

Superficial CD 34 Positive Fibroblastic Tumour – A Case report

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Abstract

Introduction: Soft tissue tumours are common and comprise a large spectrum of lesions according to the cell of origin or differentiation. These tumors can be benign, intermediate grade or frankly malignant. Superficial CD 34 positive fibroblastic tumor is a newly described entity yet to find its place in WHO classification of tumors of soft tissue and bone or other standard text books of soft tissue pathology. This tumour was first reported in 2013 by Jodi M Carter et al. by reviewing 18 cases of soft tissue tumours previously reported as unclassified, low grade sarcoma etc. After finding a specific strong diffuse CD 34 positivity in these tumours and excluding all other differential diagnoses, they suggested the name- “Superficial CD 34 positive fibroblastic tumor” for this entity. Two more similar cases were reported by Hendry Shona et al in 2015.

Case Report: We are reporting a case of a rare soft tissue tumour in a 22 year old female who presented with a nodule in the right upper thigh, having similar histology and immunohistochemical staining pattern as described in the above mentioned two publications.

Conclusion: Superficial CD 34 positive fibroblastic tumour is a new entity in soft tissue tumours. To the best of our knowledge, this is the first case in English literature to be reported from Asia. The exact biological behaviour of this lesion is to be documented by long term follow up of these patients.

Key Words: CD 34 positive tumor, Fibroblastic tumor, intermediate malignancy, superficial soft tissue tumour, subcutaneous / suprafascial plane.

Introduction

Soft tissue tumors are a vast group of lesions and each is categorised according to the cell of origin or differentiation. The large majority of soft tissue tumours are benign, with a very high cure rate after surgical excision. Malignant mesenchymal neoplasms amount to less than 1% of the overall human burden of malignant tumours [1]. In between these two groups, there is a category of tumors with intermediate malignancy. WHO

classification of tumors of soft tissue and bone, 4th edition [2] has subdivided the intermediate malignancy group into two categories -locally aggressive and rarely metastasizing. With the advent of immunohistochemistry, cytogenetics and molecular genetic analysis, significant changes have occurred in the classification and diagnosis of soft tissue tumors [3].

There are several recently characterized entities like superficial CD34 positive fibroblastic tumor,

fibrosarcoma-like lipomatous neoplasm, angiofibroma of soft tissue, low-grade sinonasal sarcoma with neural and myogenic features, malignant gastrointestinal neuroectodermal tumor, hemosiderotic fibrolipomatous

tumor, and epithelioid inflammatory myofibroblastic sarcoma [4]. Of these, a few have been included in the latest WHO blue book. Herein, we are reporting a case of a newly described entity of superficial soft tissue tumor- Superficial CD 34 positive fibroblastic tumour.

Case Report

22 year old otherwise healthy female patient presented with a swelling in the anterior aspect of upper part of right thigh since last 3 years, gradually increasing in size. On examination, a nodular swelling of size 3cm was palpated in the superficial plane, not fixed to skin. Provisional clinical diagnosis was fibroma/dermoid cyst. Ultrasonogram suggested a subcutaneous, supra fascial lesion in the upper anterior right thigh region. The underlying thigh musculature showed

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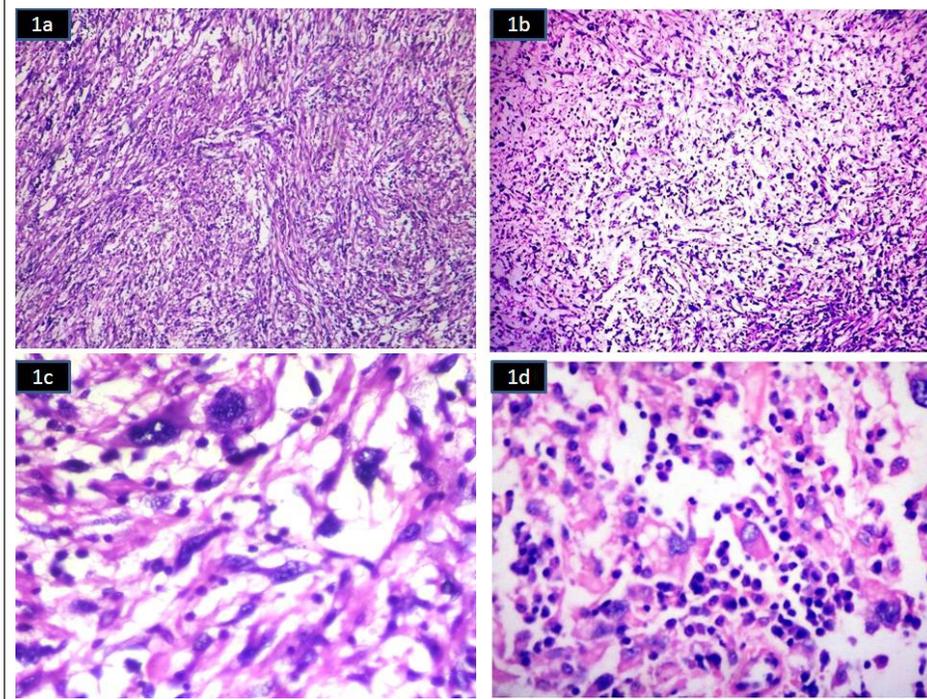


