 per cent had good result, some residual deformity present in 31% and poor result in four percent of our patients. Our study involves significantly lesser number of cases, and lesser time of follow up (two years) but there is not any significant difference in the results, except the bigger percent of poor results that involves only one patient in our series.

Of great importance is to mention the common errors in treatment of congenital clubfoot.[18] It is only illusive that the severe supination of the foot is corrected by pronation, in fact, the forefoot is pronated related to the rearfoot and it should be supinated to achieve a correction of the cavus. The foot must not be externally rotated if the calcaneus is in varus because it leads to posterior displacement of the lateral malleolus which is a sign of poorly treated clubfoot. Another common mistake, commonly taught by previous methods of conservative treatment of clubfeet is pressing the calcaneocuboid joint as a fulcrum to rotate the foot. It is necessary to use the talar head as fulcrum under which the foot underlines properly.[19] All manipulations must be followed by plaster cast immobilzation, in order to keep soft tissues stretched between manipulations. It is necessary the plaster cast to be long, from toes to the groin, otherwise, the ankle and talus will rotate. Also, the surgeon must be aware that perfect anatomical correction is not possible in most cases, but it doesn't cause any functional impairment in the future.

And last but not the least, the casting and molding of the plaster cast must not be left to the casting technician, because all of these afore mentioned errors are responsibility of the treating physician.

It is interesting to mention how the internet affected the treatment of congenital idiopathic clubfoot. It is well observed in Morcuende's work from 2003.20 Since the information for clubfoot treatment in Iowa became virtual on the internet, there'd been a dramatic increase in the number of patients in their clinic, representing profound effect in clinical practice patterns, and in the patient physician relationship. In that way the parents became aware of the Ponseti method in order to avoid extensive surgical treatment.

BIBLIOGRAPHY

1. Dobbs MB, Khan SA. The life and legacy of Ignacio Ponseti. Indian J Orthop. 2010;44(1):114.
2. Ponseti IV, Smoley EN. The classic: congenital club foot: the results of treatment. 1963. Clin Orthop Relat Res. 2009 May;467(5):1133-45. doi: 10.1007/s11999-009-0720-2. Epub 2009 Feb 14. PMID: 19219519; PMCID: PMC2664436.
3. Congenital Clubfoot: Fundamentals of Treatment, Ignacio V Ponseti, New York, Oxford University Press, 1996
4. Morcuende JA, Egbert M, Ponseti IV. The effect of the internet in the treatment of congenital idiopathic clubfoot. Iowa Orthop J. 2003;23:83-6. PMID: 14575256; PMCID: PMC1888396.
5. Green AD, Lloyd-Roberts GC. The results of early posterior release in resistant club feet: a long-term review. J Bone Joint Surg [Br] 1985;67:588–93.
6. Kite JH. Principles involved in the treatment of congenital clubfoot. J Bone Joint Surg 1939;21:595–606.
7. Nather A, Bose K. Conservative and surgical treatment of clubfoot. J PediatrOrthop 1987;7:42–8.
8. Kite JH. Nonoperative treatment of congenital clubfoot. ClinOrthop. 1972;84:29 -38
9. Dimeglio A, Bonnet F, Mazeau PH, De Rosa V. Orthopaedic treatment and passive motion machine: consequences for the surgical treatment of clubfoot. J PediatrOrthop B. 1996;5:173-180
10. Dewaele J, Zachee B, De Vleeschauwer P, Fabry G. Treatment of idiopathic clubfoot: critical evaluation of different types of treatment programs. J PediatrOrthop. 1994;3:89-95
11. Morcuende JA, Dolan LA, Dietz FR, Ponseti IV. Radical reduction in the rate of extensive corrective surgery for clubfoot using the Ponseti method. Pediatrics. 2004 Feb;113(2):376-80. doi: 10.1542/peds.113.2.376. PMID: 14754952.
12. Herzenberg JE, Radler C, Bor N. Ponseti versus traditional methods of casting for idiopathic clubfoot. J PediatrOrthop. 2002 Jul-Aug;22(4):517-21. PMID: 12131451.
13. Saini R, Dhillon MS, Tripathy SK, Goyal T, Sudesh P, Gill SS, Gulati A. Regeneration of the Achilles tendon after percutaneous tenotomy in infants: a clinical and MRI study. J PediatrOrthop B. 2010 Jul;19(4):344-7. doi: 10.1097/BPB.0b013e3283361b47. PMID: 20375912.
14. Jochymek J, Peterková T. ARE SCORING SYSTEMS USEFUL FOR PREDICTING RESULTS OF TREATMENT FOR CLUBFOOT USING THE PONSETI METHOD? ActaOrtop Bras. 2019 Jan-Feb;27(1):8- 11. doi: 10.1590/1413-785220192701189801. PMID: 30774521; PMCID: PMC6362704.
15. Laaveg SJ, Ponseti IV. Long-term results of treatment of congenital clubfoot. J Bone Jt Surg Am 1980 Jan;62(1):23e31.
16. Morcuende JA, Abbasi D, Dolan LA, Ponseti IV. Results of an accelerated Ponseti protocol for clubfoot. J Pediatr Orthop 2005 Sep-Oct;25(5):623e6.
17. Ippolito E, Farsetti P, Caterini R, Tudisco C. Long-term comparative results in patients with congenital clubfoot treated with two different protocols. J Bone Jt Surg Am 2003 Jul;85-A(7):1286e94.
18. Ponseti IV. Common errors in the treatment of congenital clubfoot. Int Orthop. 1997;21(2):137-41. doi: 10.1007/s002640050137. PMID: 9195271; PMCID: PMC3616653.
19. Kite JH (1964) The clubfoot. Grune & Stratton, New York London
20. Morcuende JA, Egbert M, Ponseti IV. The effect of the internet in the treatment of congenital idiopathic clubfoot. Iowa Orthop J. 2003;23:83-6. PMID: 14575256; PMCID: PMC1888396.

GIANT CELL TUMOR OF THE LUMBAR SPINE - one case report

ГИГАНТОЦЕЛУЛАРЕН ТУМОР НА ЛУМБАЛНИОТ ДЕЛ НА РБЕТОТ - приказ на случај

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АПСТРАКТ

Вовед:
Гигантоцелуларниот тумор во ‘рбетот е редок но најагресивниот бенигнен тумор на ‘рбетот со непредвидлив исход. Тој е редок тумор на коските и сочинува 5% од сите коскени тумори. Најмногу се јавува во долгите коски. Лумбалната локализација е многу ретка.

Материјал и методи:
Целта на овој приказ на случај е да се опише необична локализација на гигантоцелуларен тумор, дијагнозата и лекувањето. 54 годишен пациент од машки пол се пожали на болки во долниот дел на ‘рбетот кои биле присутни во последните 6 месеци. Била дијагностицирана протрузија на интервертеблалниот диск пред 5 месеци со продолжителна болка и развивање на слабост во долните екстремитети. Потоа беше примен во нашата болница. Невролошкиот преглед откри силно изразена слабост на дорзалната и плантарната флексија на стапалата. Рефлексот на Ахиловата тетива беше обострано отсутен. Нативните радиографии на лумбосакралната регија прикажаа деструктивна лезија од тежок степен на третиот лумбален прешлен. На приказот од магнетната резонанса се презентираше голема маса во регијата меѓу третиот и четвртиот лумбален прешлен. Се спроведе и SPECT/CT испитување.

Интраоперативно, се виде сиво-кафеава високоваскуларизирана маса со ерозија на ламините и телото на L3, после што се изведе ламинектомија. Следеше тотално отстранување на туморот со парцијална корпоректомија на L3 и L4, со декомпресија на нервните корени на тоа ниво. Хистолошката анализа на лезијата беше со карактеристики на коскен гигантоцелуларен тумор. Реконструкцијата беше направена со шипки и транспедикуларна фиксација на L2, L3, L4 и L5 прешлените. Постоперативниот период мина без компликации, а вертикализацијата беше спроведена на вториот постоперативен ден, со TLSO корсет.

Резултати:
Во опишаниот случај, тотална ексцизија на туморот се постигна со интрарегионален пристап и солидна реконструкција и фиксација со шипки и педикуларни шрафови. Бидејќи en-bloc ексцизија не можеше да се спроведе, неопходно е внимателно следење заради можноста од појавување на локални и далечни рецидиви.

Заклучоци:
Неопходна е хируршка интервенција и внимателно следење за рано откривање на рецидиви.

Клучни зборови:
Гигантоцелуларен тумор на ‘рбетот, задна en-bloc ексцизија на прешленското тело, радиотерапија на туморот

ABSTRACT

Objective:
Giant cell tumor (GCT) of the spine is uncommon but most aggressive benign tumor of the spine with unpredictable outcome. Giant-cell tumours (GCT) of bone are rare, and constitute 5% of all bone tumours. The majority occur in long tubular bones. The lumbar localization of a giant-cell tumor is also very rare.

