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Retrorectal Dermoid Cyst Manifested as an Extrasphincteric Perianal Fistula - Case Report

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Rezumat

Chist dermoid retrorectal manifestat ca fistulă perianăextrasfincteriană - prezentare de caz

Tumorile retrorectale sunt entități patologice foarte rare, dar bine definite, în literatura de specialitate. De asemenea, o fistula extrasfincteriană este o formă foarte rară de fistulă perianală, ceea e face cazul nostru foarte neobișnuit și rar, în special prin faptul că aceasta a fost tratată cu succes cu prima operație şi fără formarea unei stomii de protecție. Pacientul a fost tratat în spital în primă fază pentru un abces retrorectal care a generat o ruptură spontană în spațiul postanal. Din cauza drenării constante de conținut purulent din deschiderea postanală în următoarele luni, s-au efectuat RMN și fistulografie, care au înregistrat o formațiune chistică în spațiul retrorectal cu formare de fistulă ce comunică cu rectul de mai sus și complet separată de aparatul sfincterian. Ulterior pacientul a fost internat pentru tratament definitiv. Operația a fost efectuată cu pacientul înclinat, în poziție de briceag. A fost efectuată excizia completă a chistului împreună cu fistula și rectul a fost suturat în dublu strat cu suturi resorbabile ușor separate. Rana a fost lăsată deschisă şi pacientul a fost externat în ziua a 5-a postoperator. După aproximativ zece luni defecația este normală, rana este închisă și nu există semne de inflamație și secreție la nivel local.

Cuvinte cheie: fistula extrasfincteriană, chist retrorectal, excizie, sutură

Abstract

Retrorectal tumors are very rare but well defined pathological entities in the literature. Also, an extrasphincteric fistula is a very rare form of perianal fistula which makes our case a very unusual and rare one, especially by the fact that it was successfully treated with the first operation and without protective stoma formation. The patient was first treated in hospital for a retrorectal abscess that had spontaneously ruptured in the postanal space. Because of the constant drainage of the suppurative content from the postanal opening in the following months, MRI and fistulography were performed, registering cystic formation in the retrorectal space with fistulous communication with the rectum above and completely separate from the sphincter mechanism. After that the patient was admitted for definitive treatment. The operation was performed with the patient in a prone jack-knife position. Complete excision of the cyst with the fistulous communication was performed and the rectum was sutured in two layers with separate slowly absorbable sutures. The wound was laid open and the patient was discharged on the 5th post-operative day. After about ten months the defecation is normal, the wound is sealed and there are no signs of inflammation and secretion locally.

Key words: extrasphincteric fistula, retrorectal cyst, excision, suture

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Introduction

Retrorectal tumors are well defined, classified and understood pathological entities in the literature, but in practice they represent very unusual and infrequent pathology (1,2,3). The presacral or retrorectal space is a common site for embryologic remnants from which neoplasms and cysts may arise. This group of heterogeneous lesions is known as retrorectal tumors. The retrorectal space lays above the horse shoe shaped supralevator space behind the rectum and is bounded superiorly by the peritoneal reflection in communication with the retroperitoneal space, anteriorly by the fascia propria of the rectum, laterally by the lateral ligaments, ureters and iliac vessels and inferiorly by the rectosacral or Waldeyer's fascia, which passes forward from the S4 vertebra to the rectum 3-5 cm above the anorectal junction (4).

Classification

The worldwide accepted classification of retrorectal tumors is that proposed by Uhlig and Johnson, shown in *Table 1* (4,5,6,7).

Incidence

Retrorectal tumors are rare. Cleveland Clinic reports 50 cases over a 55-year period. The Mayo Clinic estimates the incidence to be about 1 in 40,000 hospital admissions. They reported 120 cases of which 66% were congenital, 12% neurogenic, 11% osseous and 11% were miscellaneous. Metastatic and inflammatory masses were excluded from this report. Stewart and al. combined reports for a total of 301 retrorectal tumors of which 63% where congenital, 8% were inflammatory, 10% neurogenic, 7% osseous and 12% miscellaneous. Sacrococcygeal teratoma is the most common retrorectal tumors in the pediatric population and occurs in 1 in 40,000 births (4).

