A case of solitary fibrous tumor of the pleura: A Pleural Fibroma
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Background:
Solitary fibrous tumors (SFTs) refer to a histologic spectrum of fibroblastic mesenchymal neoplasm. Solitary fibrous tumors may arise in serosal membranes, dura of the meninges or deep soft tissue. Solitary fibrous tumor of the pleura, specifically, are a rare occurrence with only limited number of cases reported. Patients usually present with nonspecific symptoms including dyspnea, cough, chest pain. Some patients may be asymptomatic, and tumor may be discovered incidentally. We present a case of solitary fibrous tumor of the pleura.

Case Presentation:
A 61-year female with no history of smoking, lung disease or malignancy was found to have left sided lung mass after undergoing evaluation for digital clubbing. She had presented to her primary care doctor with complaints of left arm pain, abnormal shape of her fingers and 20-pound weight loss over the last three months. She denied any chest pain, dyspnea at rest or on exertion, orthopnea chronic cough, wheezing. She had no family history of chronic lung disease but has a strong family history of inflammatory bowel disease. On exam she was found to have significant clubbing and no other abnormality. Bloodwork revealed no abnormality with a normal complete blood count, comprehensive metabolic panel, and thyroid function tests. Imaging of the shoulder revealed supraspinatus tear with mass like structure in the lung. Computed Tomography (CT) of the chest without contrast revealed a left sided mass measuring 13.6cm x 8.6cm x 9.8cm without evidence of hilar lymphadenopathy or chest wall invasion (figure 1). She was seen by pulmonology who then ordered a PET-Scan which revealed a 13.6 cm heterogeneous mass in the left lung base abutting the left heart border with mild to moderate increase FDG uptake. Detailed discussion with radiology was done and they thought it could be benign or malignant possibly a pleural-based lesion. CT guided biopsy of the lesion done, was not adequate for diagnosis. Patient subsequently underwent bronchoscopy and video assisted thoracotomy and excision of tumor successfully. Pathology revealed solitary fibrous tumor with focus of infant type necrosis, without any atypia (figure 2). Patient tolerated the procedure well and had a full recovery.

Discussion:
Solitary fibrous tumors (SFTs) of the pleura are rare malignancies which make up less than 5% of all primary pleural tumors. The incidence rate is reported to be 2.8 cases per 100,000 population.1 Presentation is usually the 6th or 7th decade and there is no gender predilection. Patients may present with cough, chest pain or dyspnea however majority of cases are asymptomatic, and tumors are usually found incidentally on thoracic x-ray or imaging for some other reason. There have been case reports of patients presenting with pierre-marie-bamberger syndrome, also known as hypertrophic osteoarthropathy (HPO) characterized by clubbing of the fingers.2 HPO is typically seen in patients with bronchogenic carcinoma. Our patient also presented with clubbing of her hands which is what prompted further work up for underlying occult malignancy. Another rare presentation is the Dooge-Potter syndrome, which is characterized as a paraneoplastic syndrome with hyperinsulinemic hypoglycemia resulting from the ectopic secretion of a prohormone of insulin-like growth factor II from a solitary fibrous tumor. Histologically, the tumors are composed of irregularly arranged fascicles consisting of spindle cells separated by collagen and are thought to originate from submesothelial mesenchymal cells, with 80% arising from the visceral pleura. Majority are benign however, approximately 30% of solitary fibrous tumors of the pleura are malignant.3 Immunohistochemistry is beneficial to differentiate SFTs from mesotheliomas and other sarcomas. SFTs is usually vimentin positive and keratin negative. In addition, CD34 is positive in most benign and malignant SFTs, whereas it remains negative for most other pulmonary tumors.4 Radiologically SFTs appear as well-defined, homogeneous, and rounded mass. CT usually demonstrates a well-delineated, homogeneous, lobulated mass of soft tissue attenuation which typically appears to be in contact with the pleural surface. Complete surgical resection is usually curative for benign pleural SFTs with a local recurrence rate of 8%. 4

References: