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Case Report

Superficial CD34 positive fibroblastic tumour: A rare mesenchymal tumor of borderline (intermediate) malignant potential

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ABSTRACT

Superficial CD34 positive fibroblastic tumor is a rare soft tissue tumor first brought to light by Jodi M Carter in 2014. Several cases have been reported since, and they have been newly included in the recent WHO classification under the fibroblastic and myofibroblastic tumors category. It is a mesenchymal neoplasm of borderline malignant potential predominantly found in middle-aged adults with a median age of 37 years and most commonly located in the lower extremities. It is a spindle cell tumor with pleomorphic epithelioid cells, low mitotic activity, and strong diffuse positivity for CD34 on immunohistochemistry. The prognosis is good post excision but needs long-term follow-up since few cases revealed recurrences and lymph node metastases. As far as we know, only 64 cases have been documented thus far. We have reported a 63-year female with essential histological features of superficial CD34 positive fibroblastic tumor with no local recurrence or metastasis to date.

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INTRODUCTION

Superficial CD34-positive fibroblastic tumor (SCPFT) was first described in 2014 by Carter et al in a report of 18 cases.[1,3] These are typically slow-growing and found in the subcutaneous planes or the superficial soft tissue.[4] This entity has been described as low-grade neoplasm under fibroblastic and myofibroblastic tumors of intermediate rarely metastasizing tumor category in the recent WHO classification 2020. Only 64 of these cases have been recorded thus far.[1,2,4]

A 63-year woman came with an eight-year history of swelling over the medial aspect of her right leg, gradually increasing in size and developing painful burning sensation in the last two months. On clinical examination, a lump approximately 5 cm × 5 cm, solid, irregular, mobile, and firm in consistency was noted on the medial aspect of the right leg, 7 cm away from the knee joint. Contrast-enhanced magnetic resonance imaging studies revealed, a hypointense lobulated lesion in the subcutaneous plane of the leg, showing intense enhancement on post-contrast T1 weighted images (fig. 1).

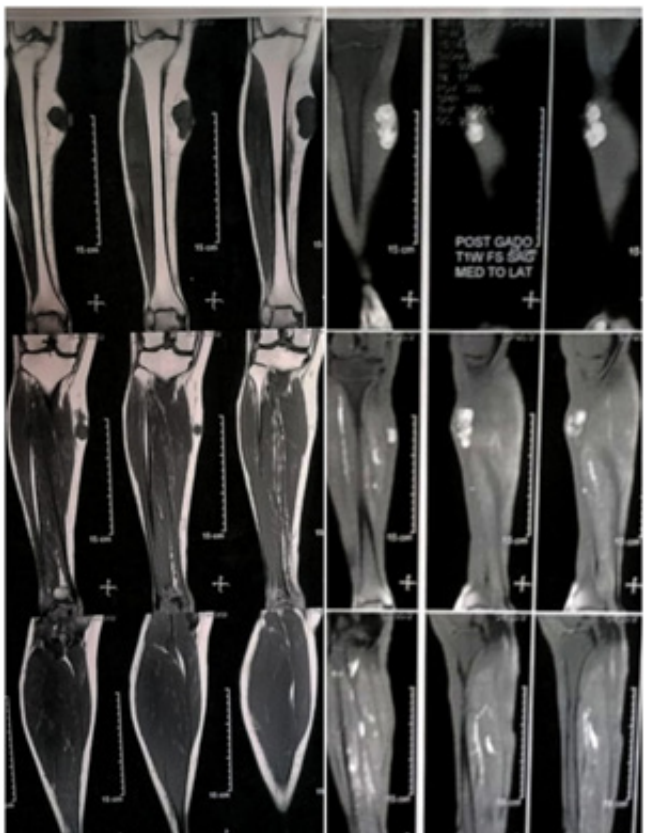

Figure 1: *T1 weighted images with post-contrast intense enhancement of the lesion*

