January 2017 Pulmonary Case of the Month

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History of Present Illness

The patient is a 53-year-old woman transferred for acute respiratory failure and hemoptysis. She has a prior history of antiphospholipid syndrome and recurrent diffuse alveolar hemorrhage (DAH). She was admitted to another hospital about 2 weeks prior to transfer with hypoxic respiratory failure which ultimately required intubation. Bronchoscopy revealed a bloody aspirate raising concerns for recurrent DAH. She was started on high-dose solumedrol and extubated after 4 days. One week later, her respiratory status decompensated and her chest x-ray showed worsening diffuse bilateral opacities concerning for recurrent DAH. She was transferred to the Mayo Clinic Arizona for further evaluation. Upon arrival, she required 50% FiO2 by face mask to maintain adequate oxygenation and was started on broad-spectrum antibiotics. Her corticosteroids were tapered to 20 mg prednisone daily.

Past Medical History, Social History and Family History

She has a history of a mitral valve replacement with a St. Jude's mechanical mitral valve and was on chronic anticoagulation with warfarin. In addition, there was a history of moderate aortic stenosis with moderate aortic insufficiency.
She had a history of diffuse alveolar hemorrhage, antiphospholipid antibody syndrome and possible systemic lupus erythematosus.

Medications

- Dapsone 100mg daily
- Ethacrynic acid 75mg daily
- Gabapentin 900mg QHS
- Lisinopril 20mg daily
- Meropenem 1g Q8 hrs
- Metoprolol 50 mg BID
- Prednisone 20mg daily
- Simvastatin 40mg QHS
- Vancomycin 1.5g Q12 hrs
- Warfarin 4mg T,F; 3mg SMWRSa

Physical Examination

- Vitals: T 36.3° C; HR 79 beats/min; BP 100/63 mm Hg; RR 26 breaths/min; SpO2 99% face mask
- Gen: no acute distress
- HEENT: hematoma on chin
- Lungs: clear to auscultation and percussion

- Cardiac: Mechanical valve click

Laboratory

- CBC: WBC 15,900 cells per microliter (mcL); Hemoglobin 9.1 g/dL; hematocrit 29%; platelet count 156,000 cells per microliter.
- Electrolytes: within normal limits.
- BUN and creatinine: within normal limits.
- Blood sugar: 220 mg/dL.

Radiography

Her initial chest x-ray is shown in Figure 1.