Figure 1:

- a- Hypercellular spindle cell area (H&E x100)
- b- Hypocellular bizarre cell area (H&E x100)
- c- Multinucleate giant cells (H&E x 400)
- d- Cells with glassy cytoplasm(H&E x 400).

present.(Figure 1d) Some cells showed prominent nucleoli and intranuclear pseudo-inclusions.(Figure 2b,c) Mitosis was seen infrequently (2-3/50hpf). Atypical mitosis was not seen. Necrosis was absent.

Hypocellular areas showed similar spindle shaped cells in a myxoid /oedematous background. Scattered cells with abundant vacuolated cytoplasm were also present. A dense collection of inflammatory cells composed mainly of plasma cells, lymphocytes and mast cells were seen admixed with tumor cells. Many plasma cells showed Russell bodies. Focal lymphoid follicle formation was seen. (Figure 2d)

Immunohistochemically, tumour cells showed diffuse strong positivity with CD34 and Vimentin. (Figure 3a,b) Immunostains for SMA (Figure 3c), EMA,CK, S100 and CD68 were negative. Ki-67 showed a low proliferative index in tumor cells (3%)

normal pinnate echo pattern. Inguinal lymph nodes were not present. Excision of the swelling was done under local anaesthesia. We received the slides and blocks of the specimen for consultation. The specimen was as per the gross description, a nodular mass measuring 3x2.5x2 cms; overlying skin was not present. Cut section showed grey white appearance. Histologically, the sections showed a neoplasm composed of spindle cells arranged diffusely and also in interlacing bundles. Hypercellular and hypocellular areas were present.(Figure 1a, b) Hypercellular area showed spindle cells arranged in interlacing fascicles. Individual cells had moderate amount of eosinophilic cytoplasm and elongated pleomorphic vesicular nucleus. Mono and multinucleated tumour giant cells and bizarre cells were noted (Figure 1c) scattered among the spindle cells. Cells with glassy cytoplasm were also

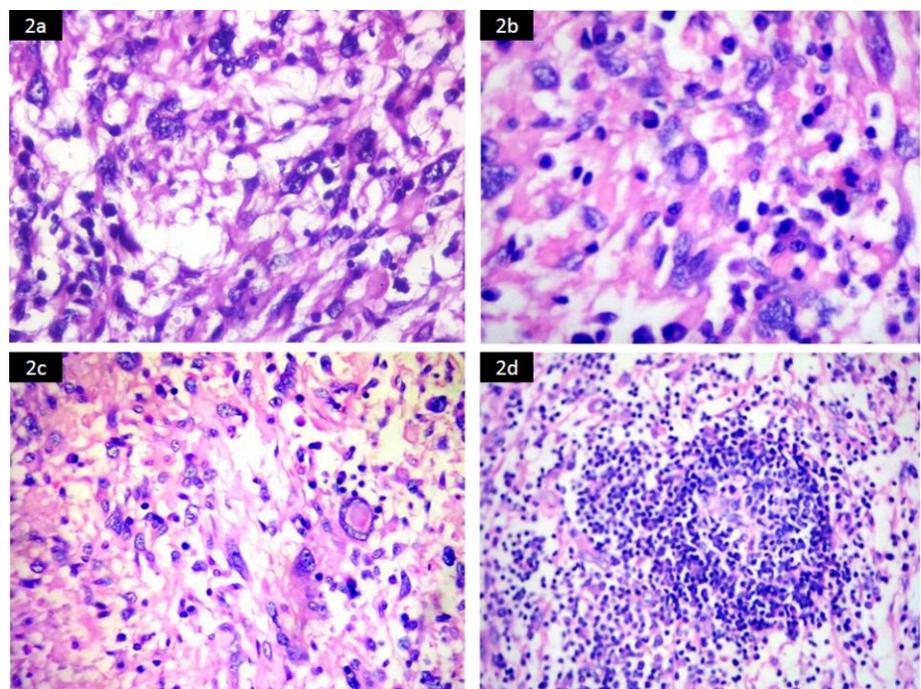


Figure 2:

- a- Cells with prominent nucleoli (H&E x 400)
- b& c - Intranuclear pseudo inclusion (H&E x 400).
- d-Lymphoplasmacytic infiltrate in the stroma with follicle formation (H&E x 400).