Table 1. *Classification of retrorectal tumors*

Congenital	**Osseous**
Developmental cysts (epidermoid, dermoid, and mucus-secreting cysts; teratoma)	Osteoma
	Osteogenic sarcoma
Chordoma	Simple bone cyst
Teratocarcinoma	Ewing's tumor
Adrenal rest tumor	Chondromyosarcoma
Anterior sacral meningocele	Aneurismal bone cyst
Duplication of rectum	Giant cell tumor
Inflammatory	**Miscellaneous**
Foreign body granuloma	Metastatic carcinoma
Perineal abscess	Liposarcoma
Internal fistula	Hemangioendothelial sarcoma
Retrorectal abscess	Lymphangioma
Chronic infectious granuloma	Extra-abdominal desmoid tumor
	Neurogenic Plasma cell myeloma
	Malignant neoplasm of unknown type
	Neurofibroma and sarcoma
	Lipoma
	Neurilemoma
	Fibroma
	Ependymoma
	Fibrosarcoma
	Ganglioneuroma
	Leiomyoma
	Neurofibrosarcoma
	Leiomyosarcoma
	Hemangioma
	Pericytoma
	Endothelioma

Epidermoid and dermoid cysts

These lesions belong in the group of developmental cysts as a part of the congenital lesions which can be found in the retrorectal space. Congenital lesions account for more than 50% of all retrorectal tumors and about two thirds of them are developmental cysts. The majority of developmental cysts is asymptomatic and may be missed on rectal examination due to low tension in the cyst. Epidermoid and dermoid cysts result from defective closure of the ectodermal tube, which results in inclusions of skin with or without accessory appendages. Both are lined with stratified squamous epithelium, well circumscribed with thin layer of connective tissue and filled with tick yellow-green fluid. The difference is that epidermoid cysts have no skin appendages, whereas dermoid cysts contain them. There is a 30% rate of infection presenting as either as retrorectal abscess or mistakenly diagnosed as perianal fistula. They can communicate with the skin characteristically presenting with a so-called "postanal dimple" (8,9).

Clinical presentation and diagnosis

The symptoms are mostly related to the size and complications. Benign lessons are usually asymptomatic and malignant lesions tend to produce symptoms. Pain mostly occurs with malignization as low back, rectal or perineal pain and if the sacral plexus is invaded it can refer to the buttocks or thighs. The pain is characteristically associated with sitting. Infections may be represented by systemic and local symptoms or in a form of recurrent perianal suppuration. Interference with pelvic outlet may lead to constipation, incontinence or dystocia (10). Disturbances to the bladder and urinary function can be due to damage to the innervation, pressure on the bladder, urethra or ureters. CNS manifestations in form of headaches and recurrent episodes of meningitis are characteristic for anterior sacral meningocele.

Examination begins with inspection of the perineal area where one should look for characteristic postanal dimple, sings of infection or signs of anal incontinence when innervation to the anal sphincters is involved. On DRE a solid mass overlain with intact mucosa should be well recognized, whereas cystic lesions may be felt as mucosal folds and may be missed if they are not infected. The exact location of the lesion, consistency, the surface and relationship to the sacrum and coccyx should be recorded. Sygmoidoscopy is the next step where the condition of the overlaying mucosa should be inspected. On

plain films the position and intactness of the sacrum and coccyx should be assessed. Bony distraction may be a sign of malignancy. Anterior sacral meningocele have a characteristic "scimitar" sacrum. With teratomas the presence of bone or even teeth has been reported. Fistulography may show communication with the rectum. Barium enema may show an extrarectal mass with anterior displacement of the rectum. CT scan is the most important diagnostic tool which shows whether the lesion is cystic or solid, and defines the surface of the tumor and the relationship to the surrounding structures such as: rectum, sacrum, coccyx, bladder, etc. Endorectal ultrasonography is a very sensitive method in assessing the rectal wall involvement and pelvic floor muscle invasion. MRI for some has become the preferred imaging modality to CT because of its superior characteristic in delineating the structures in this area. Myelography is indicated when anterior sacral meningocele is suspected (11,12).

Biopsy

The biopsy is only indicated if the lesion is inoperable because if the lesion is solid spreading of malignant cells may occur, if the lesion is cystic infection may be spread and with anterior sacral meningocele meningitis may occur. If however biopsy is performed, special consideration is made to include the site of the biopsy tract in the resected specimen. In those cases biopsy can be performed by two routes: transrectal or extrarectal-presacral, which can be CT or Ultrasonography guided (4,11).

Operation

Once the retrorectal tumor is diagnosed it should always be removed for several reasons: the lesion is or may become malignant (13); cystic lesion may become infected; the mortality rate associated with anterior sacral meningocele is 30% and the lesion may cause dystocia, which can be dangerous to the life of the mother or the fetus (10). Distal retrorectal tumors can be well managed by experienced colorectal surgeons, however more extensive lesions are best treated by a multidisciplinary team of colorectal surgeon and either orthopedist or neurosurgeon (11,12).

