

## August 2014 Pulmonary Case of the Month: A Physician's Job is Never Done

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### ***History of Present Illness***

A 75-year-old man presented with recurrent minimally productive cough, dyspnea, fatigue, low-grade fevers, and weight loss in November 2013. The patient had been treated twice as an outpatient with antibiotics in the previous 6 weeks for pneumonia.

### ***PMH, FH, SH***

The patient has a history of obstructive sleep apnea but is not compliant with his prescribed continuous positive airway pressure. He also has a history of obesity, dyslipidemia, and peripheral vascular disease.

There is no significant family history.

He is a retired brick layer with a 50 pack-year smoking history but quit a few weeks prior to admission. He drinks a case of beer/week.

### ***Physical Examination***

VS stable. There were no significant findings on physical examination.

### ***Radiography***

A chest radiograph (Figure 1) was performed.

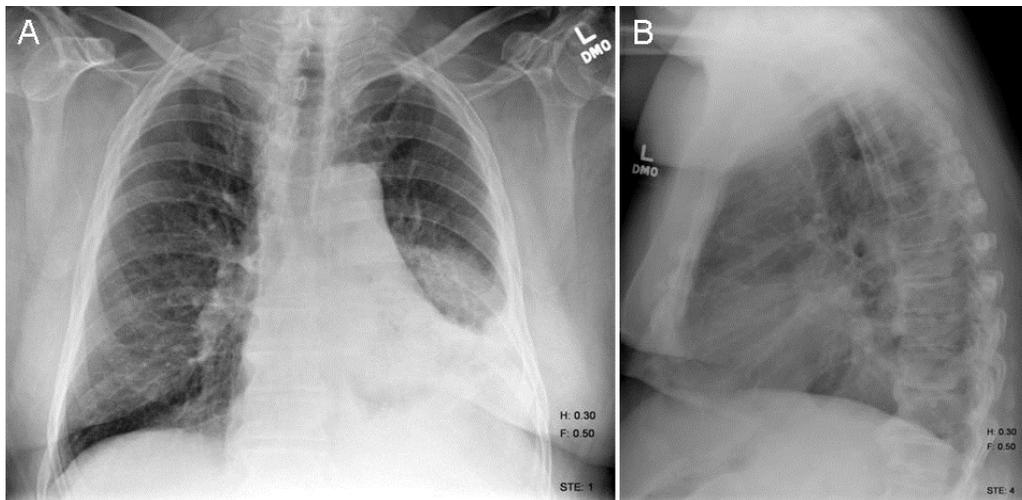


Figure 1. Admission AP (Panel A) and lateral (Panel B) chest radiograph.

What should be **done next?**

1. Bronchoscopy with bronchoalveolar lavage
2. Bronchoscopy with transbronchial biopsy
3. Needle biopsy
4. Thoracentesis
5. Video-assisted thorascopic surgery (VATS)

**Correct!**  
**4. Thoracentesis**

The chest x-ray shows a left lower consolidation and a small left pleural effusion. In a patient with a history of pneumonia the concern would be for an empyema and a thoracentesis is indicated. Bronchoscopy with bronchoalveolar lavage might also be considered. Transbronchial biopsy would not seem to add to the diagnostic yield of bronchoscopy with bronchoalveolar lavage and VATS is overly aggressive at this point. Needle biopsy is usually done for pulmonary nodules.

A thoracentesis was performed and revealed an exudate with negative bacterial cultures. He was treated with an additional course of antibiotics and discharged home. However, he returned in December, 2013 with worsening symptoms.. In addition, he was found to have pancytopenia with the WBC 2.5 million cells/mcL, hemoglobin 8 grams/dL, and platelets 99,000 cells/mcL

A repeat chest x-ray (Figure 2) and thoracic CT scan were performed.

