

## December Pulmonary Case of the Month: A Young Man with Multiple Lung Masses

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

A 28-year-old man from Tennessee has been feeling ill with malaise and weight loss for the past 3 months. He had been in the in the Palm Springs area a few weeks prior to becoming ill. He works as a musician.

### ***Past Medical History, Social History and Family History***

He has a history of Wolf-Parkinson-White syndrome and had a prior ablation procedure at age 16. He does not smoke tobacco but does smoke marijuana occasionally. Family history is noncontributory.

### ***Physical Examination***

Physical examination was unremarkable.

Which of the following are ***indicated at this time?***

1. Bronchoscopy with EBUS
2. Chest X-ray
3. VATS
4. 1 and 3
5. All of the above

**Correct!**  
**2. Chest X-ray**

His symptoms are nonspecific but are commonly seen with a number of pulmonary processes. The part that is troubling is the prolonged time frame. For this reason, a chest x-ray was performed which revealed multiple pulmonary nodules. Bronchoscopy is probably premature at this juncture and certainly video-assisted thorascopic surgery (VATS) is premature.

Which of the following are ***indicated at this time?***

1. Coccidioidomycosis serology
2. Histoplasmosis serology
3. Thoracic CT scan
4. 1 and 3
5. All of the above

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

The CT scan confirms the presence of multiple nodules (Figure 1).

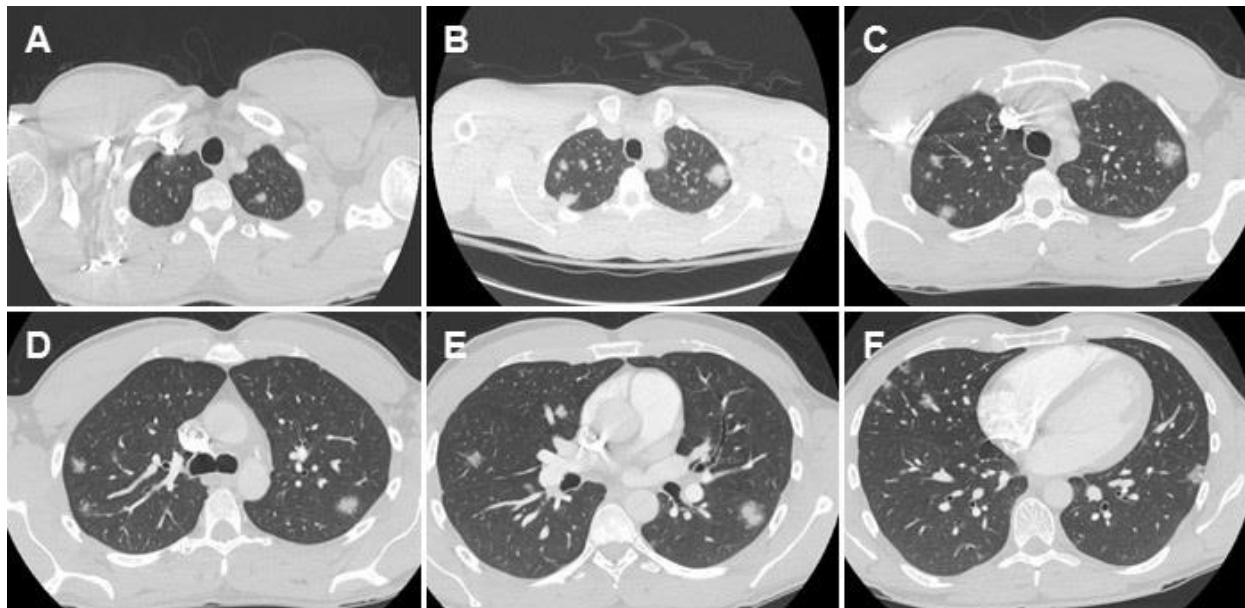


Figure 1. Representative images from thoracic CT scan in lung windows showing multiple pulmonary nodules. Soft tissue windows (not shown) showed a few marginally enlarged mediastinal lymph nodes.

The fungal infections, coccidioidomycosis and histoplasmosis, are reasonable considerations. Both can produce nonspecific symptoms in many patients over several months' duration. Histoplasmosis is common in the Midwest and Ohio River Valley (1). Coccidioidomycosis, Valley Fever, is common in the Southwest (2). The serologies for histoplasmosis and coccidioidomycosis are both drawn and sent to a reference laboratory.

Which of the following are ***indicated at this time***?

1. Bronchoscopy with EBUS
2. Pulmonary angiography
3. VATS
4. 1 and 3
5. All of the above

**Correct!**

### **1. Bronchoscopy with EBUS**

Bronchoscopy might be a simple and the least invasive way of making a diagnosis. VATS still seems overly aggressive when bronchoscopy has a reasonable expectation of making a diagnosis. Pulmonary angiography is usually reserved for pulmonary vascular disease, especially pulmonary embolism. There is no reason to suspect pulmonary embolism based on his clinical course or radiology.

A bronchoscopy with transbronchial biopsy was performed with endobronchial ultrasound (EBUS) lymph node biopsy. Both showed nonspecific findings. The EBUS showed lymphocytes at station 7 which were not abnormal. The transbronchial biopsy showed a few hemosiderin-laden alveolar macrophages with some features of organizing pneumonia. Needle biopsy was recommended but the patient refused.