Material/Method
The purpose of this report is to describe the unusual location of the giant-cell tumor, its diagnosis and treatment. A 54 year old male had low back pain for 6 months. He had a L4-5 protruded intervertebral disc 5 months previously, but his back pain continued and he developed weakness of his feet. The patient was then admitted to our hospital. The physical examination was within normal limits. On neurological examination, there was severe weakness of dorsal and plantar flexion of his feet. The ankle reflexes were bilaterally absent. Plain radiographs of the lumbosacral area showed a severe destructive lesion of the L3 vertebra. A huge mass of the region between L3 and L4 vertebra was seen in MR imaging. It was also made SPECT/CT.

Intraoperatively, the paravertebral muscles were dissected, a greybrown and highly vascular mass was seen, the lamina and body of L3 vertebra were eroded, and laminectomy was performed. Tumor was removed totally with partial corrporectomy of L3 and L4 vertebrae, decompression of the third and fourth lumbar vertebrae nerve roots. The histological appearance of this lesion was identical to that of a giant-cell tumor of bone. Reconstruction was done using pedicle screw and rod fixation in L2, L3, L4 et L5 dorsal vertebrae. There was no complication in the postoperative period, and the patient was postoperatively verticalized the second postoperative day with pre-placed TLSO corset.

Results:
In the present case, total tumor excision was achieved by intraregional approach and solid reconstruction was achieved using pedicle screw and rod fixation. As en-bloc excision could not be carried out, close follow-up is required for any sign of local and distant recurrence.

Conclusions:
Surgical intervention is mandatory. Close follow up is needed for early diagnosis of recurrences.

Keywords:
Giant Cell Tumor of the spine, GCT, dorsal vertebral body, en bloc excision, giant cell tumor, tumor recurrence radiotherapy.

INTRODUCTION

Giant cell tumour (GCT) of bone is classified by the World Health Organisation (WHO) as a benign but locally aggressive tumour that usually involves the end (epiphysis) of a long bone [1]. It most frequently occurs in young adults between 20 and 40 years of age with a slight female predominance [2–5]. GCT has a significant incidence, accounting for 5 % of all bone tumours [2]. Higher incidence has been reported for the Chinese population, in which it can be up to 20 % of all bone tumours [6].Except for the sacrum, the lumbar localisation of a giant-cell tumour is also very rare.

The most commonly used classification of Enneking and Campanacci et al. were designed to define the extent of surgery required to completely remove the tumour, which may influence the surgical resection.

One major reason to determining the ideal treatment selection of GCT, may be that the surgeons cannot accurately evaluate the severity or aggressiveness of GCT preoperatively by single clinical, radiographic, histological or demographic aspects. Thus, many factors must be considered comprehensively:

- Patients' age
- Size of GCT
- Cortical bone destruction and soft tissue involvement
- Pathological fracture
- Involvement of the subchondral bone
- Other possible prognostic factors

There are many other factors that should be taken into account when evaluating the aggressiveness or severity of GCT, such as the distance from articular surface to the tumor, the percentage of bone occupied by tumor and the anteroposterior and mediolateral diameter.

However, to our knowledge, there is no clear definition of "more," "extensive" or "severe" destruction" in grade III GCTs and the reported indications of wide resection for GCT are also heterogeneous [5, 7, 22, 23, 30, 38]. Furthermore, another dilemma of the application of the system of Campanacci et al. is that some authors declared that conservative treatments instead of wide resection should be recommended for grade III GCTs. Therefore, the role of the classifications of Enneking and Campanacci et al. in planning the initial surgical treatment is limited.

The purpose of this report is to describe the unusual location of the giant-cell tumor, its clinical and radiographic characteristics of and its magnetic resonance (MR) images.

CASE REPORT

The patient was a 53 year old man, factory worker. He had suddenly developed left L3 lumbar–crural pain 2 months before. Clinical examination showed partial loss of lumbar lordosis and pain during palpation of the L3-L4 vertebrae with paravertebral lumbar contraction. The neurological examination showed racket-shaped hypothesia of the anterior left thigh, with a minimal motor deficit in the left roots of L3 and L4 which was graded 4 on the motoricity test. Genito-sphincter results were normal. The rest of the clinical tests and the biological results were normal.

A standard anterior-posterior X-ray (Figure 1a) and lateral view (Figure 1b) associated with a myelogram (Figure 1c) showed purely osteolytic bone deficit with unclear boundaries associated with collapse of the posterolateral third lumbar vertebra and erosion of the left vertebral pedicle resulting in a "one-eyed" vertebra. This was associated with incomplete extradural uptake in the spine, which was opaque (across from L3).

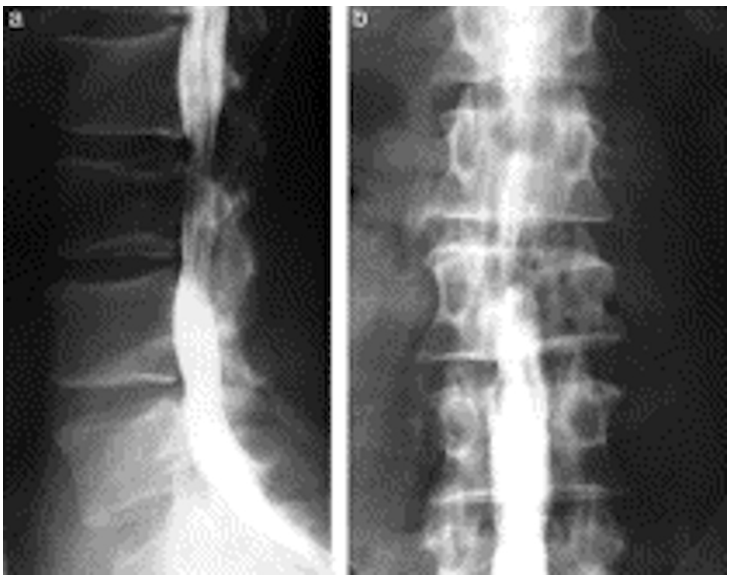


Figure 1. a. standard anterior-posterior X-ray
b. lateral view

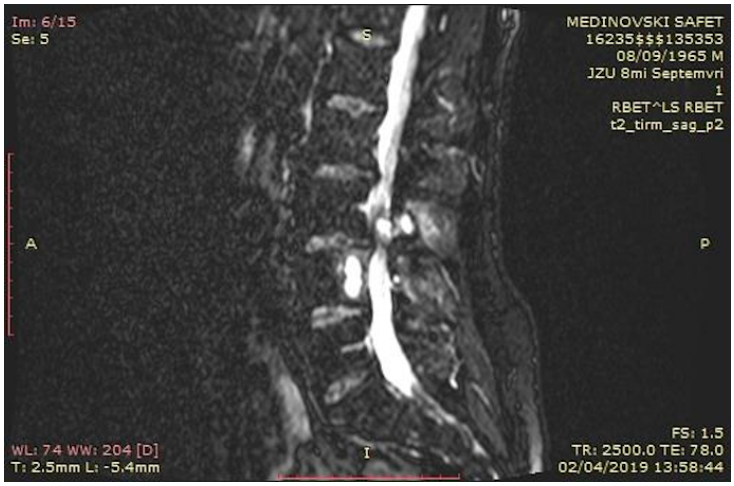


Figure 1. c. myelogram (osteolytic bone deficit with unclear boundaries and vertebral collapse, incomplete extradural uptake)

Axial CT myelogram (Figure 2a) with sagittal reconstruction (Figure 2b) confirmed the presence of well-limited osteolytic lesions of dense tissue of the hemibody of L3 associated with left anterolateral epiduritis compressing and forcing out the dural sheath.