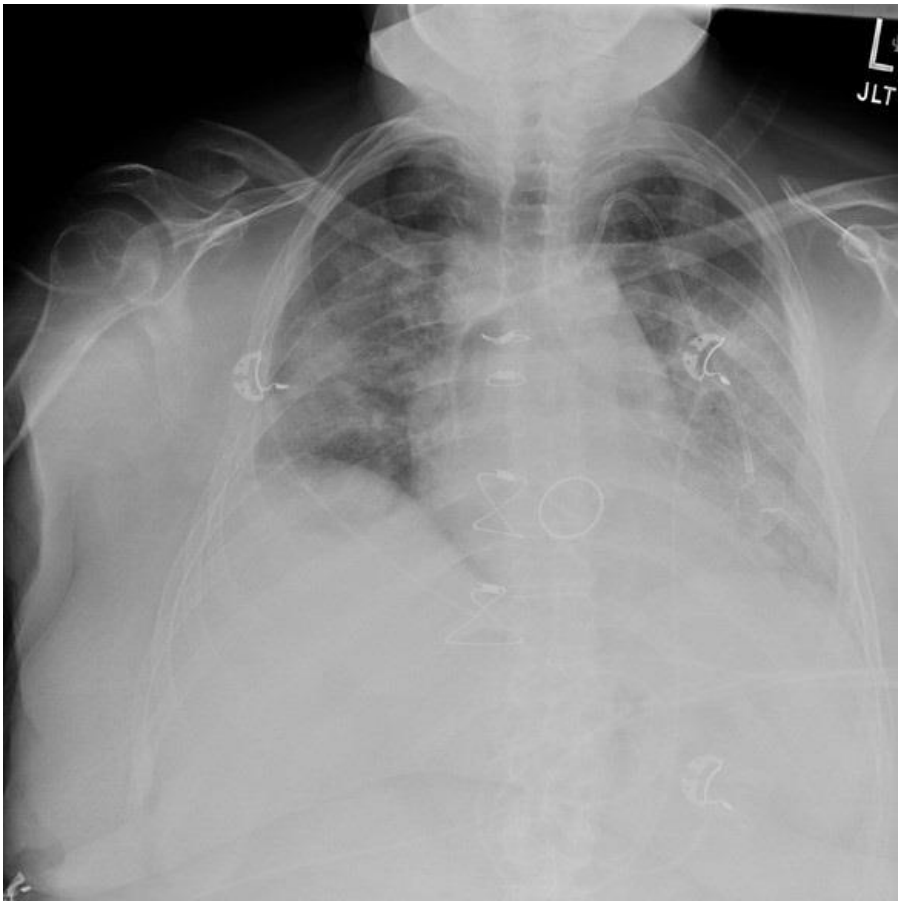

Figure 1. Initial chest radiograph.

Which of the following ***best describes*** the chest x-ray?

1. Diffuse lung consolidation
2. Previous median sternotomy
3. Previous mitral valve replacement
4. 1 and 3
5. All of the above

Correct!
5. All of the above

There are diffuse, patchy areas of lung consolidation. A ring is present which is in the area of the heart that would be anticipated with a mitral valve replacement. An aortic valve would be anticipated to be superior and to the patient's right of the mitral valve. There are wires characteristic of a previous median sternotomy present.

A thoracic CT scan was performed (Figure 2).

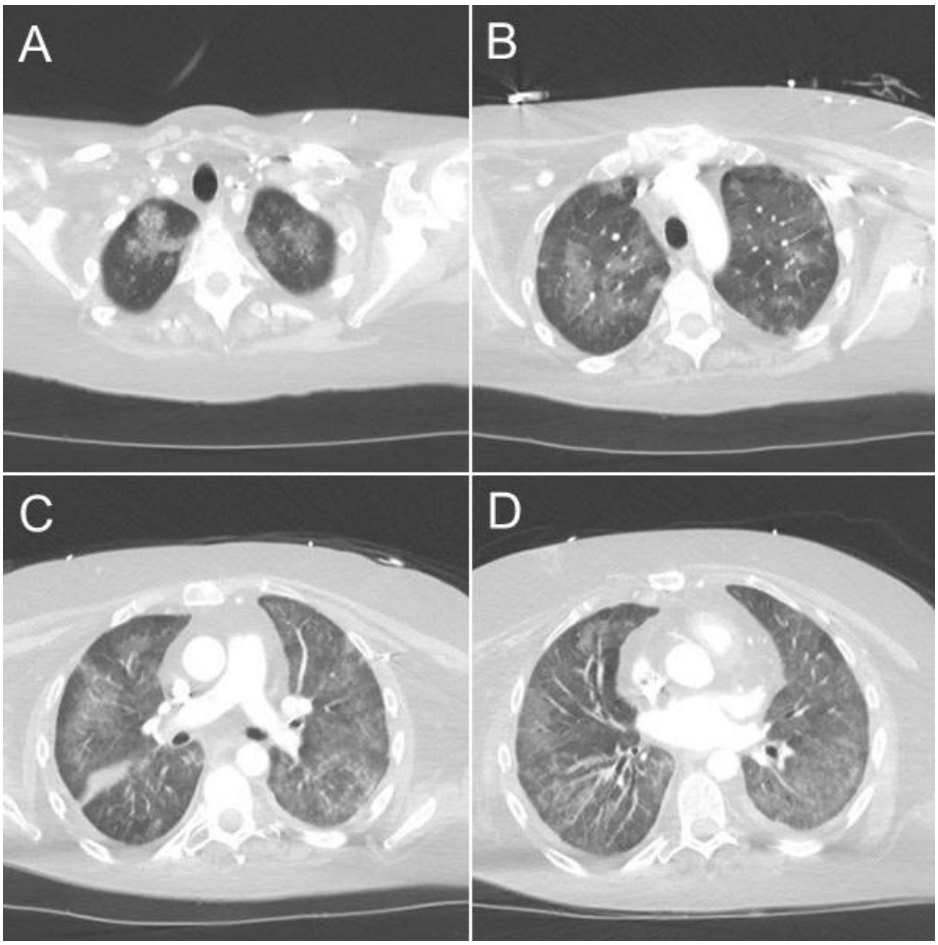

Figure 2. Representative images in lung windows from the thoracic CT scan.

Which of the following should be ***done next***?