The patient underwent wide local excision of the tumor and the specimen was received in the histopathology section. Grossly, the specimen was well-circumscribed, encapsulated, with bosselated, firm to hard in consistency. On cut section, it was homogenous and grey-white without any apparent haemorrhage or necrosis (fig. 2). The routine histological processing of paraffin embedded sections with hematoxylin-eosin staining was studied. Microscopic examination showed a circumscribed partially encapsulated spindle cell tumor with moderate cellularity, arranged in a storiform pattern with interspersed epithelioid morphology with abundant granular cytoplasm (fig. 3a). These cells were singly scattered and arranged in nests. Few cells showed intranuclear inclusions. Xanthomatous change was seen focally (fig. 3b). A few atypical cells having hyperchromatic nuclei with irregular nuclear membrane were also noted. Mitotic activity was low. Immunohistologically, the tumor showed strong diffuse positivity for CD34, focal positivity for PanCK, SMA and negative for Desmin (fig. 4). Based on these findings, the diagnosis of Superficial CD34-positive fibroblastic tumor was made. The patient was followed up for four months postoperatively and showed no signs of recurrence.

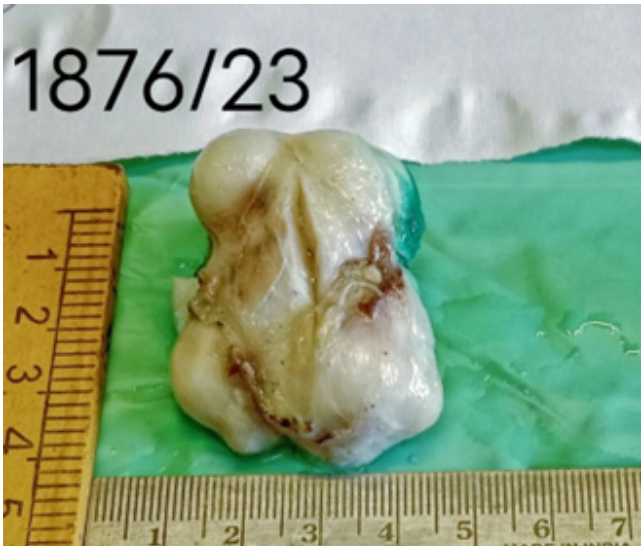

Figure 2: *Gross appearance of the tumor*

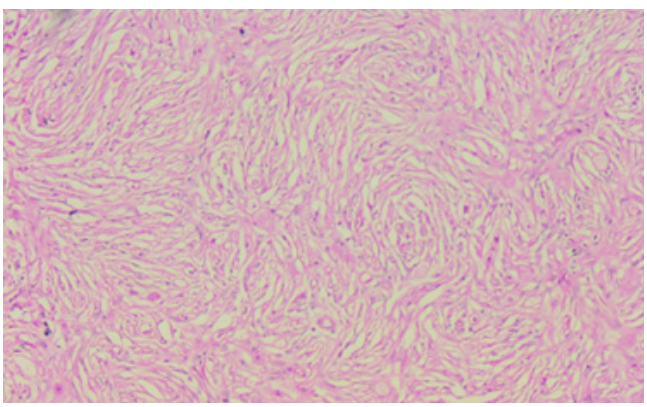

Figure 3a: *Spindle cell tumour with storiform pattern (HE stain, X10)*

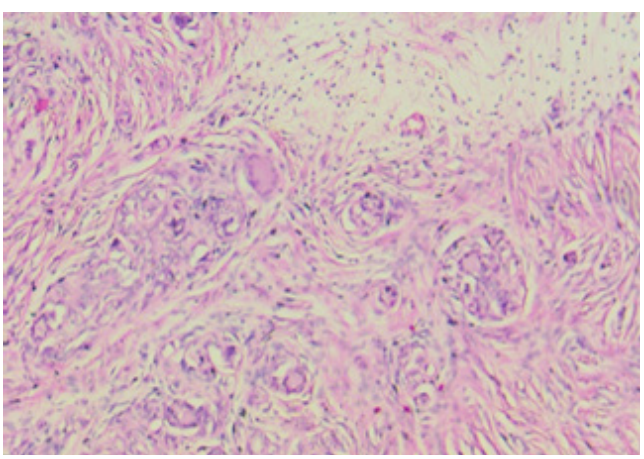

Figure 3b: *Tumour showing areas of xanthomatous change and giant cells (HE stain, X10)*

