



Superficial solitary fibrous tumor masquerading as a dermoid cyst: a case report

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Background: Solitary fibrous tumors (SFTs) are relatively rare spindle cell neoplasms uncommonly seen in dermatology practice. Initially discovered as a pleural tumor, SFTs have also been found in extra-pleural sites including the skin and soft tissues. When arising within the dermis or subcutis they are termed superficial SFTs, where they often present as solitary, unilateral, slow growing superficial masses. Histologically, they are composed of spindle cells arranged in a “patternless” pattern with hemangiopericytoma-like vessels dispersed throughout. Historically, CD34, CD99 and Bcl-2 immunohistochemical (IHC) stains were used to differentiate SFTs from other spindle cell neoplasms, however these markers are not entirely specific. Recent discovery of a disease defining NGFI-A binding protein 2 (*NAB2*)-signal transducer and activator of transcription 6 (*STAT6*) fusion gene has led to the use of STAT6 IHC staining to help verify the diagnosis of SFTs, particularly in unexpected sites.

Case Description: We report a case of a 23-year-old woman with a slowly growing lateral supra-orbital mass, clinically concerning for a dermoid cyst, which was subsequently discovered to be a SFT on pathologic examination, with the diagnosis being verified by STAT6 immunostaining.

Conclusions: SFTs are rarely encountered in dermatologic practice, however, must be kept on the differential of subcutaneous nodules, including those occurring in young adults. Due to the rarity of these tumors in clinical practice, a proposed algorithm for the approach to management of SFTs is included, guided by a validated, histology-driven, metastatic risk assessment tool, to help guide other clinicians confronted by these tumors.

Keywords: Solitary fibrous tumor (SFT); STAT6; case report

Received: 22 March 2022; Accepted: 11 July 2022; Published: 30 October 2022.

doi: 10.21037/acr-22-17

View this article at: <https://dx.doi.org/10.21037/acr-22-17>

Introduction

Solitary fibrous tumors (SFTs) are relatively rare mesenchymal neoplasms uncommonly encountered in dermatology practice. Originally described as a pleural tumor, SFTs in extra-pulmonary sites have become increasingly recognized (1,2). However, superficial SFTs (“cutaneous SFTs”) arising within the skin and soft tissue

remain uncommon, with only case reports and limited case series noted in the literature to date (1). Periorbital superficial SFTs, in particular, are amongst the rarest presentations documented (1). Clinically, superficial SFTs manifest as solitary, unilateral, slow growing, non-descript superficial masses in patients above the age of 55 years, with symptoms primarily related to mass effect (1-3). Histologically, SFTs are

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Figure 1 Solitary fibrous tumor: clinical findings. Firm subcutaneous nodule (arrow) overlying the right lateral supraorbital rim. This image is published with the patient's consent.

composed of spindle cells arranged in a “patternless” growth pattern, with a rich hemangiopericytoma-like vascular component (also referred to as “staghorn” vessels) (1,4). The “molecular hallmark” of SFTs is the NGFI-A binding protein 2 (*NAB2*)-signal transducer and activator of transcription 6 (*STAT6*) fusion gene, present in the vast majority of cases (1,5,6). A high index of suspicion in the appropriate clinical context with special attention to clinicopathologic correlation is critical to accurate diagnosis of this rare entity.

We present the case of a 23-year-old woman with a slowly growing lateral supra-orbital mass, clinically concerning for a dermoid cyst, discovered to be a superficial SFT on pathologic examination. Her age and tumor location represent unique SFT manifestations in the literature, and *STAT6* staining played a critical role in the diagnostic assessment of the lesion, further supporting the role of this immunohistochemical (IHC) stain in evaluation of these lesions. We present the following case in accordance with the CARE reporting checklist (available at <https://acr.amegroups.com/article/view/10.21037/acr-22-17/rc>).

Case presentation

A 23-year-old, African American female presented to the dermatology clinic with concerns for a slowly enlarging nodule overlying the lateral aspect of her right eyebrow. The lesion had been present for many years prior to

presentation and was causing her no discomfort. Desire for removal was based on cosmesis. Exam revealed a flesh-colored, soft, rubbery, mobile mass with no overlying skin changes, measuring 2.2 cm in diameter, overlying her right lateral supra-orbital rim (*Figure 1*).

