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Solitary fibrous tumor presenting on the scalp: a potential diagnostic pitfall

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1 | INTRODUCTION

Solitary fibrous tumor (SFT) is a rare spindle cell mesenchymal tumor originally described as a pleural tumor in 1931.¹ Since then, extrapleural locations have been well documented in the skin, soft tissue, central nervous system, liver, kidney and thyroid.² Roughly 40% of cases of extrapleural SFTs occur in subcutaneous tissue, with a predilection for the head and neck region.^{3,4} SFT occurring in the scalp is rare, with limited cases reported in the literature.^{4–10} This rare tumor presents a diagnostic challenge due to clinical and histopathologic similarities with other soft tissue tumors, such as dermatofibrosarcoma protuberans (DFSP) and spindle cell lipoma (SCL). We present a case of SFT on the scalp, and highlight the histopathological and immunohistochemical findings to support the diagnosis of SFT.

2 | CASE REPORT

A 37 year old healthy female presented with a mass on the scalp for 6 months. The mass was a soft, smooth, painless cyst-like growth on the mid-occipital scalp that was occasionally pruritic. She denied prior trauma to the area or other similar lesions. Prior to presentation, the patient had a biopsy of the mass performed by an outside dermatologist, diagnosed as a DFSP. Physical examination revealed a soft, mobile pink cystic biopsy site within an ill-defined subtle skin colored plaque encompassing 2 × 2.5 cm (Figure 1). There was no palpable head and neck lymphadenopathy. The outside biopsy was reviewed at our institution and additional immunohistochemistry and cytogenetic studies were performed. Histopathology showed a spindle cell tumor arranged in a patternless architecture (Figure 2), with alternating hypo- and hypercellular areas (Figure 3). The tumor cells displayed diffuse CD34 expression (Figure 4). The tumor cells were negative for S100, Melan A, and Sox10.

Additional evaluation showed immunohistochemical expression of STAT6 by tumor cells (Figure 5) and absence of *PDGFB* rearrangement by fluorescence in situ hybridization. Excision was performed over subsequent days until negative margins were achieved. Excision pathology showed deep infiltration of the galea and irregular tumor borders, requiring further removal of the periosteal layer. The final defect measured 4.5 cm × 3 cm and was subsequently closed with sutures.

3 | DISCUSSION

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm previously described in various organs of the body including the skin. The scalp is a rare location; previously reported cases are summarized in Table 1. These tumors varied in size, and of those with available information, all were treated with excision, with some demonstrating involvement of bone and periosteum. A common histopathological characteristic of SFT is the alternating hypo- and hypercellular areas, also seen in our case. In addition, all cases of SFT demonstrated positive CD34 expression.

SFT presents a challenging diagnosis due to its clinically and histopathologically similar appearance to other soft tissue tumors, such as dermatofibrosarcoma protuberans (DFSP) and spindle cell lipoma (SCL). SFT, DFSP, and SCL are all CD34+ tumors that can be difficult to distinguish. Our patient was initially diagnosed with a DFSP, for which she presented to our clinic for surgical management. While the most common location of DFSP is the trunk, a recent report found that 61% of deep, or subcutaneous DFSPs are located on the head.¹¹ On review of the patient's biopsy specimen, the spindle cells were seen in an alternating hypo- and hypercellular pattern, which raised suspicion for an SFT. The spindle cells of DFSP typically display storiform architecture, while in SCL they classically exhibit a "school of fish" pattern and are admixed with ropy collagen and mature adipose tissue. Additional molecular studies for this patient, including analysis of *PDGFB* rearrangement by fluorescence in situ hybridization, was negative, making DFSP less likely.

While histological patterns are important, given a small biopsy sample, as in our case, it can be difficult to clearly differentiate between spindle cell tumors. The detection of a NAB2-STAT6 fusion gene in SFT led to the reliable distinction of SFTs from other CD34+ spindle cell tumors. NAB2 is a transcriptional repressor of the early growth response (EGR) transcription factor, and its fusion with STAT6 converts the repressor into a transcriptional activator, thus driving neoplastic progression.^{12,13} This finding led to the identification of STAT6 as a highly sensitive and specific immunohistochemical marker for SFT, whereas DFSP and SCL are both negative for STAT6.³

The differential diagnosis for a subcutaneous soft tissue tumor of the scalp is broad, and clinical and histopathologic features alone may not be sufficient for diagnosis. The discovery of a specific fusion gene has been instrumental in allowing accurate diagnosis and counseling of patients with SFT. Until recently, cases of SFT of the scalp relied on histopathological features and non-specific immunohistochemical markers. The present case demonstrates the utility of molecular markers in making the accurate diagnosis. The majority of SFTs are at low risk of metastasis and surgical excision provides complete cure.

