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MYXOINFLAMMATORY FIBROBLASTIC SARCOMA

Pathology

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ABSTRACT

Myxoinflammatory fibroblastic sarcoma is a rare soft tissue tumor with most occurring in the distal extremities of adult patients with equal sex predilection. It has a high rate of local recurrence and a low rate of metastasis, it is often inadequately treated and misdiagnosed as benign tumour. In this review, the intent is to highlight salient clinicopathologic features, detail the broad microscopic spectrum including high-grade aggres sive variants and review the molecular features.

KEYWORDS

INTRODUCTION

MIFS is a rare soft tissue tumor first reported in 1998 by 3 separate investigators. Montgomery et al1 described 51 cases. They observed neoplastic cells with large, inclusion-like nucleoli within a myxoid and fibroinflammatory milieu, frequent involvement of the hands and feet, and potential for local recurrence. Because it is now known to occur in both acral and nonacral body sites, the World Health Organization has renamed it as MIFS. The genetic abnormalities detected in MIFS are the t(1;10)(p22:q24) translocation, with rearrangements of TGFBR3 and MGEA5 genes associated with increased levels of FGF8, with chromosome 3 marker/ring formation, and amplification of the VGLL3 locus.

Case Presentation

Our case concerns a 60-year-old male presented in Orthopedics Department at GMC Amritsar with swelling in the right leg for 3 years, which was painless, slowly growing, with no discharge, no other constitutional symptoms and no other co morbidities. No relevant past or family history.

The physical examination finds a subcutaneous mass, measuring 6 X 4.5 cm soft non tender mass in the anteriomedial and distal end of right leg.

On MRI, a well defined mass with cystic areas fixed in relation to the deep plane. A surgical biopsy was done by the treating physician and sent to our department.

Gross examination we received multiple grey brown soft tissue pieces, measuring 5 X3X 1.5 cm.

On histopathology examination, a multinodular lesion composed of myxoid and fibrous heterogeneous areas with intense polymorphic inflammatory infiltrate consisting of neutrophils, lymphocytes, plasma cells, eosinophils and few histiocytes were seen. The tumour was composed of epithelioid or fusiform cells with large dispersed cells, with bizarre nuclei with nucleoli, resembling viral inclusions or Reed-Sternberg cells with tumour giant cells.

DISCUSSION

Myxoinflammatory fibroblastic sarcoma presents as a painless, slowly growing mass. It typically affects adult patients with an average age around 40 years. However, the age range includes both young children and old aged. The sex ratio is 1:1. It has a distinct propensity for acral and dorsal soft tissue locations. The dorsal foot/ anterior ankle region is a characteristic site. However, MIFS can occur at various body sites including proximal extrem ities, trunk, and head and neck. Clinically, MIFS may resemble a benign lesion such as synovitis or tenosynovial giant cell tumor. Grossly, MIFS is lobulated and varies from gelatinous to fleshy to firm, often being heterogeneous in color and texture. It usually occurs in subcutaneous adipose tissue where it infiltrates along fibrous septa and fascial planes. The diagnostic challenges include, its broad histologic spectrum for example, the proportions of myxoid lobules , fibroinflammatory areas, inclusion-like nuclei and presence of a heavy inflammatory infiltrate that obscures the neoplastic cells. The spectrum of neoplastic cells found in MIFS varies within and among tumors. The most consistent histologic feature include a subpopulation of cells displaying marked nuclear pleomorphism with enlarged spindle cells with fibrillary cytoplasm, large epithelioid cells with abundant eosinophilic cytoplasm degenerated cells with smudged chromatin, neoplastic cells filled with mucoid vacuoles, so called pseudolipoblasts found within myxoid areas and large histiocytoid cells with emperipolesis, usually containing intracytoplasmic neutrophils. Within myxoid lobules, the neoplastic spindle and epithelioid cells often interconnect with one another to form strands, complex networks and discohe sive sheets forming a so-called dilapidated brick wall pattern. The inflammatory infiltrate usually consists of lymphocytes and lymphoid aggregates. However, substantial numbers of neutrophils, eosinophils, macrophages, and plasma cells are often present. The mitotic rate is generally low and necrosis is uncommon. The importance of correctly diagnosing MIFS is that it can be mistaken for either an inflammatory process or another malignant tumor such as Hodgkin lymphoma or a more aggressive sarcoma such as myxofibrosarcoma. In a series of high-grade MIFSs, Michal et al5 reported 18 patients with available follow-up. Nine (50%) developed metastases, including 7 who died. Most tumors in this series were situated in proximal locations with only 1 tumor in a distal extremity.

