SKIN

BRIEF ARTICLE

A Rare Case of Superficial CD34+ Fibroblastic Tumor: A Case Report

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ABSTRACT

A superficial CD34+ fibroblastic tumor (SCPFT) presents as a diagnostic conundrum due to its rarity and histologic similarity to other spindle cell neoplasms. SCPFT often presents as an inconspicuous, subcutaneous nodule with a predilection for the lower extremities in patients aged 20-40 years old. Herein, we describe a rare case of an SCPFT in a 61-year-old male who presented to the dermatology clinic for evaluation of a growth on his right neck that had been present for 5 years. On clinical examination, he had a well-circumscribed 1-cm mobile, subcutaneous nodule, suspicious for a lipoma. An excision was performed, and the specimen was sent for dermatopathology analysis which revealed a subcutaneous tumor composed of large, epithelioid to spindled, markedly pleomorphic cells with intranuclear pseudoinclusions and some with prominent nucleoli. Intermixed were large, atypical cells with ample bubbly cytoplasm, many of which contained hemosiderin, reminiscent of xanthomatous cells. Immunohistochemistry staining revealed strong and diffusely positive staining for CD34, vimentin, and CD10 with weak positivity for CD45 and CD68. Staining was negative for S100, PRAME, MART1, SOX10, AE1/3, desmin, STAT6, and p63. Given these findings, a diagnosis of SCPFT was made. SCPFT is a newly described entity in the family of CD34-positive spindle cell neoplasms and can be included in the differential for other pleomorphic soft tissue neoplasms. Given the low risk for local recurrence and metastasis of this tumor, complete surgical excision is mainstay. Our patient underwent successful surgical excision with negative margins and had no recurrence after one year.

INTRODUCTION

A superficial CD34+ fibroblastic tumor (SCPFT) is a rare, recently recognized mesenchymal neoplasm characterized by its clinical appearance and histologic pleomorphism. SCPFT often presents as a subcutaneous nodule with a predilection for the lower extremities in middle-aged individuals.[1] Due to its subtle clinical presentation and significant histologic overlap with both benign and malignant spindle cell tumors, SCPFT remains a diagnostic conundrum. Awareness of its key distinguishing histopathologic and immunohistochemical features is essential to avoid misdiagnosis, particularly in patients with a prior history of malignancy. We present a case of SCPFT arising in the cervical region in a patient with prior melanoma, emphasizing the need for a broad differential

diagnosis in subcutaneous spindle cell lesions.

CASE REPORT

A 61-year-old man with a history of melanoma in situ in his right axilla presented to the dermatology clinic with a subcutaneous growth on the right lateral neck. The lesion had been slowly enlarging over approximately five years and was asymptomatic. On physical examination, it appeared as a well-circumscribed, mobile, subcutaneous nodule measuring 1 cm. Given its clinical appearance, the lesion was initially suspected to be a lipoma, and the patient proceeded with surgical excision for definitive diagnosis and management.

Histopathologic examination of the excised specimen revealed a well-demarcated subcutaneous tumor composed of large epithelioid to spindled cells with marked nuclear pleomorphism, occasional intranuclear pseudoinclusions, and some with prominent nucleoli. Interspersed among these were large atypical cells with abundant

