International Journal of Research in Medical Sciences
Kupati S et al. Int J Res Med Sci. 2026 Mar;14(3):1207-1210
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20260646

Case Report

Superficial CD34-positive fibroblastic tumor presenting as a retroauricular mass in an elderly male: a case report

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Received: 20 January 2026
Revised: 17 February 2026
Accepted: 18 February 2026

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ABSTRACT

Superficial CD34-positive fibroblastic tumor (SCPFT) is a rare, recently recognized fibroblastic neoplasm of the skin and subcutaneous tissue, characterized by distinctive histomorphology and strong CD34 expression. It predominantly affects middle-aged and elderly individuals and typically follows an indolent clinical course. Due to its rarity and morphological overlap with other spindle cell neoplasms, accurate diagnosis can be challenging. We report a case of SCPFT in a 73-year-old male presenting with a slowly growing, painless retroauricular mass of three years' duration. Histopathological examination revealed a well-circumscribed lesion composed of spindle and epithelioid cells with bizarre nuclei but lacking mitotic activity or necrosis. Immunohistochemistry demonstrated strong, diffuse CD34 positivity with negativity for STAT6, SOX10, β-catenin, S100, desmin, and smooth muscle actin, supporting the diagnosis of SCPFT. This case highlights the importance of recognizing SCPFT at uncommon anatomical sites and underscores the role of immunohistochemistry in differentiating it from other spindle cell tumors. Awareness of this entity is essential to ensure appropriate management and avoid overtreatment.

Keywords: Superficial CD34-positive fibroblastic tumor, CD34, Spindle cell tumor, Fibroblastic neoplasm, Retroauricular mass

INTRODUCTION

Superficial CD34-positive fibroblastic tumor (SCPFT) is a rare mesenchymal neoplasm arising in the superficial soft tissues, including the dermis and subcutis.[1] It was first characterized as a distinct entity in 2014 and has since been incorporated into the world health organization (WHO) Classification of soft tissue and bone tumours (5th edition).[3] SCPFT is considered a low-grade fibroblastic tumor with characteristic histological and immunophenotypic features, most notably strong and diffuse CD34 expression.[1,2]

Clinically, SCPFT typically presents as a slow-growing, painless superficial mass. The lower extremities are most frequently involved, followed by upper extremities and trunk.[2] Head and neck presentation is rare, and retroauricular localization is distinctly uncommon.

Histologically, SCPFT shows spindle to epithelioid cells arranged in fascicles with nuclear pleomorphism and bizarre hyperchromatic nuclei.[1] Despite cytological atypia, mitotic activity is minimal or absent[1,3]. A mixed inflammatory infiltrate is often present.[2]

Given its rarity and overlapping features with dermatofibrosarcoma protuberans, solitary fibrous tumor, and atypical fibroxanthoma, accurate diagnosis requires careful histopathological and immunohistochemical assessment.[1,2]

Given its rarity and overlapping features with other spindle cell tumors such as dermatofibrosarcoma protuberans,

solitary fibrous tumor, and atypical fibroxanthoma, SCPFT remains a diagnostic challenge. This report documents a rare case of SCPFT arising in the retroauricular region of an elderly male, expanding the anatomical spectrum of this tumor and emphasizing the importance of careful histopathological and immunohistochemical evaluation.

CASE REPORT

A 73-year-old male presented with a slowly enlarging swelling behind his left ear, which had been present for approximately three years. The lesion was painless and not associated with discharge, ulceration, or systemic symptoms. There was no history of trauma, radiation exposure, or similar lesions elsewhere on the body. The patient's medical history was otherwise unremarkable.

On physical examination, a firm, well-defined, mobile nodule measuring approximately 1 cm was noted in the retroauricular region. The overlying skin appeared normal, with no erythema or ulceration. Regional lymph nodes were not palpable. Based on the clinical impression of a benign soft tissue lesion, an excision biopsy was performed.

Gross findings

The excised specimen consisted of a grey-white, firm tissue fragment measuring 10×8×6 mm. The cut surface was solid and homogeneous, without areas of hemorrhage or necrosis.

Microscopic findings

Histopathological examination revealed a well-defined lesion located in the superficial soft tissue, with focal infiltration into the surrounding tissue. The tumor was composed predominantly of spindle-shaped cells arranged in short fascicles and bundles, interspersed with epithelioid cells. Many tumor cells exhibited enlarged, irregular, and bizarre hyperchromatic nuclei with glassy chromatin. Despite the striking nuclear atypia, mitotic figures were not identified. No necrosis or atypical mitosis was seen.

The stroma showed a mixed inflammatory infiltrate comprising lymphocytes, plasma cells, and occasional mast cells. The overall morphology suggested a low-grade neoplasm with atypical cytological features.

Immunohistochemistry

Immunohistochemical analysis demonstrated strong, diffuse cytoplasmic positivity for CD34 in tumor cells. The tumor cells were negative for STAT6, SOX10, β-catenin, CD10, S100 protein, desmin, and smooth muscle actin, effectively excluding solitary fibrous tumor, neural tumors, fibromatosis, melanocytic lesions, and smooth muscle neoplasms. Pan-cytokeratin showed focal patchy positivity.

Based on the combined histomorphological features and immunoprofile, a diagnosis of SCPFT was rendered.

Figure 1: Clinical picture of a patient showing retro auricular swelling.