Due to clinical concern for an underlying dermoid cyst, imaging was performed to rule out connection to the central nervous system. Computed tomography showed a well-circumscribed subcutaneous nodule without connection to underlying structures; additionally, no pathologic regional lymph nodes were noted. Surgical excision was subsequently performed, and the nodule was sent for pathologic evaluation.

Histologic sections revealed a spindle cell neoplasm arranged in a “patternless” pattern, with hemangiopericytoma-like vessels dispersed throughout (*Figure 2*). The lesion stained diffusely positive for Bcl-2 and CD34, with patchy weak staining for NKIC3, CD68, and Factor XIIIa, as well as a low Ki-67 proliferation rate. Additional IHC staining for *STAT6* was obtained and showed diffuse nuclear positivity, confirming the diagnosis of a superficial SFT (*Figure 3*). Risk-stratification classified the patient's tumor as low risk for metastasis (*Table 1*) (7). Complete physical examination and review of systems was unremarkable at post-surgical follow-up and subsequent encounters, with no evidence of recurrence at one year follow-up.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

SFTs are uncommon spindle cell neoplasms of fibroblastic/myofibroblastic lineage originally described as arising within the pleural cavity (1,2). Extra-pleural SFTs have been subsequently reported in various other locations, including in the skin and its immediate subcutis, where they are termed superficial SFTs (1,8). Case series suggest that superficial SFTs most commonly present on the head and neck of middle-aged females (1). Typically, they are removed for cosmetic reasons, often being mistaken clinically for a benign adnexal neoplasm or cyst, as was the case in our patient. Radiological imaging is generally non-specific,