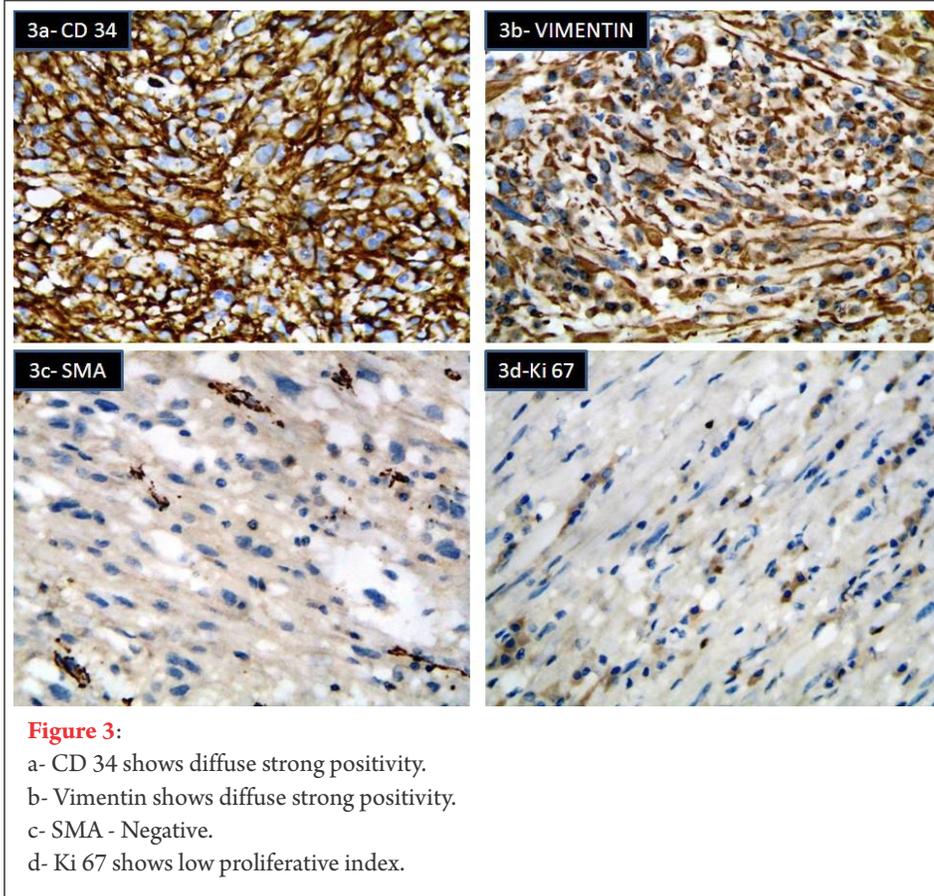


Figure 3:

- a- CD 34 shows diffuse strong positivity.
- b- Vimentin shows diffuse strong positivity.
- c- SMA - Negative.
- d- Ki 67 shows low proliferative index.

(Figure 3d). Thus, excluding the possibilities of other spindle cell neoplasms, a diagnosis of Superficial CD 34 positive fibroblastic tumour, a mesenchymal soft tissue tumour of intermediate malignancy, was made.

Discussion

Superficial CD 34 positive fibroblastic tumour is a newly described soft tissue neoplasm, first described by Jodi M Carter et al [5] in 2013. In their retrospective study, they recognised 18 cases of soft tissue tumour which showed diffuse strong positivity for CD 34 and did not fit into any of the conventional types of soft tissue tumours by histology or immunohistochemical staining pattern. These authors proposed the name Superficial CD 34 positive fibroblastic tumour for these types of soft tissue tumours. In their study, the age group varied from 20-76 years with a slight male predominance. Tumour size

ranged from 1.5 to 10 cms and thigh was the most common site though it is reported to occur in any superficial site. The present case was that of a 22 year old female patient with a superficial swelling in the thigh with a size of 3cms in greatest dimension, very similar to the cases in the first reported case series.

