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*Case report*

# NEGLECTED CONDITION: NODULAR FASCIITIS AND OUR CASE

# ЗАНЕМАРЕНА СОСТОЈБА: НОДУЛАРЕН ФАСЦИИТ И ПРИКАЗ НА НАШ СЛУЧАЈ

Elizabeta Mirchevska Zhogovska[1], Slavica Kostadinova Kunovska[2], Tomislav Jovanoski[1], Igor Peev[1], Boro Dzonov[1], Lazo Noveski[1], Margarita Peneva[1], Magdalena Bogdanovska Todorovska[2] and Lena Kakasheva-Mazhenkovska[3]

[1]University Clinic for Plastic and Reconstructive Surgery [2]Institute of Pathology, [3]Institute of Histology and Embriology, Faculty of Medicine, Ss. Cyril and Methodius University, Skopje, Republic of North Macedonia

**Abstract**

In 1955, nodular fasciitis was identified for the first time. Other names are infiltrative fasciitis, pseudosarcomatous fasciitis, and pseudosarcomatous fibromatosis. A quickly expanding lesion is the most typical sign of nodular fasciitis, and roughly half of the cases are accompanied by discomfort or pain. Depending on where the lesion is located, there are three basic types of nodular fasciitis: subcutaneous, intramuscular or fascial with intradermal and intravascular as unusual subtypes. We present an unusual case of a 52-year-old male with a 10+ year history of a tumor in the right gluteal region associated with pain during sitting in the last few months. The tumor may be grossly fibrous, myxoid, or even cystic and histopathologically, the tumor can be hypercellular and may imitate a sarcoma. Ultrasound can be helpful, but MRI is more accurate. However, the various histologic features make this tumor diagnostically nonspecific even on MRI with several differential diagnoses including fibrosarcoma, neurofibroma, and small fibrous histiocytoma. Fine-needle aspiration and histologic features may correlate well, but biopsy is typically necessary for a conclusive diagnosis.

**Keywords: fasciitis, pseudosarcomatous fasciitis, surgical treatment**

**Апстракт**

Во 1955 година, за прв пат беше идентификувана состојбата нодуларен фасцитис. Други имиња за оваа состојба се инфилтративен фасциитис, псевдосаркоматозна фасциитис и псевдосаркоматозна фиброматоза. Лезија која брзо се шири е најтипичен знак за нодуларен фасциитис, а приближно половина од случаевите се придружени со непријатност или болка. Во зависност од тоа каде се наоѓа лезијата, постојат три основни типови на нодуларен фасциитис: поткожен, интрамускулен или фасцијален со интрадермален и интраваскуларен како невообичаени подтипови. Прикажуваме необичен случај на 52-годишен маж со 10+ годишна историја на тумор во десната глутеална регија поврзана со болка при седење во последните неколку месеци. Туморот може да биде фиброзен, миксоид па дури и цистичен а хистопатолошки може да биде хиперцелуларен при што може да имитира сарком. Ултразвукот може да биде корисен, но магнетна резонанца е попрецизен. Сепак различните хистолошки карактеристики го прават овој тумор дијагностички неспецифичен дури и на магнетна резонанца со неколку диференцијални дијагнози вклучувајќи ги и фибросарком, неурофибром и малиген фиброзен хистиоцитом. Тенкоиглената аспирациона биопсија и хистолошките карактеристики може добро да корелираат, но ексцизионата биопсијата обично е неопходна за конечна дијагноза.

**Клучни зборови: нодуларен фасциит, псеудосаркоматозен тумор, хируршки третман**

## Introduction

In 1955, nodular fasciitis was identified for the first time. Other names are infiltrative fasciitis, pseudosarcomatous fasciitis, and pseudosarcomatous fibromatosis [1]. It comprises benign, self-contained fibroblast growth with an unknown cause, not related to age, race or gender. Most of the patients are under the age of 50 (85%), while only 5% are over the age of 70 [2], located mostly at the upper extremities (48%) and torso (20%). Lower extremity (15%), neck and face (17%)

*Correspondence to:* Elizabeta Mirchevska Zhogovska, University Clinic for Plastic and Reconstructive Surgery, 1000 Skopje, R. N. Macedonia; E-mail: elizabetamircevska@yahoo.com

are additional sites [3,4], while retroperitoneum [5], hand and foot [6] are examples of uncommon locations. It typically occurs as solitary lesion. Lesions can range in size between 5 to 100 mm, but 71% are 20 mm or smaller, with the majority being under 40 mm [2]. A quickly expanding lesion is the most typical sign of nodular fasciitis, and roughly half of the cases are accompanied by discomfort or pain [1]. Less common, numbness, paresthesia, and shooting pain indicate peripheral nerve compression [7].
Depending on where the lesion is located, there are three basic types of nodular fasciitis: subcutaneous, intramuscular or fascial with intradermal and intravascular as unusual subtypes [8].

