September 2013 Imaging Case of the Month

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Clinical History

A 61-year-old man presented with a 6-month history of easy fatigability and worsening shortness of breath. The patient noted that his difficulty breathing improved somewhat when lying flat. 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. The chest radiograph shows basal predominant fibrotic lung disease
2. The chest radiograph shows large lung volumes with cystic change
3. The chest radiograph shows multiple nodules
4. The chest radiograph shows tubular opacities in the lower lobes bilaterally
5. The chest radiograph shows upper lobe peripheral consolidation and reticulation

Correct!

4. The chest radiograph shows tubular opacities in the lower lobes bilaterally

The frontal chest radiograph shows normal lung volumes with a normal heart size, but there are bilateral lower lobe tubular-shaped opacities (Figure 2).

Figure 2. Frontal chest radiography shows bilateral lower lobe faintly seen tubular-shaped opacities (arrows). The heart size, central pulmonary arteries, and lung volumes appear normal.

There is no evidence of architectural distortion, reticulation, or honeycombing- hallmarks of fibrotic lung disease. No evidence of consolidation or cystic change is apparent.

Which of the following is an **appropriate consideration** among the differential diagnostic possibilities for the appearance of the patient's chest radiograph?

1. Allergic bronchopulmonary aspergillosis
2. Anomalous systemic arterial supply to the lower lobes
3. Hereditary hemorrhagic telangiectasia
4. Post-infectious bronchiectasis
5. All of the above

Correct!
5. All of the above

The tubular nature of the bilateral lower lobe opacities suggests an abnormality involving one of two anatomical elements within the thorax that normally exhibit a tubular shape- vessels or airways. Each of the above choices is related to either the pulmonary vasculature or airways, and therefore any of the choices should be considered among the differential diagnostic possibilities for the opacities on the chest radiograph.

The patient underwent contrast-enhanced thoracic CT (Figure 2) for further characterization of the abnormalities seen at chest radiography.

Figure 3. Axial enhanced thoracic CT displayed in soft tissue (A-E) and lung windows (axial, F, sagittal, G and H).

Which of the following statements regarding this CT examination is **most accurate**?

1. The thoracic CT shows anomalous systemic arterial supply to the lower lobes bilaterally originating from the cranial abdominal aorta
2. The thoracic CT shows that the tubular opacities on the chest radiograph represent bronchial impaction
3. The thoracic CT shows that the tubular opacities on the chest radiograph represent dilated and tortuous peripheral pulmonary vessels
4. The thoracic CT shows that the tubular opacities on the chest radiograph represent innumerable pulmonary arteriovenous malformations
5. The thoracic CT shows that the tubular opacities on the chest radiograph are artifactual, probably created by atelectasis and superimposition of overlapping structures

Correct!

3. The thoracic CT shows that the tubular opacities on the chest radiograph represent dilated and tortuous peripheral pulmonary vessels

The enhanced thoracic CT shows numerous dilated and tortuous vessels extending to the subpleural regions of lung. Note that rather large vessels can be seen in direct contact with the visceral pleura, particularly well seen on the images shown in lung windows (Figure 3, Panels F-H). These structures clearly enhance and therefore are vessels, not bronchi; therefore, bronchial impaction and an airway etiology for the tubular abnormalities seen on the chest radiograph are excluded. There is no large, anomalous vessel emanating from the cranial abdominal aorta supplying the lower lobes, as can be seen with anomalous systemic arterial supply to the lower lobes. Peripheral pulmonary arteriovenous malformations remain a consideration, but the characteristic morphology at CT that allows their recognition- a vascular nidus supplied by one or more enlarged pulmonary arteries and drained by an enlarged pulmonary vein- is not seen on the images provided. Rather, a diffuse enlargement of peripheral pulmonary arteries and veins is seen.

What is the **appropriate next step** for the evaluation / management of this patient?

1. ^{99m}Tc macroaggregated albumin (MAA) scanning
2. Echocardiographic bubble study
3. Shunt fraction measurement with the 100% oxygen method
4. Any of the above
5. None of the above

Correct!
4. Any of the above

The enlarged peripheral pulmonary vessels can be associated with conditions that produce right-to-left pulmonary shunting, and such shunting could explain the patient's complaints. Measuring the shunt fraction using the 100% oxygen method, ^{99m}Tc -MAA scanning, and an echocardiographic bubble study are all methods that could be used to detect the presence of intrapulmonary right-to-left shunting.