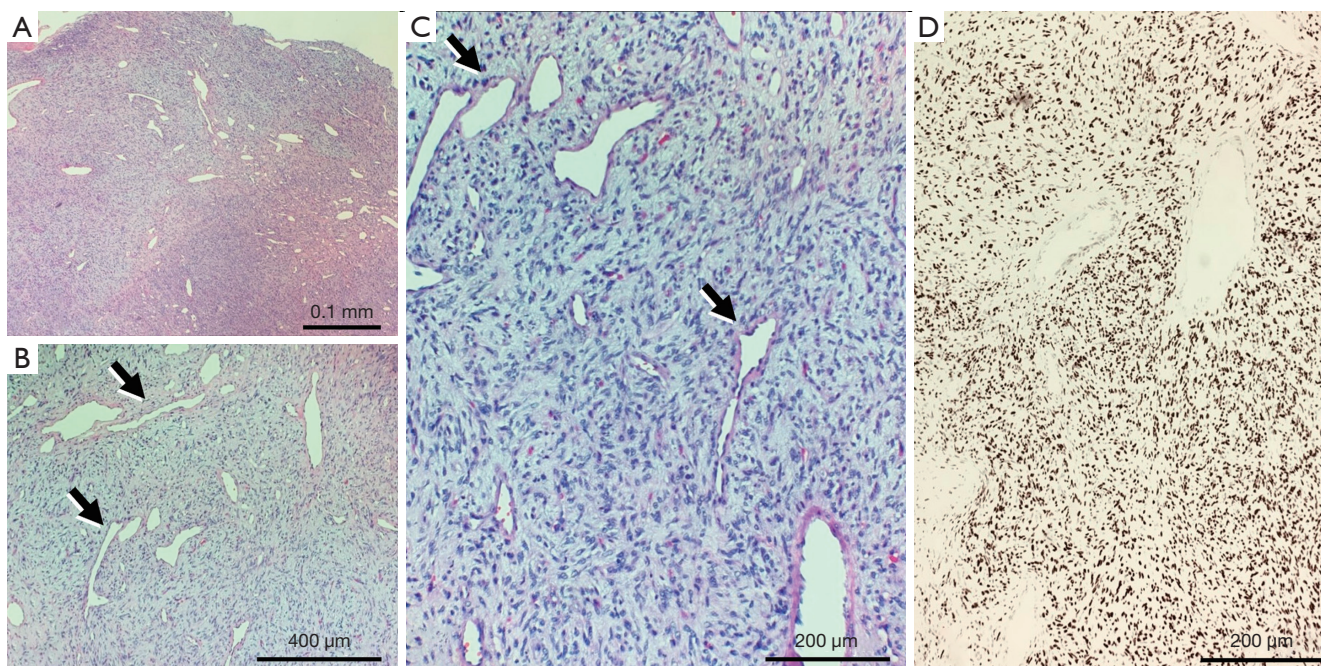


Figure 2 Solitary fibrous tumor: histologic findings. Dermal neoplasm consisting of spindle cells arranged in a “patternless” pattern featuring hemangiopericytoma-like vessels (arrows) dispersed throughout shown at both low (A,B) and high power (C) (hematoxylin and eosin, H&E, $\times 2.5$, $\times 10$, and $\times 20$). The spindle cell population exhibits diffuse nuclear STAT6 positivity (D) (STAT6 immunohistochemistry $\times 20$). Note that the vascular endothelial cells do not stain positive for STAT6 (D). STAT6, signal transducer and activator of transcription 6.

showing a solid nodular mass with well circumscribed borders—making the diagnosis of SFT largely one based on histology (2). Our case aligns with previous reports of superficial SFTs in several aspects including female gender, head/neck location, non-specific imaging findings, and slow growth over several years; however, several aspects are unique. Our patient reported onset in childhood and presented with a lateral supraorbital location, a presentation that may lead to the misdiagnosis of a dermoid cyst. A distinguishing feature that can be elicited by history would be the exact time of onset, with majority of dermoid cysts apparent in early childhood, whereas our case presented in later childhood.

On histologic analysis SFTs are primarily comprised of spindled cells in a “patternless” pattern admixed within a variable amount of thick keloidal collagen and prominent hemangiopericytoma-like vessels (1,8). The differential diagnosis of spindle cell neoplasms is broad, including both benign and malignant entities, and has been previously reviewed (1,6,9). Differentiating superficial SFTs from more aggressive spindle cell neoplasms (e.g.,

dermatofibrosarcoma protuberans, malignant peripheral nerve sheath tumor) may be challenging and IHC staining is often required for a definitive diagnosis. Historically, the combination of typical morphology with IHC staining for CD34, CD99, and Bcl-2 was required for the diagnosis of SFT (6). Limitations to specificity were acknowledged with this diagnostic profile, though, and the recent discovery that SFTs uniformly possess a *NAB2-STAT6* fusion gene which correlates with STAT6 IHC staining has revolutionized the diagnostic assessment of these tumors—especially in clinical settings where this entity is unexpected and other spindle cell tumors may be more common (2,4-6,10). Due to these findings, we suggest that STAT6 be added to the panel of immunoperoxidase stains utilized by pathologists when confronted with a spindle cell neoplasm of unknown lineage.

Due to the low incidence of disease, there is limited data to guide prognostic discussions and treatment recommendations for these tumors. Considered tumors of intermediate biologic potential, SFTs have shown the potential to recur and metastasize (1). As such, complete