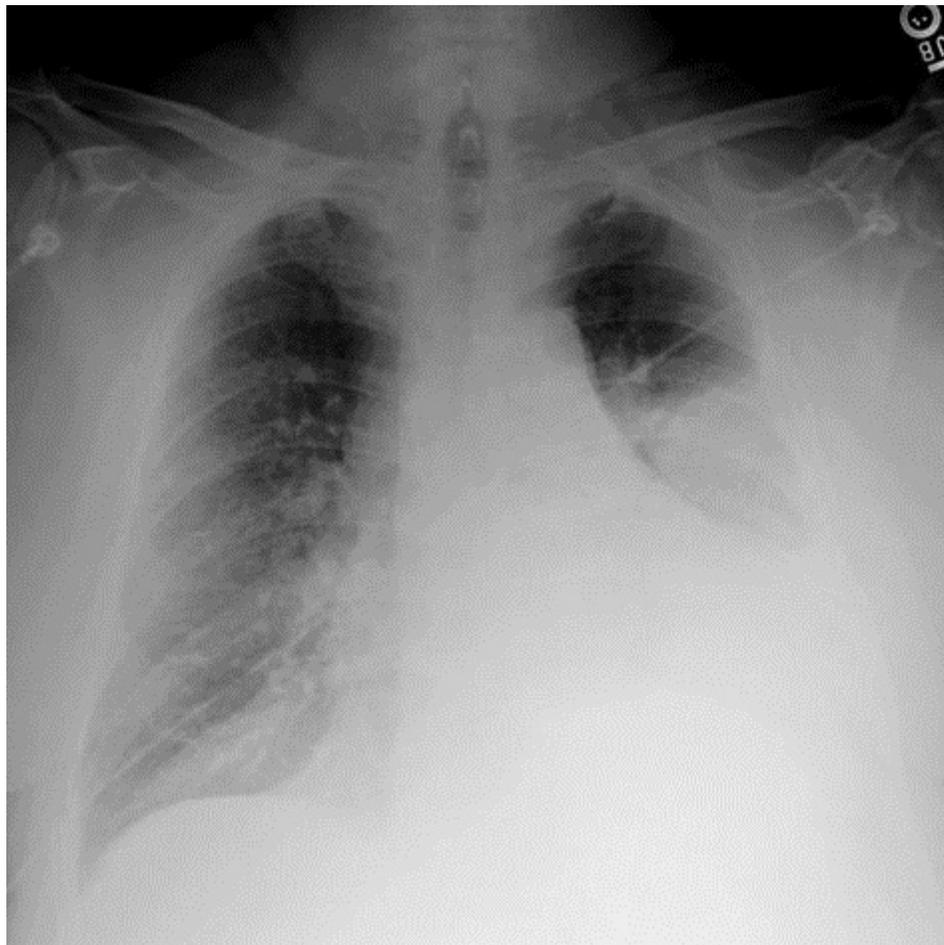


Figure 2. Repeat chest-ray at December 2013 admission.

