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Case Report

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Cytohistomorphological Features of Superficial CD34-Positive Fibroblastic Tumor

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ABSTRACT

Introduction: SCPFT is a rare, low-grade mesenchymal neoplasm first described by Carter et al. in 2014. Clinically, it presents as a slow-growing, painless, well-circumscribed mass in superficial soft tissue over lower extremities and trunk of adults. It is characterized by spindle to epithelioid cells with eosinophilic cytoplasm, mild nuclear atypia, and rare mitotic activity. The tumor shows diffuse and characteristic strong CD34, while being negative for S100, Desmin, and smooth muscle actin (SMA). Cytological descriptions of SCPFT are scarce in the literature, posing a diagnostic challenge on fine-needle aspiration.

Aim/Objective: To describe the cytomorphological features of a rare case of Superficial CD34-positive fibroblastic tumor (SCPFT) and emphasize its diagnostic considerations on fine needle aspiration cytology.

Case Report: A 16-year-old girl presented with a slow-growing, painless swelling over the posterior aspect of the right arm for two years. Clinical examination revealed a mobile, well-circumscribed mass measuring 3 cm x 3 cm. Fine needle aspiration cytology was performed, followed by histopathological examination and immunohistochemical analysis for confirmation.

Results: Cytology smears were moderately cellular, showing loose cohesive clusters and scattered singly dispersed spindle to epithelioid cells. The tumor cells exhibited moderate to abundant eosinophilic cytoplasm, occasional vacuolation, vesicular nuclei, and prominent nucleoli. Focal nuclear pleomorphism was observed, with occasional bizarre and multinucleated cells, but no mitotic figures. The background showed a mixed inflammatory infiltrate. Differential diagnoses considered were solitary reticulohistiocytoma, atypical fibrous histiocytoma, alveolar soft part sarcoma, and atypical fibroxanthoma. Histopathological examination and immunohistochemistry confirmed the diagnosis of Superficial CD34-positive fibroblastic tumor.

Conclusion: Superficial CD34-positive fibroblastic tumor is a rare, low-grade neoplasm of mesenchymal neoplasm with overlapping cytological features. Recognition of its characteristic cytomorphology, along with histopathology and immunohistochemistry, is crucial for accurate diagnosis and appropriate patient management.

Key Words: Low-grade mesenchymal neoplasm, Fibroblastic, Epithelioid cells, CD34 positive, FNA, Histopathology, Immunohistochemistry

INTRODUCTION

SCPFT is a rare, recently recognized low-grade mesenchymal neoplasm first described by Carter et al. in 2014. [1] It typically arises in the superficial soft tissue, involving the dermis and subcutis, most often over the lower extremities and trunk of young to middle-aged adults, with no significant sex predilection. [2,3] Clinically, it presents as a slow-growing, painless, well-circumscribed mass.[2,3]

Histopathologically, SCFT is characterized by spindle to epithelioid cells with eosinophilic cytoplasm, mild nuclear atypia, and rare mitotic activity. The tumor shows diffuse and strong CD34 positivity and may exhibit focal keratin reactivity, while being negative for S100, Desmin, and smooth muscle actin (SMA). Cytological descriptions of SCPFT are scarce in the literature, posing a diagnostic challenge on fine-needle aspiration (FNA).

Because SCPFT may mimic a range of benign and malignant spindle cell tumors, including dermatofibrosarcoma protuberans, atypical fibroxanthoma, and epithelioid sarcoma, recognizing its cytomorphologic features is essential for accurate preoperative diagnosis and appropriate management.

In this report, we present the cytological findings of a case of SCPFT, highlighting its distinguishing features and discussing the differential diagnosis on cytology.