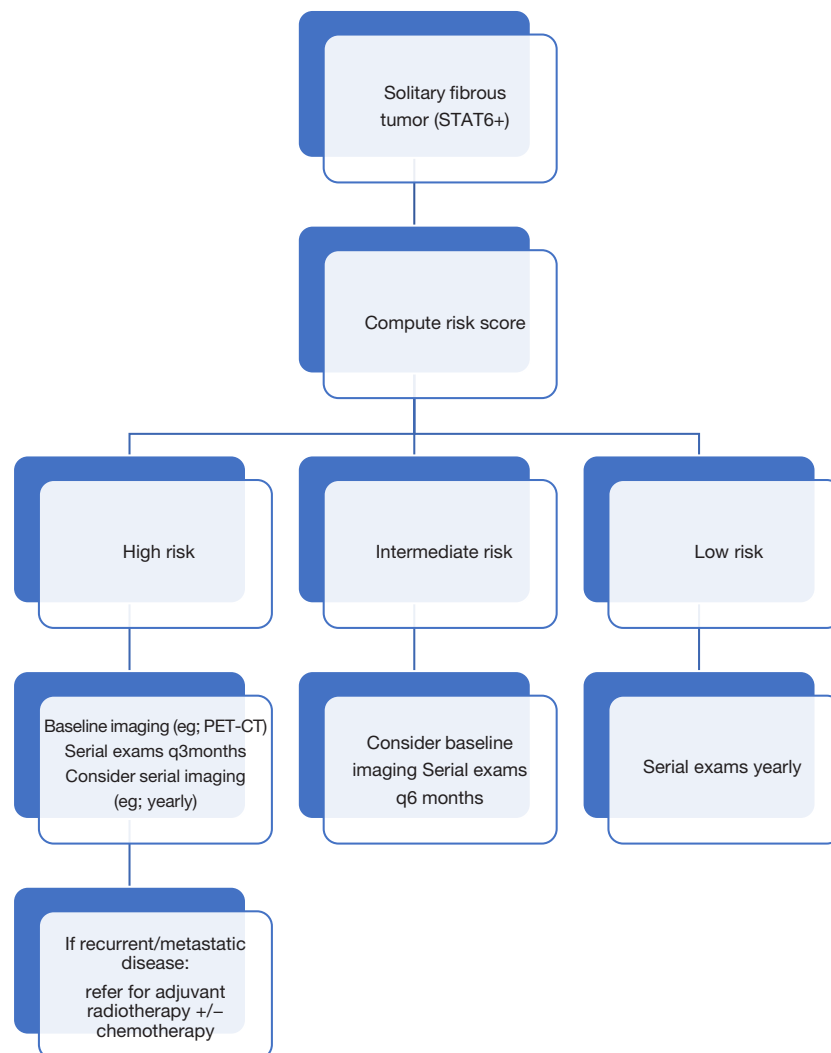


Figure 3 Proposed approach to management of superficial solitary fibrous tumor. Complete removal of tumor is advised following pathologic diagnosis if partial biopsy is obtained. SFT risk score is calculated via the model proposed by Demicco *et al.* (see *Table 1*). Serial examinations should include at a minimum: evaluation of the primary excision site, regional lymphatic examination, and review of systems. Further studies (e.g., imaging, laboratory evaluation) are guided by examination findings as well as the review of systems. If at any time metastatic or recurrent disease is suspected, prompt referral to radiation and/or medical oncology for management is recommended. STAT6, signal transducer and activator of transcription 6; PET, positron emission tomography; CT, computed tomography; SFT, solitary fibrous tumor.

resection is generally recommended (1,3). Histology is a helpful but imperfect method of predicting biologic behavior, with increasing tumor size, patient age, mitotic rate, and presence of necrosis correlating with high-risk tumors (*Table 1*) (7,10). Location of the tumor may also play a role in predicting behavior of SFTs (e.g., superficial versus deep soft tissue). The largest case series to date evaluating

specifically superficial SFTs, found no evidence of recurrence or metastasis in all cases, with a median follow-up time of 16 months (1). This suggests that superficial SFTs behave more indolently compared to their deeper counterparts; whether this is due to easier accessibility (e.g., for tumor extirpation prior to sustained growth) or other factors remains to be determined. Long term follow-up

Table 1 Modified four-variable risk stratification model for development of metastasis in solitary fibrous tumors (adapted with permission from Demicco *et al.* 2017) (7)

Risk factor	Score	Present case
Age (years)		
<55	0	0
≥55	1	
Tumor size (cm)		
<5	0	0
[5–10)	1	
[10–15)	2	
≥15	3	
Mitotic count (mitoses/10 high-power fields)		
0	0	0
1–3	1	
≥4	2	
Tumor necrosis		
<10%	0	0
≥10%	1	
Risk class	Total score	
Low	0–3	0
Intermediate	4–5	
High	6–7	

after complete excision is critical to monitor for recurrence and/or metastasis; a proposed approach to management of superficial SFTs is included (*Figure 3*). Importantly, our case showed no high-risk features and has not recurred nor metastasized at one year follow-up, further validating the risk-stratification model proposed by Demicco *et al.* (7).

In conclusion, we present the case of a young adult female with a slow growing supraorbital superficial SFT for which recognition of key histopathologic features and STAT6 staining played a critical role in diagnosis. The young age of onset and lateral supraorbital location were atypical for SFT leading to an initial misdiagnosis of dermoid cyst, confirming the importance of histologic analysis for these lesions. Understanding that this uncommon tumor can present in skin and soft tissues and has a unique IHC pattern is important for accurate and timely diagnosis.

Acknowledgments

Funding: None.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at <https://acr.amegroups.com/article/view/10.21037/acr-22-17/rc>

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <https://acr.amegroups.com/article/view/10.21037/acr-22-17/coif>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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doi: 10.21037/acr-22-17

Cite this article as: Foss MG, Dunn C, Marks E, Nathoo R. Superficial solitary fibrous tumor masquerading as a dermoid cyst: a case report. *AME Case Rep* 2022;6:34.