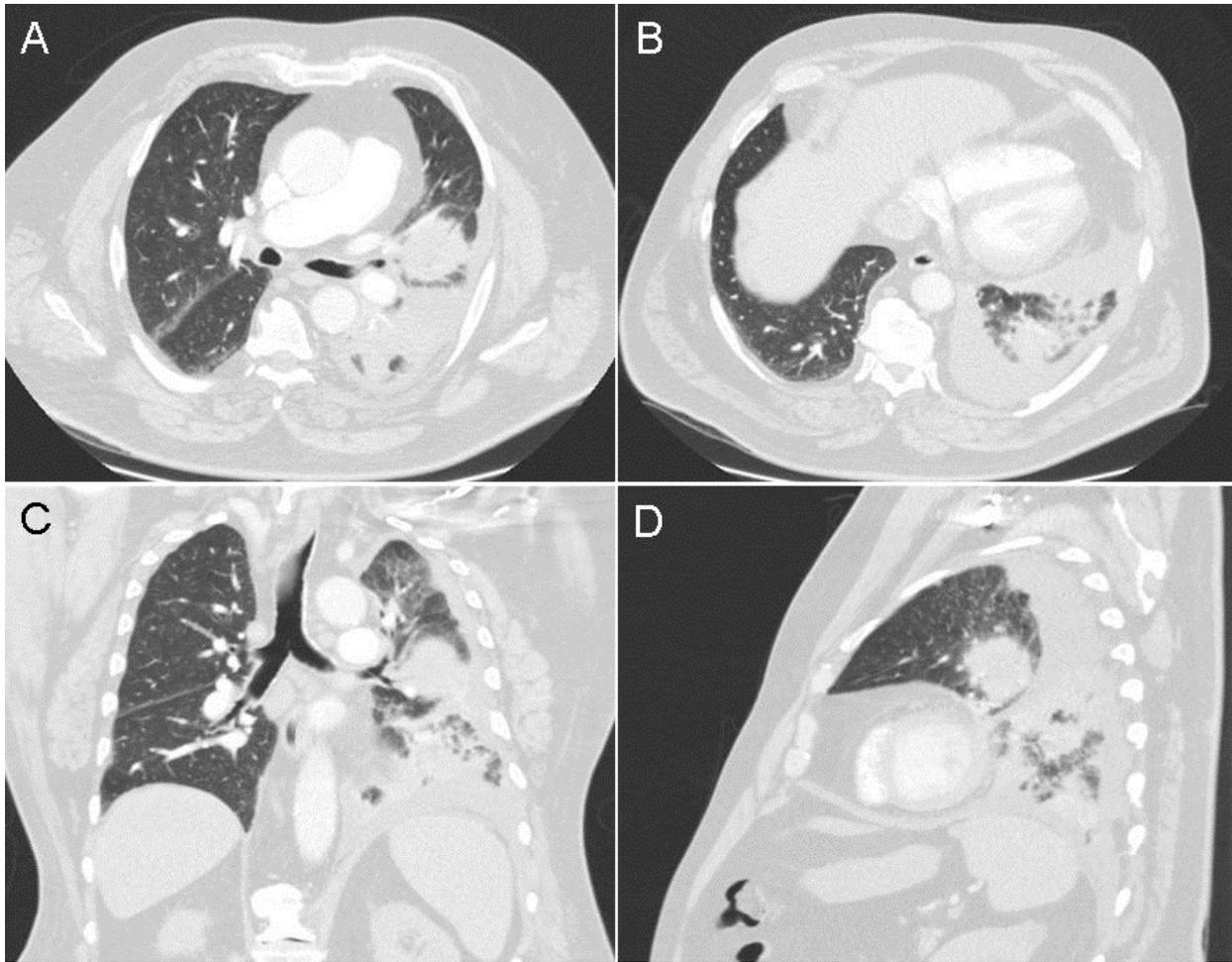


Figure 3. Representative images from the thoracic CT scan in lung windows.

What should be ***done next?***

1. Bronchoscopy with bronchoalveolar lavage, transbronchial biopsy, and brushing
2. More antibiotics adding antifungals
3. Needle biopsy
4. Repeat thoracentesis
5. Video-assisted thorascopic surgery (VATS)

**Correct!**

## **2. Bronchoscopy with transbronchial biopsy**

The patient's course is not compatible with a typical outpatient community-acquired pneumonia. At this juncture we favored a bronchoscopy with a transbronchial biopsy, brushing and lavage. However, needle biopsy and VATS are reasonable considerations.

The bronchoscopic evaluation was negative for any infectious etiology (bacterial, fungal, mycobacterial, viral) and otherwise was unrevealing for etiology. We next elected for CT-guided core needle biopsy which showed patchy involvement of the distal airways and adjacent alveoli filled by fibromyxoid plugs of granulation tissue (Figure 4).