1. Bronchoscopy
2. Video-assisted thorascopic surgery (VATS)
3. Cardiac ultrasound
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

The patient has at least 2 reasons for hemoptysis-a connective tissue disease and mitral valve disease. Usually, hemoptysis indicates a bronchial process such as bronchitis or lung cancer. However, there is no clinical signs to suggest either of these two common processes. It was felt that a transesophageal echocardiogram (TEE) and bronchoscopy would likely both be necessary and both procedures were simultaneously performed

The TEE showed a left ventricular ejection fraction of (LVEF) 74% with a mitral valve diastolic mean Doppler gradient of 23 mm Hg and a right ventricular systolic pressure (RVSP) 57 mm Hg.

Bronchoscopy showed blood within the airways. After washing to remove the blood, the bronchial mucosa was hyperemic with dilated submucosal veins (Figure 3).

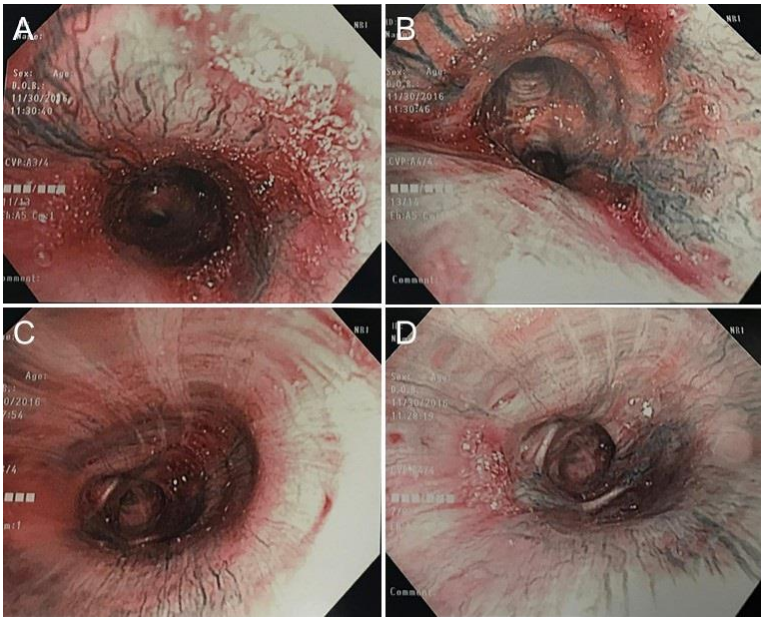

Figure 3. Representative bronchoscopic images showing bronchial hyperemia and mucosal vein dilation.

Bronchoalveolar lavage resulted in a bloody return which did not clear.

What is the most likely ***cause*** of her hemoptysis?

1. Acute infectious bronchitis
2. Diffuse alveolar hemorrhage secondary to connective tissue lung disease
3. Mitral stenosis
4. Occult lung cancer
5. None of the above

Correct!
3. Mitral stenosis

The cardiac ultrasound showed a markedly elevated mitral valve gradient of 23 mm Hg with an elevated RVSP of 57 mm Hg. A gradient of >10 mm Hg is considered severe. Furthermore, the ultrasound showed a stationary lateral mitral valve leaflet secondary to a thrombus. This resulted in functional mitral stenosis.

Patients with chronically elevated left atrial pressures from mitral stenosis have engorged pulmonary veins (1). The pulmonary veins transmit the excess pressure into the bronchial venous plexus which transmit the pressures to the bronchial veins. This results in the intensely hyperemic mucosa with dilatation of submucosal veins as seen in our patient.

Hemoptysis, regardless of the amount, is usually considered an indication for bronchoscopy, especially when it is unexplained and/or recurrent (2). Massive hemoptysis has no standard definition but occurs when the amount or the rate of bleeding becomes potentially life-threatening. Patients with an intact cough reflex usually are able to clear their airways, but when bleeding is >500 ml in 24 hours or >100 ml in an hour, bleeding may overwhelm the patient's ability to expectorate the blood (3). Urgent bronchoscopy should be performed in unstable patients. It can be used to facilitate the introduction of balloon-tip catheters into the bleeding bronchus for tamponade of the hemorrhagic artery, protecting the contralateral lung or nonbleeding bronchi from blood aspiration. Endobronchial tamponade should only be used as a temporary measure until a more specific treatment is instituted. In stable patient computed tomography should be ordered before any bronchoscopic exploration.

References

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