Case Report

Malignant solitary fibrous tumor of the thigh in a young female: A rare case report

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ABSTRACT

Solitary fibrous tumor (SFT) is a mesenchymal origin tumor mainly confined to pleura and peritoneum. The extrapulmonary location is rare in incidence up to the level that fewer than 40 cases are described in literature in soft parts of extremities most of which are benign in nature with a mean age of presentation 52 years. The extrapulmonary SFT with malignant features at young age is an extremely rare mesenchymal neoplasm. A 20 years old female presented with a mass in the left thigh. The positron emission tomography scan performed, showed a hypermetabolic lesion at medial side of the left thigh. Subsequently, *en bloc* excision of mass was done along with adductor longus and brevis. Based on the histomorphology and immunohistochemical markers, diagnosis of malignant SFT was made. We report this case because of its rarity and to add on to literature and list of differential diagnosis of soft tissue tumors in the extremities.

Key words: Malignant, solitary fibrous tumor, thigh

INTRODUCTION

The extrapulmonary solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm. Only few cases affecting the extremities have been described so far. Case reports of SFTs of the thigh with atypical features on histopathology are very uncommon.[1] The mean age of presentation is 52 years with no gender predilection. Clinically, it usually presents as a painless slow-growing mass. Poor prognostic factors include large tumor size, extrathoracic location and malignant features on histology. Complete surgical excision with free surgical margins is the best modality of treatment as well as the most important prognostic factor. Very late relapses and metastasis (up to 30 years after the first surgery) have been documented, for which reason the clinical follow-up of these patients should be maintained indefinitely.[2]

CASE REPORT

We report a case of malignant SFT of left thigh in a 20 years female. The patient presented with a large mass in thigh and enlargement of femoral lymph nodes. Tru-cut biopsy of the mass was done which suggested a mesenchymal tumor of possibly malignant behavior. Radiological investigations revealed a soft tissue swelling that on positron emission tomography-scan showed a hypermetabolic subcutaneous soft tissue nodule at medial aspect of the left thigh with ulceration of overlying skin. No other metabolically active disease was found elsewhere in the body [Figure 1]. *En bloc* excision of mass was done along with adductor longus and brevis and the whole specimen was sent for histopathological examination. Grossly, an encapsulated firm tissue mass of size 17 cm × 11 cm × 6 cm was received [Figure 2]. On external surface of the mass, muscles were attached. Cut surface revealed a gray-white area that appeared to be merging with the muscle fibers. On histopathological examination, the tumor appeared well-circumscribed and revealed patternless proliferation of fibroblast-like cells with abundant eosinophilic cytoplasm and variable cellularity embedded in a collagenous and myxoid matrix. Foci of necrosis were also observed. Some areas revealed hemangiopericytoma-like pattern. The maximum mitotic activity observed was 35–40/10 high power fields [Figure 3]. No muscle infiltration was observed as such. Immunohistochemistry revealed tumor positivity for CD34, vimentin and focal positivity for NSE. The epithelial membrane antigen (EMA) and smooth muscle marker (SMA) were negative [Figure 4]. Based on the histomorphology and immunohistochemical markers, diagnosis of malignant SFT was made.

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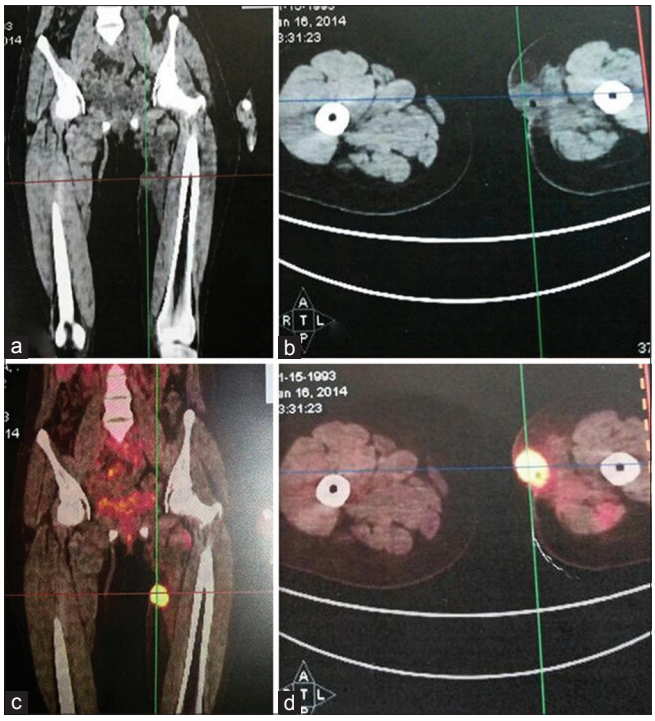