Posterior approach through parasacrococcigeal midline, curvilinear or horizontal incision with the patient in prone jack-knife position is indicated for low lesions or infected cysts (14,15).

Abdominal approach through transverse or midline incision with the patient in lithotomy position is indicated for high lesions (above S4 on the imaging technics or when the upper border cannot be assessed on DRE) (4, 11) and for extra spinal neurogenic neoplasms.

Combined abdominal and posterior approach or abdominosacral approach is indicated in large high lesions that could not be resected from above or when rectal involvement necessitates abdominoperineal resection.

Transrectal and intersphincteric approach can be used in selected patients (16).

Adjuvant therapy

Radiotherapy is the only modality that is feasible for palliation used chiefly with inoperable soft tissue sarcomas or with chordomas. There are no satisfactory chemotherapy regimens for retrorectal tumors (11).

Case report

We present a 35 year-old female patient who was first admitted to hospital with severe perianal and rectal pain, local signs of inflammation (tumor, dolor, calor, rubor and functio laesa) systemic signs of inflammation (fever, leucocytosis) and typical so-called postanal dimple from which there was a leakage of suppurative exudate or pus. The DRE was very painful and soft tumor can be felt in the retrorectal space with intact mucosa over it. The symptoms were present for about one week and the patient had no trouble before in her life. The leaking pus started two days before admission when she felt stool release. At first the condition was understood as a retrorectal, pre-coccigeal abscess which had spontaneously ruptured in the postanal region. The patient was started on broad spectrum antibiotics and incision with evacuation of the pus from the retrorectal space was performed through the spontaneous opening in the postanal dimple. Penrose drain was installed in the precoccigeal space which was changed daily along with daily washings with antiseptic solutions. After seven days of such treatment the local and systemic signs of inflammation were subsided as well as the suppuration and the patient was discharged. On controls the patient was complaining of occasional perianal pain. On inspection of the perianal region there were two openings: anal and epithelized postanal opening, which created a picture of a double anus (Fig. 1).

On bimanual examination using DRE with probing of the postanal opening the probe goes deep in the postanal precoccigeal and presacral region. There is constant drainage of serosanguineous fluid from the postanal opening as a sign of a fistula formation with irritation of the perianal skin and persisting tumor in the pre-coccigeal, presacral space on DRE with partial compression of the anorectum. In the meantime MRI and fistulography were performed, showing cystic tumor in the retrorectal, presacral space with minor communication with the rectum above and completely separate from the

