December 2015 Imaging Case of the Month

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Clinical History: An 80-year-old woman with a history of polycythemia vera (12 years), migraines, hypertension, and gastroesophageal reflux disease presented with complaints of declining functional status due to worsening shortness of breath over 3-4 weeks' duration. She also complained of occasional palpitations. No history of fever, cough, chest pain, or hemoptysis was elicited. A frontal chest radiograph (Figure 1) was performed.

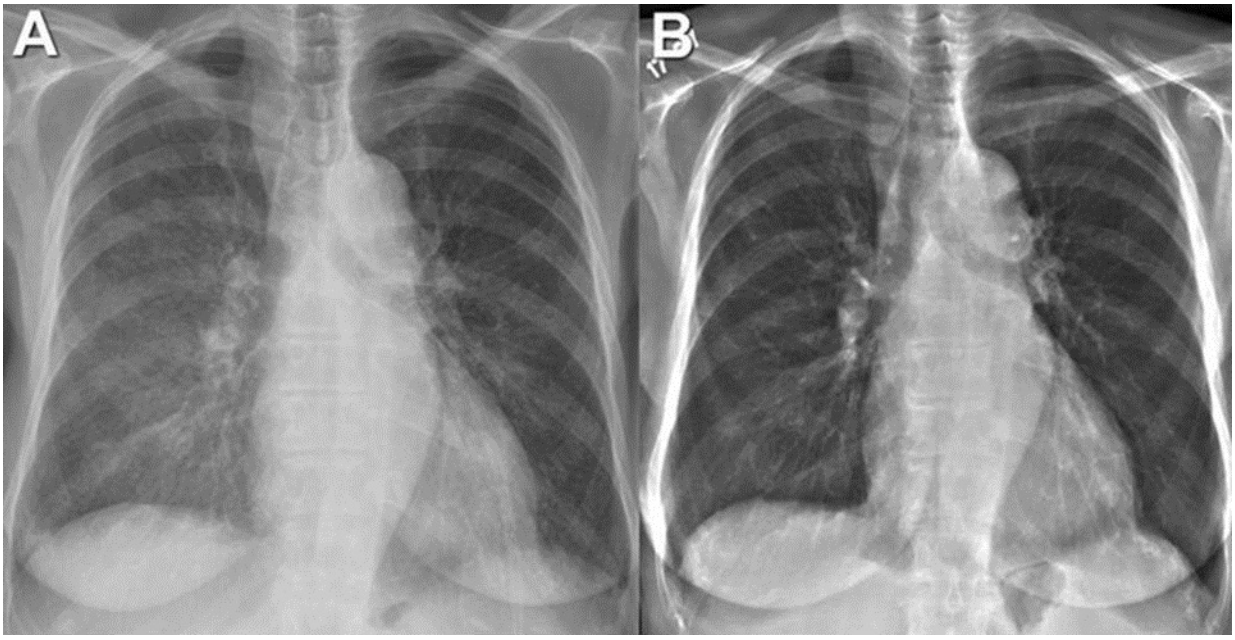

Figure 1. Panel A: Frontal chest radiograph obtained at presentation, when the patient complained of worsening shortness of breath. Panel B: 3 years earlier.

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

1. The frontal chest radiograph shows abnormal mediastinal contours
2. The frontal chest radiograph shows fairly diffuse, bilateral, increased opacity
3. The frontal chest radiograph shows layering pleural effusions bilaterally
4. The frontal chest radiograph shows multifocal consolidation
5. The frontal chest radiograph shows no abnormal findings

Correct!

2. The frontal chest radiograph shows fairly diffuse, bilateral, increased opacity

The presentation chest radiograph (Figure 1A) shows fairly diffuse, right-greater-than-left, bilateral increased attenuation, without clear air bronchograms. Note that the pulmonary vessels are visible through the areas of increased attenuation- an appearance often referred to as “ground-glass opacity” (although this term is more commonly applied to increased attenuation at CT that does not obscure the bronchial walls and vessels). Note that this appearance contrasts with *consolidation*, in which increased lung attenuation obscures the bronchial walls and pulmonary vessels, and is often associated with air bronchograms- no consolidation is evident on the chest radiograph. The mediastinal contours appear normal. Layering pleural effusions can produce a ground-glass appearance at chest radiography, but this situation is typically observed in supine patients, and the patient is in upright position for the chest radiograph in Figure 1A, and no pleural liquid is evident.

