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Case Report

Acral Myxoinflammatory Fibroblastic Sarcoma of the Foot: A Case Report

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Abstract

Myxoinflammatory fibroblastic sarcoma is a rare low grade malignant soft tissue tumor, mainly affecting middle-aged persons, and located in the distal extremities. The absence of clinical and radiological signs specific to this tumor makes it confused with many benign lesions and leads to inappropriate treatment. Though it is classified as a low grade tumor, it is reported to have an important infiltrative character responsible for a considerable rate of local recurrence. The therapeutic management of this tumor remains a subject of debate, there are no formal standard treatment protocols. In this presentation, we aimed to report the case of a patient with myxoinflammatory fibroblastic sarcoma of the foot treated successfully by surgery with adjuvant radiotherapy.

Keywords: *Acral Myxoinflammatory Fibroblastic Sarcoma (AMIFS); Radiotherapy; Foot*

Introduction

Acral myxoinflammatory fibroblastic sarcoma (AMIFS) is a rare, low-grade sarcoma occurring mostly in the subcutaneous tissues of distal extremities of young adults [1]. The therapeutic management of these tumors remains a subject of debate, there are no formal standard treatment protocols. The difficulty lies in obtaining a complete excision, because of its tendency to grow in infiltrative patterns, which may partly account for their tendency to repeated local recurrence. We report the case of an adult in his forties who present myxoinflammatory fibroblastic sarcoma of the foot through which we illustrate the difficulties of the therapeutic management of this tumor.

Patient and Observation

A 42-year-old woman with medical history of hypertension and depression presented in our outpatient department of Orthopedics with a main complaint of a slow-growing mass in the right dorsal foot evolving for a year. There was no history of trauma. she did not present a fever nor a deterioration of the general state. Physical examination revealed a swelling of soft consistency of 3 cm long axis that is movable with respect to the deep plane adhering to the surface plane, sitting on the dorsal surface of the first ray of the right foot. The mass was painless without local inflammatory signs or satellite lymphadenopathy.

Magnetic resonance imaging (MRI) revealed a homogenous mass in the subcutaneous tissue in the dorsal right foot. The mass intimately abuts the superficial fascia of the anterior tibial tendon without extension deep to the fascia. There was a nodular mass showing a low signal in T1 (Figure 1a), a high signal in T2 (Figure 1c) and gadolinium enhanced T1 homogenous enhancement (Figure 1b). There was no bone lysis.

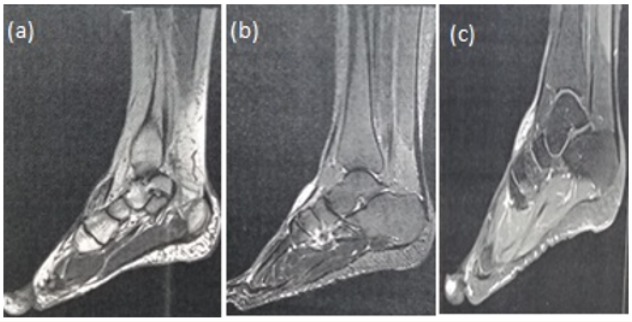

Figure 1: MRI showing homogenous soft tissue mass. T1 weighted image (a) shows subcutaneous nodular lesion in low signal, where it is enhanced by gadolinium (b). T2 weighted image (c) shows a high signal lesion.

Clinically and radiologically, the patient was thought to have a benign condition at the time of the initial evaluation. A marginal excisional biopsy was done.

Anatomopathology

The resection piece has a nodular outer surface (Figure 2). It was inked and included in full.

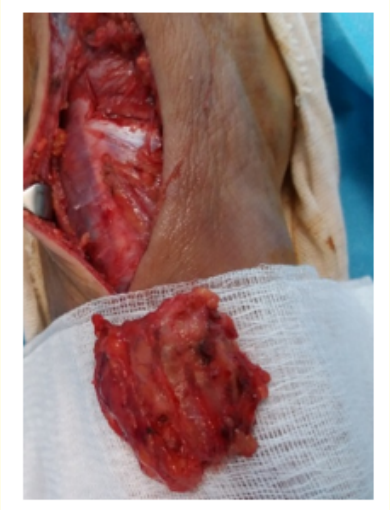

Figure 2: Piece of resection.

