August 2016 Imaging Case of the Month

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Clinical History: A 47 year-old white man presented with a history of worsening shortness of breath over the past few months. When questioned, the patient indicated that his activity tolerance had probably been slow decreasing over a relatively long period of time, but had become more pronounced recently.

Laboratory data, include white blood cell count and serum chemistries were within normal limits. Oxygen saturation on room air was 93%.

Frontal chest radiography (Figure 1) was performed.

Figure 1. Frontal and lateral chest radiography.

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

1. The frontal chest radiograph appears normal
2. The frontal chest radiograph shows abnormal mediastinal contours
3. The frontal chest radiograph shows abnormally increased lung volumes
4. The frontal chest radiograph shows bilateral linear and reticular abnormalities
5. The frontal chest radiograph shows pneumothorax

Correct!

3. The frontal chest radiograph shows abnormally increased lung volumes

The cardiac and mediastinal contours appear normal, and there is no evidence of abnormal linear or reticular opacities to suggest an infiltrative disorder of the interstitium. No pneumothorax is present. However, the frontal chest radiograph is not normal- the lung volumes are abnormally increased. The increased lung volumes are evidenced by not only the abnormally large cephalocaudad appearance of the thorax, but also the increased retrosternal clear space and flattening of the diaphragms, the latter best seen on the lateral chest radiograph (Figure 2).

Figure 2. Frontal and lateral chest radiography shows large lung volumes, evidenced by flattening of the diaphragms, increased retrosternal clear space (line), and visualization of slips of the diaphragm (arrowheads). Focal nodular opacity is also present at the left lateral base.

The patient was treated for presumed community-acquired pneumonia, given the focal opacity at the left base. Several months later, he returned with continued complaints of shortness of breath. A repeat chest x-ray was performed (Figure 3).

Figure 3. Repeat frontal chest radiography.

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

1. The repeat frontal chest radiograph appears normal
2. The repeat frontal chest radiograph remains unchanged
3. The repeat frontal chest radiograph shows new pneumomediastinum
4. The repeat frontal chest radiograph shows progression of the previously seen left base opacity
5. The repeat frontal chest radiograph shows that the lung volumes have now normalized

Correct!

2. The repeat frontal chest radiograph remains unchanged

The frontal chest radiograph shows that the left base opacity persists but is unchanged, and the lung volumes remain large- essentially, the same findings detailed previously persist unchanged from prior. No evidence of pneumomediastinum, pneumothorax, or other abnormal gas collection is seen.

Which of the following is the **most appropriate** next step for the evaluation of this patient?

1. Pulmonary function testing
2. ⁶⁸Ga-citrate scintigraphy
3. Thoracic CT
4. 1 and 3
5. None of the above

Correct!
4. 1 and 3

Both thoracic CT and pulmonary function testing are appropriate for this patient. The large lung volumes suggest obstructive pulmonary disease, and both pulmonary function testing and thoracic CT are very useful for the assessment of suspected obstructive lung disease. ^{68}Ga -citrate scintigraphy is occasionally useful for diffuse inflammatory lung disease detection, but has no role for the evaluation of obstructive lung disease.

The patient underwent unenhanced thoracic CT (Figure 4).

Figure 4. Axial thoracic CT displayed in lung windows.

Which of the following is **correct** regarding the description of the thoracic CT findings?

1. Thoracic CT shows enlarged peripheral pulmonary arteries suggesting pulmonary hypertension
2. Thoracic CT shows multifocal cystic airway dilation
3. Thoracic CT shows smooth interlobular septal thickening suggesting increased pressure pulmonary edema
4. Thoracic CT shows extensive low attenuation throughout the lung parenchyma
5. Thoracic CTA shows segmental areas of air trapping

Correct!

4. Thoracic CT shows extensive low attenuation throughout the lung parenchyma

The thoracic CT shows multifocal areas of low attenuation throughout the lung parenchyma, but the low attenuation areas do not represent true cysts- they do not have “walls” as cysts typically possess. Rather, the low attenuation in the apices has a somewhat “punched-out” appearance, whereas the basal low attenuation is more diffuse appearing. In the bases, at first glance, the lung parenchyma may appear relatively normal. However, on closer inspection, there are relatively fewer vessels seen in the bases than expected, and the vessels appear small, stretched, attenuated, and more separated from each other than usual. The airway walls may appear somewhat thickened, but they are not abnormally dilated. The peripheral pulmonary arteries do not appear enlarged; rather, they actually appear smaller than normal. There is no evidence of interlobular septal thickening to suggest increased pressure edema. Air trapping is typically difficult to diagnosis on inspiratory scans alone. Usually one reviews post-expiratory scans to observe the lack of appropriate increase, or the paradoxical decrease, in lung parenchymal attenuation on post-expiratory imaging, compared with inspiratory imaging, to diagnose air trapping. However, one can occasionally suggest the presence of air trapping on an inspiratory examination when mosaic perfusion is present- areas of inhomogeneous lung attenuation, often with a geographic appearance. When air trapping produces mosaic perfusion (as opposed to the much rare vascular obstructive causes, like pulmonary hypertension, pulmonary emboli, and obliterative thrombotic microangiopathy), the vessels often appear smaller in the more lucent regions and somewhat larger in the denser regions of lung], and areas of distinct lobular low attenuation may also be evident. In this case, the low attenuation in the bases appears lobular, but it is diffuse rather than geographic in distribution.

Additional history was performed and the patient admitted to smoking 2-3 cigarette packs per day for the previous 15 years.

Based on the information thus far, what is the **most likely diagnosis?**

1. Bronchiectasis
2. Bronchiolitis obliterans
3. Chronic obstructive pulmonary disease due to cigarette smoking
4. Cystic lung disease
5. None of the above

Correct!

5. None of the above

Certainly choice 1- chronic obstructive pulmonary disease due to cigarette smoking- is contributing to the patient's shortness of breath by accounting for the appearance of the centrilobular emphysema seen in the apices on the patient's thoracic CT. However, cigarette smoking-induced emphysema may not entirely account for the diffuse lobular low attenuation in the bases. The thoracic CT shows no evidence of bronchiectasis or cystic lung disease, so these choices are not correct. Bronchiolitis obliterans often appears as an area of segmental airway thickening and dilation with focal low attenuation in the lung parenchyma subtended by the abnormal bronchi, also accompanied by a small ipsilateral pulmonary artery (this is often the appearance of Swyer-James syndrome)- this morphology is not present on the thoracic CT. Bronchiolitis obliterans may also be more diffuse, typically appearing as mosaic perfusion and multifocal lobular low attenuation on inspiratory imaging, proven to reflect air trapping on post-expiratory imaging; however, this appearance is not consistent with the morphology seen at this patient's thoracic CT. Therefore, another process may account for the patient's presentation.

At this point, which of the following tests would be **most useful for** establishing the **diagnosis** for this patient?

1. ^{99m}Tc -MAA ventilation – perfusion scan
2. Anterior abdominal wall fat pad biopsy
3. Serum α_1 -protease inhibitor level
4. Serum angiotensin-converting enzyme levels
5. Thoracic MRI with contrast – enhanced MR angiography

Correct!

3. Serum alpha₁-protease inhibitor level

An anterior abdominal wall fat pad biopsy may be used to assess for the presence of amyloidosis, but that diagnosis is not a consideration for this patient's presentation and imaging. Biopsy of fat could potentially detect panniculitis as well, which could be suggestive of a particular diagnosis for this patient, but this would merely constitute indirect information, and there is a choice provided above that is capable for directly suggesting a diagnosis for this patient. ^{99m}Tc-MAA ventilation – perfusion scanning is typically used to assess for suspected pulmonary embolism and is occasionally used to determine relative perfusion to each lung, often as part of a pre-operative assessment prior to thoracic surgery. The ventilation portion of this study would be capable of showing air trapping, but given the thoracic CT appearance, airflow obstruction is certainly present, so ^{99m}Tc-MAA ventilation – perfusion scanning is unlikely to add new, useful information. Thoracic MRI performed with intravenous contrast would provide little information to that already known with thoracic CT, and, in fact, would be largely incapable of yielding useful information regarding the lung parenchyma. A serum angiotensin converting enzyme level can be used to suggest the presence of lung disease, as in the case of sarcoidosis, but normal levels would be non-contributory, whereas elevated levels would provide little useful direction towards establishing a diagnosis.

The patient's serum level of alpha₁-protease inhibitor level was found to be diminished- 7 µmol (normal values range from 20 - 48 µmol; emphysema rarely seen with levels greater than 11 µmol). Based on this finding, he underwent further phenotypic testing, which disclosed the Pi ZZ (homozygotic) phenotype of alpha₁-protease Inhibitor deficiency.

Diagnosis: Panlobular emphysema due to alpha₁-protease inhibitor deficiency

References

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