## Case report

We present an unusual case of a 52-year-old male with a 10+ year history of a tumor in the right gluteal region associated with pain during sitting in the last few months. Based on clinical examination and history, the tumor was diagnosed as sebaceous cyst. On physical examination, a 50 x 60 mm firm, subcutaneous, soft tissue tumor with pink to brownish discoloration of the perilesional skin was noted (Figure 1).

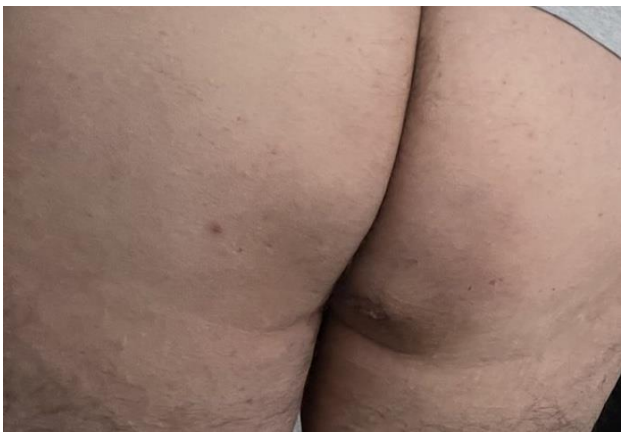

**Fig. 1.** Tumor in the right gluteal region

Lesion was attached to the underlying tissue that was seen during the examination. Due to its superficial localization, MRI, ultrasound or aspiration biopsy were not performed and the patient was admitted for one-day surgery. Longitudinal excision was done and the tumor was completely removed; the wound was closed and the sample was sent for histopathological analysis, as it was seen that macroscopically it did not look like a sebaceous cyst.
Histopathological examination revealed well-circumscribed tumor node in the subcutaneous tissue, composed of spindle cells with storiform pattern, minor nuclear pleomorphism, and foci of myxoid change and collagen deposition in the extracellular matrix (Figure 2).

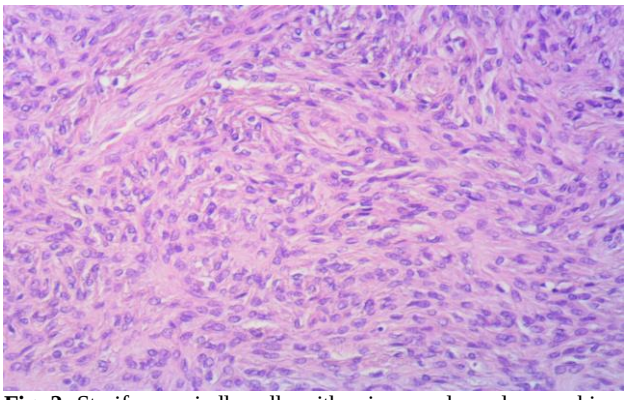

**Fig. 2.** Storiform spindle cells with minor nuclear pleomorphism and collagenous extracellular matrix (HeEo, x100)

Immunohistochemical analyses showed a focal positivity for smooth muscle actin (Figure 3), and negative staining for CD34 (Figure 4), S-100, Desmin and Caldesmon. The proliferative index on the staining with Ki67 was low, mainly <5%, with foci of up to 15% (Figure 5).

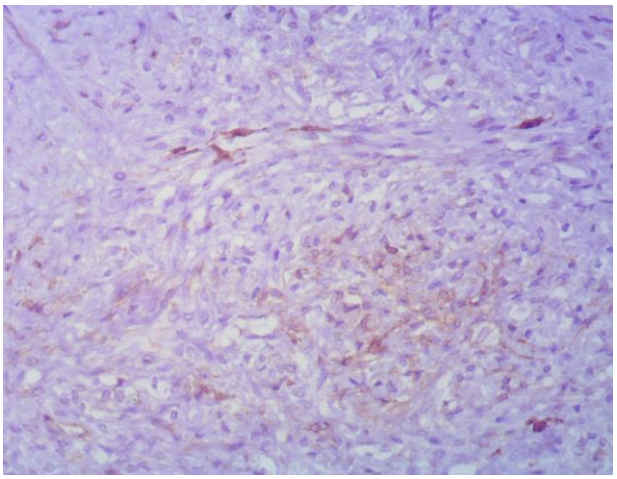

**Fig. 3.** Focal positivity for SMA, x100

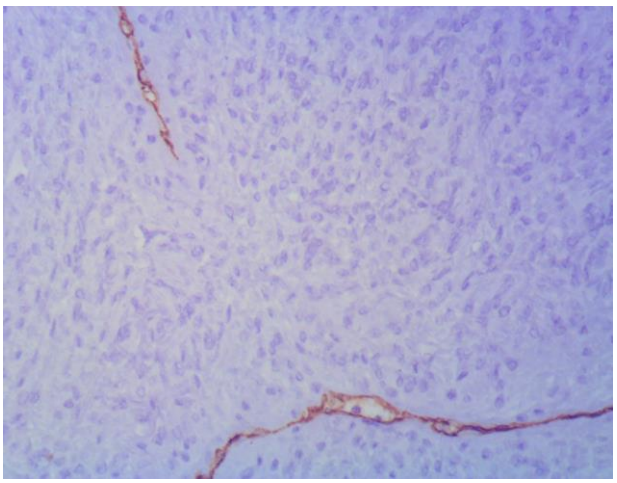

**Fig. 4.** CD34 negative tumor cells, x100

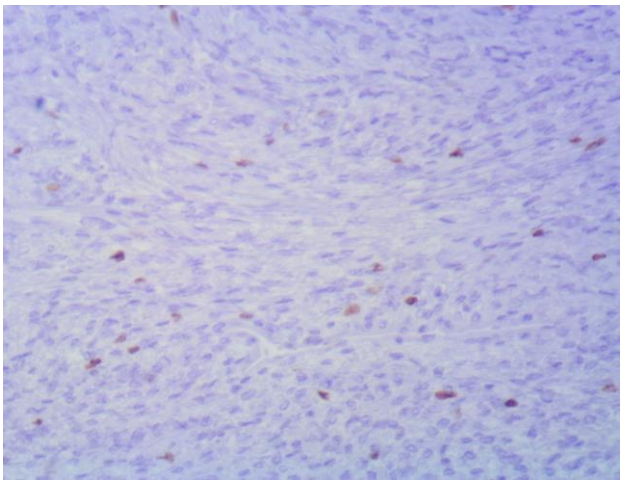

**Fig. 5.** Low proliferative index (Ki67, x100)

## Discussion

The tumor may be grossly fibrous, myxoid, or even cystic. According to some researchers, the type and quantity of extracellular matrix correlate with the maturity of the lesion: an early lesion has a higher proportion of myxoid tissue, whereas an older lesion has a higher proportion of fibrotic tissue. Histopathologically, the tumor can be hypercellular and may imitate a sarcoma since it is made up of plump fibroblasts that are organized in short bundles and fascicles across a myxoid stroma. In other situations, there is greater fibrosis and reduced cellularity. In nodular fasciitis, there is no intralesional hemosiderin deposition [9]. Ultrasound can be helpful, but MRI is more accurate. There are a few diagnostic imaging publications detailing the MRI features of nodular fasciitis, despite the opinion that the disease diverse histologic features make its appearance on MRI nonspecific. According to Meyer *et al.* [10], who presented three cases, nodular fasciitis is typically well-defined. On T1-weighted imaging, their two mucoid and cellular intramuscular instances seemed slightly inhomogeneous and hyperintense to muscle tissue; on T2-weighted images, they appeared generally homogeneous with signal intensity greater than the fatty tissue. On all pulse sequences, the subcutaneous lesion, which had a fibrous histology, was hypointense. There was no perilesional tissue swelling. Meyer *et al.* concluded that the lesion's MRI appearance reflected its overall shape. The nodular fasciitis contrast-enhancing pattern was not described. Although rim enhancement was found [13-15], later reported cases demonstrated that the contrast-enhancing appearance of nodular fasciitis is typically homogenous [9,11,12].
There are several radiologic differential diagnoses for nodular fasciitis because its diagnostic signs are nonspecific. These include fibrosarcoma, sarcoidosis, dermatofibroma, neurofibroma, aggressive fibromatosis, neuroma and malignant fibrous histiocytoma. Fine-needle aspiration and histologic features may correlate well [16], but biopsy is typically necessary for a conclusive diagnosis.
Excision is the basis of therapy, while some researchers have also recommended monitoring [17] and corticosteroid injections in the tumor [18]. Relapse of the lesion is extremely uncommon, occurring in 1-2% of patients, and is frequently discovered shortly after excision [1,19,20].
There are a few clinical and radiologic characteristics that make the diagnosis of nodular fasciitis less likely: lesions in patients over 70 years old, tumors in the hand or foot, more than one lesion or tissue edema around the tumor; deposition of hemosiderin in the lesion seen on MRI and lesions that reoccur. The main concern is the similarity in clinical presentation and microscopic appearance between nodular fasciitis and sarcoma [12,21,22].

## Conclusion

Nodular fasciitis needs to be recognized since, due to its rapid growth, rich cellularity, strong mitotic activity, and loosely confined form, it is sometimes mistaken as a sarcoma. Further research is required to establish the disease benign nature because large lesions are sometimes misinterpreted for malignant lesions. To overcome the difficulties of diagnosing nodular fasciitis, magnetic resonance imaging is required in addition to histology and immunohistochemistry. In any case, careful clinical follow-up is essential.

*Conflict of interest statement.* None declared.

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