Figure 1: Computed tomography scan showing soft tissue mass (a and b) while positron emission tomography scan revealing a metabolically active lesion at medial side of left thigh (c and d)

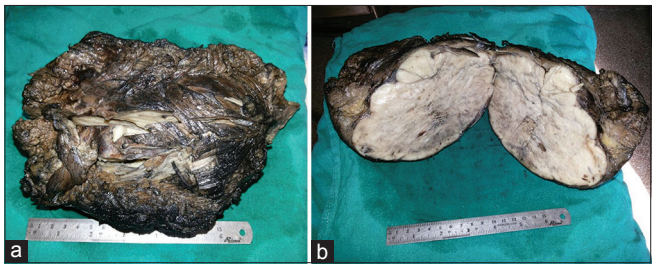

Figure 2: (a) Gross photomicrograph of tumor mass with attached muscles, (b) cut section revealing a circumscribed gray-white tumor mass

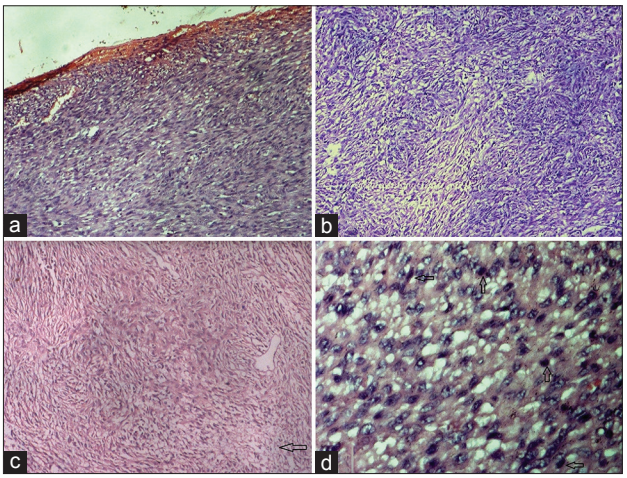

Figure 3: H and E sections showing a circumscribed cellular tumor area revealing patternless proliferation of spindle cells embedded in a collagenous stroma (a and b; ×100) with areas of necrosis (arrow head c; ×200) and high mitotic activity (arrow head d; ×400)