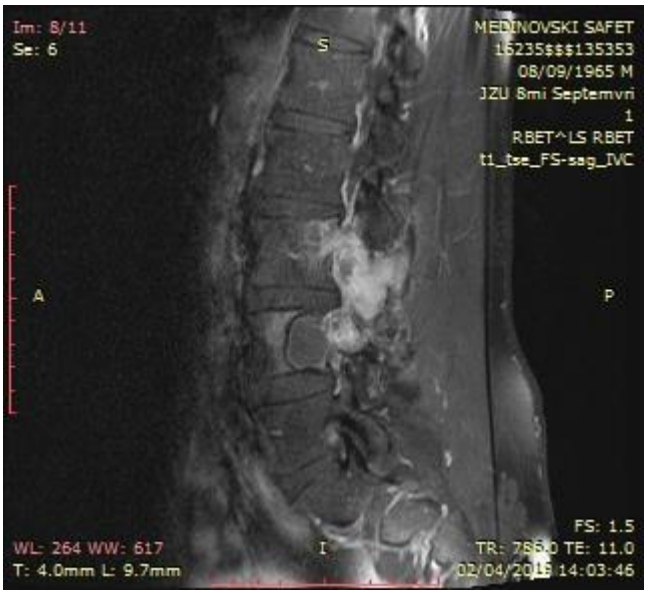


Figure 2a: axial CT Scan: localized osteolysis of the body and posterior arch with invasion of the canal;

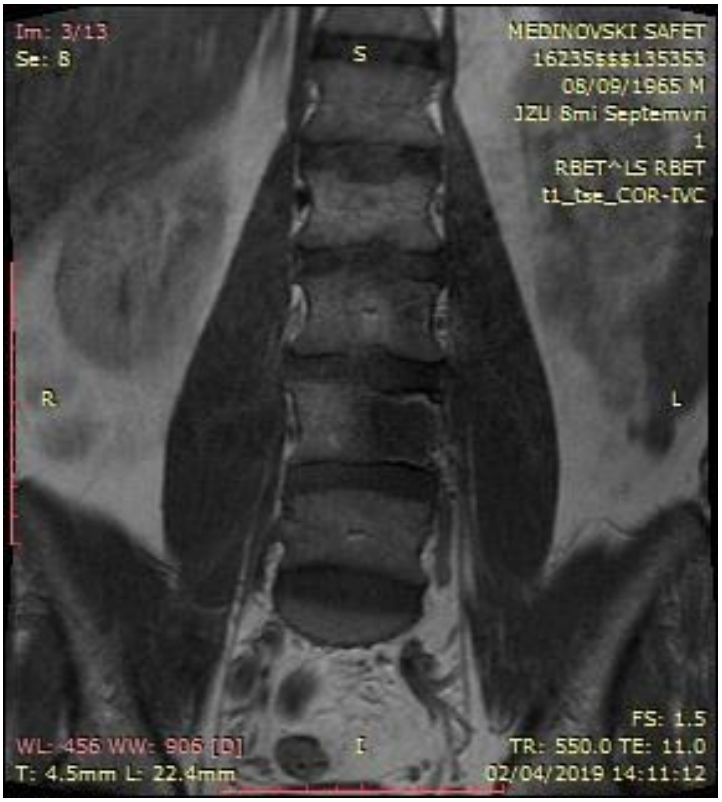


Figure 2b: b: sagittal CT Scan: compression of the dural sheath

MRI (Figure 3a,b) results confirmed the CT Scan results and showed a high intensity signal on T2 and low intensity on T1 sequences with moderate, heterogeneous enhancement of the left hemibody of the L3 vertebra. Invasion of the anterior epidural space and compression of the dural sheath was also seen.

Figure 3

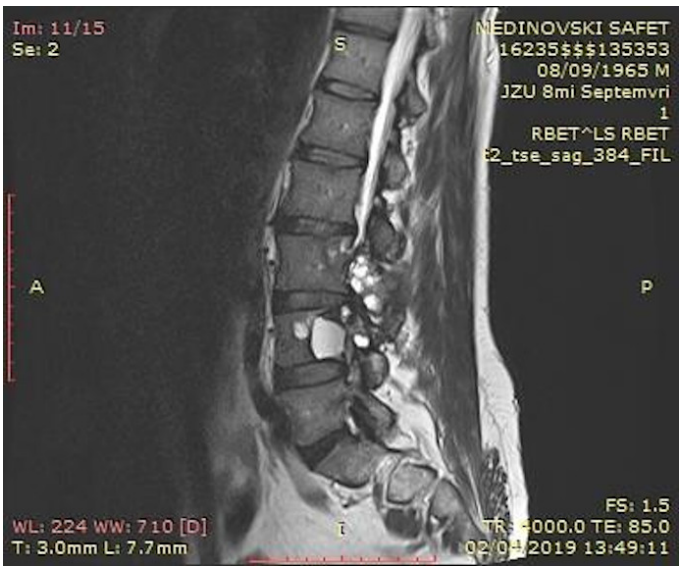


Figure 3

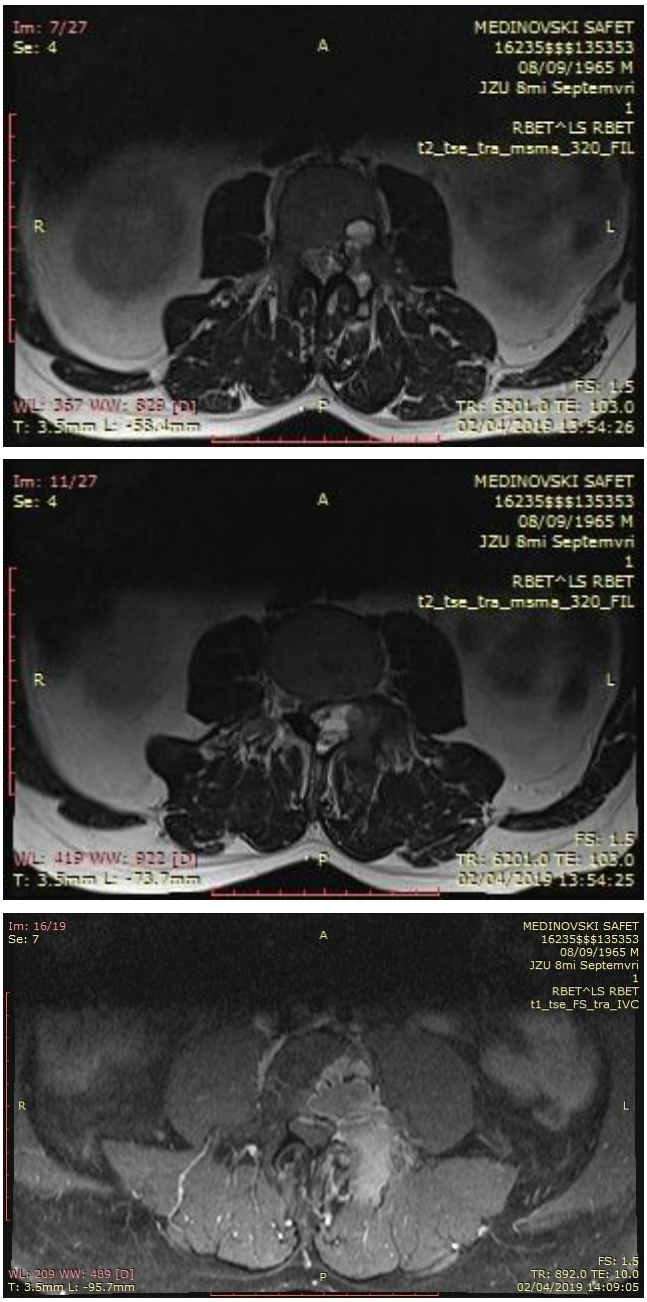


Figure 3:
a: MRI: high intensity signal of the lesion on T2–weighted sequence
b:low intensity signal on T1 sequence and moderate heterogenous enhancement of the hemibody of the left L3 vertebra, invasion of the anterior epidural space and compression of the dural sheath.

CT scan guided puncture biopsy confirmed the presence of Sanerkin's grade 1 GCT [7].

As a next examination we were doing CT-guided needle biopsy may help confirm the diagnosis, which helps in treatment planning.

In a needle biopsy, a hollow needle is inserted through the skin into the tumor.

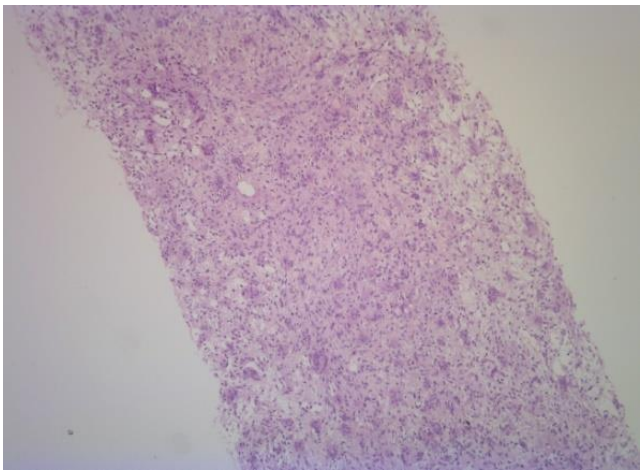


Figure 4. Microscopic slide of core biopsy specimen (H&E staining, x100)