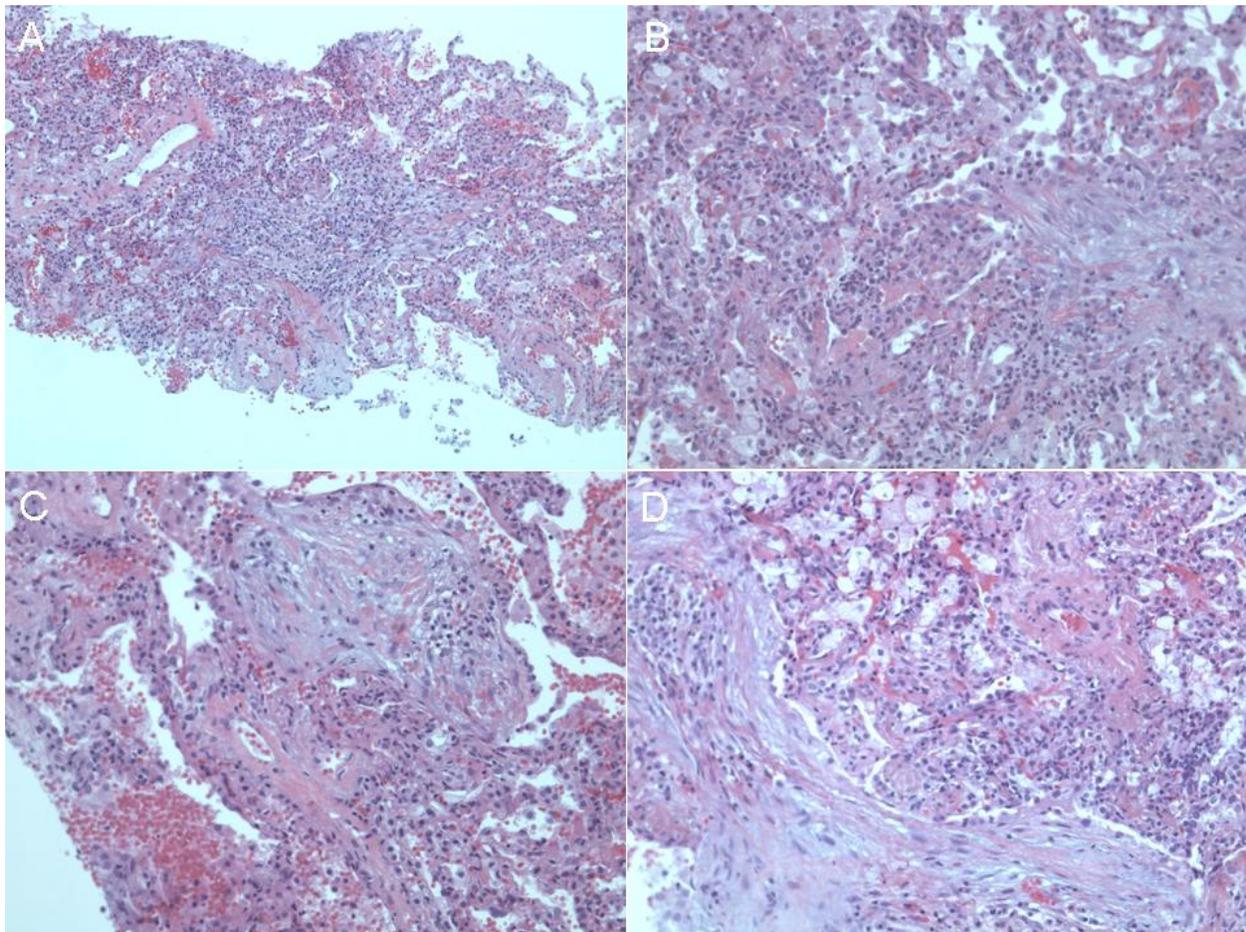


Figure 4. Panel A: Low power view of core biopsy. Panels B-D: Higher power view.

Neutrophils and eosinophils were infrequent and no remodeling or honeycomb changes were observed. The histology was interpreted as fragments of alveolar sac with organizing pneumonia with BOOP-like regions, which is consistent with cryptogenic organizing pneumonia (COP) (1).

Which of the following **is/are true** with cryptogenic organizing pneumonia?

1. Dramatically responds to corticosteroids
2. Has been associated with many diseases including infections, malignancy, radiation and drugs
3. Is the idiopathic form of bronchiolitis obliterans organizing pneumonia (BOOP)
4. Usually presents with the subacute dyspnea and cough and patchy airspace consolidation
5. All of the above

**Correct!**  
**5. All of the above**

The patient was treated with steroids with a prolonged taper. Follow-up in clinic two months later showed almost complete resolution of his consolidation and effusion (Figure 5).

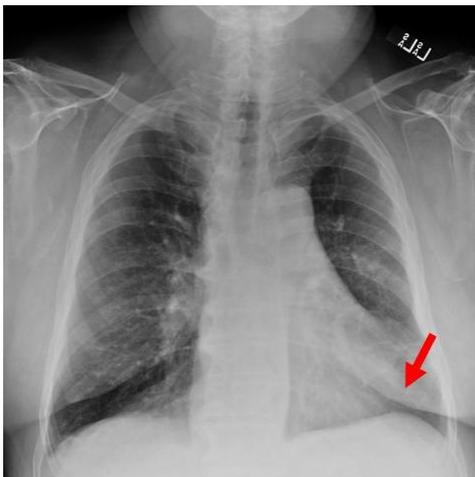


Figure 5. Repeat chest radiograph taken in February 2014. Note clearing in the left lung (red arrow).

He felt better but not great and his pancytopenia had not improved. A hematology consult was ordered as an outpatient.

He was again seen in clinic after two months. He felt "terrible". He had quit taking the corticosteroids due to "bruising easily". His chest radiograph was worse (Figure 6).



Figure 6. Chest radiograph taken in April 2014. Note reappearance of consolidation and effusion in the left lung (red arrow).

He was admitted to the hospital with worsening leukopenia and neutropenia, severe anemia and thrombocytopenia. A hematology/oncology consult was obtained and a peripheral blood leukemia/lymphoma panel was ordered and was normal. He was transfused PRBCs and the corticosteroids were restarted and he is discharged the following day with outpatient hematology follow up.