Microscopically, the closely intermixed inflammatory component in a spindle and pleomorphic cell population led us first to the differential of inflammatory myofibroblastic tumor (IMT). But, the histology of other cell components was not typical. Moreover, the CD 34 will never be strong, diffuse positive in IMT. Immunohistochemically, most cases of IMT are positive for smooth muscle and muscle-specific actins. But the present case was SMA negative. The large bizarre cells with prominent nucleoli and intra nuclear inclusions raised the possibility of a myxoinflammatory fibroblastic

sarcoma in which virocyte-like cells are seen. But location of this tumour entity is usually in the distal areas like acral region – in toes, fingers, wrist and ankle, although it has been reported in other locations also like forearm, arm, and thigh [6]. In a study of 44 cases of myxoinflammatory fibroblastic sarcoma by Meis-Kindblom et al [7], the neoplastic cells showed strong positivity for vimentin (25 of 25), focal positivity for CD68 antigen (17 of 25) and focal positivity for CD34 (7 of 25). Dermatofibrosarcoma protuberans was another differential which also can show strong diffuse CD 34 positivity. But the location in the dermal plane and the typical storiform histology differentiates it from the CD 34 positive fibroblastic tumour. Because of the presence of large cells with prominent nucleoli and the spindle cells with an inflammatory component, a misdiagnosis of inflammatory malignant fibrous histiocytoma may be done. But superficial location, CD 68 immunomarker negativity, strong CD 34 positivity and very low proliferative index in the present tumour should exclude this possibility. Myxofibrosarcoma and fibromyxosarcoma though can occur in superficial location, were not considered because by definition these need more than 50% myxoid stroma [8]. Hemosiderotic fibrolipomatous tumor/early pleomorphic hyalinizing angiectatic tumor and classical pleomorphic hyalinizing angiectatic tumor did not come into the differentials, since significant hemosiderosis or hyalinised ectatic vessels were not present in our case. A spindle cell epithelial neoplasm was ruled out with CK and EMA negativity. SMA was also negative ruling out a smooth muscle neoplasm. To determine the proliferative activity, a Ki 67 immunostain was done which

showed a low proliferative index of 3% suggesting that it is not a high grade sarcoma.

The histology in the present case was exactly like the cases reported by Jodi M Carter et al [5] with spindle and bizarre cells arranged in hyper/hypocellular pattern. The bizarre cells showing glassy cytoplasm, prominent nucleoli, intra nuclear eosinophilic inclusions were present in the present case also. And our case showed a strong diffuse positivity for CD 34 too. Molecular study for TGFBR3 and/or MGEA5 rearrangements was done by Jodi M Carter in a few of their cases and found to be negative ruling out a histologically similar tumor myxoinflammatory fibroblastic sarcoma. We have not done molecular study in our case. The degree of atypia shown by superficial CD34-positive fibroblastic tumor exceeds that typically seen in myxoinflammatory fibroblastic sarcoma and the latter do not show diffuse strong CD 34 expression [5].

Two more similar cases were reported by Hendry Shona et al [9] in 2015 which also showed all these features. Only 20 cases have been reported so far the world over. To the best of our knowledge, this is the first case to be reported from Asia in English literature.

Conclusions

New entities of soft tissue tumours with specific immunohistochemistry, cytogenetics and molecular genetics pattern are being described which have a particular behavior. CD34 positive fibroblastic tumour is one such new entity. The behaviour of the lesion is said to be of intermediate grade. However, more cases need to be described to correctly categorise this tumour into WHO subgroups of locally aggressive or locally metastasizing. The challenge in the diagnosis of this tumour is the myriad of histological patterns seen in the same tumour. Diffuse, strong CD 34 positivity combined with negativity for other markers helps to clinch the diagnosis. With the reporting of our case, we have data of 21 cases of CD 34 positive fibroblastic tumour in the literature. Awareness of this specific entity and reporting of more such cases will help to learn more about this entity and find place in the next edition of WHO classification of tumours of Soft tissue and Bone and other text books of soft tissue pathology.

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Conflict of Interest: NIL

Source of Support: NIL

Informed consent of the patient was taken for publication

How to Cite this Article

Jayalakshmy P.S, Feroze M, Merin J, Freena R. Superficial CD 34 Positive Fibroblastic Tumour – A Case report. Journal of Bone and Soft Tissue Tumors Sep-Dec 2016;2(3):10-13.