March 2017 Imaging Case of the Month

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Clinical History: A 69-year-old man presented with long-standing complaints of dyspnea, progressing to dyspnea at rest, associated with some dysphagia to solids. He also noted symptoms consistent with exertional stertor (a respiratory sound characterized by heavy snoring or gasping). His past medical history was remarkable only for hypertension controlled with medication.

Laboratory data, include white blood cell count, coagulation profile, and serum chemistries were within normal limits. Oxygen saturation on room air was normal.

Frontal chest radiography (Figure 1) was performed.

Figure 1: Frontal chest radiography.

Which of the following statements regarding the chest radiograph is **most accurate**?

1. Frontal chest radiography shows a cavitary lung mass
2. Frontal chest radiography shows an abnormal mediastinal contour
3. Frontal chest radiography shows multiple small nodules
4. Frontal chest radiography shows peribronchial and mediastinal lymphadenopathy
5. Frontal chest radiography shows pleural effusion

Correct!

2. Frontal chest radiography shows an abnormal mediastinal contour

The frontal chest radiograph shows an abnormal contour along the left superior mediastinum- there is a convex leftward “bulge” cranial to the left heart border and left hilum (Figure 2).

Figure 2. Frontal chest radiography shows an abnormal contour (arrows) along the left superior mediastinum.

There are no pulmonary nodules, nor is there evidence of dependent pleural liquid. The hilar and peribronchial regions appear normal and no mediastinal lymph node enlargement is present. No cavitary lung mass is present.

Which of the following choices represents the **most likely** structure giving rise to the abnormal finding at chest radiography?

1. The esophagus
2. The heart
3. The mediastinum
4. The pleura
5. The thoracic aorta

Correct!
3. The mediastinum

The left mediastinum is the most likely source of the contour abnormality detected at chest radiography. In general, when a smoothly contoured soft tissue lesion shows extensive contact with the mediastinum, with no lung visible between the medial margin of the lesion and the mediastinum itself, and the center of a circle extrapolated from the lesion's margins triangulates within the mediastinum, a mediastinal origin should be suspected. It is technically possible that the lesion at chest radiography arises from the medial left superior pleura, but a mediastinal location is more likely. The thoracic aorta shows a normal contour and appears normal in caliber. While the contour abnormality along the left superior mediastinum appears to be in close proximity to the aortic arch, the aortic arch itself can be seen "through" the abnormality and therefore is probably unrelated to that abnormality. The contour abnormality does not contact the heart and therefore does not arise from this structure. The esophagus is an unlikely origin for the lesion. Often gas or an air-fluid level will be evident if the cause of a contour abnormality at chest radiography is due to a dilated esophagus, but no gas or air-fluid level is present in this circumstance. An esophageal mass of this size, in this location, while technically possible, would be very unlikely.

The frontal chest radiograph (Figure 1) shows which **chest radiographic sign**?

1. The "atoll" sign
2. The "cervico-thoracic" sign
3. The "ground-glass halo" sign
4. The "hilum overlay" sign
5. The "incomplete border" sign

Correct!
2. The “cervico-thoracic” sign

The “cervico-thoracic” sign describes the appearance of a mediastinal lesion near the thoracic inlet. At the thoracic inlet, the anterior first ribs and clavicle demarcate the anterior aspect of the lung, whereas the posterior lung extends more cranially (Figure 3).

Figure 3. Thoracic inlet anatomy. Sagittal volume-rendered neck CT shows the relationship of the clavicle (arrow), anterior first rib (single arrowhead) and posterior first rib (double arrowhead), and lung apex (curved arrow). The trachea and lungs appear blue in this image. The first rib defines the junction of the neck and mediastinum at the thoracic inlet. Note that the posterior portion of the first rib (double arrowhead) is more cranially located than the anterior portion of the first rib (single arrowhead), with the clavicle (arrow) also positioned anteriorly and relatively caudal to the posterior portion of the first rib. Similarly, the posterior portion of the lung at the apex also extends more cranially than the anterior portion of the lung. This anatomical arrangement explains the “cervico-thoracic” sign, in which anteriorly located mediastinal lesions will lose their sharp interface with lung at about the level of the clavicle, whereas posteriorly located mediastinal lesions will maintain this sharp interface more cranially, to the level of the posterior first rib.

Therefore, a lesion in the anterior mediastinum will show a sharply demarcated contour to the level of the first ribs and clavicle, above which such a lesion will enter the neck, thereby losing contact with the lung, resulting in “disappearance” of the abnormal contour. In contrast, a lesion in the posterior mediastinum will maintain contact with the lung to the level of the posterior first rib, and therefore the abnormal contour on the

chest radiograph will extend more cranially in the presence of a posterior mediastinal lesion (Figure 4C) compared with an anterior mediastinal lesion.

