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Myxoinflammatory Fibroblastic Sarcoma Presenting as a Painful Mass of the Left Great Toe: A Case Report

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Abstract **Case Report**

Myxoinflammatory fibroblastic sarcoma is a rare soft tissue tumor that occurs predominantly in the distal extremities. Its nonspecific clinical presentation and morphological heterogeneity can make diagnosis challenging. We report the case of a 50-year-old woman who presented with a painful, 3.5-cm mass of the left great toe that had been present for four months. Imaging initially suggested a tenosynovial giant cell tumor, with no evidence of bone destruction on radiography. The mass was excised with preservation of the great toe. Histological examination revealed a multinodular proliferation with myxoid and fibrous stroma, spindle and pleomorphic cells with prominent nucleoli, and a polymorphous inflammatory infiltrate. These findings favored a diagnosis of myxoinflammatory fibroblastic sarcoma. The surgical margins were negative, and no local recurrence was observed after 12 months of follow-up. This case highlights the importance of careful pathological examination of persistent acral masses and continued surveillance after excision because of the risk of local recurrence.

Keywords: Myxoinflammatory fibroblastic sarcoma; great toe; soft tissue tumor; histopathology.

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INTRODUCTION

Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare soft tissue tumor described in the late 1990s. It occurs predominantly in adults and most often affects the distal extremities, particularly the hands and feet [1,2]. MIFS typically presents as a slowly growing mass that may clinically resemble a benign lesion. Diagnosis relies primarily on histological examination, which shows a variable combination of myxoid and fibrous areas, atypical tumor cells, and an inflammatory infiltrate. The lack of a specific immunohistochemical profile and the diversity of reported molecular abnormalities can further complicate the diagnostic process [3,4]. We report the case of a woman with a painful mass of the left great toe whose histopathological features favored MIFS.

CASE REPORT

A 50-year-old woman presented with a painful nodular mass of the left great toe that had been present for four months. The lesion measured 3.5 cm in greatest dimension, was tender on palpation, and appeared fixed to the overlying skin and underlying tissues. Anteroposterior and lateral radiographs of the foot showed no evidence of bone destruction. Ultrasonography revealed an infiltrative soft tissue mass. Based on the imaging findings, a tenosynovial giant cell tumor was initially suspected. The mass was surgically excised, preserving the great toe. On gross examination, the specimen consisted of a partially circumscribed, lobulated nodular mass with gelatinous and myxoid areas and hemorrhagic changes. The lobules were separated by fibrous-appearing bands (Figure 1). Histological examination revealed a multinodular mesenchymal proliferation with myxoid and fibroblastic components (Figure 2). It contained spindle cells and large pleomorphic cells with atypical nuclei and prominent nucleoli, some resembling Reed–Sternberg cells. Multinucleated cells were also observed (Figure 3). The background contained a polymorphous inflammatory infiltrate, including neutrophils, eosinophils, and histiocytes. The proliferation infiltrated the adjacent adipose tissue (Figure 4). Based on the location and the overall morphological findings, a diagnosis of MIFS was favored. The surgical margins were reported as negative. No local recurrence had been reported at the 12-month follow-up.

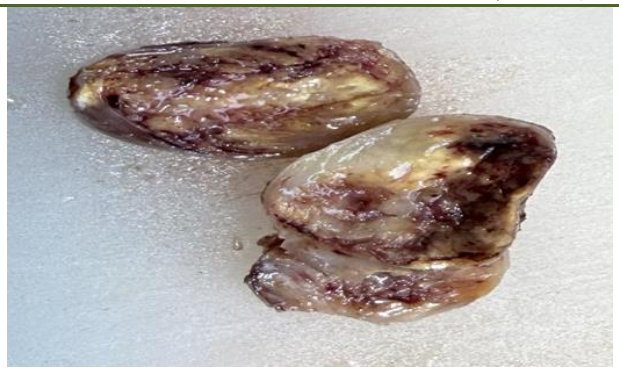


Figure 1: A multinodular, grayish-white lesion, exhibiting gelatinous areas that correspond to fibrous and myxoid zones, which are the site of hemorrhagic changes

