January 2013 Pulmonary Case of the Month: Maybe We Should Call GI

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History of Present Illness

A 55 year old man from Arizona was undergoing a renal transplant evaluation because of polycystic kidney disease. He was referred for an abnormal chest x-ray. He was a nonsmoker and there were no respiratory symptoms.

PMH, FH and SH

He has a long history of polycystic kidney disease, hypertension, gout, and a history of a kidney stone. He is a life-long nonsmoker. There is no significant family history including polycystic kidney disease. He works as a border patrol agent and is originally from Honduras. His present medications include:

- Allopurinol
- Amlodipine
- Atenolol
- Hydralazine
- Sodium bicarbonate

Physical Examination

His blood pressure is elevated at 142/84, but otherwise his physical examination is unremarkable.
Chest X-ray

His chest X-ray is below (Figure 1).

Figure 1. PA (Panel A) and lateral (Panel B) chest x-ray.

The chest x-ray was interpreted as showing bilateral lower lobe nodules.

Which of the following is appropriate?

1. Obtain old chest x-rays for comparison
2. Spiral CT for pulmonary embolism
3. Coccidioidomycosis serology
4. A + C
5. All of the above
Old x-rays should be compared to the most recent x-ray to determine the age of any lesion(s). This would be a very unusual presentation both clinically and radiographically for a pulmonary embolus. In areas such as Arizona where Valley Fever is endemic a coccidioidomycosis serology is indicated when evaluating most chest processes.

In our patient old chest x-rays did not show the lower lobe lesions and a coccidioidomycosis serology was negative. To better define the lesions a chest CT was done (Figure 2).

Figure 2. Representative images from the thoracic CT scan.

What is the next most appropriate step?
1. Pulmonary angiography
2. Bronchoscopy
3. Needle biopsy
4. Video-assisted thorascopy (VATS)
5. Coil embolization
Correct!

1. Pulmonary angiography

There is an obvious vascular abnormality present on the thoracic CT scan. However, it is unclear what abnormality is present and pulmonary angiography is needed to further define the lesions.

Pulmonary angiography was performed (Figure 3).

Figure 3. Static views of the pulmonary angiogram with matching negative images.

What abnormality is present on the pulmonary angiogram?

1. Pulmonary embolus
2. Pulmonary AV malformation
3. Pulmonary varices
4. Pulmonary artery aneurysm
5. Anomalous pulmonary venous return
3. Pulmonary varices

A filling defect is not present as would be seen with a pulmonary embolus. A large feeding pulmonary artery is not present as would be seen in pulmonary AV malformation (1). The lesion appears to be in the pulmonary venous circulation rather than the arterial circulation as abnormalities were only noted on the delayed venous phase of the pulmonary angiogram. There is no evidence of anomalous pulmonary venous return which occurs when pulmonary venous blood enters the systemic circulation or the right heart (1).

Pulmonary varices are dilatation of the pulmonary veins like varicose veins in the systemic circulation. These are very rare with only 71 cases reported in 1988 (2).

Which of the following are true regarding pulmonary varices?
1. Generally are asymptomatic and considered benign
2. Most occur as a result of congenital cardiac disease
3. Most frequently occur in the left lower lobe
4. Need to be coil embolized to prevent massive hemoptysis
5. Require surgical resection to prevent massive hemoptysis
Correct!
1. Generally are asymptomatic and considered benign

Pulmonary varices usually occur in the third to sixth decade of life, are asymptomatic and considered benign. However, complications such as hemoptysis, recurrent infection, and cerebral emboli have been reported (2). They may be congenital or acquired. When acquired they often occur as a result of increased pulmonary venous pressure such as seen with mitral valve disease, coarctation of the aorta, or pulmonary venous stenosis. Most are located in the right lower lobe (RLL, 60%) with only 4% located in the LLL. To my knowledge there are no prior reports of bilateral lower lobe varices or an association with polycystic kidney disease.

References