Solitary Fibrous Tumor, Not Always the Pleura, Not Always Benign

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Introduction

Solitary fibrous tumors (SFTs) are fibrotic mesenchymal neoplasms including lesions historically known as localized mesothelioma, subserosal fibroma, and hemangiopericytoma. Immunohistochemical profiling, and detection of the NAB2-STAT6 fusion gene, enabled the integration of these labels into the same entity.

The incidence is ubiquitous at 1 case/million people/year, most commonly in the pleura, where they were initially described.

They preferentially emerge from serosal surfaces or from the dura mater. Only 10% arise from deep soft tissues.

The most common presenting clinical symptoms are chest pain, cough, and shortness of breath.

Early studies reported that 78-88% of SFTs were benign. However, recent data suggests that SFTs are unpredictable, and that existing methods for differentiating benign vs. malignant, poorly correlate with disease prognosis. However, the most aggressive subtype, the dedifferentiated SFT (DD-SFT), is consistently malignant and comprises less than 1 percent of SFTs.

Here we present an extrathoracic DD-SFT in an otherwise asymptomatic patient. We describe the histologic, immunohistochemical, and genetic features in order to increase awareness this rare aggressive subtype.

Case Summary

45-year-old male, no prior significant medical history presented with a rapidly growing non painful palpable exophytic right thigh mass. CT of the right extremity revealed a 8 x 7.4 x 7.0 cm heterogenous mass in the antero-medial lower thigh abutting but not invading the underlying muscle. He was referred to an oncologic surgeon with the presumptive diagnosis of sarcoma. A radical excision was performed.

Results

Histologic examination shows a biphasic zonal tumor with hypocellular and hypercellular areas, hemorrhage and necrosis (**Figure 1**). A magnified view of the hypocellular areas reveals short spindle cells with mild atypia, abundant collagenous stroma and prominent dilated blood vessels (**Figure 2**) consistent with a conventional Solitary Fibrous Tumor. The hypercellular areas **(Figure 3**) consisted of highly atypical spindle and polyhedral cells with numerous mitotic figures consistent with a high grade sarcoma. There was a sharp transition between the two distinct components (**Figure 4** upper half conventional SFT, bottom half High grade sarcoma. Immunohistochemistry shows the tumor cells are positive for STAT6 (**Figure 5**), BCL-2 (Figure 6) and CD34 only in the conventional SFT (**Figure 7**). Negative stains included pankeratin, EMA, CAM5.2, SOX-10, S-100, MART-1 and CD99. MDM2 FISH was negative. RNA sequencing studies detected a NAB2::STAT6 [inv (12;12)(12q13.3;12q13.3)] gene fusion.

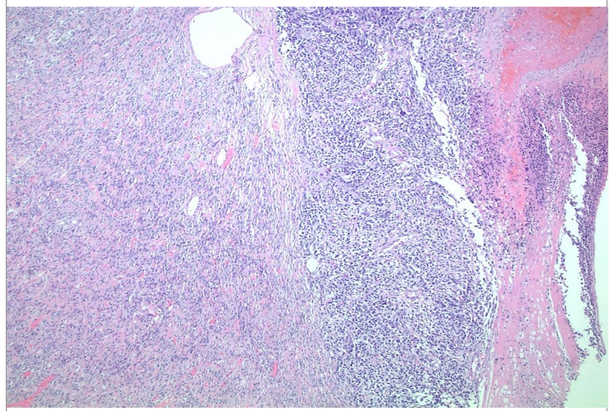

Figure 1: Biphasic tumor with hypocellular (left) and hypercellular (center) areas in addition to abundant hemorrhage and necrosis (right).

Results (Continued)

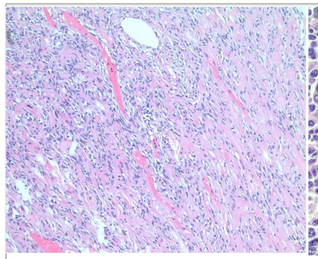

Figure 2: H&E 40x. Consistent with Conventional SFT.

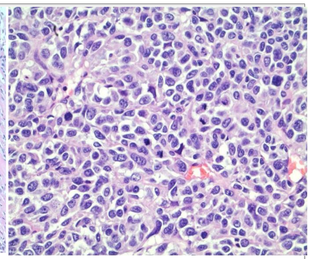

Figure 3: H&E 400x Consistent with high grade sarcoma/DD-SFT.

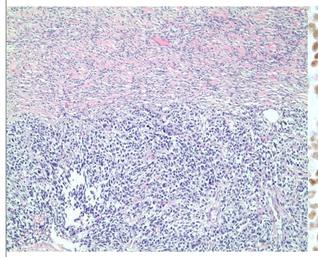

Figure 4: H&E 40x
Top: conventional SFT,
Bottom: DD-SFT.

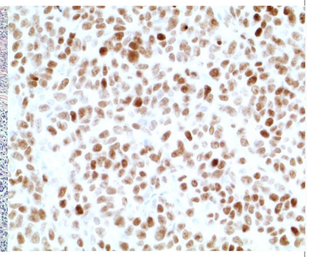

Figure 5: STAT6 400x
Positive (brown) staining

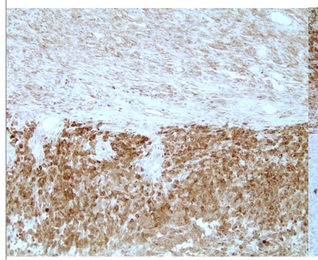

Figure 6: BCL-2 40x
Positive (brown) staining

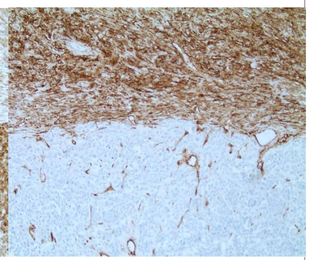

Figure 7: CD34 40x
Positive (brown) in conventional SFT (top half) component.

Discussion

The fusion of NAB2 to STAT6 replaces a repressor domain with a transactivation domain and correlates with high nuclear STAT6 staining in SFTs. Most morphologic variants, do not change the prognosis. However, the DD-SFT subtype does change the prognosis. This subtype is characterized by a sharp transition from conventional SFT morphology, to a zone resembling high-grade sarcoma and loss typical SFT staining. There are no molecular markers to predict an SFTs metastatic potential without also observing the distribution of those markers on histological images.

Conclusions

SFTs are rare neoplasms that were traditionally regarded as benign pleural lesions. However, they can arise anywhere.. They are difficult to predict and the recent incorporation of labels such as hemangiopericytoma has further complicated the benign/malignant categorization, but the subtype of DD-SFT is readily regarded as aggressive once it is correctly identified. Needle biopsies may suggest SFT, but full excision with complete histological with IHC evaluation is currently still needed to illuminate the subtype. SFTs express characteristic markers, but nuclear STAT6 is the most definitive. Once SFT is confirmed, correlating a loss of staining, with a transition to high grade sarcoma morphology can establish diagnosis of the DD-SFT.

References

To view the complete list of references, please point your phone's camera at the QR code to the right and click on the link that appears.