Extra-Pleural Dermal Solitary Fibrous Tumor on the Posterior Shoulder: A Case Report.

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ABSTRACT

- Mesenchymal in origin, solitary fibrous tumors (SFTs) are primarily seen within the pleura of the lung or in serosal-lined body cavities. Constituting 1-2% of all soft-tissue tumors, solitary fibrous tumors are rare entities, especially when found in extra-pleural locations. Diagnosis requires tissue sampling and staining for immunohistochemical markers. Management of these tumors is based on wide-local excision with histologically negative margins. If negative margins cannot be surgically achieved, adjuvant therapies including radiation have been described.
- In this report, we describe a 74-year-old male with an extra-pleural dermal solitary fibrous tumor. We present the clinical course, surgical procedure, histopathologic features, as well as discuss the treatment options. We also review the published literature reports of dermal solitary fibrous tumors.
- With extra-pleural manifestations of solitary fibrous tumors seldom reported in the literature, it is our hope that reporting these unusual instances will raise awareness of such disease manifestations and allow for earlier diagnosis and treatment.

INTRODUCTION

- SFTs are a spindle-cell neoplasm that is mesenchymal in origin
- Generally benign, they are well-circumscribed, mobile, and painless
- If malignant, the most common site of metastasis is the lungs [1]
- Most commonly found in pleural of lung and/or on serosal surfaces
- Associated symptoms can include hypoglycemia, arthralgia, osteoarthritis, and clubbing [1]
- Most commonly affect adults from 40 – 70 years old [1]
- Overall all prevalence low, representing 1-2% of all soft-tissue tumors with extra-pleural SFTs representing 0.6% of all soft tissue tumors [4]
- Fewer than 850 cases of extra-pleural SFTs have been described in the medical literature worldwide
- Diagnosis requires sampling and subsequent histopathologic analysis
  - Immunohistochemical (IHC) staining can be used to identify gene expression of *CD34, bcl-2, CD99,* and *STAT6* and are used as reliable surrogates for detection and diagnosis of SFTs [2]
  - Cytogenetic features concerning for metastatic disease include:
    - Diameter > 5 cm, significant pleomorphism, atypia, high cellularity, tumor cell necrosis, & > 4 mitotic figures /10 HPF [3]
- Work-up includes staging with computed tomography
- Benign tumors are managed with wide-local excision, while metastatic tumors may require radiation or anti-angiogenic medical therapies
  - 5 year-survival for benign tumors ranges from 89-100% [4]
- Because of malignant potential, SFTs require long-term surveillance

CASE REPORT

- 74-year-old male was evaluated by Dermatology for a mass on his shoulder
  - Located on posterior shoulder over right trapezius muscle
  - Present for last 3 years, but rapidly increasing in size in last 2 months
  - Painless and without any associated symptoms
  - 3 x 3 cm palpable, non-tender mobile mass noted
- Significant past medical history included actinic keratoses
- Incisional biopsy performed by Dermatology and pathology showed:
  - Spindle-cell tumor without necrosis, but up to 6 mitoses/10 HPF
  - IHC positivity for *CD34 & CD99*
  - Pathology reviewed at *Johns Hopkins University – Department of Pathology* and diagnosis confirmed with IHC positivity for *STAT6*

Figure 1. A representative routine H&E section at 20x magnification showing mitosis and low-grade atypia

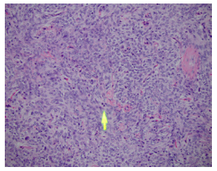
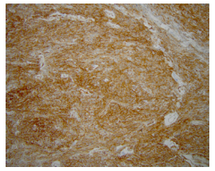


Figure 2. A representative IHC stain at 10x magnification showing positivity for *CD34*



- Patient evaluated by Surgical Oncology:
  - Well-healing incisional biopsy without satellitosis
  - CT chest, abdomen, and pelvis negative for metastatic disease
  - Referral to Multidisciplinary Cutaneous Oncology Clinic without recommendation for neoadjuvant therapy
- Wide-local full-thickness excision performed with 1 cm margins
  - Trapezius muscle taken for oncological boundary of safety
  - Defect reconstructed with local rotational-advancement flap
  - Permanent pathology revealed positive deep margins
- After discussion, patient elected to forgo secondary surgery and pursue radiation therapy with plans for 30 total treatments

Figure 3. A representative IHC stain at 10x magnification showing positivity for *STAT6*

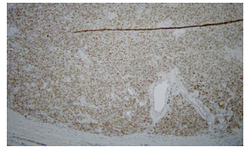
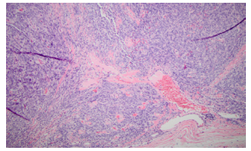


Figure 4. Rrepresentative routine H&E section at 10x magnification showing vessels & perivascular tumor growth



DISCUSSION

- Pathologic diagnosis of SFTs requires recognition of histologic features coupled with supportive positivity of IHC stains
- Similar to their pleural counter parts, this spindle-cell neoplasm showed evidence of characteristic gaping and bifurcating staghorn vessels (Figure 4) and alternating areas of hyper and hypocellularity (Figure 5)

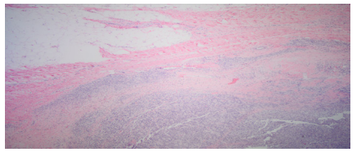


Figure 5. A representative H&E section at 4x magnification showing alternating areas of hypo and hypercellularity

- Hyalinization of vessels is a subtle feature to make the diagnosis
- Key IHC positive staining for *CD34* (Figure 2) is indicative of perivascular cells, the putative origin of SFTs [2, 3]
- *CD99* and *bcl-2* can also show variable staining, but these stains are not lineage specific [2]
- A recurrent paracentric inversion involving chromosome 12q13 has been identified in SFTs in the pleura and soft tissues that results in *NAB2-STAT6* gene fusion [7]
  - These fusion results in oncogenic overexpression of activation factor STAT6 that drives tumor proliferation
  - Identification of *STAT6* by IHC confirms the diagnosis (Figure 3)
- Reported recurrence and metastasis rates range from 10-37% [9]
- Failure to remove all tumor capsule by wide-local excision has a higher rate of recurrence, but can be minimized with adjuvant radiation therapy
- In this case, the patient was found to have positive margins, but elected to forgo additional surgery and to pursue adjuvant radiation therapy
- With extra-pleural SFTs seldom reported in the literature, it is our hope that presenting our experience will raise awareness of the disease and allow for earlier diagnosis and treatment of such tumors

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