However, there is difficulty in predicting the behavior of individual tumors, which has important implications when counseling patients. Demicco et al. developed a risk stratification model to predict cases at higher risk for metastasis. The first model included age, tumor size, and mitotic figures.¹⁴ This model was validated in a recent study, and tumor necrosis was added as a fourth variable, which increased its predictive power. In this four-variable model, no metastases were seen in low-risk cases, moderate-risk cases had a 7% risk of metastasis at 5 and 10 years, and high-risk cases had a 49% risk of metastasis at 5 years.¹⁵ Complete surgical excision remains the recommended treatment for these tumors. Positive margins are common in head and neck SFTs, in which approximately 25–36% locally recur; therefore, long term followup is also advised.^{14,16}

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FIGURE 1.
Clinical presentation. Soft subcutaneous plaque on the mid-occipital scalp

FIGURE 2.
Hematoxylin-eosin stain, 50× magnification. Excision specimen showing a spindle cell tumor arranged in a patternless architecture

FIGURE 3.
Hematoxylin-eosin stain, 200× magnification. Spindle cells with alternating hypo- and hypercellular areas

FIGURE 4.
CD34 immunostain diffusely highlights tumor cells, 200× magnification

FIGURE 5.
Immunohistochemistry for STAT6 shows nuclear expression in tumor cells, 200×
magnification

Summary of cases of SFT on the scalp in the literature, including the case presented in this report

TABLE 1

Case number	Authors, Year	Age/Sex	Size (cm)	Treatment	Follow-up	Histopathological Features	Immunohistochemical Findings
1	Okamura et al, 1997 ⁵	37/F	N/A	Local excision	No recurrence in 10 months	Interlacing fascicles of uniform spindle cells, hypocellular areas with hyalinized collagen, areas of hemangiopericytoma-like pattern.	(+): CD34, vimentin, collagen IV (-): CD68, desmin, actin, SMA, cytokeratin, EMA, S-100
2	Cowper et al, 1999 ⁶	38/M	4	Wide excision	No recurrence in 6 months	Well-circumscribed biphasic spindle cell and vascular neoplasm, superficial portions comprised of bland spindle cells in storiform pattern, deeper portions with "staghorn" vascular channels in hemangiopericytoma-like pattern.	(+): CD34, vimentin (-): cytokeratin, EMA, SMA, desmin, factor XIIIa, S-100
3	Ramdial et al, 2001 ⁷	2.5/M	15.5	Wide excision with bone and dura	No recurrence in 18 months	Alternating hyper- and hypocellular areas arranged in various patterns, including haphazardly, storiform pattern, and hemangiopericytoma-like foci.	(+): CD34 (-): desmin, S-100
4	Erdag et al, 2007 ⁴	58/M	N/A	Local excision	No recurrence, length of follow-up not specified	Circumscribed, nodular growth pattern, spindle cells with cellular areas, hyalinized stroma, 1 mitosis per 10 HPF fields.	Tested markers (does not specify which were positive in scalp case): CD34, CD99, bcl-2, SMA, factor XIIIa. (-): keratin, EMA, desmin, S-100
5	Rizk et al, 2013 ⁸	2/M	N/A	Excision with periosteum	No recurrence in 12 months	Fusiform cell proliferation of variable cellularity with a central hyalinization zone.	(+): CD34 (-): SMA, desmin, h-caldesmon, S-100
6	Omori et al, 2014 ⁹	64/M	5	Wide excision	No recurrence in 3 months	Spheroid, well-circumscribed tumor with alternating hypercellular and fibrous hypocellular areas. Spindle-shaped cells without atypical nuclei in short interlacing fascicles, mixed with small vessels and interstitial fibrous tissue.	(+): CD34, bcl-2, CD99 (-): EMA, SMA, desmin, S-100
7	Feasel et al, 2018 ¹⁰	81/F	N/A	N/A	N/A	Classic patternless architecture and hemangiopericytic vasculature 8,11, cellular type with irregular fascicles of spindled cells with staghorn-like vessels and variable collagen 7,9,10, myxoid 10.	(+): CD34 7-11, STAT6 (8-10, 11 [*]) (-): S-100, cytokeratins (does not specify if negative cases were on scalp or elsewhere)
8		55/F	1.8				
9		64/F	2				
10		31/F	3				
11		35/F	N/A				
12	Our case	37/F	2.5	Staged excision	No recurrence in 5 months	Spindle cell tumor with alternating hypo- and hypercellular areas.	(+): CD34, STAT6 (-): S-100, Melan-A, Sox10

M, male; F, female; EMA, epithelial membrane antigen; SMA, smooth muscle actin; HPF, high-power microscopic fields

^{*} NAB2-STAT6 rearrangement was identified by next-generation sequencing.