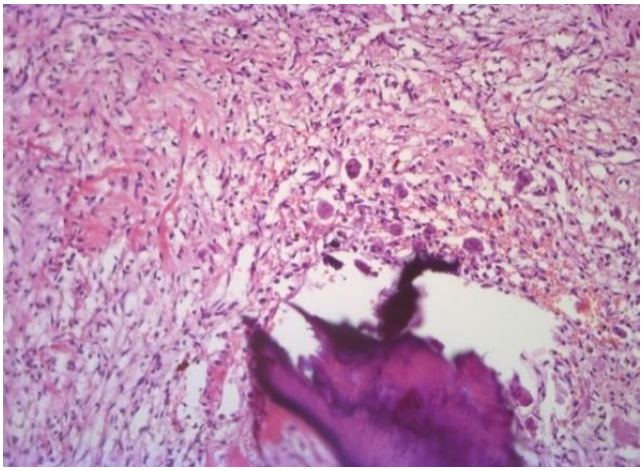


Figure 5. Microscopic slide of the operative material specimen (H&E stainingx100)

For histopathological analysis, two specimens were obtained with CT guided core biopsy from the L4 vertebral body. The biopsies were measuring 1,5cm and 1,8cm in length each. Microscopic analysis of the slides revealed neoplastic proliferation composed from numerous osteoclast-like multinucleated giant cells, intermixed with spindle and round to oval mesenchymal mononuclear cells (Fig.4). The case was signed out as Giant cell tumour of bone, NOS (Osteoclastoma). The analyses of the specimens obtained afterwards with surgical procedure confirmed our initial diagnoses of Giant cell tumour of bone, with more clear characteristics and infiltrative features towards surrounding bone and cartilage. There were erosions towards subchondral bone plate and destruction of the cortex, with emphasize of the rich vascularization, acute haemorrhage and focal discrete sediment from hemosiderin (Fig.5).

Three-phase skeletal scintigraphy was performed as well as a SPECT / CT study in the region of interest with an osteotropic 99mHz-MDP radiotracer. (Figure 6)

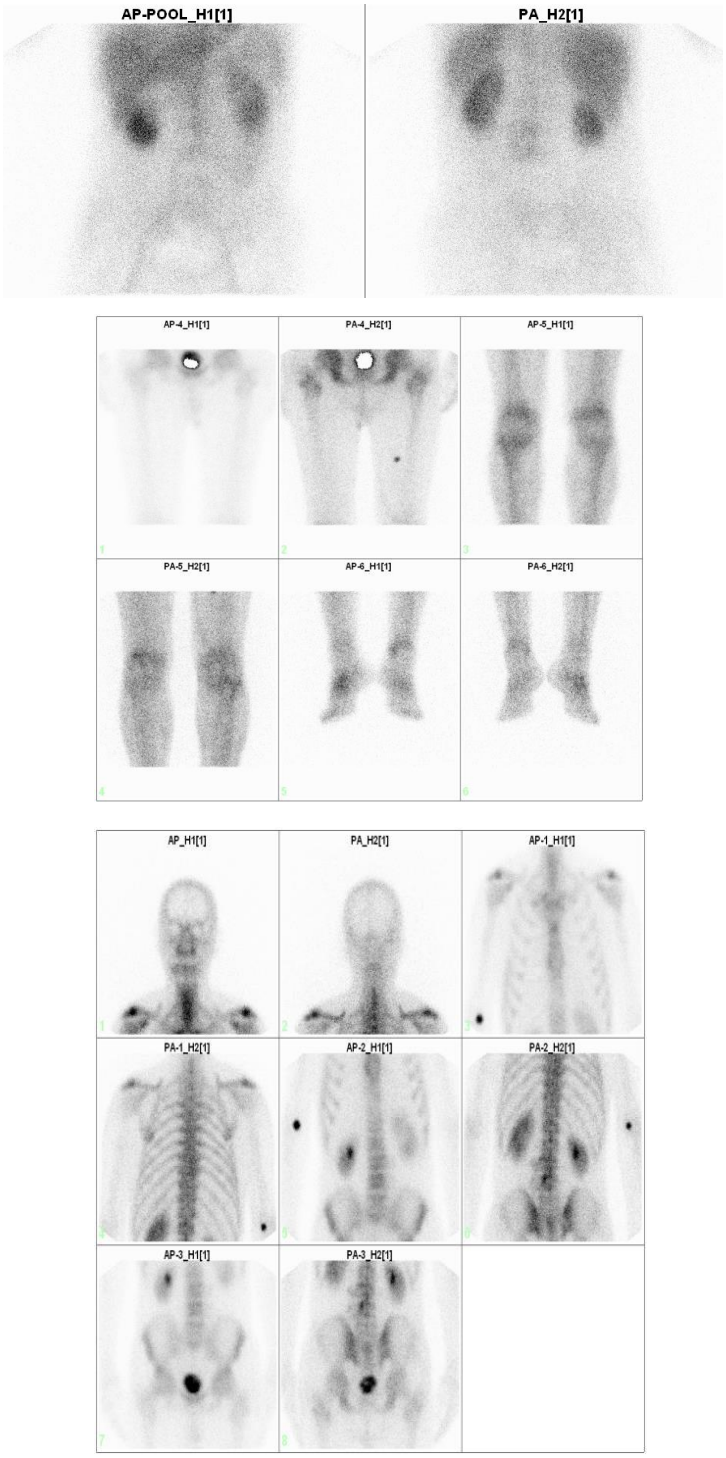


Figure 6: Radionucleide bone scan with Tc-MDP

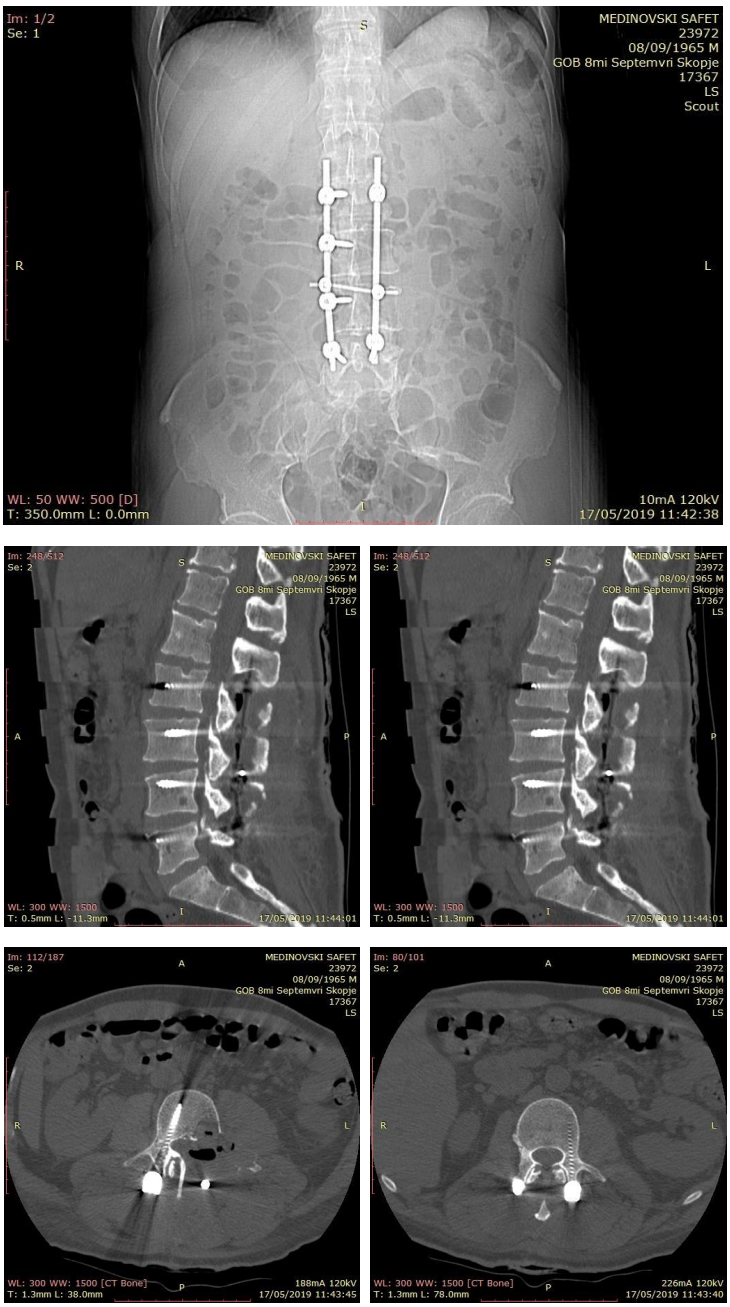
In the pulse phase, in the paravertebral lumbar projection, the left shows a more intense (probably soft) blood formation that is superimposed with the lumbar segment of the spine.

Late scintigrams show photoperiods of vertebral bodies and lateral aspects to the left of L3 and L4 due to osteolysis and destruction by infiltrative growth of soft tissue tumors positioned paravertebrally to the left.

The rest of the skeleton's findings are without pathological accumulations of the tracer.