Differential Diagnosis

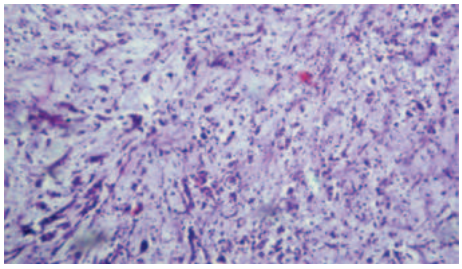
- Hemosiderotic fibrolipomatous tumour of bone
- Epithelioid Sarcoma
- Hodgkin Lymphoma
- Myxofibrosarcoma
- Pleomoarphic Liposarcoma
- Rosai-Dorfman Disease Tenosynovitis

Prognosis and Treatment

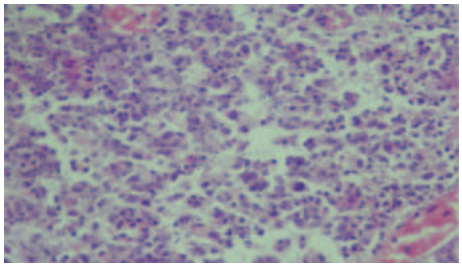
Myxoinflammtory fibroblastic sarcoma is locally aggres sive. It recurs 22% to 67% of the time, yet metastasizes in only 2% of cases, mostly to regional lymph nodes. However, pulmonary and widespread metastases do occur. Atypical histologic features have been implicated as negative prognostic indicators. Myxoinflammtory fibroblastic sarcoma is best treated by complete surgical excision.

CONCLUSION

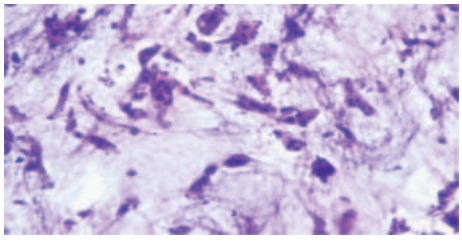
Myxoinflammatory fibroblastic sarcoma is a rare soft tissue sarcoma that tends to occur in the distal extremities in adults patients. It may be mistaken for an inflammatory process on clinical grounds and is often undertreated. Its heterogeneous histologic findings and high inflammatory background make the diagnosis challenging for pathologists. In general, MIFS has a high potential for local recurrence and low potential for metastasis, consistent with a low-grade sarcoma. Recently, high-grade, aggressive variants have been reported. As discussed above, the molecular findings are inconsistent, in some, but not all, MIFSs. Additional clinicopathologic and molecular studies should help to further define the spectrum and classification of this tumor.



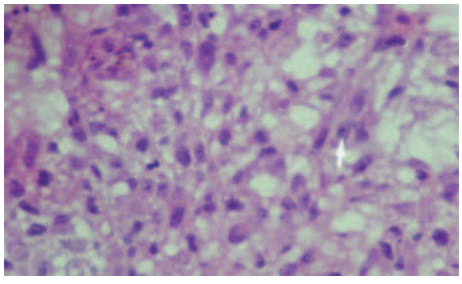
Low Power Photomicrograph Showing Inflammatory and Atypical Cells in the Background of Fibromyxoid Stroma.



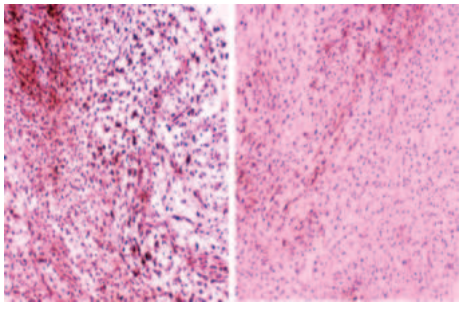
High Power Photomicrograph Showing Atypical Cells.



Low Power Photomicrograph Showing Chronic Dense Inflammatory Infiltrate



High Power Photomicrograph Showing Inflammatory Infiltrate Along with Fibroblasts in the Background of Myxoid Stroma.



Low Power Photomicrographs Showing Positive IHC Markers: CD68 and SMA.

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