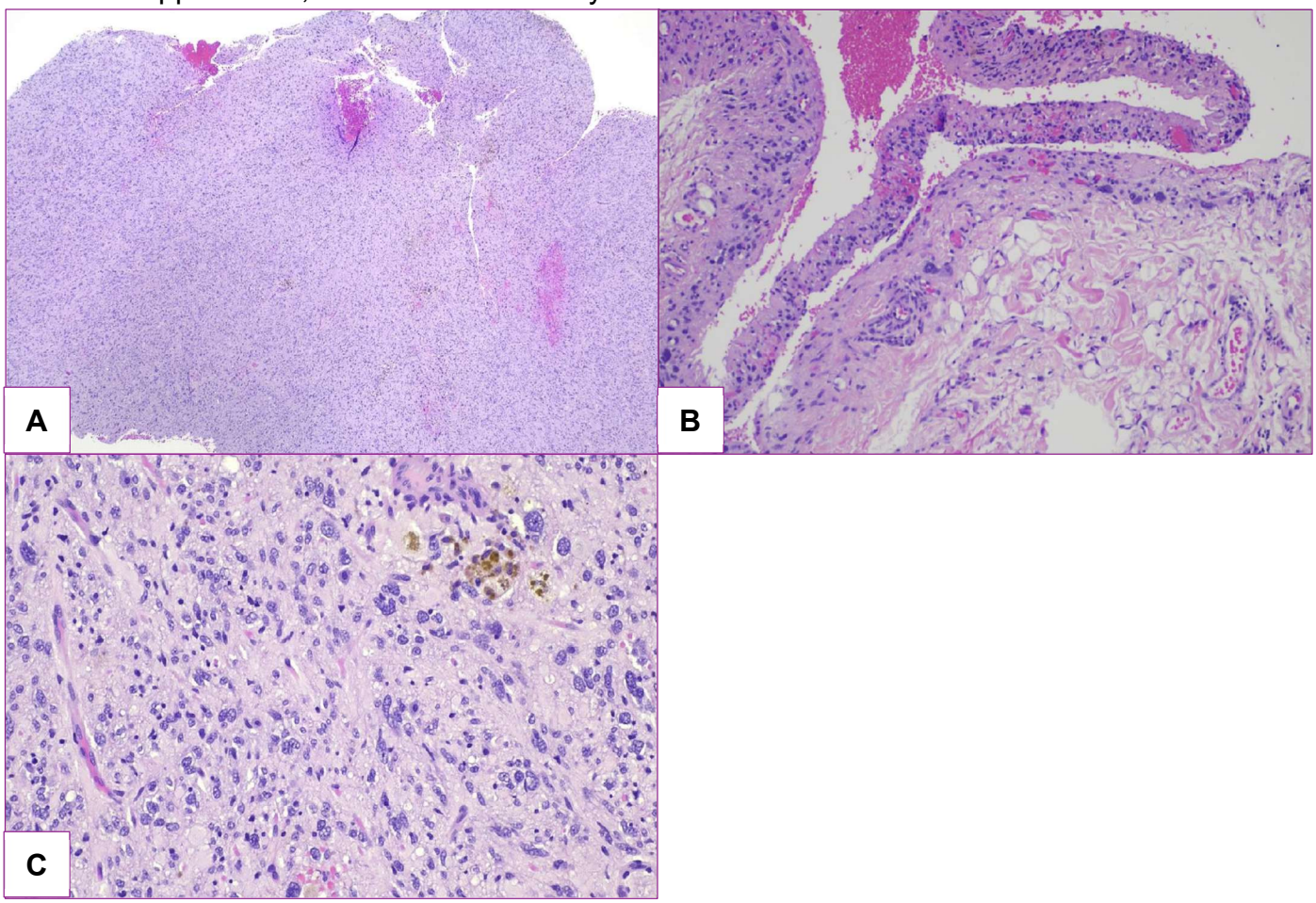

Figure 1A-C. Large, epithelioid to spindled, markedly pleomorphic cells with intranuclear pseudoinclusions and some with prominent nucleoli. Intermixed were large, atypical cells with ample bubbly cytoplasm, many of which contained hemosiderin, reminiscent of xanthomatous cells.

vacuolated cytoplasm containing hemosiderin pigment, morphologically reminiscent of xanthomatous cells (**Figure 1A-C**). No connection to the overlying epidermis or dermis was identified.

Immunohistochemical staining showed strong, diffuse positivity for CD34, vimentin, and CD10 with weak positivity for CD45 and CD68. The tumor cells were negative for S100, PRAME, MART1, SOX10, AE1/3, desmin, STAT6, and p63 (**Figure 2A-H**). This immunoprofile, in combination with the characteristic histologic features, supported the diagnosis of SCPFT, a rare low-grade mesenchymal neoplasm.

and no recurrence was observed at the one-year follow-up visit. Given the patient's prior melanoma history and the unusual location of the tumor, this case highlights the importance of histologic and immunophenotypic evaluation in distinguishing SCPFT from more aggressive cutaneous or metastatic entities.