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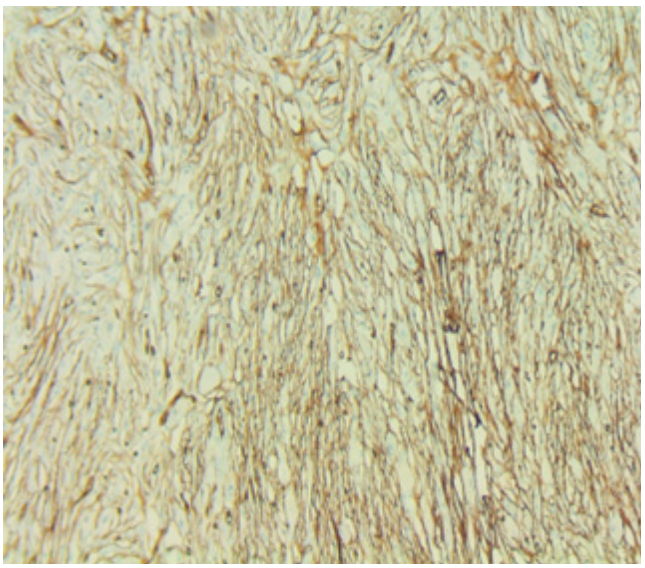

Figure 4: *Strong diffuse positivity for CD34 (IHC)*

DISCUSSION

SCPFT is a recently discovered rare mesenchymal tumor of intermediate malignant potential.[3,5] After Jodi M Carter described this tumor in 2014 in 18 cases, around 64 cases have been reported till now.[3]

The oldest patient was 55 years, while the youngest SCPFT instance documented involved an 8-year child. According to 16 case studies, the median age group for SCPFT was 42.5 years; however, the patient is 63 years in our case. Studies also indicate that men are more likely to have the condition.[8] It is a slow-growing tumor but other stories indicate rapid growth.[4] As in our instance, SCPFTs are more frequently identified in superficial soft tissue and are more common in areas of the lower extremity.[1] We report our experience of two decades with a distinctive, previously undescribed low-grade fibroblastic tumor of the superficial soft tissues. Eighteen cases were identified within our consultation files, previously coded as 'low-grade sarcoma, not further classified' and 'malignant fibrous histiocytoma, low grade'. The tumors occurred in adults (median age 38 years, range 20–76 years SCPFTs can be painless or can be associated with pain.[8] In the current case, there was initially no pain but a burning sensation gradually developed over two months.[9] Other symptoms like hyperpigmentation, and redness on the overlying skin, were absent.[4,6]

Histological features include spindle cells arranged in a classic storiform pattern and cells with granular cytoplasm.[7] Tumor cells appear stellate-shaped with interspersed epithelioid morphology having abundant granular cytoplasm.[1,4,7] Other features like pleomorphic cells, xanthomatous cells, intranuclear inclusions, sparse inflammatory cells (lymphocytes), and hemosiderin pigment are noted. Low mitotic activity (0-1 mitoses/50 HPF) is present.

The morphological differentials include Myxoinflammatory fibroblastic sarcoma (MIFS) and Dermatofibrosarcoma Protuberans (DFSP). In our case, the absence of the infiltrative nature of the tumor and myxoid background ruled out MIFS and DFSP. Characteristic pseudolipoblasts and marked nuclear pleomorphism with atypia did not constitute most of the tumor hence myxofibrosarcoma (MFS) and undifferentiated pleomorphic sarcoma (UPS) were ruled out. Immunohistologically, the tumor showed diffuse and strong positivity for CD34 like all other cases reported to date. SCPFTs have shown focal positivity invariably for PanCK and approximately 25% of previous cases showed focal positivity for SMA consistent with the current case. Almost all previous cases and current case were negative for desmin.

Considering all the clinical, histological, and immunohistochemical studies, our case is that of SCPFT. However, recent studies show the role of CD34, SynCAM3, and cyclin D1 help distinguish SCPFT from histological mimics like MIFS, MFS, UPS, etc also overlapping features of SCPFT with PRDM10-rearranged soft tissue tumor need further evaluation by molecular studies for PRDM10 gene status.[3]

A total of six cases of all the cases reported in the literature showed recurrence hence a close long-term follow-up becomes mandatory.[3]

CONCLUSION

In summary, the borderline malignant potential of SCPFT necessitates further study in order to avoid confusion with other histological mimics and, consequently, prevent the rarest chances of recurrence in such patients. Additionally, it is best to refrain from over-treatment given the favorable prognosis despite the substantial pleomorphism on the histological pattern.

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