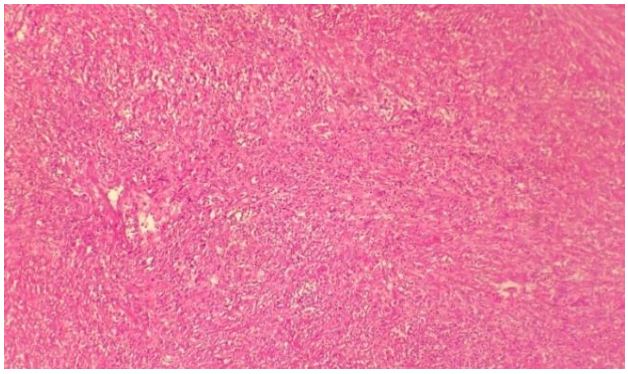


Figure 2: Histological image showing a multinodular lesion composed of variable myxoid and fibrous/hyalinized areas with an associated dense inflammatory infiltrate : Hematoxylin and eosin staining (H&E) Gx10

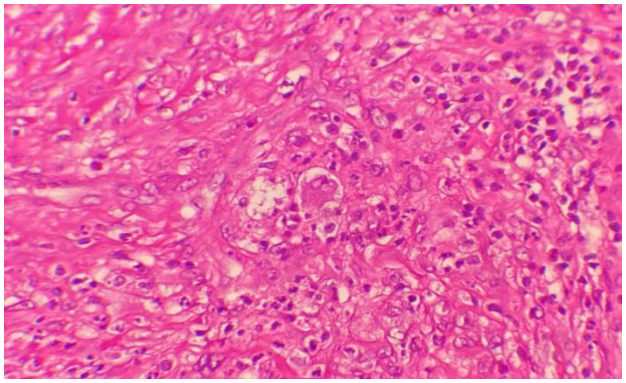


Figure 3: Histological image showing tumor cell population is composed of spindle-shaped epithelioid cells with scattered large cells with odd nuclei and prominent nucleoli resembling viral inclusions (virocyte-like cells) or cells Reed-Sternberg: Hematoxylin and eosin staining (H&E) Gx 40

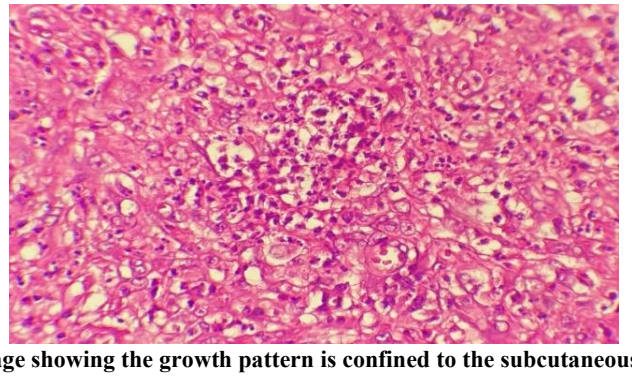


Figure 4: Histological image showing the growth pattern is confined to the subcutaneous tissue with polymorphic inflammatory cells including neutrophils, lymphocytes, plasma cells, histiocytes and eosinophils: Hematoxylin and eosin staining (H&E) Gx40