On histologic findings, the architecture was vaguely multinodular with the alternation of cellular areas with foci of hyalinized collagen fibers and myxoid areas. The tumor has infiltrative margins in subcutis (Figure 3). It was containing numerous inflammatory cells and scattered by, large, bizarre tumor cells with vesicular nuclei, prominent inclusion-like nucleoli, and abundant eosinophilic cytoplasm, which was homogeneous to vacuolated resembling Reed Sternberg-like cells (Figure 4).

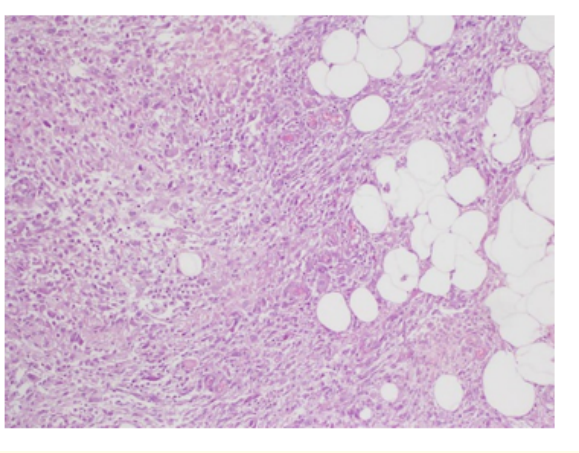

Figure 3: Tumors cells infiltrating subcutaneous tissue.

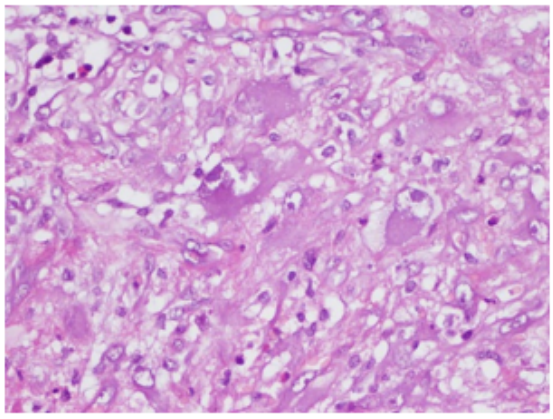

Figure 4: Mixed inflammatory cell infiltrate comprising Reed Sternberg-like cells.

Tumor cells were strongly immunoreactive for vimentin and variably positive for CD68 and CD34.

On the basis of histopathology and immunohistochemistry, a final diagnosis of AMIFS was made. The margins of resection were tumorous in places, which required a re-excision.

The complement of resection included the inferior retinaculum extensor as well as the sheath of the anterior tibialis and the long extensor of the hallux, which resulted in a significant loss of substance requiring a rail plastic surgery (Figure 5).

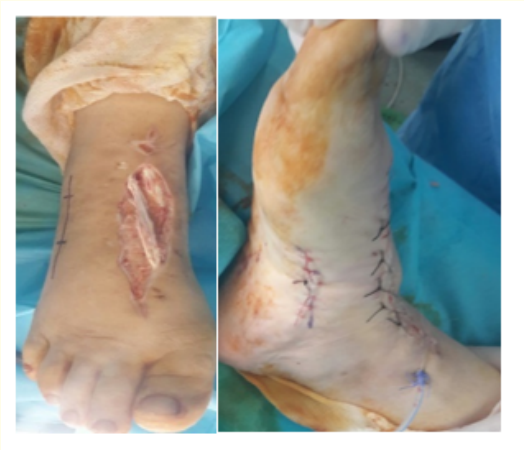

Figure 5: MManagement of loss of skin substance. Loss of skin substance after re-excision (a), a rail plastic surgery (b).

The Patient had neoadjuvant radiation therapy.

At 12 months follow-up the evolution was favorable with no evidence of local recurrence at physical examination and at magnetic resonance imaging. A CT scans of the chest, abdomen, and pelvis revealed no evidence for metastatic progression of the tumor.

Discussion

AMIFS is an extremely rare form of sarcoma first described in 1998 by Meis-Kindblom and Kindblom [1], and further characterized that same year [2]. The lesion occurs primarily in middle-aged adults and near equally in both genders.

