CASE REPORT

Myxoinflammatory fibroblastic sarcoma of the thigh: A morphologic diversity

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ABSTRACT

Myxoinflammatory fibroblastic sarcoma (MIFS)/acralmyxoinflammatory fibroblastic sarcoma (AMFS) is a rare, painless, low-grade neoplasm which commonly occurs in the extremities. It has a distinctive morphology and can be a diagnostic challenge, simulating inflammatory conditions as well as neoplastic conditions. They are low-grade sarcomas with a protracted clinical course, a high rate of local recurrence and a low rate of metastasis. We report a case of proximal MIFS in a 50-year-old woman who presented with a mass in the thigh.

Key words: Bizarre cells, myxoinflammatory fibroblastic sarcoma, thigh

INTRODUCTION

Myxoinflammatory fibroblastic sarcoma (MIFS)/acral myxoinflammatory fibroblastic sarcoma (AMFS) is a rare, distinct low grade tumor of modified fibroblasts, which characteristically occurs in the distal extremities and has a propensity for local recurrence.[1] Although, initially described in the acral sites, it has now been increasingly recognized in the proximal soft tissues of the limbs. This tumor has only been recently described and the differential diagnosis is broad. It can often be mistaken for various reactive fibroinflammatory processes and tumors with higher metastatic potential, the management of which is different.[2,3] The prominent inflammation and fibrosis seen histologically in MIFS simulate a reactive process. The presence of myxoid foci and scattered bizarre cells which are occasionally multivacuolated may cause confusion with malignant fibrous histiocytoma and liposarcoma. A correct diagnosis is important to avoid unnecessary procedures and for proper clinical management.[2]

CASE REPORT

A 50-year-old woman presented with a mass in the left thigh of 15 days duration. The mass was painless and smooth with ill-defined borders. Fine needle aspiration yielded only adipose tissue fragments. Wide excision was done. Grossly, the mass was grey pink and measured 6×3.5×2 cm. The outer surface was nodular. Cut section showed grey-white to grey-yellow areas. Histological examination showed a well circumscribed, multinodular tumor having myxoid and hyalinized areas [Figure 1]. A heterogeneous population of round to oval cells with eccentric nuclei and clear cytoplasm, pleomorphic multinucleated giant cells with bizarre vesicular nuclei and inflammatory infiltrate consisting of lymphocytes and plasma cells were seen. Lipoblast-like cells, large cells with vesicular unilobed and bilobed nuclei, prominent macronucleoli and abundant cytoplasm resembling Reed Sternberg-like cells and virocyte-like cells were also noted [Figure 2]. On immunohistochemistry (IHC), tumor cells were positive for vimentin and negative for epithelial membrane antigen (EMA), cytokeratin (CK), CD30, CD68, CD34 and HMB45. On the basis of histopathology and IHC, a final diagnosis of MIFS was made. The margins of the tumor were free. Patient continues to remain disease free after two years of surgical excision with no evidence of recurrence or metastasis.

DISCUSSION

MIFS was first described in 1998, almost simultaneously by three different teams under three different names.[4,5] Since then about 100 cases have been reported.[5] Clinically

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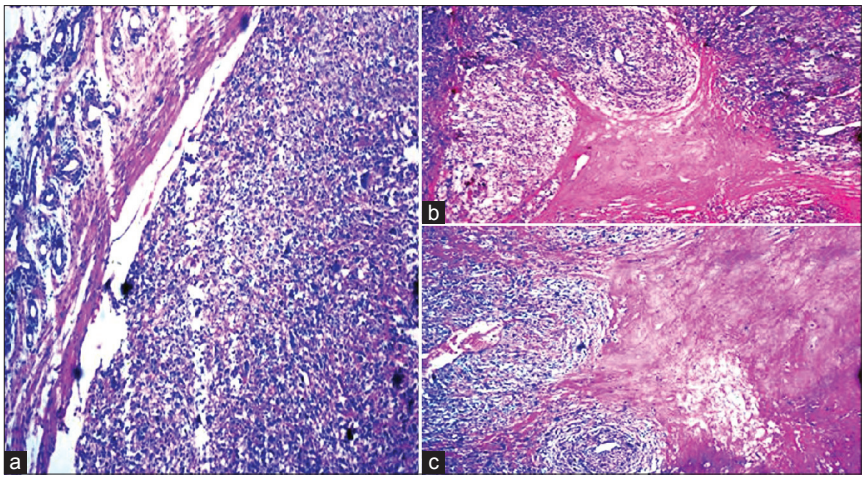

Figure 1: (a) Well circumscribed tumor (H and E, ×40). (b) Nodules and hyalinised areas (H and E, ×100). (c) Fibro-myxoid areas (H and E, ×40)