The patient first underwent decompression with laminectomy, partial corporectomy of verthebrae L3 - L4 and emptying the tumor by curettage and suction and discectomy of L3 - L4. Macroscopically excision of the tumor was considered complete. Marked peridural bleeding was controlled with careful hemostasis.

After that we were doing L2 – L5 osteosynthesis as well as a transpedicular fixation. (Figure 7)



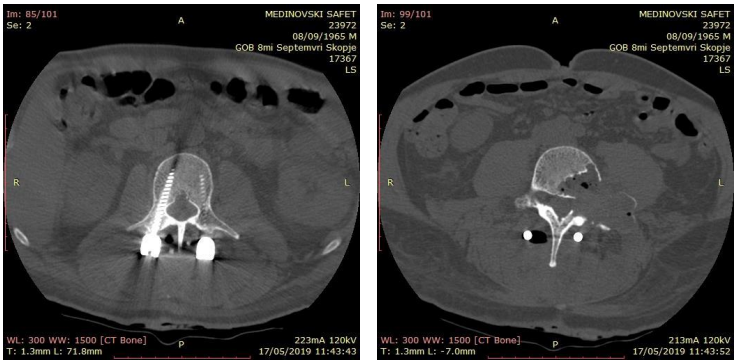


Figure 7. a. Postoperative standard X-rays b. Postoperative CT

Histological confirmation of the diagnosis requires a surgical biopsy [2] or a CT scan guided puncture biopsy, whose reliability is 85% [15]. In this report we performed a surgical biopsy at the same time as decompressive laminectomy. A CT scan guided puncture biopsy is safe [16], and was performed to confirm tumor recurrence. In most cases, the histological examinationconfirms the diagnosis of GCT and excludes the main differential diagnoses, in particular aneurysmal cyst [1]. Sanerkin is the reference classification for the histological grading of a bone GCT [1, 17]. Grade I is the benign form of the disease, while grade III is osteosarcoma, and grade II is a borderline form.

DISCUSSION

Spinal GCTs are rare; according to the Mayo clinic, they represent 6.5% of bone GCTs [6] and 1–9% of bone GCTs according Bedwell et al. [8]. The largest worldwide series of 31 cases of giant cell tumors from different centers was reported in 1977 by Dahlin et al. [2]. In 1993, Sanjay et al. [6] reported 24 cases of spinal giant cell tumors from cases in the Mayo Clinic between 1955 and 1989. Although this lesion often only involves one vertebra, Kos et al. [9] published a case of multifocal thoracic and sacral spine GCT and Erdogan et al. [4] published a case of GCT in the sixth cervical vertebra. The different reported cases often occurred in patients between 20 and 30 years old and especially in women [2, 6, 10, 11].

Spinal pain with or without radiculalgia is the most frequent cause for consulting [3, 6, 10]. However, the diagnosis of GCT is often made after a neurological deficit has developed [2, 3, 6, 10]. Standard X-ray usually shows an osteolytic lesion, without peripheral bone condensation, of even density, which follows the intervertebral disc. If no pedicle is identified on anteroposterior X-ray views (“one-eyed” vertebra), this suggests invasion of the posterior arch. Unlike an aneurysmal bone cyst, GCT usually develops in the vertebral body then invades the posterior arch [10]. In the case reported here, a CT myelogram was performed because a rapid MRI was impossible. CT scan is useful to evaluate bone degeneration because of the excellent spatial resolution. A precise evaluation of any epidural invasion associated with medullary compression can also be obtained [12]. At present, MRI is the gold standard for evaluating locoregional invasion in spinal GCT, to determine the size of the tumor and look for intracanal extension. The signal is usually mixed, with a low intensity signal on T1 and a high intensity signal on T2-weighted mages [10, 13, 14].

Treatment of these tumors must take into account three problems: mechanical because of the extensive osteolysis of the vertebral body, neurological and tumoral with the risk of recurrence [2, 6, 11]. Treatment of spinal GCT is usually surgical [1, 2, 3, 5, 6, 10, 11]. The possibilities of extratumoral surgery are extremely limited [18]. An isolated lesion of the vertebral body can be treated by total spondylectomy by the anterolateral approach [18, 19, 20]. Unfortunately, extension into one of the two pedicles makes extratumoral resection impossible [18]. Partial spondylectomy, corporectomy or resection of the posterior arch is a viable option in well-circumscribed lesions [2, 6, 11, 18, 19, 20, 21, 22, 23]. In a GCT of the second lumbar vertebra which had invaded the medullary canal, Li et al. [3] performed wide en bloc resection of the tumor, including the vertebral body and the psoas muscle followed by a laminoarthrectomy by resection of the right root of L2. No recurrence had occurred at 13 years of follow-up. Several vertebral reconstruction procedures have been used. Lafarge et al. [2] filled bone defects with autologous grafts alternating with slices of allograft strengthened with transversal screws and screw plate osteosynthesis. Li et al. [3] used fibular grafts to strengthen vertebrae above and below with compression screws. Smartis et al. [19] performed posterior resection and short-termosteosynthesis, then anterior corporectomy with a cage implant for filling, then a posterior approach for pedicular reconstruction. The use of adjuvant radiotherapy is considered to be a factor favoring the development of sarcoma in an estimated 10% of cases [6]. It can be indicated in inoperable GCT [10, 24], incomplete GCT resections, recurrent GCT [6] or as adjuvant therapy to surgery [21, 24]. The role of biphosphonates in the prevention of recurrent bone GCT was confirmed in a

study by Tse et al. [25]. Its efficacy in spinal forms was reported by Fujimoto et al. [26] but in association with radiotherapy.

Bleeding during surgery of spinal GCT is a severe complication, which can make it impossible to complete the surgical procedure [11]. Preoperative embolisation can prevent this complication and reduce the size of the tumor, facilitating resection [27].

Recurrent GCT after surgery is a serious complication, and treatment is a problem. Most authors believe that it is due to marginal surgical resection [3, 28]. Sanjay et al. [6] reported 10 cases of recurrence in 24 spinal GCT. According to Campanacci et al. [29], 90% of recurrence developed in the first three years after surgery. He noted that recurrence had not occurred in total spondylectomy 13 years after surgery. In our report, recurrence developed 5 years after surgery at a stage of neurological compression. Follow-up visits ought to have included MRI imaging to detect recurrence as early as possible. The complication in our report is mainly explained by insufficient resection, which was limited to simple anterior curettage. Recently, Junming et al. [21] published a series of 22 cervical spine GCTs. The rate of recurrence with subtotal spondylectomy was 71% while for total spondylectomy it was only 7.7%.

CONCLUSION

GCTs of the lumbar spine are rare and their clinical and radiographic characteristics are not specific. MRI is indispensible to evaluate local extension and especially to identify nerve compression. If the vertebral body and the posterior arch are affected, curettage of the lesion is insufficient to prevent tumor recurrence. This occurred in the present report, where a total spondylectomy should have been performed to minimize this risk.

REFERENCES

1. Mertens F. Pathology and genetics of tumours of soft tissue and bone. Lyon: International Agency for Research on Cancer Press; 2002. pp. 309–313. [Google Scholar]
2. Niu X, Zhang Q, Hao L, Ding Y, Li Y, Xu H, Liu W. Giant cell tumor of the extremity: retrospective analysis of 621 Chinese patients from one institution. J Bone Joint Surg Am. 2012;94(5):461–467. 10.2106/ JBJS.J.01922. [PubMed] [CrossRef] [Google Scholar]
3. Jeys LM, Suneja R, Chami G, Grimer RJ, Carter SR, Tillman RM. Impending fractures in giant cell tumours of the distal femur: incidence and outcome. IntOrthop. 2006;30:135–138. doi: 10.1007/s 00264-005-0061-z. [PubMed]Fletcher CDM, Unni KK,
4. Kivioja AH, Blomqvist C, Hietaniemi K, Trovik C, Walloe A, Bauer HC, Jorgensen PH, Bergh P, Follerrecommended in intralesional surgery of giant cell tumors: a Scandinavian Sarcoma Group study of 294followed for a median time of 5 years. ActaOrthop. 2008;79(1):86–93.10.1080/17453670710014815 [PubMed] [CrossRef]
5. Rooney RJ, Asirvatham R, Lifeso RM, Ali MA, Parikh S. Giant cell tumour of bone. A surgical approach to grade III tumours. IntOrthop. 1993;17(2):87–92. doi: 10.1007/BF00183548. [PubMed]
6. Sung HW, Kuo DP, Shu WP, Chai YB, Liu CC, Li SM. Giant cell tumor of bone: analysis of two hundred and eight cases in Chinese patients. J Bone Joint Surg Am. 1982;64:755–761. [PubMed]
7. Giant cell tumour of bone.Thomas DM, Skubitz, CurrOpinOncol. 2009 Jul; 21(4):338-44. [PubMed]
8. Tomeno B., Forest M. Tumeurs à cellule géantes Cahiers d'enseignement de Sofcot 1990;38:31-50
9. Laffargue P.H., Cotton A., Cortet B., Lecomte-Houcke M., Decoulx J. Tumeur à cellules géantes du rachis: à propos d'un cas ActaOrthBelg 1997 ; 63 : 28-34
10. Li Y.H., Kour A.K., Pho R.W.H. Giant cell tumor of lumber vertebra Clinic Orthop 1998;353:218-2221
11. Erdogan B., Aydin M.V., Sen O., Sener L., Bal N., In Y. Giant cell tumor of the sixth cervical vertebrae with close the vertebral artery ClinNeurosci 2005 ; 12 : 83-85
12. Takeda N., Kobayashi T., Tandai S., Matsuno T., Shirado O., Watanabe T., Minami A. Treatment of giant cell tumor and spine with curettage and argon beam coagulator OrthopSci 2009 ; 14 : 210-214
13. Sanjay B.K.S., Sim F.H., Unni K. McLeod RA Klassen RA. Giant cell tumors of the spine. J Bone Joint Surg Br 199 154
14. Sanerkin N.G. Malignancy, agressiveness and recurrence in giant cell tumor of bone Cancer 1980 ; 46 : 1641-1649
15. Bidwell J.K., Young J.W.R., Khalluff E. Giant cell tumor of the spine: computed tomography appearance and review literature J ComputTomogr 1987 ; 11 : 307-311

