September 2016 Imaging Case of the Month

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Clinical History: A 64-year-old woman presents with a several month history of slowly worsening shortness of breath and dry cough, with worsening exercise limitation. Laboratory data, include white blood cell count and serum chemistries were within normal limits. Oxygen saturation on room air was 93%.

Frontal and lateral chest radiographs (Figure 1) were performed. Previous frontal and lateral chest radiographs, performed 7 years prior to presentation, are shown for comparison.

Figure 1. Frontal (A) and lateral (B) chest radiography. Frontal (C) and lateral (D) chest radiography performed 7 years prior are shown for comparison.

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

1. Frontal and lateral chest radiography appears normal
2. Frontal and lateral chest radiography bilateral linear and reticular abnormalities
3. Frontal and lateral chest radiography shows abnormally increased lung volumes
4. Frontal and lateral chest radiography shows bilateral hilar enlargement
5. Frontal and lateral chest radiography shows developing pleural thickening

Correct!

2. Frontal and lateral chest radiography bilateral linear and reticular abnormalities

Frontal and lateral chest radiography at presentation is arguably normal. However, with the benefit of prior chest radiography for comparison, a slight increase in basal, peripheral reticulation and linear opacity, with a slightly coarser and more prominent appearance to the interstitium, is appreciable. The changes are subtle, and occasionally changes in technology, such as installation of a new chest radiography unit or introduction of new imaging post-processing algorithms, can account for such changes. Nevertheless, the slight change may be of importance in a patient with worsening respiratory symptoms. The hilar contours and lung volumes appear normal. No pleural abnormalities are evident.

The patient's past medical history was significant for rheumatoid arthritis. She was a lifelong non-smoker. Physical examination revealed slightly decreased bibasilar breath sounds on auscultation.

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

1. Repeat frontal chest radiography
2. Ventilation - perfusion scintigraphy
3. Flexible fiberoptic bronchoscopy
4. Pulmonary function testing
5. Right heart catheterization

Correct!

4. Pulmonary function testing

Repeat chest radiography is unlikely to be of benefit for short-term follow up of a slowly progressive clinical abnormality. Ventilation-perfusion scintigraphy is commonly used for the assessment of acute or chronic thromboembolic disease and occasionally for differential lung function assessment prior to surgical resection. This study could be of some benefit for this patient, given her complaint of slowly progressive shortness of breath, which could be a manifestation of chronic thromboembolic disease; however, ventilation-perfusion scintigraphy is not the best choice among those listed. Flexible fiberoptic bronchoscopy and right heart catheterization could prove useful, but are needlessly invasive at this point of the patient's evaluation. The most appropriate next step is obtaining pulmonary function testing.

The patient underwent pulmonary function testing, which showed increased residual volume (171% predicted) and a slightly increased total lung capacity (106% predicted). No significant response to bronchodilator inhalation was seen. The diffusion capacity for carbon dioxide was normal. The forced expiratory volume in one second was severely decreased at 34% predicted.

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

1. ⁶⁸Ga-citrate scintigraphy
2. Flexible fiberoptic bronchoscopy
3. High-resolution chest CT
4. Right heart catheterization
5. Unenhanced thoracic CT

Correct!

5. Unenhanced thoracic CT

While thoracic CT would be appropriate for the evaluation of this patient, given the presence of a significant obstructive abnormality at pulmonary function testing, it is important to include post-expiratory imaging to assess for air trapping as part of the CT protocol. Routine thoracic CT does not include post-expiratory imaging, but high-resolution chest CT (HRCT) does, so high-resolution chest CT is the best choice among those listed. ^{68}Ga -citrate imaging can be used for diffuse lung diseases, but typically in the context of alveolitis, not obstructive lung disease. Flexible fiberoptic bronchoscopy and right heart catheterization could be of benefit for this patient, but are invasive and HRCT results could inform which of these invasive procedures may be indicated..

The patient underwent unenhanced HRCT (Figures 3 and 4).

Figure 2. Axial inspiratory HRCT displayed in lung windows.

Figure 3. Axial expiratory HRCT displayed in lung windows.

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

1. HRCT shows multifocal bilateral areas of lobular low attenuation on inspiratory imaging and air trapping on post-expiratory imaging
2. HRCT shows multifocal bronchiectasis
3. HRCT shows multifocal ground-glass opacity
4. HRCT shows numerous small nodules consistent with a miliary pattern
5. HRCT shows patchy areas of peribronchial consolidation

Correct!

1. HRCT shows multifocal bilateral areas of lobular low attenuation on inspiratory imaging and air trapping on post-expiratory imaging

HRCT shows bilateral inhomogeneous lung attenuation, with areas of both increased attenuation and decreased attenuation. Note that the areas of low attenuation have a somewhat geographic and lobular appearance, and that these particular areas have relatively small-appearing pulmonary vessels compared to the areas of higher attenuation. Furthermore, the areas of low attenuation fail to appropriately increase, and paradoxically actually decrease, in attenuation on post-expiratory imaging- this finding is characteristic of air trapping. The areas of relatively increased appearing attenuation do not represent actual ground-glass opacity- they are merely more normal lung contrasted against the areas of low attenuation, which represent mosaic perfusion. No nodules are present. The large airways show normal wall thickness and caliber- no bronchiectasis is seen and no consolidation is evident.