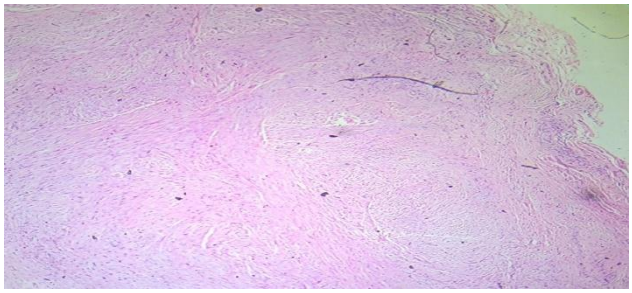

Figure 2: H and E 10× showing well defined lesion with areas of infiltration.

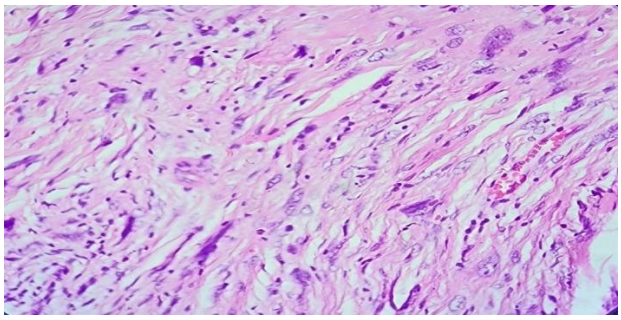

Figure 3: H and E 40× showing bizarre spindle to epithelioid cells with glassy chromatin.

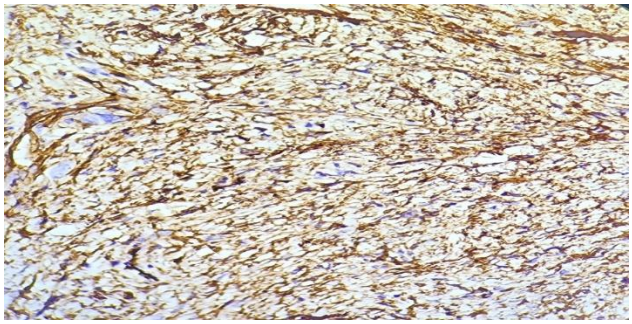

Figure 4: Immunohistochemistry CD34 Strong diffuse positivity.

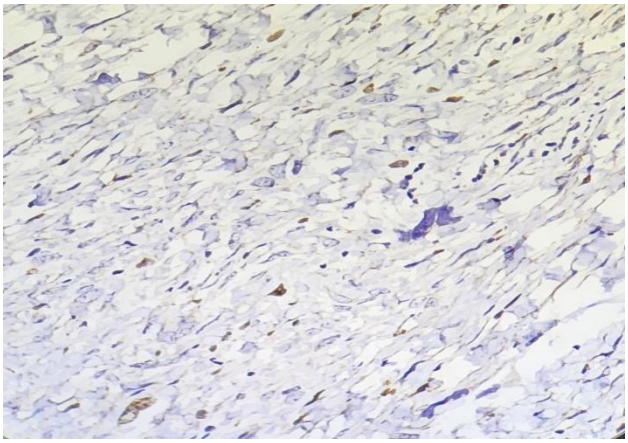

Figure 5: Immunohistochemistry PanCK patchy positivity.

DISCUSSION

SCPFT is an uncommon fibroblastic neoplasm with distinctive pathological features and an indolent clinical course. Since its initial description by Carter et al fewer than a hundred cases have been documented making it a relatively unfamiliar entity for practicing pathologists.[1,3] The tumor predominantly affects middle-aged and elderly adults, consistent with our patient.[3]

Recognition of SCPFT is important because its cytological atypia may raise concern for malignancy, potentially leading to overtreatment.

Most cases arise in the lower extremities, whereas head and neck involvement remains uncommon.[3-11] The retroauricular location in this case expands the anatomical spectrum of SCPFT.

Histologically, SCPFT demonstrates marked nuclear pleomorphism but lacks significant mitotic activity or necrosis. The inflammatory background is a characteristic feature.[1-3]

Immunohistochemically, strong CD34 positivity is the defining feature. Other markers including desmin, SMA, S100, SOX10, ERG, and STAT6 are negative.[1,3] Patchy cytokeratin positivity has been reported in up to 68% of cases.[5,15]

The differential diagnosis includes:Dermatofibrosarcoma protuberans (CD34 positive but storiform pattern).[12] Solitary fibrous tumor (STAT6 positive). Atypical fibroxanthoma (high mitotic rate, CD34 negative).[12]

Recent molecular studies have identified recurrent PRDM10 rearrangements in SCPFT, supporting its classification as a distinct entity.[9-15]

SCPFT is considered a tumor of intermediate behavior with very low recurrence risk. Complete surgical excision remains the treatment of choice.[3]

CONCLUSION

Superficial CD34-positive fibroblastic tumor is a rare, low-grade fibroblastic neoplasm that can present at uncommon anatomical locations, including the retroauricular region, as demonstrated in this case. Despite its striking cytological atypia, the tumor follows an indolent clinical course with minimal risk of recurrence when adequately excised. Accurate diagnosis relies on recognition of its characteristic histomorphology and immunophenotype, particularly strong CD34 positivity with absence of lineage-specific markers. This case contributes to the growing body of literature on SCPFT and emphasizes the importance of awareness of this entity to ensure appropriate diagnosis, avoid overtreatment, and improve understanding of its clinicopathological spectrum.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kupati S, Lakkundi S. Superficial CD34-positive fibroblastic tumor presenting as a retroauricular mass in an elderly male: a case report. Int J Res Med Sci 2026;14:1207-10.