16.Kos C.B., Taconis W.K., Fidler M.W., Ten Velden Jos J.A.M. Multifocal giant cell tumor in thespine Spine 1997 ; 22 : 821-822
17.Ozaki T., Liljenqvist U., Halm H., Hillman A., Gosheger G., Winkelmann W. Giant cell tumor of the spine Clinic Or 401 : 194-201
18. Shimada Y., Hongo M., Miyakoshi N., Kasukawa Y., Ando S., Itoi E., Abe E. Giant cell tumor of fifth lumbar vertebrae: two case reports and review of the literature Spine 2007 ; 7 : 499-505
19.Zimmer W.D., Berquist T.H., McLeod R.A., Sim F.H., Pritchard D.J., Shives T.C., Wold L.E., May G.R. Bone tumors: magnetic resonance imaging versus computed tomography Radiology 1985 ; 155 : 709-718
20.Colombo N., Berry I., Norman D. Vertebral tumors New York: Raven Press (1992). p. 445–487.
21.Saint Amour T.H.E., Hodges S.C., Laakman R.W., Tamas D.E. MRI of the spine New York: Raven Press (1994). p. 5
22. Vieillard M.H., Boutry N., Chastanet P., Duquesnoy B., Cotten A., Cortet B Rentabilité de la ponction-biopsiepercutan diagnostic des tumeursosseusesvertébrales et périphériques Rev Rhum 2005 ; 72 : 59-67
23.Murphy W.A., Destouet J.M., Gilula L.A. Percutaneous skeletal biopsy: a procedure for radiologists. Results, review, a recommandations Radiology 1981 ; 139 : 545-549
24.Donthineni R., Boriani L., Ofluoglu O., Bandiera S. Metastatic behaviour of giant cell tumour of the spine Intorthop 20 501
25.Enkaoua E., Saillant G. Tumeursdu rachis et dusacrum Techniques chirurgicales orthopédie. Encycl.MédChir : (1999). p. 44165 5
26.Samartzis D., Foster W.C., Padgett D., Shen F.H. Giant cell tumor of the lumbar spine: operative management via spot segment, 3-column reconstruction with pedicle recreation SurgNeurol 2008 ; 69 : 138-141
27.Shimizu K., Ido k, Fujio K., Ttanaka K., Nakamura T. Total spondylectomy and spinal shortening for giant-cell tumor of spine Lancet 1996 ; 3 : 277-346
28.Junming M.,Cheng Y.,Dong C.,Jianru X.,Xinghai Y., Quan H., Wei Z., Mesong Y., Dapeng F., Wen Y., Bin N.,Lia Giant cell tumor of the cervical spine: a series of 22 cases and outcomes Spine 2008;33: 280-288
29.Fidler M.W. Surgical treatment of giant cell tumours of the thoracic and lumbar spine: report of nine patients Eur Spin
30.Shikata J., Yamamuro T., Shimizu K., Kotoura Y. Surgical treatment of giant–cell tumors of the spine Clinic Orthop 1
31.Caudell J., Ballo M., Zagars G.K., Lewis V.O. Radiotherapy in the management of the giant cell tumors of bone Int J RadiatOncol Phys 2003 ; 57 : 158
32.Tse L.F., Wong K.C.,Kumta S.M., Huang L., Chow T.C., Griffith J.F. Bisphosphonates reduce local recurrence in extremity giant cell tumor of bone: a case-control study Bone 2008 ; 42 : 68-73
33.Fujimoto N., Nakagawa K., Seichi A., Terahara A., Tago M., Aoki Y., Hosoi Y., Ohtomo K. A new bisphosphonate treatment option for giant cell tumors Oncol Rep 2001 ; 8 : 643-647
34.Broaddus W.C., Grady M.D., Delashaw J.B., Erguson R.D.G. Preoperative superselective arteriolar embolisation: a ne respectability of spinal tumors Neurosurg 1990 ; 27 : 755-759
35.Shimada Y., Hongo M., Miyakoshi N., Kasukawa Y., Ando S., Itoi E., Abe E. Giant cell tumor of fifth lumbar vertebra review of the literature Spine 2007 ; 7 : 499-505
36.Campanacci M., Baldini N., Boriani S., Sudanese S. Giant cell tumor of bone J Bone joint Surg Am 1987; 69: 106-114
37.Recurrence of giant-cell tumors of the long bones after curettage and packing with cement. O'Donnell RJ, Springfield DS, Motwani HK, Ready JE, Gebhardt MC, Mankin HJ J Bone Joint Surg Am. 1994 Dec; 76(12):1827-33. [PubMed] [Ref list]
38.Giant cell tumor of bone: risk factors for recurrence.Klenke FM, Wenger DE, Inwards CY, Rose PS,Sim FHClinOrthopRelat Res. 2011 Feb; 469(2):591-9. [PubMed] [Ref list]
39.Balke M, Schremper L, Gebert C, Ahrens H, Streitbuerger A, Koehler G, Hardes J, Gosheger G. Giant cell tu treatment and outcome of 214 cases. J Cancer Res ClinOncol. 2008;134(9):969–978. doi: 10. 1007/s00432-008-0[CrossRef][GoogleScholar][Ref list]
40.Giant-cell tumor of bone. Campanacci M, Baldini N, Boriani S, Sudanese A J Bone Joint Surg Am. 1987 Jan; 69(1):106-14. [PubMed] [Ref list]
41.Conservative treatment of Campanacci grade III proximal humerus giant cell tumors. Lackman RD, Crawford EA, King JJ, Ogilvie CM ClinOrthopRelat Res. 2009 May; 467(5):1355-9. [PubMed] [Ref list]
42.Joint salvage for pathologic fracture of giant cell tumor of the lower extremity. Deheshi BM, Jaffer SN, Griffin AM, Ferguson PC, Bell RS, Wunder JS Clin Orthop Relat Res. 2007 Jun; 459():96-104. PubMed] [Ref list]

RARE TENDON SHEATH GIANT CELL TUMOR OF THE THUMB – CASE REPORT

РЕТКА ЛОКАЛИЗАЦИЈА НА ТЕТИВЕН ГИГАНТОЦЕЛУЛАРЕН ТУМОР НА ПАЛЕЦОТ-ПРИКАЗ НА СЛУЧАЈ

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АПСТРАКТ

Giant cell tumor на тетивите е бениген мекоткивен тумор. Во литературата е опишано дека се јавува кај луѓе од 30 до 50 годишна возраст, без доминантност кај машкиот или женскиот пол. Giant cell tumor е бениген тумор кој има потенцијал да направи рецидив. По радикално хируршко отстранување, според литературата, кај 15% - 44% од пациентите има појава на рецидив на истата локација. Според процентот на застапеност, овој тип на тумор е вториот најчест тумор од бенигните тумори на дланката. Најчесто се јавува на прстите од дланката. Во овој труд, презентираме хируршки третиран пациент со giant cell tumor на тетивата, на палмарната страна во ниво на дисталната фаланга на десниот палец.

Клучни збоорови: гигантоцелуларен тумор на тетивната обвивка, палец

ABSTRACT

Giant cell tumor of the tendon sheath is benign soft tissue tumor. The literature shows it affects people between age of 30 to 50 years with no gender predominance. Giant cell tumor is benign tumor which sometimes has potential to reappear. After radical surgical removal, according to the literature, 15%-44% of the patients have recurrence of the tumor on the same location. This type of tumor is usually located on the fingers, and resembles the second most common benign tumor of the hand. In this report we present a surgically treated patient with giant cell tumor of the tendon sheath in the palmar part of the distal phalanx of the right thumb.