As observed in our case and other published cases, this tumor generally presents as an ill-defined, slow growing, painless mass that has appeared over the course of several months.

AMIFS mainly occur in the subcutaneous tissues of the distal extremities and has a predilection for the dorsal aspect of the hands and feet [1,2].

Grossly, the tumor has a median size around 3 cm and mucoid character and is usually poorly circumscribed [3,4].

On the microscopic plane the lesion contain three major characteristics [5]:

1. Multinodular architecture, alternating densely cellular and myxoid hypocellular areas.
2. Mixed inflammatory infiltrate.
3. Bizarre giant and lipoblast-like cells.

Although the immunohistochemical findings are non-specific, immunohistochemistry is used for accurate diagnosis of the tumor [6]. In the present case all the features were noted.

The pathogenesis of MIFS is unknown. The two main hypotheses advanced in this area are the inflammatory hypothesis [1] and the genetic hypothesis [7,8].

Given the histological features of AMIFS (i.e. myxoid, inflammatory and atypical features), the differential diagnosis for this lesion is broad. It includes most of the benign and malignant myxoid lesions and can range from an infectious inflammatory process to malignant myxofibrosarcoma.

The absence of pathognomonic clinical signs, as well as the lack of signs of imaging specific to AMIFS, make this tumor clinically managed as a benign lesion and leads to inappropriate treatment. Silver., *et al*. [9] reported the case of a patient originally presenting with a diagnosis of chronic abscess, treated initially with antibiotics and steroids. In another report Lang., *et al*. [10] described patients initially diagnosed with various benign conditions including: ganglion cyst, giant cell tumour of the tendon sheath and benign tumor of the hand.

An institutional case series of five patients along with review of the literature in 2013 identified 138 cases following a MEDLINE literature review of cases from 1997 to 2012 reported that 74% of cases underwent surgery, with clinical diagnosis of benign disease [11]. These mimic the scenario in the present report.

Wide surgery excision is the gold standard in the treatment of these tumors. However, the difficulty in obtaining local control is an unanimous problem. In fact seen its infiltrative character and its proximity to certain elements such as tendons, ligaments and vas-

culo-nervous elements makes obtaining a wide excision limited in certain directions. Therefore, when wide excision fails to preserve a function extremity, amputation may be an option.

The risk of local recurrence described in the early published reports range from 22% to 67% [1,2]. Baldini., *et al*. [12] had concluded that the histologic resection margin status less than 1 cm was the only statistically significant predictor for local recurrence for patients with soft-tissue sarcoma treated with surgery alone.

Radiotherapy may have a role in supplementing local control, particularly those with positive surgical margins. At the Center for Sarcoma and Connective Tissue Oncology at Massachusetts General Hospital (Boston, USA), Tejwani., *et al*. [13] reported excellent local control with preoperative radiotherapy in 14 patients with only one recurrence. Knowing that these data were from a specialized sarcoma center and the diagnosis of AMIFS was known preoperatively.

In his study for patients with soft tissues sarcoma Baldini., *et al*. [12] deduced certain conditions where treatment with wide excision alone is recommended, they include patients with (1) primary presentation of disease, (2) histologic resection margins of more than 1 or 2 cm in all directions, (3) sites of disease such that a local recurrence would not preclude function-sparing salvage surgery and RT and (4) the ability and willingness to comply with close follow-up.

This tumor, long regarded as a low grade lesion, recent literature review has shown its invasive and locally disruptive character associated with a high rate of local recurrence as well as increasingly reported cases of metastasis [11,14]. Which prompts the revision of the low-grade nature of this tumor.

Conclusion

AMIFS is an extremely rare sarcoma and may easily be confused with many benign lesions. However we should keep in mind slowly growing malignancies when the lesion is located close to the synovial regions of the extremities. AMIFS should be included among differential diagnosis in order to avoid misdiagnosis and ensure adequate management.

Competing Interests

The authors declare no competing interest.

Authors' Contributions

All the authors have contributed to this manuscript in ways that comply to ICMJE authorship criteria. All the authors have read and approved the final version of the manuscript.

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