Navigating the Diagnosis and Management of Pleural Solitary Fibrous Tumors: A Case Study

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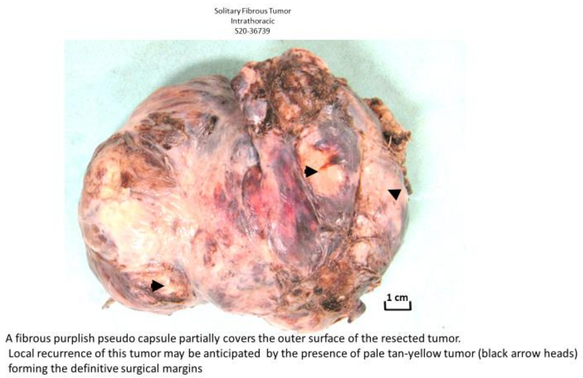
Introduction

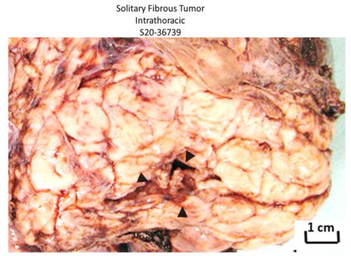
Solitary Fibrous Tumors (SFTs) are rare primary neoplasms of mesenchymal origin which can be found in various tissues, including the mediastinum, visceral and parietal pleura, peritoneum, and meninges. Pleural SFTs can present with non-specific pulmonary symptoms such as cough, shortness of breath, and chest pain. Diagnosis of these tumors is made by imaging and clinical features, but a biopsy with histological confirmation is necessary to guide management. MRI can help distinguish the relationship between the SFT and the surrounding structures, but overall, CT is a better diagnostic imaging modality.



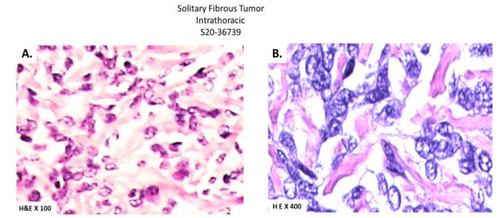
A fibrous purplish pseudo capsule partially covers the outer surface of the resected tumor. Local recurrence of this tumor may be anticipated by the presence of pale tan-yellow tumor (black arrow heads) forming the definitive surgical margins

Case Description

The patient is a 69-year-old female who presented with two weeks of exertional shortness of breath, dry cough, and nail clubbing. A CT chest revealed a moderate left sided pleural effusion, a collapsed consolidated lung, and a “huge” left lung mass, initially attributed to either a sarcoma or a pleural fibrous tumor. A thoracentesis and a follow-up bronchoscopy both failed to yield a diagnosis, but a subsequent CT guided IR lung biopsy showed spindle cell proliferation mixed with keloidal collagen, consistent with a solitary fibrous tumor.



On serial section, firm tan-yellow tumor nodules form the vast majority of this tumor. The aggressive nature of this tumor is grossly evidenced by its infiltration of adjacent normal tissue and zones of necrosis (black arrow heads).



A. Under low power the diffusely infiltrating bland-appearing tumor cells lie between abundantly-produced dense pink-red collagen. B. In high power views, although mitoses are rare, intermediate to significant pleomorphism characterizes the large vesicular tumor nuclei. These dysplastic changes are consistent with malignant potential and are most prominent in areas of necrosis.

A month later, the patient underwent excision of the approximately 25 cm thoracic mass, along with wedge resection of the left lower lung. Nine months later, on surveillance imaging recommended by our tumor board, the patient had metastatic disease, which was confirmed via lung nodule biopsy showing the spindle cell neoplasm. At our oncologist's behest, with the goal of pursuing palliative and survival benefit, the patient began chemotherapy with Dacarbazine and Doxorubicin, which provided short-lived improvement. This was followed by Panzopanib after a recent case series showed tumor burden benefit, yet the cancer progressed. She was transiently on Sunitinib and Temozolomide, also providing little benefit. Currently, the patient is on Trabectedin and is closely being followed by her medical oncologist.

Discussion

Currently, the standard of treatment for localized SFT is radical surgical removal, with or without radiotherapy. This case highlights the importance of early recognition and diagnosis of a solitary fibrous tumor in the pleural space, particularly as they can cause rapid respiratory decline. It also highlights the challenges of finding long-term systemic treatment options for recurrent metastatic disease that maintain remission with minimal side effects. A trial evaluating the efficacy of Panzopanib found that 58% of patients had a partial response, 39% had stable disease, and 3% experienced progressive disease at the 18-month follow up. This treatment response was also limited due to side effects. Furthermore, this study demonstrated that Trabectedin combined with either Ponatinib or Dasatinib had synergistic effects, suggesting two potential new treatment strategies for pleural SFT in the future. Further randomized controlled clinical trials and analytical research are needed to identify effective treatments for SFT.

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