Key words: giant cell tumor of the tendon sheath, thumb.

INTRODUCTION

Giant cell tumoris benign soft tissue tumor. The literature shows it affects people between ages of 30 to 50 years with no gender predominance. Giant cell tumoris benign tumor, which sometimes has potential of recurrence. After surgical removal of the tumor, literature reports that 15%-44% of the patients have recurrence of the tumor on the same location.[1]Soft tissue mass or bump associated with local swelling are typical clinical symptoms.[2] Local palpable pain on the lesion may occur. This type of tumor is usually found in the hand, although the literature describes other localizations also, like the ankle or the foot.[3] Most common localization of the tumor is the dorsum of the index finger, after that is the thumb and the rarest localization of the hand is the little

Ifinger.[4] The giant cell tumor is the second most common benign tumor of the hand.

In this report we present a surgically treated patient with giant cell tumor of the tendon sheath in the palmar part of the distal phalanx of the right thumb.

CASE REPORT

We present a 47 year old male, right hand dominant, with five month mild pain in the right thumb. The patient reported no sign of trauma. The clinical examination showed palpable painful soft tissue mass (bump) in the proximal part of the right thumb (Figure 1). Radiographs were made (Figure 2) and the patient was scheduled for needle biopsy. The results of the biopsy referred a giant cell tumor. The patient was admitted in our clinic for excision of the tumor.

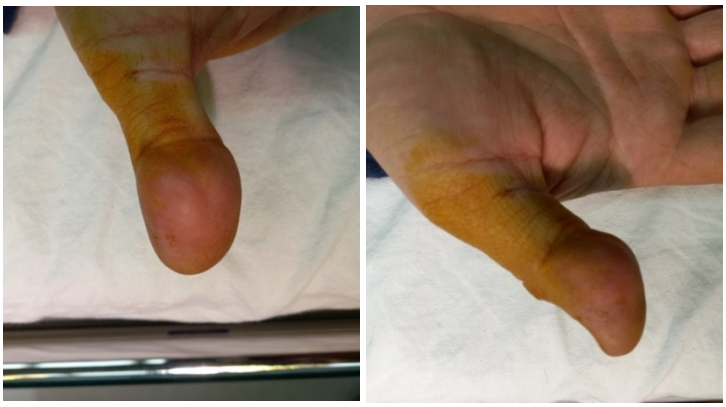

Figure 1: Soft tissue mass of the right thumb

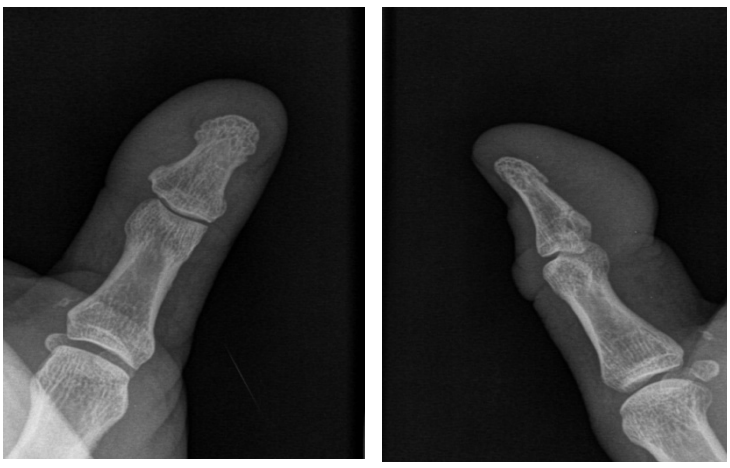

Figure 2: Native radiographs of the right thumb

In local anesthesia skin incision was made. A folded surgical glove as Tourniquet was used. Stuffing of the subcutaneous tissue was made and identification of the local anatomical structures was performed. (Figure 3). The tumor was carefully dissected and separated from the local tissue, than was radically removed (Figure 4).

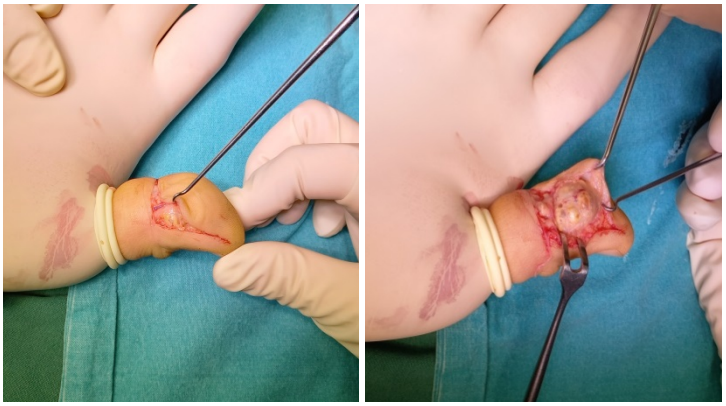

Figure 3: Surgical approach and dissection of the tumor

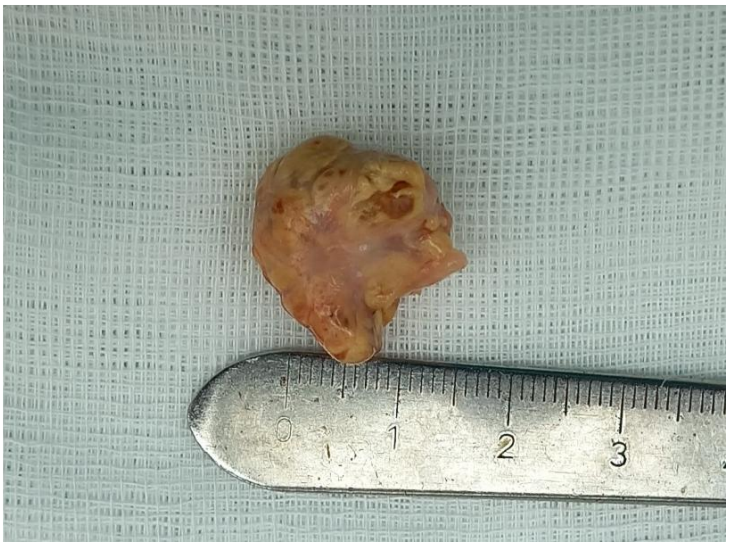

Figure 4: Dimensions of the radically removed tumor

Curette was taken to clear and remove the suspicious tissue in the vicinity of the tumor. Hemostasis was meticulously performed and the surgical wound was irrigated. The wound edges were approximated and closed with intradermal stiches. Dressing of the surgical wound was done and the tumor with all of the tissue samples were sent to pathohistology (Figure 5).

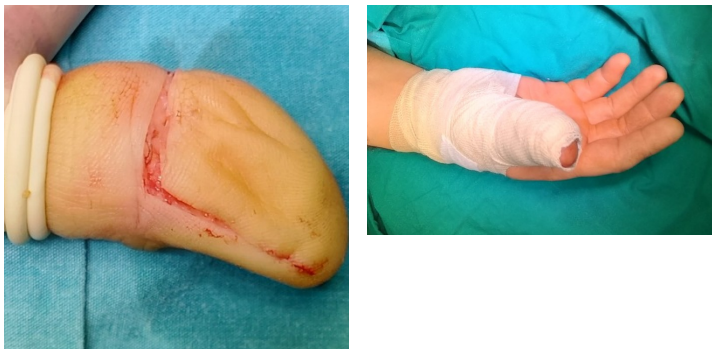

Figure 5: Intradermal closing of the wound and dressing

DISCUSSION

Giant cell tumor of the tendon sheath is relatively rare tumor with predominant localization on the hand. It is a slow growing benign tumor that can develop for months and years. The literature shows it affects people between ages of 30 to 50 years with no gender predominance. Giant cell tumor of the tendon sheath is benign tumor, that sometimes have potential of recurrence. The surgical treatment is recommended, although incomplete tumor resection can result in tumor recurrence. Recurrence after surgical excision has been reported as high as 15–45%. The literature reports that recurrence of giant cell tumor is much higher at the thumb localization, because of the anatomical structures like: the neuro-vascular bundles that are quite close to tumor margins and the local soft tissue compartments.

Depending of the localization of the giant cell tumor, preservation of the flexor and extensor tendons, as well as the digital arteries and nerves should be performed. Because of the pseudo capsule of the tumor, the giant cell tumor can be easily radically removed.

CONCLUSION

Giant cell tumor of the tendon sheath is benign soft tissue tumor, most frequently located in the hand. In this case study we present a 47 year old male with giant cell tumor in the palmar part of the distal phalanx of the right thumb, which was treated surgically, with radical extirpation. Operative treatment is treatment of choice for giant cell tumors, although incomplete resection of the tumor may result in tumor reappearance in the same location.

