May 2012 Pulmonary Case of the Month: Things Are Not Always as They Seem

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History of Present Illness

A 69 year old woman was seen for side effects of corticosteroids. She is a winter visitor to Arizona. She was hospitalized in March 2008 with increased dyspnea and cough and had an abnormal CT chest. A VATS lung biopsy was performed. The pathology of the lung biopsy interpreted as bronchiolitis obliterans. She was started on prednisone 60 mg/day.

Subsequently, she returned to Minnesota and was seen by rheumatologist with a diagnosis made of possible rheumatoid arthritis. She was treated with methotrexate (12.5 mg weekly) and continued prednisone at 20 mg/day from 2008 to 2011. At that time a question was raised of methotrexate lung toxicity and it was stopped but she continued on prednisone 20 to 40 mg/day. She is currently having issues with steroid side effects and seen for a second opinion.

PMH, SH and FH

She has a history of knee and other joint pains. She had knee replacement surgery in Jan 2008 with worsening of her dyspnea and cough. She has a history of diabetes which was apparently induced by the corticosteroids. Her current medications include prednisone 20 mg/day, insulin, metformin, lovastatin. She is a former smoker with 25 pack-years but quit 25 years ago. She has no family history of lung disease.

Physical Examination

She was an obese woman appearing somewhat Cushingoid in no acute distress. On chest auscultation she had diminished breath sounds but no crackles or wheezes. Examination of her joints revealed no abnormalities. The remainder of her physical examination was normal.

Chest X-ray

Her chest x-ray was interpreted as normal.
Which of the following are indicated?
1. Pulmonary function testing
2. Pulmonary CT scanning
3. Rheumatologic evaluation
4. Repeat of open lung biopsy
5. All of the above
1. Pulmonary function testing
2. Pulmonary CT scanning
3. Rheumatologic evaluation

Pulmonary function testing is indicated to assess the severity of her lung disease, especially since her chest x-ray is normal. Pulmonary CT scanning is indicated to further define her chest disease. Rheumatologic evaluation is indicated to assess the activity of her rheumatoid arthritis. Repeat of the open lung biopsy would be overly aggressive at this juncture.

Her pulmonary function testing revealed moderate restrictive disease with a moderate decrease in DLco.

Spirometry

- FVC 62% of predicted
- FEV1 69% of predicted
- FEV1/FVC 86%
- FEF 25-75 89% of predicted
- TLC 63% of predicted
- DLco 47% of predicted

CT scanning was interpreted as showing scattered areas of ground glass opacities and bronchial wall thickening (Figure 1).

Figure 1. A representative lung window from thoracic CT scanning. (Click here for a movie of the CT scan)
Rheumatologic evaluation revealed the following:
  Rheumatoid factor (RF): negative
  Cyclic citrullinated peptide antibody (CCP): negative
  Erythrocyte sedimentation rate: 3 mm/hr
  Hand Films: no destructive changes

What is the next most logical step in her evaluation?
  1. Bronchoscopy with bronchoalveolar lavage
  2. Review the previous open lung biopsy
  3. Repeat the open lung biopsy
  4. Thoracic PET scanning
  5. Increase her prednisone to 40 mg/day
Correct!

2. Review the previous open lung biopsy

Although bronchoscopy with bronchoalveolar lavage is not wrong, it is unclear what would be gained at this point. Repeating the open lung biopsy is again overly aggressive. PET scanning is usually indicated for evaluation of the metabolic activity of lung nodules.

Multiple lung pathologies are associated with rheumatoid arthritis, however, the evidence for rheumatoid arthritis in this patient is weak. Therefore, increasing the prednisone is not indicated. Review of the previous open lung biopsy seems the most reasonable approach.

The previous open lung biopsy was interpreted as showing nodular foci of fibrinous and organizing pneumonitis accompanying chronic small airways disease. There are also areas of granulation tissue within the respiratory bronchioles (Figure 2).

![Figure 2. Low power view (Panel A) with higher power views (Panel B and C).](image-url)
Which of the following most likely explains the patient’s lung pathology?

1. Rheumatoid arthritis
2. Drug reaction
3. Tuberculosis
4. Granulomatosis with polyangitis
5. Chronic aspiration pneumonitis
5. Chronic aspiration pneumonitis

The pathology is consistent with bronchiolitis obliterans and all the diagnoses listed are associated with bronchiolitis obliterans (2). The diagnosis of rheumatoid arthritis is suspect. The drugs she is presently taking were not being taken at the time of the open lung biopsy. There is no evidence for tuberculosis although this remains a possibility with the patient chronically immunosuppressed. Granulomatosis with polyangitis was formerly known as Wegner’s granulomatosis and can be a very difficult diagnosis, especially when limited to the lungs. However, the clinical course is not compatible with this diagnosis.

Chronic aspiration was suggested by the pathologists because of the histological pattern. This seems to be increasingly recognized and can present with diffuse alveolar damage, bronchiolitis and/or organizing pneumonia (3). Chronic aspiration is not always associated with food particles on biopsy. Although usually thought of as a diagnosis where loss of consciousness or decreased gag reflex is present, 40% had gastroesophageal reflux as their only predisposing factor in a recent series (3).

The patient denied any symptoms associated with chronic aspiration. However, on barium swallow she had a hiatal hernia with free reflux to the upper esophagus in the recumbent position. Her steroids are being tapered, she is being treated for her gastroesophageal reflux and she feels well.

Subclinical aspiration should be considered as a possible cause of small airways disease that can appear similar to bronchiolitis obliterans. Pathology can be suggestive, even in the absence of food or other particulate matter. This case illustrates an Osler quote, “Listen to the patient, he is telling you the diagnosis.”. The patient was telling us that she did not have rheumatoid arthritis and so other diagnosis needed to be considered.

References