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

1. Bronchiolitis obliterans
2. Congenital cystic pulmonary airway malformation
3. Congenital lobar overinflation / emphysema
4. Emphysema
5. None of the above

Correct!

1. Broncholitis obliterans

Congenital lobar overinflation / emphysema is rare in adults, and often represents as a lobar area of low attenuation, possibly with airway thickening and dilation, rather than multifocal bilateral areas of lobular low attenuation and air trapping. Furthermore, one of the oldest reported patients with congenital lobar overinflation / emphysema was aged 37 at diagnosis- substantially younger than this patient. Congenital cystic pulmonary airway malformation typically presents as a focal low attenuation areas with variably-sized thin-walled cysts, rather than the multifocal bilateral areas of lobular low attenuation and air trapping seen at thoracic CT in this patient. Emphysema may present as upper lobe areas of circumscribed low attenuation without walls (centrilobular emphysema), more confluent and often lower lobe areas of lobular low attenuation, with stretched and attenuated pulmonary vessels, creating the appearance of lung parenchymal “simplification” (panlobular emphysema), and/or discrete areas of upper lobe predominant subpleural thin-walled cysts or bullae (paraseptal emphysema)- all of these appearances are distinct from the CT pattern present in this patient. Finally, this patient is a life-long non-smoker.

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

1. Bronchoscopic biopsy
2. Cervical mediastinoscopy
3. Medical pleuroscopy
4. Open surgical lung biopsy
5. Percutaneous transthoracic needle and core biopsy

Correct!

1. Bronchoscopic biopsy

Among the choices listed, bronchoscopic biopsy is the most appropriate, although the transbronchial biopsy diagnostic yield for small airway disorders is inferior to surgical lung biopsy techniques. Percutaneous transthoracic needle and core biopsy is a very useful procedure for obtaining lung tissue for diagnosis, but is typically only employed for focal lung disorders, not diffuse lung diseases. Cervical mediastinoscopy is useful for the evaluation of the mediastinum, particularly inferior to the carina, but would be of little use for a patient with diffuse low attenuation pulmonary parenchyma at HRCT. Medical pleuroscopy would not be useful for this patient as no pleural abnormalities are present. Open surgical lung biopsy would surely establish a diagnosis for this patient but is overly invasive at this point, and could be considered if transbronchial bronchoscopic biopsy is unrevealing and thoracoscopic lung biopsy cannot be performed.

The bronchoscopic transbronchial biopsy revealed non-specific chronic inflammation but was not diagnostic of a particular disorder. The patient subsequently underwent surgical thoracoscopic lung biopsy, and the material retrieved at this procedure showed patchy, peribronchiolar fibrosis surrounding small airways narrowing and obliterating the bronchiolar lumen. The histopathologic features were diagnostic of bronchiolitis obliterans (constrictive bronchiolitis, obliterative bronchiolitis).

Diagnosis: Bronchiolitis obliterans

References

1. Barker AF, Bergeron A, Rom WN, Hertz MI. Obliterative bronchiolitis. *N Engl J Med*. 2014 May 8;370(19):1820-8. [\[CrossRef\]](#) [\[PubMed\]](#)
2. Boisselle PM, Lee KS, Ernst A. Multidetector CT of the central airways. *J Thorac Imaging*. 2005 Aug;20(3):186-95. [\[CrossRef\]](#) [\[PubMed\]](#)
3. Pipavath SN, Stern EJ. Imaging of Small Airway Disease (SAD). *Radiol Clin North Am*. 2009 Mar;47(2):307-16. [\[CrossRef\]](#) [\[PubMed\]](#)
4. Ryu JH, Myers JL, Swensen SJ. Bronchiolar disorders. *Am J Respir Crit Care Med*. 2003 Dec 1;168(11):1277-92. [\[CrossRef\]](#) [\[PubMed\]](#)
5. Kreiss K. Occupational causes of constrictive bronchiolitis. *Curr Opin Allergy Clin Immunol*. 2013 Apr;13(2):167-72. [\[CrossRef\]](#) [\[PubMed\]](#)
6. Parambil JG, Yi ES, Ryu JH. Obstructive bronchiolar disease identified by CT in the non-transplant population: analysis of 29 consecutive cases. *Respirology*. 2009 Apr;14(3):443-8. [\[CrossRef\]](#) [\[PubMed\]](#)
7. Sasieta HC, Nichols FC, Kuzo RS, Boland JM, Utz JP. Congenital Lobar Emphysema in an Adult. *Am J Respir Crit Care Med*. 2016 Aug 1;194(3):377-8. [\[CrossRef\]](#) [\[PubMed\]](#)