DISCUSSION

MIFS preferentially affects the soft tissues of the distal extremities. In the series of 104 cases reported by Laskin *et al.,* it most often presented as a solitary, painless mass, with a median size of 2.4 cm [2]. The location in the great toe and the size of 3.5 cm in our patient are consistent with the reported spectrum of this tumor. Pain is a less common presenting symptom, although it has been described. In the series of 73 cases reported by Suster *et al.,* three patients presented with pain [4]. Chahdi *et al.,* also described MIFS involving a finger in an adult, illustrating its acral presentation [5]. These clinical features remain nonspecific and do not reliably distinguish MIFS from other soft tissue lesions. Imaging helps define the location and extent of the lesion, but its findings alone are insufficient to establish the diagnosis. In a retrospective study of 29 patients, Kumar *et al.,* described imaging features that could resemble those of either benign or malignant lesions [6]. Tateishi *et al.,* reported poorly circumscribed masses that could involve tenosynovial structures on magnetic resonance imaging [7]. In our patient, radiography showed no bone destruction, and ultrasonography revealed an infiltrative soft tissue mass. The initial imaging diagnosis was tenosynovial giant cell tumor. This finding highlights the importance of correlating imaging with histological examination. The histological appearance of MIFS is heterogeneous. It combines variable proportions of myxoid and fibrous areas, spindle or epithelioid cells, and an inflammatory infiltrate. Some tumor cells are large, with atypical nuclei and prominent nucleoli resembling those of Reed–Sternberg cells or virocytes. Multivacuolated cells, described as pseudolipoblasts, may also be present [2–4]. The five-case series reported by Sakaki *et al.,* illustrates the morphological diversity of this tumor and the diagnostic challenges it may pose [8]. Representative sampling is therefore useful because characteristic features may vary across different areas of the lesion. In our case, the multinodular architecture, myxoid and fibrous stroma, polymorphous inflammatory infiltrate, and pleomorphic cells with prominent nucleoli supported the diagnosis of MIFS. Infiltration of the adjacent adipose tissue was also observed. The differential diagnosis depends on the predominant component in the tissue examined. A lesion with a prominent inflammatory infiltrate may suggest an inflammatory or reactive process, whereas abundant myxoid matrix may raise consideration of other myxoid mesenchymal tumors, including myxofibrosarcoma [3,9]. In the present case, imaging had suggested a tenosynovial giant cell tumor. However, the morphological features of the excised lesion did not favor that diagnosis. Distinction requires assessment of the overall architecture, tumor stroma, and cell populations rather than reliance on a single feature. Cytological examination may also help guide the diagnostic process in selected cases. Wickham *et al.,* reported a case in which fine-needle aspiration suggested the diagnosis, which was subsequently established on the excision specimen [10]. This observation illustrates the potential value of initial sampling while emphasizing the importance of histological examination for comprehensive assessment. Immunohistochemistry can help investigate differential diagnoses, but no single marker confirms MIFS. In their study of 73 cases, Suster *et al.,* observed variable expression of several markers, supporting interpretation in conjunction with the morphology and clinical context [4]. Molecular abnormalities, including rearrangements involving *TGFBR3* and *OGA* and alterations involving *BRAF*, have also been studied in this tumor [4,11]. Their presence or absence should be interpreted cautiously and does not replace pathological examination. No immunohistochemical or molecular results were available in the data provided for our case. Complete excision is an important part of management. In the series by Laskin *et al.,* 30 of the 59 patients with available follow-up experienced at least one local recurrence. Complete initial excision was associated with a lower recurrence rate [2]. This finding comes from a selected series and cannot precisely predict the risk for an individual patient. Most conventional MIFS have a low metastatic potential, although aggressive forms have been described. Michal *et al.,* reported a series of 23 tumors selected for high-grade features [12]. The outcomes of that series should not be extrapolated to all MIFS, and the data available in our case do not establish a high-grade tumor. In our patient, the surgical margins were reported as negative, and no local recurrence had been observed after 12 months. This follow-up remains limited. Continued surveillance is warranted given the risk of local recurrence reported in the literature [2,3].

CONCLUSION

This case illustrates the diagnostic challenge posed by a painful mass of the great toe. The combination of myxoid, fibroblastic, and inflammatory features with atypical cells favored a diagnosis of MIFS. Despite negative reported margins and no recurrence at one year, continued follow-up is warranted.

DECLARATIONS

Ethics Approval and Consent to Participate: This work has respected all the rools of medical ethics and has been elaborated by all the authors.

Availability of Material and Data: All data is available in the Moulay Ismail Military Hospital, Meknès Morocco

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