The patient underwent ^{99m}Tc -MAA scanning (Figure 4).

Figure 4. ^{99m}Tc -Macro aggregated albumin scintigraphy.

Which of the following **best describes** the results of this examination?

1. Homogeneous lung perfusion indicates that the study is low probability for pulmonary embolism
2. The study is normal
3. Inhomogeneous lung perfusion suggests the possibility of pulmonary hypertension
4. The study shows systemic tracer embolization
5. 1 and 4
6. 3 and 4

Correct!
5. 1 and 4

The ^{99m}Tc -MAA study does show relatively homogeneous tracer distribution within the lungs, indicating a low likelihood of pulmonary embolism; however, this is not the most interesting or relevant finding on this examination. Rather, the study shows tracer activity within the brain (Figure 4, Panel B) and the kidneys (Figure 4, Panel C), which indicates systemic tracer embolization, and is diagnostic of right-to-left shunting. Such shunting does not localize the level of the shunt- it could be intracardiac or extracardiac- but it does indicate a physiologically significant right-to-left shunt is present.

An echocardiographic bubble study was performed and the findings were consistent with the presence of a pulmonary shunt. Measurement the shunt fraction using the 100% oxygen method indicated a shunt fraction of 20%.

What is the **appropriate next step** for the evaluation / management of this patient?

1. Catheter pulmonary angiography and embolotherapy
2. Expectant management and treatment with supplemental oxygen
3. Lower lobe surgical pulmonary arterial ligation
4. Lung transplant
5. Vasodilator therapy

Correct!

1. Catheter pulmonary angiography embolotherapy

The thoracic CT has already shown enlarged pulmonary arteries, indicating a target for embolotherapy, and the physiologic derangement necessitating treatment has been well documented. There is no adequate thoracic surgical approach for this patient's condition, and the patient's hypoxemia cannot be corrected with inspired oxygen. The patient does not have pulmonary hypertension and vasodilator therapy is not the correct choice.

The patient subsequently underwent catheter pulmonary angiography (Figure 5).

Figure 5. Catheter pulmonary angiography performed in the right (A) and left (B) lower lobes.

Catheter pulmonary angiography identified large, dilated, tortuous lower lobe pulmonary arteries bilaterally, but without a clear vascular nidus to suggest typical pulmonary arteriovenous malformation. The largest, abnormal lower lobe pulmonary arteries were embolized using coils (Figure 6), with good results.

Figure 6. Catheter pulmonary angiography with embolotherapy performed in the left lower lobe shows coil embolization of the enlarged, tortuous vessels, resulting in occlusion of these vessels.

Review the patient's thoracic CT (Figure 2) again. Based in the information thus far, what is the patient's **diagnosis**?

1. Bechet syndrome
2. Hepatopulmonary syndrome
3. Hereditary hemorrhagic telangiectasia
4. Pseudoscimitar syndrome
5. Pulmonary vasculitis

Correct!

2. Hepatopulmonary syndrome

In addition to the dilated tortuous pulmonary vessels, the inferior-most images of the thoracic CT shows a nodular liver contour and perihepatic ascites, consistent with cirrhosis and portal hypertension. These findings provide the proper context for the pulmonary vascular abnormalities and allow the diagnosis of hepatopulmonary syndrome. As noted previously, the pulmonary vascular findings do not show the typical morphology of pulmonary arteriovenous malformations and therefore hereditary hemorrhagic telangiectasia is not the proper choice. Pulmonary vasculitis does not fit the constellation of clinical and physiologic data provided thus far, and, when pulmonary vascular findings are evident in patients with pulmonary vasculitis, a “beaded” appearance of the arteries, with areas of dilation mixed with stenosis, may be seen, rather than the diffuse dilation present in this case. Pseudoscimitar syndrome refers to a condition in which an anomalous right pulmonary vein, often referred to as a “meandering” pulmonary vein, pursues an unusual course, but ultimately drains into the left atrium, or drains into both the left atrium and the inferior vena cava. Dextroposition of the heart and hypoplasia of the affected thorax are present, and thus the findings closely resemble true scimitar, or hypogenetic lung, syndrome. Bechet syndrome is associated with pulmonary arterial aneurysm development, but not diffusely ectatic, tortuous pulmonary vessels and intrapulmonary right-to-left shunting.

Diagnosis: Hepatopulmonary syndrome

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