The patient’s medications included hydroxurea and diltiazem. She was a non-smoker. Physical examination showed some mild, symmetric lower extremity edema and elevated jugular venous pressure as well as hepatosplenomegaly. The patient’s heart rate varied between 80 and 120, with respirations approaching 20 / minute. Her oxygen saturation on room air was 90%, increasing to 96% on 2 liters oxygen by nasal cannula, but decreasing to 88% with exercise. The patient’s white blood cell count was persistently elevated at 20 – 50 x 10^9 / L with a neutrophilic predominance and a few immature forms. The platelet count was 189,000 10^3 / mcL. Her hemoglobin was diminished at 10.4 gm/dL. Other laboratory data abnormalities were noted as well, including sodium and potassium levels of 153 and 5.3 mmol/L, respectively, and creatinine of 1.9 mg/dL. The patient subsequently underwent thoracic CT (Figure 2).

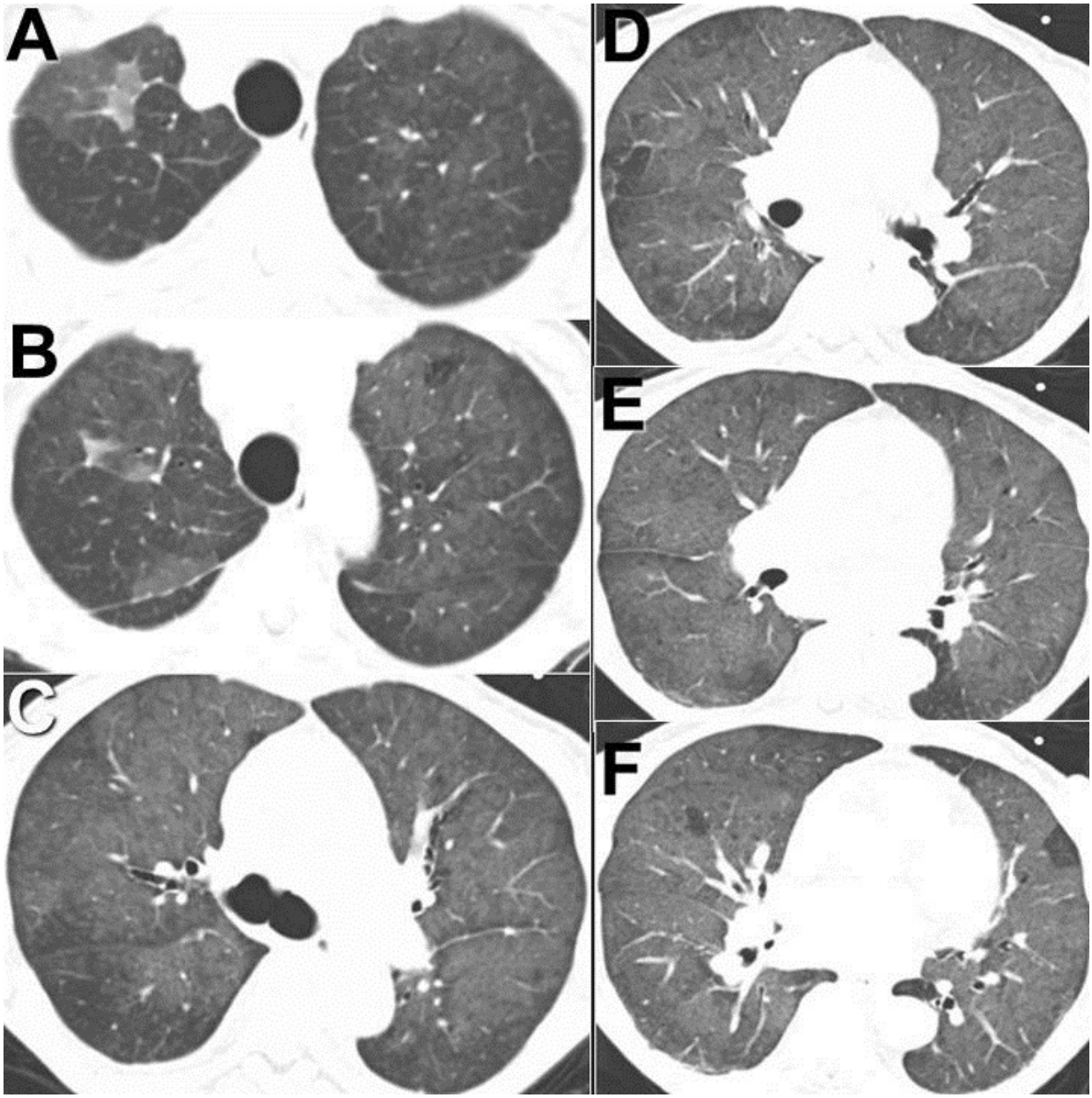

Figure 2. Thoracic CT scan displayed in lung windows.

Which of the following regarding this pulmonary CTA examination is ***correct***?

1. The thoracic CT shows “crazy paving”
2. The thoracic CT shows multifocal areas of consolidation
3. The thoracic CT shows multifocal ground-glass opacities
4. The thoracic CT shows multifocal reticulation and architectural distortion suggesting fibrotic lung disease
5. The thoracic CT shows normal findings

Correct!
3. The thoracic CT shows multifocal ground-glass opacities

The thoracic CT shows multifocal areas of ground-glass opacity bilaterally, but without features to suggest fibrotic lung disease (such as traction bronchiectasis, architectural distortion, reticulation, and / or honeycombing). Additionally, there is no evidence of "crazy paving", typically defined as "thickened interlobular septa and intralobular lines