DISCUSSION

Malignant SFT of the thigh is a rare mesenchymal neoplasm. SFT is mainly confined to pleura and peritoneum. Recently, this tumor type has been reported in numerous sites including the liver, orbit, nasal passages, meninges, pericardium, tunica vaginalis, testis, skin, respiratory tract and thyroid.[2] So far, <40 cases have been described in literature in soft parts of extremities.[3] This tumor occurs in a wide age range with a mean age of presentation of 52 years and no sex predilection as such. They present clinically as slow-growing painless mass and may reach a size of more than 10 cm leading to compression symptoms. Rarely, it may cause paraneoplastic syndromes such as hypoglycemia due to the production of an insulin-like growth factor.[4] Most of the SFTs of the extremities documented are benign and aggressive behavior has been noted in only 10–20% cases. Generally, these tumors are well-circumscribed, and on histomorphology, their appearance varies from hypocellular fibrous lesions to less fibrous hypercellular neoplasms. Fibrous SFTs have a heterogenous microscopic appearance with alternate cellular and fibrous areas along with the presence of ramified medium sized hyalinized thick-walled vessels. At the other end, hypercellular lesions reveal monotonous histology with less intervening fibrosis and relatively thin-walled blood vessels. Cell population consists of round-to-spindle cells with a fascicular, storiform or fibrosarcoma-like arrangement. Tumor cell nuclei usually have a vesicular appearance due to open nuclear chromatin. Foci of chronic inflammation, myxoid change, and mast cells are commonly observed. Mitosis and necrosis are uncommon features. Earlier the hypercellular SFTs were reported as hemangiopericytomas. Histological variants such as giant cell SFT fibroma and fat forming SFT have been described.[5] Immunohistochemically, SFT commonly expresses CD34, CD99 and bcl-2, EMA. Occasional expression of SMA has also been seen. They are usually negative for S-100 protein, desmin and cytokeratins.[6] Immunohistochemically, malignant SFT tends to show reduced CD34 reactivity.[7] Distinctive histological criteria for malignant SFT have been variably described and consist of infiltrative nature of tumor, cytological atypia, mitotic count more than 4/10 high power fields and presence of necrosis. In our case, mitotic count was quite high that is, 35–40 per HPF and necrotic foci were also seen which suggested the malignant behavior of the tumor. The behavior of SFT is unpredictable.[8] The correlation between morphology of the tumor and prognosis of the tumor is poor. Some histologically benign appearing tumors may behave as a malignant one and vice-versa.

Differential diagnosis consists of various benign and malignant lesions such as fibromatosis, fibrosarcoma,

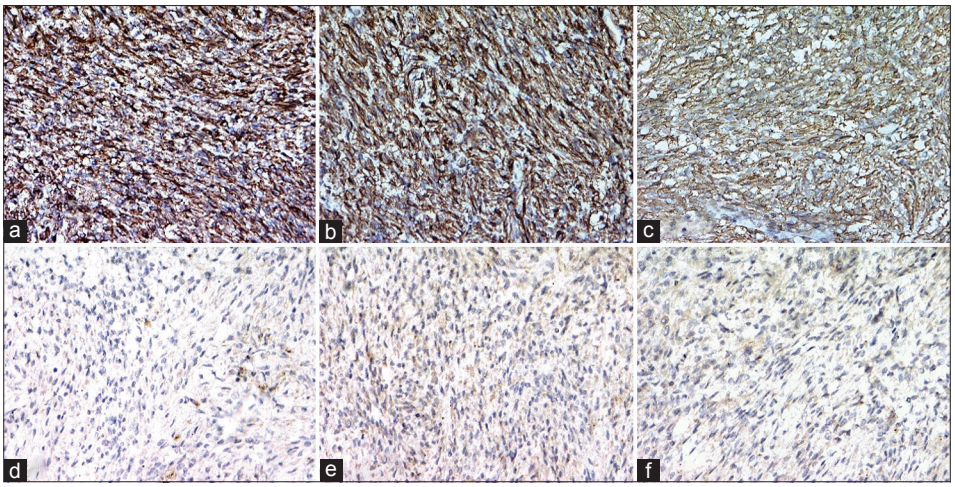

Figure 4: Immunohistochemistry expression in tumor cells (a) vimentin, (b) bcl-2, (c) CD34 – positive and (d) CK, (e) desmin, (f) smooth muscle marker – negative

malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma and dermatofibrosarcoma protuberans.[2] Synovial sarcoma is a close differential diagnosis specially when SFTs express EMA; however, synovial sarcoma can be differentiated as it rarely expresses CD34. Excision of the lesion with free surgical margins is the most important prognostic factor as incomplete resection is highly prone for recurrence. Other poor prognostic factors include extrathoracic location,[9] large tumor size >10 cm and malignant appearing histopathology.[10] These tumors are generally resistant to radiotherapy and chemotherapy. Combination therapy with temozolomide and bevacizumab has recently emerged as a potentially promising regimen for SFTs.[11]

CONCLUSION

Extrapleural SFT of an extremity is a rare entity. Varied histological features and uncommon locations often lead to confusion in the accurate diagnosis. Immunohistochemistry helps in reaching a definitive diagnosis. It should be considered cautiously in the differential diagnosis of soft tissue tumors in the extremities.

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