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CASE REPORT

A 16-year-old female presented with painless swelling in the right arm for two years. On clinical examination, a mobile swelling was noted on the posterior aspect of the right arm, the lesion was cutaneous. FNAC was performed, and smears examined showed a moderately cellular smear, plump spindle cells arranged in loose cohesive clusters as well as singly scattered. Anisonucleosis was moderate to focally marked, with a moderate amount of cytoplasm, vacuolated cytoplasm noted at places, along with nuclear vacuolisation, which was seen occasionally, admixed with multinucleated giant cells and mixed inflammatory infiltrate in the background. Based on the clinicomorphological picture, the following possible differentials were considered. Solitary reticulohistiocytoma, Atypical fibrous histiocytoma, atypical fibroxanthoma, alveolar soft part sarcoma, and Juvenile xanthogranuloma, [5]

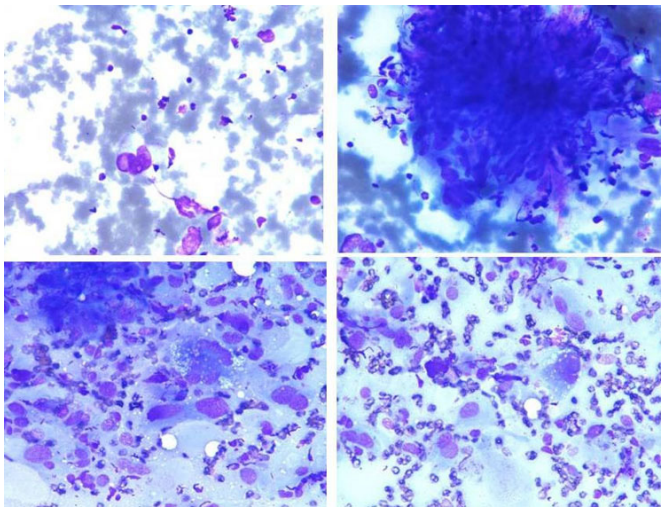

Figure 1: May-Grunwald Giemsa Smear show (A) Intranuclear vacuolisation (x400). (B) Moderately cellular smear composed of plump spindle cells. (C) Pleomorphic cell admixed with few multinucleated giant cells (x200). (D) Cells show cytoplasmic vacuolisation in the mixed inflammatory background (x200).

Immunocytochemistry (ICC) was performed on the available cytological material. On IHC, the neoplastic cells exhibited diffuse cytoplasmic positivity for vimentin, focal positivity for CD68, and a low Ki-67 proliferative index. The tumor cells were negative for epithelial membrane antigen (EMA) and Desmin. Based on the combined cytomorphologic and immunocytochemical features, a final diagnosis of Solitary Reticulohistiocytoma was rendered.

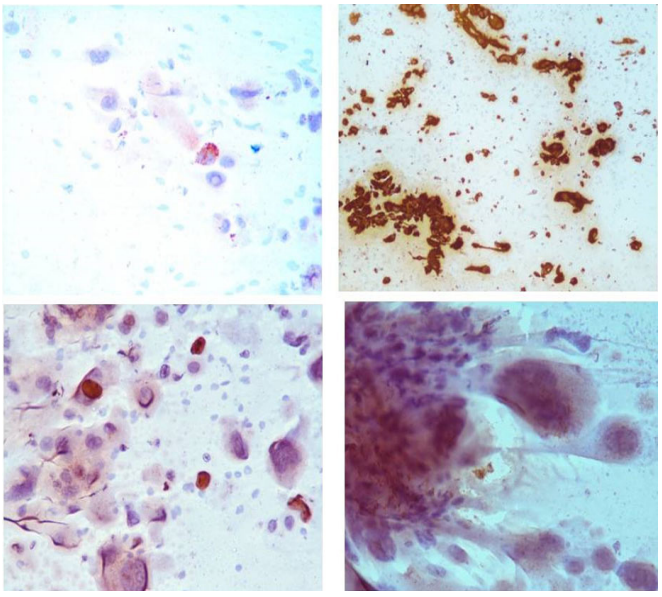

Figure 2: Immunocytochemistry (ICC). (A) Low Ki67 proliferation index (x400). (B) Diffuse Vimentin positivity (x400). (C) Focal CD68 positivity (x400). (D) Epithelial membrane antigen (EMA) and Desmin negative (x400).

The excised tumor was well-circumscribed and smooth cut surface. The H&E-stained section showed an encapsulated tumor, with proliferating spindle cells arranged in storiform and fascicular pattern admixed with few multinucleated giant cells. The spindle cells showed moderate to focal marked degree of pleomorphism, abundant granular to glassy cytoplasm. Despite focal marked pleomorphism, mitotic activity was negligible.