superimposed on a background of ground-glass opacity, resembling irregularly shaped paving stones. The crazy-paving pattern is often sharply demarcated from more normal lung and may have a geographic outline." The differential diagnosis for the "crazy paving" pattern includes a fairly broad spectrum of disorders, but the pattern is often seen in patients with pulmonary alveolar proteinosis. While there is multifocal ground-glass opacity on this patient's CT, there is no interlobular septal thickening associated with the ground-glass opacity, and therefore the "crazy paving" pattern is not present. The areas of increased attenuation on the CT are best described as "hazy increased opacity of lung, with preservation of bronchial and vascular margins"; in other words, *ground-glass opacity*. The terms ground-glass opacity contrasts with *consolidation*, typically defined as "homogeneous increase in pulmonary parenchymal attenuation that obscures the margins of vessels and airway walls," often with air bronchograms; this imaging pattern is not present on the CT images in Figure 1.

The patient underwent echocardiography which showed an ejection fraction of 55% with severe bi-atrial enlargement and left ventricular hypertrophy, and increased echogenicity affecting the left ventricular muscle, suggesting the possibility of cardiomyopathy.

Which of the following represents an ***appropriate consideration*** for the condition affecting this patient?

1. Hydrostatic pulmonary edema
2. Hypersensitivity pneumonitis
3. Lymphocytic interstitial pneumonia
4. Multifocal pulmonary hemorrhage
5. All of the above

Correct!
5. All of the above

All of the aforementioned entities may present on thoracic CT with multifocal ground-glass opacities, on occasion largely unassociated with other abnormalities (see Figure 3).