The “hilum overlay” sign is present when a mass overlies the hilum, but the hilum can still be seen “through” the mass - this implies that there is still aerated lung parenchyma around the vessels of the hilum, so the mass cannot reside in this area and must reside anterior or posterior to the hilum (Figure 4A).

Figure 4. Imaging signs. A: “Hilum overlay” sign. Note that the vessels of the left hilum (arrowhead) can be “seen through” the mass (arrows) projected over the left hilum. The pulmonary artery lies medial to the anterior mediastinal mass, which represented thymic neuroendocrine malignancy. B: “Incomplete border” sign. Note the circumscribed inferior margin (arrowhead) of the opacity overlying the right upper lobe, with the cranial margin of this opacity “fading” indistinctly. The cause of the finding was multiloculated pleural effusion. C: “Cervico-thoracic sign. Note how the lateral margin of the lesion (arrowhead) can be readily seen to the level of the posterior 1st costovertebral junction, consistent with a posterior mediastinal lesion; the abnormality was due to a schwannoma. D: “Ground-glass halo” sign. Note several lung nodules with central, solid foci of opacity surrounded by ground-glass opacity, due to hemorrhagic metastases. E: “Atoll” sign. Note focal opacity with central ground-glass opacity surrounded by consolidation, due to organizing pneumonia.

The hilum overlay sign was originally conceived to distinguish an enlarged heart and pulmonary artery from a mediastinal mass. It was noted that the proximal portions of the pulmonary arteries in the hilar regions typically lie just lateral to the heart border or may overlap the lateral heart border, even when cardiomegaly is present. Occasionally an anterior mediastinal mass can simulate cardiomegaly, but such masses cannot lie medial to the pulmonary artery since this position is occupied by the heart and pericardium; therefore, anterior mediastinal masses will overlap the pulmonary artery as it exists the hilum, and the pulmonary artery will be seen “through” the mass. The “incomplete border” sign (Figure 4B) is present when a lesion shows a circumscribed margin on one side, and an obscured, or “fading” margin on the other side- this configuration is typical of extraparenchymal lesions, such as those arising from the pleura or chest wall. The “ground-glass halo” sign (Figure 4D) is a CT sign that represents a solid, nodular area of consolidation surrounded by ground-glass opacity, and is typical of a hemorrhagic lesion, often encountered in the setting of invasive fungal infection in severely immunocompromised patients. The “atoll” sign, or “reverse ground-glass halo” sign (Figure 4E), is also an imaging finding seen at CT in which a nodular focus of ground-glass opacity is surrounded by a partial or complete ring of consolidation, is often encountered in patients with organizing pneumonia.

Which of the following pathological conditions is **least likely** to account for the lesion seen at chest radiography?

1. Bronchogenic cyst
2. Left subclavian artery aneurysm
3. Neurogenic tumor
4. Thymic mass
5. Thyroid goiter

Correct!

3. Neurogenic tumor

The chest radiograph (Figure 1) shows that the abnormal contour extends cranially to just above the anterior first rib and clavicular margins suggesting a relatively anterior location, based on the foregoing discussion. The one incorrect choice listed- neurogenic tumor- typically arises from the posterior mediastinum, and therefore would be expected to create a contour that would extend more cranially, to the level of the posterior 1st costovertebral junction. The other choices listed are located relatively anteriorly in the superior mediastinum and therefore would be correct considerations for the appearance of the abnormality at chest radiography. Bronchogenic cyst would be a consideration here, although bronchogenic cysts are more commonly encountered in the middle mediastinum rather than the anterior mediastinum. While a posterior mediastinal localization for bronchogenic cysts is unusual, neurogenic tumors most commonly present in the posterior mediastinum.

Which of the following would be **most useful** for the evaluation of this patient?

1. ¹⁸FDG-PET scanning
2. Comparison to prior chest radiographs
3. Neck CT
4. Pulmonary function testing
5. Video-assisted thoracoscopic biopsy

Correct!
3. Neck CT

All of the choices listed would provide useful information for this patient, as would thoracic MRI. Surgical biopsy is, however, premature at this point. Similarly, ^{18}F FDG-PET is a bit premature, as the decision to obtain this study for the investigation of indeterminate lesions at chest radiography is often best informed following CT characterization. Prior chest radiographic comparison would prove useful here, particularly if the lesion was shown and unchanged on remote priors. However, in this case, this particular patient has progressive symptoms that could be related to this lesion, and therefore further investigation is warranted.