He comes to the emergency room 2 months later with worsening weakness and pancytopenia and is admitted to the intensive care unit. His chest x-ray and CT scan are again worse (Figure 7).

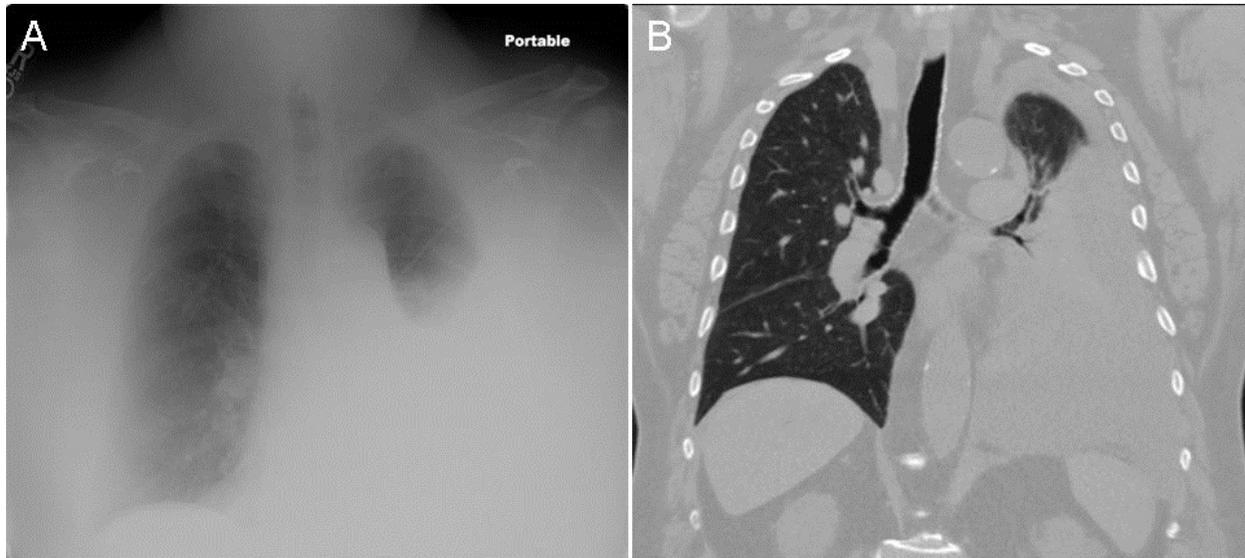


Figure 7. Chest radiograph and representative image from the thoracic CT scan done in June 2014.

What should be **done at this time?**

1. Begin cyclophosphamide
2. Bone marrow aspiration and biopsy
3. Bronchoscopy with bronchoalveolar lavage
4. Repeat the peripheral blood leukemia/lymphoma panel
5. Video-assisted thorascopic surgery

**Correct!**

## **2. Bone marrow aspiration and biopsy**

The pulmonary diagnosis seems well established although the patient could have a superimposed pneumonia. A repeat bronchoscopy with bronchoalveolar lavage was performed but was negative for infection. Following Sutton's law the money appears to be in the bone marrow which was diagnostic of acute myelogenous leukemia. A repeat peripheral blood leukemia/lymphoma panel was ordered and returned about a week later showing 2% blasts detected suggestive of low grade myelodysplastic syndrome (highlighting how inaccurate this test can be compared to bone marrow biopsy). However, the patient was now so weak he elected for hospice and no therapy and subsequently passed away.

Although rare, his leukemia was probably "driving" his organizing pneumonia (2). In retrospect, the assumption that the diagnosis of COP was sufficient was wrong and the search for an underlying disease was not aggressively pursued early on in the patient's course. Eventually the fact that a serious illness was underlying the organizing pneumonia became very evident, but unfortunately was too late. For COP, a physician's job is not done until an underlying disease has been excluded

### ***References***

1. Larsen BT, Colby TV. Update for pathologists on idiopathic interstitial pneumonias. Arch Pathol Lab Med. 2012;136(10):1234-41. [\[CrossRef\]](#) [\[PubMed\]](#)
2. Daniels CE, Myers JL, Utz JP, Markovic SN, Ryu JH. Organizing pneumonia in patients with hematologic malignancies: a steroid-responsive lesion. Respir Med. 2007;101(1):162-8. [\[CrossRef\]](#) [\[PubMed\]](#)