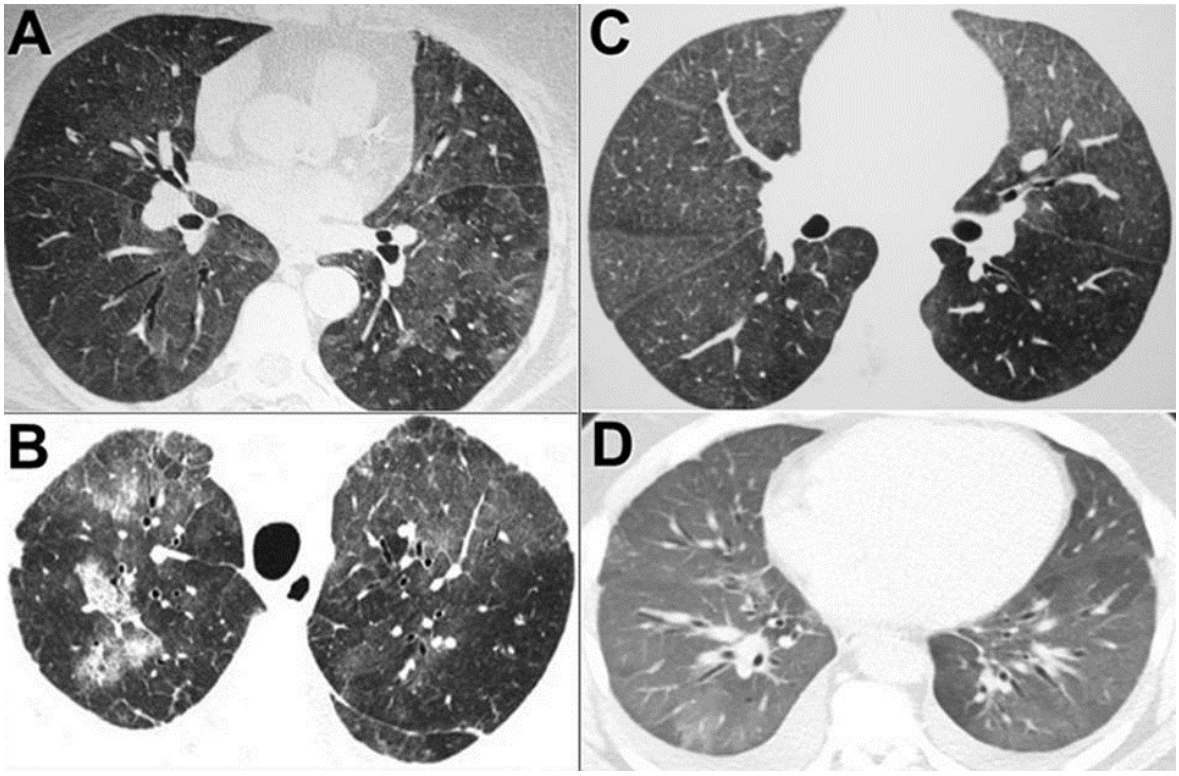

Figure 3. Various pulmonary disorders presenting with multifocal ground-glass opacity. Panel A: hypersensitivity pneumonitis; Panel B: lymphocytic interstitial pneumonia; Panel C: diffuse pulmonary hemorrhage in a patient with systemic lupus erythematosus; Panel D: hydrostatic pulmonary edema.

Hydrostatic pulmonary edema often will manifest with additional findings, such as bronchial wall thickening, pleural effusion, and interlobular septal thickening, but need not present in this fashion in every case; furthermore, the patient's cardiac abnormalities shown at echocardiography raise this possibility.

Which of the following is the ***next most appropriate step*** for the patient's condition given the data thus far?

1. ^{18}FDG-PET scan
2. Bronchoscopy with transbronchial biopsy
3. Mediastinoscopy
4. Percutaneous transthoracic fine needle aspiration biopsy
5. Surgical lung biopsy

Correct
2. Bronchoscopy with transbronchial biopsy

Bronchoscopy with transbronchial biopsy is the most appropriate next step for the evaluation of this patient among the choices lists above. Surgical lung biopsy would certainly provide tissue sufficient to establish a diagnosis, but is needlessly invasive at this point, given that less invasive methods have not yet been exhausted and may still provide an answer. In general, ^{18}FDG-PET scanning is most useful for the diagnosis and staging of lung cancer metastatic disease, and for the evaluation of focal pulmonary opacities, but not diffuse lung diseases. Similarly, percutaneous transthoracic fine needle aspiration biopsy is not typically employed for diffuse lung diseases, and is instead more commonly used to diagnose focal pulmonary lesions. Finally, mediastinoscopy is useful for diagnosing mediastinal lesions (typically in the middle mediastinum in the pretracheal region), but no such abnormality is present in this patient.

The patient underwent bronchoscopy with transbronchial biopsy without complication. Bacterial, viral, and fungal cultures were negative and endobronchial examination showed no abnormalities. Bronchial lavage showed cell count and differential of 27% polymorphonuclear cells, 5% lymphocytes and 68% monocytes – macrophages. The transbronchial biopsy specimen showed tissue containing islands of erythroid precursors, myeloid series, and both mature and immature megakaryocytes, consistent with extramedullary hematopoiesis.

Diagnosis: Extramedullary hematopoiesis in the lungs

References

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