The patient underwent enhanced neck CT (Figure 5).

Figure 5. Axial (A) and coronal (B) enhanced neck CT.

Which of the following is the **most appropriate description** of this patient's CT findings?

1. The enhanced neck CT shows a lesion arising from the esophagus
2. The enhanced neck CT shows destruction of the tracheal wall
3. The enhanced neck CT shows metastatic lymphadenopathy at the left thoracic inlet
4. The enhanced neck CT shows that the lesion is a simple cyst
5. The enhanced neck CT shows the lesion reflects a left subclavian artery aneurysm

Correct!

2. The enhanced neck CT shows destruction of the tracheal wall

The enhanced neck CT shows a mass centered in the left paratracheal region at the thoracic inlet. The mass is associated with destruction of the left lateral tracheal wall (Figure 6).

Figure 6. Axial (A) and coronal (B) enhanced neck CT shows a soft tissue mass centered in the left paratracheal region at the thoracic inlet, accounting for the chest radiographic findings. The lesion shows faint, nodular internal enhancement (arrowhead) and is associated with destruction of the left lateral wall of the trachea (arrow).

The mass minimally contacts the anterior margin of the esophagus, but the bulk of the lesion is centered anterior to the esophagus and therefore probably does not arise from the esophagus itself. A large focus of metastatic lymphadenopathy is technically possible, but an isolated metastatic nodal mass centered in the left paratracheal region at the thoracic inlet in the setting of otherwise normal chest radiography would be distinctly unusual. The mass shows some faint internal enhancement, indicating that the lesion is not a simple cyst. The enhancement, however, is not suggestive of aneurysm, and the contact of the lesion with the left subclavian artery is minimal, making pseudoaneurysm unlikely as the origin of this lesion.).

Which of the following choices is the **least appropriate** consideration for the differential diagnosis of this lesion?

1. Carcinoid tumor
2. Chondrosarcoma
3. Lymphoma
4. Squamous cell carcinoma
5. Thyroid neoplasm

Correct!
1. Carcinoid tumor

The mass certainly could reflect a thyroid malignancy extending caudally from the thyroid gland in the neck into the left superior mediastinum. The lesion could arise from the left lateral tracheal wall, given the destruction of this portion of the cartilaginous trachea- therefore, chondrosarcoma is a consideration, albeit rare. The most common primary malignancy of the trachea- squamous cell carcinoma- is a consideration as well. An isolated large nodal mass from lymphoma is also a possibility. While carcinoid tumors are not uncommon airway neoplasms, they are not commonly located in the cranial trachea, and they commonly grow mostly within and fill the airway lumen, rather than destroy the airway wall and extend into surrounding tissues.

Which of the following would be **most useful** for further evaluation of this patient?

1. Bronchoscopy
2. Mediastinoscopy
3. Percutaneous fine needle aspiration biopsy
4. Upper endoscopy
5. Video-assisted thoracoscopic surgical lung biopsy

Correct!
1. Bronchoscopy

All of the procedures listed would be capable of performing tissue sampling for this lesion, but clearly bronchoscopy allows the most direct and least invasive route to establish a diagnosis.

The patient underwent bronchoscopy (Figure 7).

Figure 7. Bronchoscopy shows a mass along the left lateral tracheal wall extending into and narrowing the tracheal lumen.

Biopsies obtained during this procedure indicated the diagnosis of chondrosarcoma. The patient elected to undergo combined endoluminal resection using cupped forceps and a KTP laser with a partial sternotomy (Figure 8) to address the extraluminal component.

Figure 8. Image during the open resection through partial sternotomy shows the lesion.

A stent was placed through the affected area at the completion of the procedure (Figure 9).

Figure 9. Bronchoscopic image at the completion of the endoluminal resection portion of the procedure following stent placement.

The final pathological diagnosis was low-grade tracheal chondrosarcoma (Figure 10).

Figure 10. Resected low-grade chondrosarcoma. A: Resected gross specimen measured 6.5 cm. B: Low-power histopathological specimen shows a pink-staining, think, fibrous capsule encompassing the majority of the surface of the lesion, extending deeply into the lesion creating distinct lobules composed of hyaline cartilage. C: High-power histopathological specimen shows typical chondrocytes with moderately atypical nuclei and partial cytoplasmic vacuolization with a low mitotic index. An abundance of cartilaginous matrix between cells is consistent with a low-grade lesion.

Diagnosis: Tracheal chondrosarcoma

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