Table 1: Differential diagnosis of a superficial soft tissue tumor on cytomorphology

Tumor entity	Age	Site	Cytological Features	IHC
Solitary Reticulohistiocytoma	Young adults	Head, Neck, and Trunk	Moderately cellular smear; large histiocytoid cells (polygonal to round with abundant eosinophilic to finely granular cytoplasm) singly and in loose aggregates with occasional multinucleated giant cell in the inflammatory and scattered histiocytes in the background	strong and diffusely positive for CD 68, Positive for CD163
Atypical fibrous histiocytoma	Adults (3rd & 4th decade	Distal extremities	Moderate to highly cellular; spindle, epithelioid/histiocyte-like cells; moderate nuclear pleomorphism, moderate eosinophilic, occasional vacuolated cytoplasm; occasional giant cells; background fibrous fragment	Positive for Vimentin And CD68

Table 1: ***(Continued)***

Juvenile Xanthogranuloma	Young children	Head, neck, and trunk	Moderate to highly cellular; foamy histiocytes; Touton giant cells with vacuolated cytoplasm, absence of atypia, mitoses, or necrosis	Diffuse Positive for CD68 And Factor XIIIa
Atypical fibroxanthoma	Elderly	Sun-exposed sites (head and neck)	Highly cellular smear; pleomorphic spindle and epithelioid cells; bizzrae multinucleated giant cells; marked nuclear pleomorphism, moderate to abundant eosinophilic, occasionally vacuolated cytoplasm; frequent atypical mitoses, necrosis, hemorrhage, inflammatory background	Strong positivity for Vimentin, Often diffusely positive for CD10 and variable positivity for CD68
Alveolar soft part sarcoma	Young adults	Distal extremities	Moderately to highly cellular smears; loosely cohesive clusters, acinar or alveolar pattern; large polygonal cells; abundant granular to clear cytoplasm; central to eccentric vesicular nuclei with prominent nucleoli	TFE3 strong nuclear positivity, Vimentin positive, CD34 negative

IHC was performed and was found to be diffusely positive for vimentin and CD34. Desmin, smooth muscle actin (SMA), EMA, and S100 came out to be negative. Ki67 index was low, and CD68 was focally positive. Thus, ruling out all the differentials considered earlier both on cytology and histology.

The definitive diagnosis was superficial CD34-positive fibroblastic tumor (SCPFT). Follow-up until date revealed no evidence of local recurrence, regional lymphadenopathy, or distant metastasis.

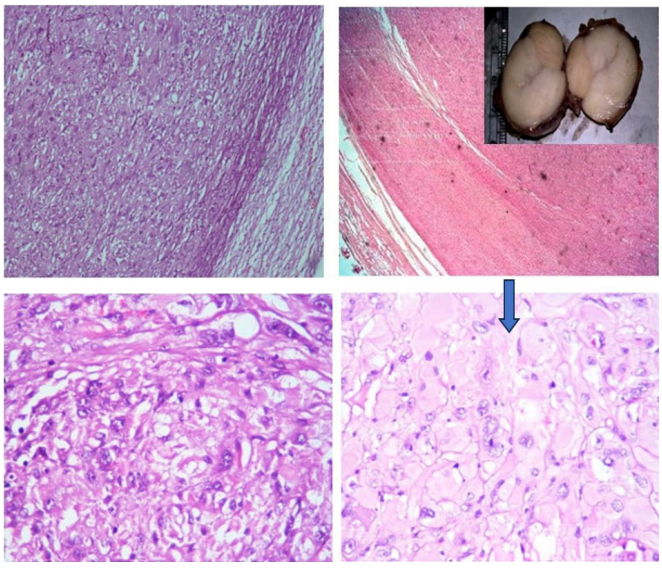

Figure 3: Haematoxylin and Eosin-stained section show (A&B) Encapsulated lesion showing marked pleomorphism along with occasional multinucleated giant cells (x100). Inset- encapsulated lesion measuring 4 cm x 3 cm x 2.5cm (C) Pleomorphic cells admixed with inflammatory cells (x100). (D) Cells show glassy cytoplasm (arrow).