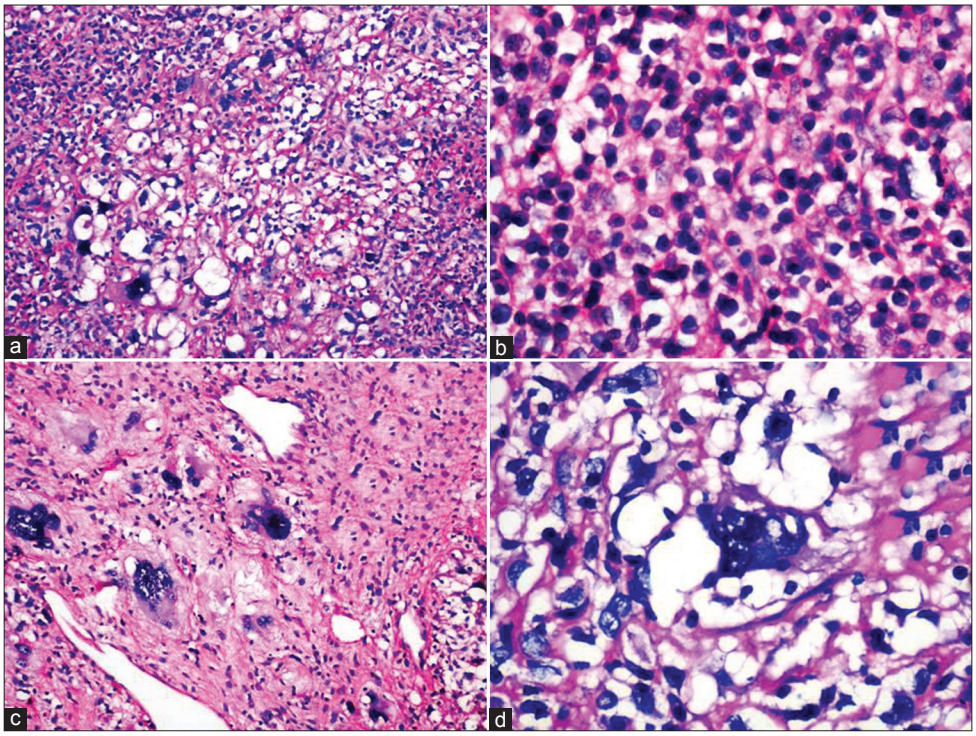

Figure 2: (a) Lipoblast-like cells (H and E, ×100). (b) Mixed inflammatory cell infiltrate, comprising predominantly of lymphocytes and plasma cells (H and E, ×400). (c) Virocyte-like cells (H and E, ×400). (d) Reed Sternberg-like cells (H and E, ×400)

it develops in patients of all ages and presents as a mass in the distal extremities, most frequently in the fingers and usually within the subcutaneous tissue.[6] It is now known that this tumor can occur more proximally and the term 'acral' has been dropped in 2002 WHO classification.[7] MIFS is currently included in the WHO classification of tumors of soft tissue under fibroblastic/myofibroblastic tumors. Most patients are adults with a peak incidence in the fourth and fifth decades of life. Male and female patients are equally affected. The most common presentation is a slow growing painless mass.[8] The upper extremities are affected about twice as often as lower.[9] In the present case, the patient was a 50-year-old woman with a slow growing painless mass in the thigh.

Three characteristic microscopic features have been described in the literature, which include:

- A somewhat multinodular overall architecture, alternating densely cellular and myxoid hypocellular areas
- Mixed inflammatory infiltrate and
- Bizarre giant and lipoblast-like cells.

The immunohistochemical phenotype is nonspecific; the tumor cells express vimentin in all the cases.[5] In the present case all the features were noted.

Identification of the atypical bizarre fibroblastic component as the manifestation of the malignant nature of this lesion is vital to correct diagnosis, and it is important to attend to the myxoid and hyalinized areas, inflammatory infiltrate, lipoblast-like cells and Reed Sternberg-like cells as features of MIFS.[6]

The differential diagnosis depends on the predominance of inflammatory, myxoid and bizarre cell components and is between non-tumoral lesions and tumoral lesions such as giant cell tumor, inflammatory fibrosarcoma, inflammatory myofibroblastic tumor, or myxofibrosarcoma.[5] Inflammatory myofibroblastic tumor and inflammatory fibrosarcoma commonly occur in the abdomen and thorax in contrast to MIFS. Cells of MIFS are more bizarre and lack histochemical features of myofibroblasts. Benign myxoid lesions lack large bizarre cells as found in MIFS. Hodgkin's disease can be considered by the presence of Reed Sternberg-like cells; however the cells in MIFS are CD15 and CD30 negative.[1]

Treatment consists of complete wide excision and checking the healthy status of the remaining edges.[3] Although MIFS consists of highly bizarre pleomorphic cells, only three cases of metastasis have been reported. However, local recurrence ranges from 22% to 67%. No associated deaths due to the disease have been reported.[5]

Morphological familiarity with MIFS is an important factor in arriving at a correct diagnosis, as it shares many features with benign as well as highly aggressive neoplasms. The role of IHC is limited and is useful in ruling out other differential diagnosis. Prognosis depends on initial surgical management, which should consist of complete surgical excision despite the probable functional impact. The possibility of metastatic potential and recurrence of this tumor must be dealt more cautiously.

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