Fibrous Tumor of the Pleura

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Our Patient: 39-year old asymptomatic female
Mass on Chest X-ray

Well-circumscribed, ~10cm mass, in the apex of the right upper lung lobe
Our Patient Presentation

• 39 year-old woman, asymptomatic, with no significant past medical history and no recent trauma.

• Differential includes pleural tumor, chronic empyema, schwannoma, lymphoma, infectious causes, Pancoast tumor, metastases.
Our Patient: C+ Chest CT
Mass on Axial Images

Pleural borders of the soft-tissue mass,
Enhanced vessels within the mass
Superior Vena Cava
Aortic Arch

The reconstructions that follow this axial image further confirm the location of the soft tissue mass, and portray its relation to lung fissures, structures of the mediastinum and the airways.
Our Patient: **Mass** on Sagittal and Coronal Chest CT

- **Horizontal fissure** between superior and middle lobe of the right lung
- **Oblique fissure** between middle and inferior lobe of the right lung

Branching of trachea into the right and left main bronchus at the **carina**

View of **left atrium** at entrance of pulmonary veins
A Brief Review of Lung Anatomy

Note the relation of the lung fissures and hilar contents to the lobes of the right and left lung.
A Brief Review of Pleural Anatomy

Note the divisions of the visceral and parietal pleura as they would appear on a frontal chest x-ray and on axial images from CT chest scan.
Benign pleural disorders

- Pleural disorders broadly divided into:
  - Effusions
  - Thickening
  - Masses
  - Pneumothorax
- Normal amount of pleural fluid in healthy people is 2-10ml
- Parietal pleura supplied by systemic capillary vessels - drains into right atrium via the azygous, hemiazygous, and internal mammary veins
- Visceral pleura supplied by pulmonary arterial capillaries and runs mainly into the pulmonary veins
- Lymphatic vessels play major role in clearance of pleural fluid - drainage mainly through parietal pleural lymphatics.
Fibrous Tumors of the Pleura

- Localized pleural tumors fall into one of two categories: fibrous tumors of the pleura or lipomas.
- Pathologic characteristics of solitary fibrous tumor of the pleura first described by Klemperer and Rabin in 1931.
- ~ 800 cases reported in the literature between 1931 and 2002 and has been referred to as localized mesothelioma, localized fibrous tumor, fibrous mesothelioma, or a pleural fibroma.
- Rare neoplasm: incidence is 2.8 per 100,000 registered hospital patients and accounts for 8% of benign pathologic diseases of chest.
Clinical Presentation of Fibrous Pleural Tumors

• Usually found incidentally by chest radiography
• Greatest occurrence in fourth to sixth decade, no hx of asbestos exposure, relatively equal gender distribution (some report slight female preponderance)
• Presenting symptoms (~50-60% are symptomatic): intrathoracic symptoms (dyspnea, chest pain, hemoptysis), systemic symptoms (hypoglycemia, hypertrophic osteoarthropathy), nonspecific symptoms (fever, weight loss, fatigue)
• Most behave as slowly growing, painless masses although large tumors may also give rise to compression symptoms.

• Associated hypertrophic pulmonary osteoarthropathy and episodic hypoglycemia (due to production of insulin-like growth factor) may be present in 4-5% of cases.

• 80% arise from visceral pleura and 20% from parietal pleura
• Calcification present in ≤5%, central necrosis is common in the larger tumors.
• Behavior is unpredictable with ~10-15% behaving aggressively so follow-up is mandatory
Representative Pathology Specimen

- Well-circumscribed
- Often pedunculated
- Remarkable blood supply

- Treatment of choice is complete surgical resection
- No established systemic therapy for malignant form

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http://www.emedicine.com/RADIO/topic448.htm
Imaging features of Pleural Fibroma on CXR

- Solitary round or ovoid mass that has **sharp interface** with lung parenchyma and is **lacking pleural effusion** in most cases
- Depending on location may be difficult to differentiate from other pathologies
- Change in position with respiration or gravity is particularly suggestive of a pedunculated pleural fibroma

Note how the blending of mass with left mediastinal contour on PA view and projection into retrosternal region on the lateral film make it mimic an anterior mediastinal mass

Dr. Paul Stark. Imaging of Pleural Plaques, Thickening and Tumors. UpToDate. Online16.1
http://utdol.com/online/content/topic.do?topicKey=int_pulm/7040&selectedTitle=2~2&source=search_result#1
Imaging features of Pleural Fibroma on CT

- Homogenous soft tissue lesion (intermediate attenuation similar to muscle and attributable to high collagen density and vascular nature), often abutting chest wall. Heterogeneity may be related to necrosis and hemorrhage. Heterogeneous pattern of enhancement after IV contrast media infusion.
- Smaller fibromas typically form obtuse angle, larger can form acute angle to pleural surface.
- No pathognomonic radiologic features to differentiate benign from malignant; however malignant lesions are typically larger than 10cm and are more likely to be associated with central necrosis and a large pleural effusion.
Imaging Features of Pleural Fibroma on MRI

- In general, role somewhat limited. Advantages include ability image directly in axial, sagittal, and coronal planes. Slightly superior to CT in fluid characterization.
- Typically exhibit low-intermediate signal intensity on both T1 and T2.
- Areas of necrosis, or myxoid degeneration may show higher signal on T2-weighted sequences (and correspond to regions of decreased attenuation on CT scans).
- After gadolinium administration, intense homogenous enhancement is typical, reflecting vascularity of tumor.

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Imaging Features of Pleural Fibroma on Ultrasound

- Ultrasound:
  - Little role in the work-up.
  - May be used to guide procedures.
  - Fibrous tumor tends to demonstrate homogenous low echogenicity in general (unless it has calcifications).

Future: Role of FDG-PET scan. Case report of high FDG uptake noted in portion of mass that had malignant features histopathologically, thus may be helpful to determine presence of malignancy preoperatively.
Returning to our Patient
Initial CXR

BIDMC, PACS

BIDMC, PACS
Our Patient: Post-operative CXR
Full surgical excision of mass was performed
Our Patient: Pathology Report

Gross pathology (no photo available) showed smooth, glistening, tan pink and encapsulated mass measuring up to 11.1 cm.

Low-power shows well-circumscribed border of tumor with adjacent normal lung. Also characteristic “staghorn” shaped vessels.

High-power further elucidates the characteristic spindle cells of a solitary fibrous tumor of pleura.

Diffusely positive for CD34 (not shown).

Confirmed diagnosis of solitary fibrous tumor of pleura.
References:

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