DISCUSSION

SCPFT is a newly described entity in the family of CD34-positive spindle cell neoplasms with a limited number of cases reported in the literature.[2] SCPFT was

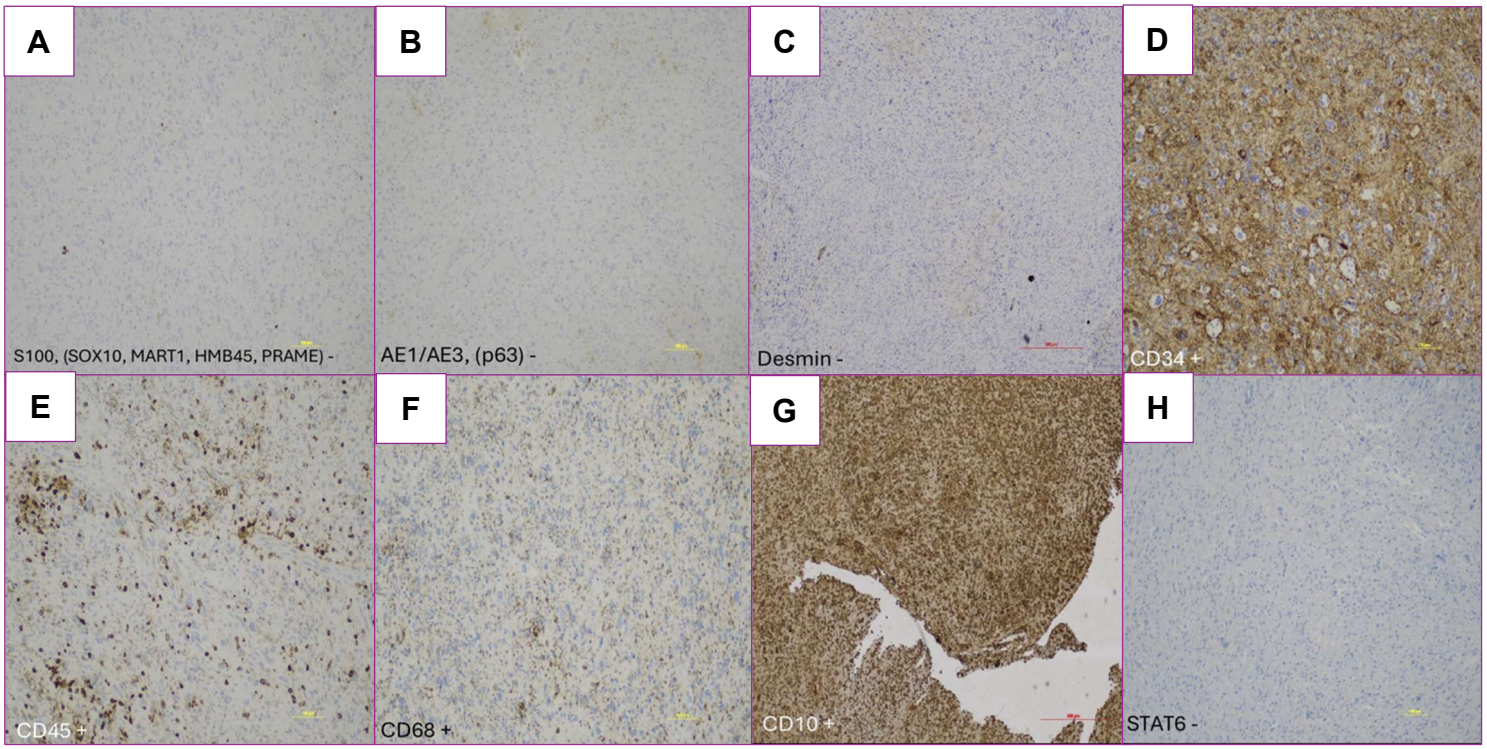

Figure 2A-H. Immunohistochemistry staining revealed strong and diffusely positive staining for CD34 and CD10 with weak positivity for CD45 and CD68. The tumor cells were negative for S100, PRAME, MART1, SOX10, AE1/3, desmin, STAT6, and p63.

SCPFT generally demonstrates indolent behavior with minimal risk for local recurrence or metastasis. Complete surgical excision is considered curative. In this case, the lesion was removed with clear margins,

formally recognized for the first time as a distinct entity of intermediate malignancy in the 2020 World Health Organization classification of soft tissues and bone tumors, following earlier clinicopathologic studies that

characterized the tumor as a low-grade mesenchymal neoplasm of intermediate malignancy.[3] Thorough clinical examination is essential in accurate diagnosis of this tumor. The neoplasm typically has a predilection for the lower extremities; however, cases have been reported in the trunk, upper limbs, and head region. Few reports have described SCPFT arising in the cervical region. Our case highlights the importance of maintaining a high index of suspicion for this neoplasm in atypical anatomic sites.

SCPFT shares morphological features with many benign and malignant soft tissue tumors histologically, which often makes the diagnosis challenging. Most cases exhibit a sheet-like infiltrate of spindle cells with abundant eosinophilic cytoplasm and nuclear pleomorphism, characteristics that are also seen in other mesenchymal neoplasms like myxofibrosarcoma, malignant peripheral nerve sheath tumors, and liposarcomas.[2,4] Unlike SCPFT, many of these other mesenchymal neoplasms lack CD34 immunoreactivity, which can serve as a useful distinguishing feature in the differential diagnosis. Recent studies have proposed that PRDM10-rearranged soft tissue tumors may represent the same entity as SCPFT. These tumors are typically characterized by a vesicular cytoplasm, low mitotic activity, and strong CD34 immunoreactivity, further supporting their overlap with SCPFT. However, some variants can be less well-defined and exhibit pleomorphic nuclei within a background of fibrous and myxoid stroma. A notable inflammatory infiltrate is also typically present in these cases, helping distinguish this entity from SCPFT.[2] A broad differential is especially critical in patients with a known history of melanoma, as in our patient.

Immunohistochemical staining is essential in distinguishing SCPFT from morphologic mimickers. While there is slight variability in the staining sensitivity of SCPFTs in the literature, the tumor consistently expresses diffuse and strong CD34, with focal positive staining of cytokeratin, which is seen in over 70% of cases.[2,5] However, cytokeratin expression is variable, and complete negativity for AE1/3, as observed in the present case, has been reported and does not exclude the diagnosis.[6] Vascular endothelial markers like CD31 and FLI-1, myogenic markers like SMA and desmin, as well as several melanin markers (S-100, HMB45, Melan-A) and SOX10 are typically negative.[5,7]

The preferred management of SCPFT is complete surgical excision with histologically confirmed negative margins. This approach is supported by the tumor's typically indolent clinical course and the low risk for local recurrence and metastasis.[3] Given the tumor's low mitotic rate, minimal cytologic atypia beyond its pleomorphic appearance, and intermediate malignant potential, there is no current role for adjuvant therapies like radiation or chemotherapy. Nevertheless, due to the relative novelty of this entity, close clinical follow-up is advised.

This case contributes to the expanding clinicopathologic spectrum of SCPFT, illustrating an uncommon cervical presentation in a patient with a prior history of melanoma, which broadened the initial diagnostic differential but is not known to contribute to SCPFT pathogenesis. Accurate diagnosis relied on histologic features and a focused immunohistochemical profile, most notably strong diffuse CD34 positivity and negative staining for melanocytic markers, allowing for distinction from more aggressive neoplasms, including metastatic melanoma. The tumor was successfully treated with a

complete surgical excision with negative margins and has not had a recurrence to date, consistent with the indolent behavior reported in the literature. Although SCPFT is now recognized as a distinct entity, further study of PRDM10 rearrangements in tumors with overlapping histologic features may refine diagnostic classification and elucidate its distinctive biology.

Conflict of Interest Disclosures: None

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