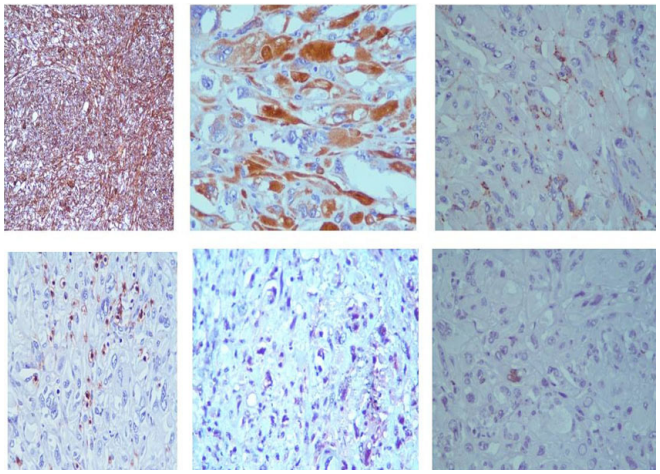

Figure 4: Immunohistochemistry (IHC) (A) Diffuse CD34 positivity, both cytoplasmic and membranous (x400). (B) Diffuse Vimentin positivity (x400). (C) Focal CD68 positivity (x400). (D) Residual lymphocytic cells show focal CD10 positivity (x400). (E) Desmin, Myogenin, S100 negative (x400). (F) Low Ki67 proliferation index (x400).

DISCUSSION

Superficial CD34-positive fibroblastic tumor (SCPFT) is a recently recognized entity in the 2020 WHO classification of soft tissue and bone tumors, characterized as a low-grade fibroblastic neoplasm with a distinctive immunophenotypic and genetic profile.[4] Although its histological and molecular features are increasingly well documented, descriptions of its cytomorphology remain sparse, reflecting both the rarity of the lesion and its relatively recent delineation as a separate entity. Our case contributes to the growing body of literature on the cytologic features of SCPFT.

Clinically, SCPFT typically presents as a slowly growing, superficial mass in the distal extremities of young to middle-aged adults. Its behaviour is indolent, with complete surgical excision being curative in most cases.[5] Recurrence is rare, and metastasis exceedingly uncommon. Awareness of its cytologic features is important to prevent both overdiagnosis as a high-grade sarcoma and underdiagnosis as a benign spindle cell lesion.

Cytologically, SCPFT aspirates are typically of moderate cellularity, comprising spindle to epithelioid cells arranged singly and in loose aggregates. The cells often display moderate to abundant eosinophilic cytoplasm, with nuclei showing vesicular chromatin, vacuolization, and prominent nucleoli. These features impart a degree of nuclear atypia that may raise concern for malignancy. However, mitotic figures are infrequent, and necrosis is absent.[6]

The principal diagnostic challenge in cytology lies in distinguishing SCPFT from other superficial spindle cell tumors. Dermatofibrosarcoma protuberans (DFSP) may be closely mimicked due to shared CD34 positivity. However, DFSP tends to exhibit more uniform, bland spindle cells and lacks the epithelioid morphology and prominent nucleoli characteristic of SCPFT.[6] Other differentials include solitary reticulohistiocytoma and atypical fibrous histiocytoma, which are diffusely positive for CD68, while focal in SCPFT. CD1a-positive tumor (LCH) and Juvenile xanthogranuloma may be included in the differentials based on clinical features, though LCH shows typical nuclear grooves, while Juvenile xanthogranuloma shows diffuse positivity for CD68. Atypical fibroxanthoma and pleomorphic rhabdomyosarcoma, which demonstrate marked pleomorphism and atypical mitoses, features that are not seen in SCPFT.[7]

Immunohistochemistry demonstrates diffuse CD34 and vimentin positivity with focal CD68 expression, while SMA and S100 are negative. Thus, a combination of cytomorphology, histopathology, and immunophenotype leads to a correct diagnosis.

CONCLUSION

In conclusion, SCPFT is a rare, low-grade neoplasm of mesenchymal origin with bizarre cytomorphology. Accurate diagnosis of this tumor holds substantial clinical and pathological significance, and to avoid overtreatment. The diagnostic process, when integrating with cytomorphological evaluation with immunocytochemistry analysis, is expected to become increasingly refined and reliable.

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Authors' Contribution:

Dr Neelam Sood- Concept, Diagnosis Editing and Final Version

Dr Kamini Singh – Drafting, Review of Literature

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