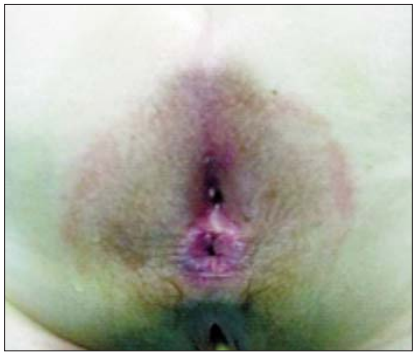

Figure 1. *Double anus*

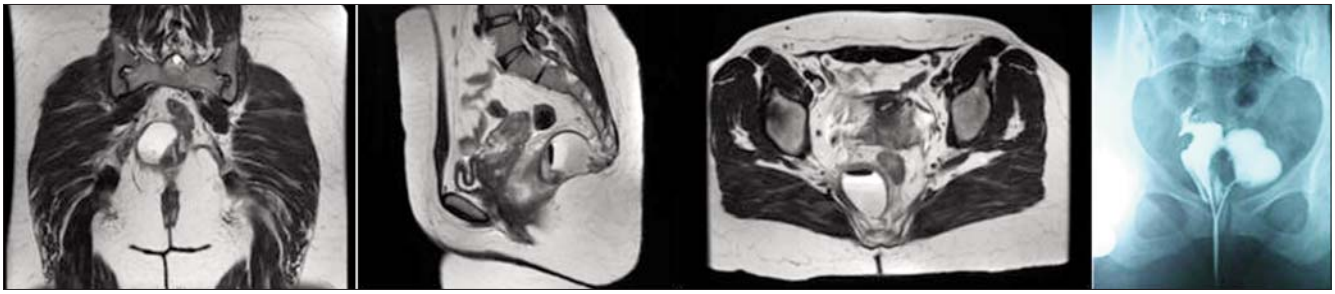

Figure 2. *MRI and fistulography*

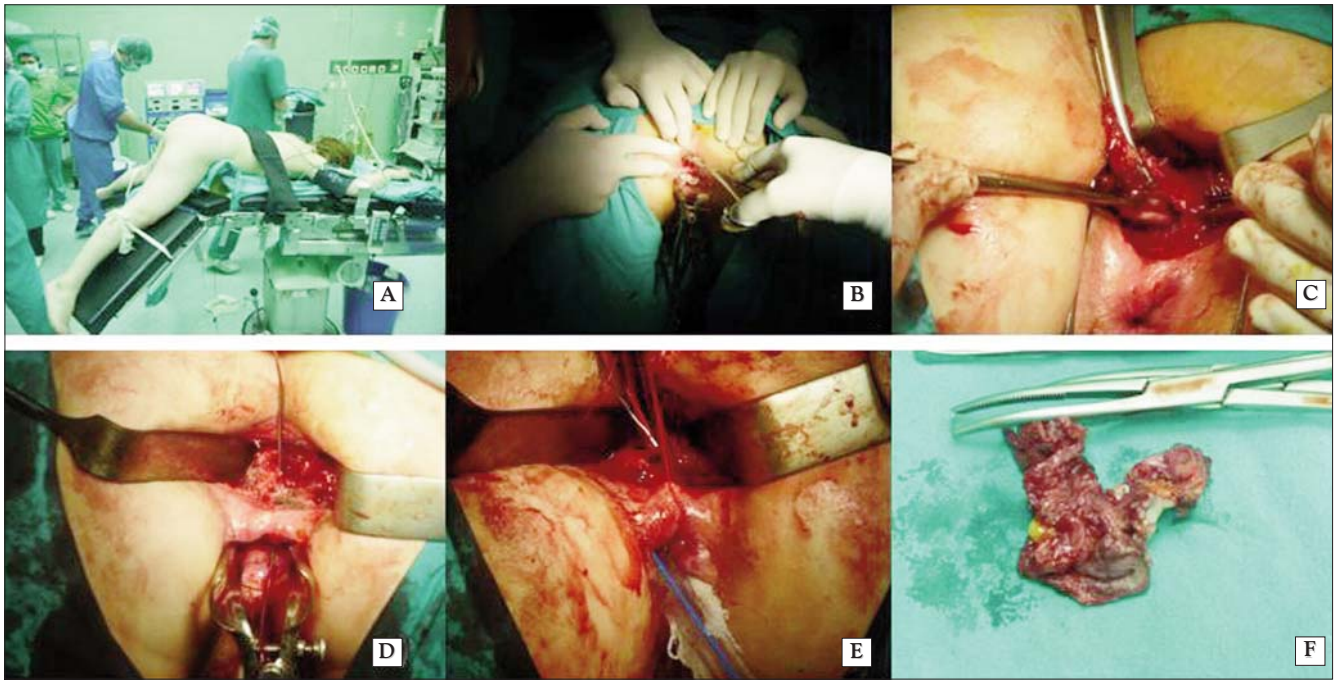

Figure 3. *Course of the operation*

sphincter mechanism in the form of an extrasphincteric fistula (Fig. 2).

About two months after the first hospitalization the patient was directed to the Clinic of Digestive Surgery at the Clinical Centre – Skopje for definitive treatment.

Preoperatively the colon was prepared conventionally and prophylactic antibiotic regimen was started. In the operating room the patient was put in prone jack-knife position (Fig. 3A) and on explorative anoscopy the internal opening of an extrasphincteric fistula in the anorectum about 3 cm above the midline posterior crypt of Morgagni was registered. The operation was started with excision of the perineal opening of the cyst (Fig. 3 B). The cyst was about 14 cm long and was liberated completely along with its capsule (Fig. 3 C, F), which enabled the visualization of the fistulous canal that led to the internal opening in the rectum (Fig. 3 D, E). The canal was excised and the rectum was sutured in two layers: first only the mucosa and then the muscle layer with separate slowly absorbable sutures (17,18).

The residual space after removal of the cyst was laid open and treated with daily washings with antiseptic solutions. The patient was discharged on the 5th postoperative day. The pathohystological finding at our Institute of Pathology is: cysta epitelialis congenita inflamata with all characteristics for dermoid cyst included in the text of the finding (17). After ten months the process of defecation is normal, the wound is sealed without signs of inflammation and secretion locally, although the postanal dimple still exists (Fig. 4).