Thorough physical examination and patient history, as well as detailed preoperative plan are needed for best patient outcome.

REFERENCES

1. Zheng, Shaoling, et al. “Giant Cell Tumor of Tendon Sheath and Tendinopathy as Early Features of Early Onset Sarcoidosis.” Frontiers in Pediatrics, vol. 7, 15 Nov. 2019, 10.3389/fped.2019.00480.
2. Mavrogenis, Andreas F., et al. “Giant Cell Tumor of Soft Tissue: A Rare Entity.” Orthopedics, vol. 42, no.4, 1 July 2019, pp. e364–e369, pubmed. ncbi.nlm.nih.gov/31323108/#:~:text=Giant%20cell %20 tumor %20 (GCT)%20of,10.3928/01477447-20190624-04. Accessed 22 Jan. 2022.
3. Suresh, S., and HosamZaki. “Giant Cell Tumor of Tendon Sheath: Case Series and Review of Literature.” Journal of Hand and Microsurgery, vol. 02, no. 02, 5 Sept. 2016, pp. 67–71, 10.1007/s12593-010-0020-9.Accessed 22 Jan. 2022.
4. G Chir, and Vol. Introduction Giant Cell Tumor of Tendon Sheath: Study of 64 Cases and Review of Literature. Vol. 34, 2013, pp. 149–152. Accessed 22 Jan. 2022.

ИНСТРУКЦИИ ЗА АВТОРИТЕ

Општи информации

Македонскиот гласник за ортопедија и трауматологија е званична публикација на македонското друштво на ортопеди и трауматолози (МАДОТ). Апстрактите на трудовите се двојазични, т.е. на македонски и на англиски јазик, а текстот на англиски јазик. Гласникот е специјализирано медицинско списание со цел да обезбеди информации за новини, научни достигнувања, клинички искуства и дискутабилни теми на полето на ортопедијата и трауматологијата, како и на граничните специјалности. Ракописи можат да се доставуваат од следните области: оригинални теми, прикази на случаи, теми за дискусија, критики, дијагностички, терапевтски и технички иновации, терминолошки дискусии, импресии од научни состаноци, информации за меѓународни состаноци, коментари за нови книги, писма до издавачот и тн.

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Секој труд кој што ги задоволува гореспоменатите инструкции е подложен на рецензија и оцена од страна на издавачкиот одбор. Процесот на рецензија и конечно прифаќање или одбивање на одреден труд трае од 4 до 6 недели. Потоа, авторите се информираат за одлуката на издавачкиот одбор.

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Текст

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Големината на оригиналниот труд не треба да надмине 8 страници (исклучувајќи ги илустрациите), додека за приказите на случаи треба да се напишат до 3 страници. За ревијални трудови дозволени се 12 страници. Страниците треба да бидат последователно нумерирани, почнувајќи од **насловната страница**. Секој труд треба да почне со насловна страница. Таа го содржи насловот на трудот, името на авторот или авторите и името на институцијата на англиски и македонски јазик. Доколку авторите на еден труд им припаѓаат на различни институции, на крајот на името и презимето на секој автор, треба да има мал реден број кој ќе ја означува институцијата од која доаѓа. Истото важи и за наведените институции под имињата на авторите, со тоа што пред името на секоја институција се става реден број кој одговара на редниот број на соодветниот автор. Името и точната адреса на авторот

Text

The language and style of the submitted manuscripts should suit the requirements of the literary **English language**, conformable to the typical characteristics of the orthopedic and traumatologic terminology. The Editorial Board will highly appreciate the efforts of the authors to submit their texts in proper English (reviewed by authorized censor).

The size of the original articles should not exceed 8 pages (excluding the illustrations) and as for the case reports - up to three pages. For literature reviews the size of 12 pages is admissible. All the pages should be consecutively numbered, beginning with the **title page**. Each manuscript should begin with a title page.

This page carries the title of the article, the names of the authors and institutions in Macedonian and in English. The numeration of the authors should correspond with the numeration of the institution. The name and the exact address of the author, to whom the correspondence is to be sent, should be typed in Macedonian and in English at the bottom of the title page. To facilitate communication, the author has to submit her/his phone and fax number, as well as her/his e-mail address.

до кого ќе се испраќа соодветната кореспондеција, треба да бидат назначени на долниот дел од насловната страница, на македонски и на англиски јазик. Заради олесување на комуникациите, авторот треба да достави телефонски број и е-маил адреса.

По насловната страница, треба да следи нова страница со **апстракт** на македонски и англиски јазик, со приближно 250 зборови, вклучувајќи ги накусо проблемот, материјалот и методите, резултатите и заклучоците од поднесениот труд. На крајот на секој апстракт следуваат 3 до 5 **клучни зборови**, дадени по азбучен ред. Тие треба да ги претставуваат најкарактеристичните делови од трудот. Изборот на клучните зборови треба да соодветствува на Индекс Медикус.

Текстот на секој труд треба да ги содржи следните наслови: **вовед, материјал и методи, резултати, дискусија и заклучок.** Во воведот треба да се наведе целта на студијата и краток осврт на проблемот во постојната и значајна литература. Во материјалот и методите треба да се вклучи детален опис на испитуваните пациенти, експерименталните животни, тестови и сл.,како и точно да се презентираат применетите методи во испитувањето. Особено внимание треба да се посвети на статистичките методи кои се употребуваат во анализата на резултатите. Резултатите треба да бидат претставени точно и во детали, бидејќи овој дел во суштина е срцето на трудот. Дискусијата содржи интерпретација на добиените резултати во споредба со постојното ниво на знаење и резултати од други автори. Заклучоците го генерализираат научното и практичното значење на публикацијата.

Трудовите да се достават во електронска форма или на ЦД, подготвени во Microsoft Word.

Во подготовката на текстовите на македонски јазик да се користи **тастутура со македонска поддршка** а соодветно за текстовите на англиски јазик да се користи тастатура со англиска поддршка. При подготовка на трудот да се користат фонтовите Arial или Times New Roman. Фонтот Arial да се користи за пишување на насловите, поднасловите, насловот на табелите и насловот на илустрациите, додека за останатиот дел од текстот да се користи фонтот Times New Roman.

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Following the title page, there should be a new one with the **abstract** in Macedonian and in English. The size of it should be approximately 250 words, including in brief the problem, material and methods and results and conclusions. At the end of each abstract, 3 to 5 **key words** should follow in alphabetical order representing the most characteristic points of the manuscript. The selection of key words should suit the requirements of Index Medicus.

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Tables and Figures must have their number and title. Table number and title should be typed over the table, and the number and title of the figures should be typed below them. The titles and content of the tables and figures should be short and clear. They should include all necessary explanations in order to be understood without reading the covered text.

All figures (graphs, schemes, drawings and pictures) must be numbered as figure 1, figure 2, figure 3 etc. All figures attached to the text must be sent on a CD together with the text as separate files (graphs created in Excel, with extension XLS; schemes, drawings and pictures with extension JPG or TIF). The resolution of the figures must be at least 200 DPI.

табелата, а бројот и насловот на илустрациите (графикони, слики, цртежи и скици) се пишуваат под нив. Насловите и содржината на табелите и илиустрациите треба да се кратки и јасни. Тие треба да ги содржат сите потребни објаснувања.

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Литература

Листата на референци треба да биде доставена на одделна страница. Во трудот треба да бидат вклучени само цитираните извори. Авторите треба да бидат нумерирани по редот на појавувањето во текстот. Таму каде што има повеќе автори на еден труд, се наведуваат по азбучен ред на презимето. Авторите со кирилско писмо се пишуваат пред оние на латиница. Се препорачува следниот редослед: презиме на авторот/авторите, иницијал од името/имињата, име на трудот, име на публикацијата, година на публикација, број на том и број на прва и последна страница. Не се стават точки по иницијалот од името. Во текстот, бројот на цитираната референца треба да биде даден во заграда или по името на авторот или по односниот пасус.

Пример:
1. Hollenberg GM, Adams MJ, Weinberg EP. Sonographic appearance of nonoperatively treated Achilles tendon ruptures. Skeletal Radiology, 2000; 29: 259-264.

References

The list of references should be submitted on a separate sheet of paper. Only the sources cited in the text should be included. The authors should be listed according to the order of appearance in the text by their family names in alphabetical order. The authors with Cyrillic alphabet should precede those with Latin alphabet. The following schedule is recommended: the surname of the author/authors, the initial of the name/ names, title of the abstract, name of the publication, year of issue, Chapter number (if any) and the first and last page of the cited part of the book or journal. There is no dot after the name initial. In the text, the number of the cited reference should be given in brackets after the name/s of the author/s or the respective passage.

Example:
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