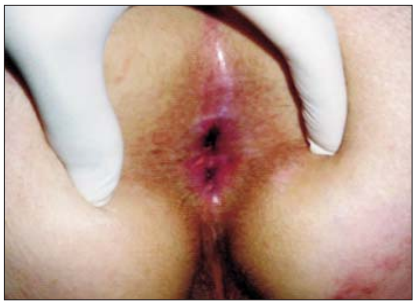

Figure 4. *Current condition*

Conclusion

Retrorectal tumors are difficult for treatment as well as for diagnosis, where even puncture biopsy is not recommended, and along with the fact that they may be complicated by some of the worst forms of perianal fistulas like in our case, they should be treated in specialized institutions by experienced surgeons from the moment of diagnosis to the definitive surgical treatment.

Reference

1. Jackman RJ, LeMon Clark III P, Smith ND. Retrorectal Tumors. JAMA. 1951;145(13):956-62.
2. Jao SW, Beart RW, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors Mayo clinic experience, 1960–1979. Diseases of the Colon & Rectum. 1985;28 (9):644-52.
3. McCarty RB. Presacral Tumors. Ann Surg. 1950;131(3):424-32.
4. Gordon PH, Nivatvongs S. Principles and Practice of Surgery for the Colon, Rectum and Anus. 3rd ed. Informa Healthcare USA, Inc.; 2007.
5. Uhlig BE, Johnson RL. Presacral tumors and cysts in adults. Dis Colon Rectum.1975;18(7):581-96.
6. Lin HY, Huang YH, Tang TN, Chiu AW. Presacral Schwannoma: A Case Report. JTUA. 2004;15(1):24-7.
7. Cerullo G, Marrelli D, Rampone B, Miracco C, Caruso S, Marianna DM, et al. Presacral ganglioneuroma: A case report and review of literature. World J Gastroenterol. 2007;13(14): 2129-31.
8. Erkan N, Agdeniz S, Polat AF, Yildirim M. Retrorectal Dermoid Cyst. Chir Gastroenterol. 2006;22:55-7.
9. Dahan H, Arrivé L, Wendum D, le Pointe HD, Djouhri H, Tubiana J. Retrorectal Developmental Cysts in Adults: Clinical and Radiologic-Histopathologic Review, Differential Diagnosis, and Treatment. Radio Graphics. 2001;21:575-84.
10. Sobrado CW, Mester M, Simonsen OS, Justo CR, deAbreu JN, Habr-Gama A. Retrorectal tumors complicating pregnancy. Report of two cases. Dis Colon Rectum. 1996;39(10):1176-9.
11. Glasgow SC, Dietz DW. Retrorectal Tumors. Clin Colon Rectal Surg. 2006;19(2):61-8.
12. Hassan I, Wietfeldt ED. Presacral tumors: diagnosis and management. Clin Colon Rectal Surg. 2009;22(2):84-93.
13. Krivokapic Z, Dimitrijevic I, Barisic G,Markovic V, Krstic M. Adenosquamous carcinoma arising within a retrorectal tailgut cyst: report of a case. World J Gastroenterol. 2005;11(39):6225-7.
14. Aranda-Narváez JM, González-Sánchez AJ, Montiel-Casado C, Sánchez-Pérez B, Jiménez-Mazure C, Valle-Carbajo M, et al. Posterior approach (Kraske procedure) for surgicaltreatment of presacral tumors. World J Gastrointest Surg. 2012;4(5):126-30.
15. Abel ME, Nelson R, Prasad ML, Pearl RK, Orsay CP, Abcarian H. Parasacrococcygeal approach for the resection of retrorectal developmental cysts. Dis Colon Rectum. 1985;28(11):855-8.
16. Serra Aracil X, Gómez Díaz C, Bombardó Junca J, Mora López L, Alcántara Moral M, Ayguavives Garnica I, et al. Surgical excision of retrorectal tumour using transanalendoscopic microsurgery. Colorectal Dis. 2010;12(6):594-5.
17. Whiteford MH, Kilkenny III J, Hyman N, Buie WD, Cohen J, Orsay C, et al. Practice parameters for the treatment of perianal abscess and fistula-in-ano (revised). Dis Colon Rectum. 2005; 48(7):1337-42.
18. Rubtsov ML, Lurin IA, Shudrak AA, Neshaĭ VS, Ihnatchenko OV. Method of the surgical treatment of nonspeciphicextrasphincteric rectal fistula. Klin Khir. 2005;(1):19-20. Ukrainian