Which of the following are **indicated at this time?**

1. Begin fluconazole while awaiting the coccidioidomycosis serology
2. Begin itraconazole while awaiting the histoplasmosis serology
3. CT/PET scanning
4. 1 and 3
5. All of the above

**Correct!**

- 1. Begin fluconazole while awaiting the coccidioidomycosis serology**
- 2. Begin itraconazole while awaiting the histoplasmosis serology**

The most likely diagnoses are histoplasmosis or coccidioidomycosis and there is little that makes one more likely than the other at this juncture. He was begun on itraconazole. Itraconazole is more commonly used for histoplasmosis and fluconazole for coccidioidomycosis although there is not strong clinical evidence to support the drug preference in either disease. Both the histoplasmosis and coccidioidomycosis serologies eventually returned negative but the itraconazole was continued because both serologies have around 70% sensitivity (1,2). CT/PET scanning is usually used for lung cancer which seems unlikely in a 28-year-old nonsmoker and PET positivity has been reported with both active histoplasmosis and coccidioidomycosis.

His thoracic CT scan was repeated three months later (Figure 2).

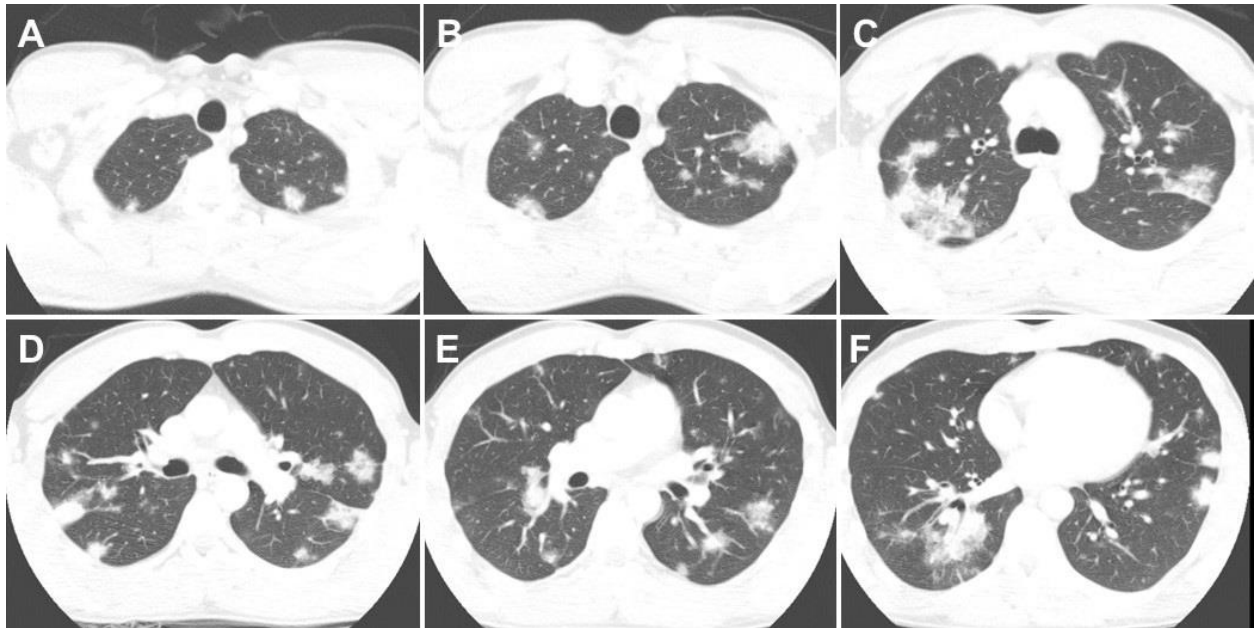


Figure 2. Representative images from the repeat thoracic CT scan in lung windows performed about 3 months after the first CT can (Figure 1).

What is the **best interpretation** of the repeat CT scan?

1. The masses are homogenous with well-defined margins
2. The masses are mostly calcified
3. The masses now show cavitation
4. There has been progression and enlargement of the lung masses
5. There has been regression and shrinking of the lung masses

**Correct!**

**4. There has been progression and enlargement of the lung masses**

Clearly the nodules have enlarged. There is some inhomogeneity of the nodules but none are clearly cavitated. The margins are indistinct and irregular. Clinically the patient was feeling no better and his weight loss had continued.

What is ***indicated at this time?***

1. Needle biopsy of the lung
2. Press for a VATS biopsy
3. Repeat the bronchoscopy
4. Repeat the histoplasmosis and coccidioidomycosis serologies
5. Switch from itraconazole to fluconazole

**Correct!**  
**2. Press for a VATS biopsy**

With progressive disease a diagnosis needs to be made. It seems unlikely that switching antifungals, repeating the bronchoscopy or repeating the fungal serologies will be successful. Needle biopsy is a consideration but it was felt that obtaining a definitive diagnosis should be done. The patient agreed to the VATS biopsy which was performed.

The biopsy showed areas of tumor cells and hemorrhage (Figure 3A). Immunohistochemistry for erythroblast transformation specific related gene (ERG) was strongly positive within the tumor cells confirming the vascular origin of the tumor cells (Figure 3B). Pleural involvement was also seen.

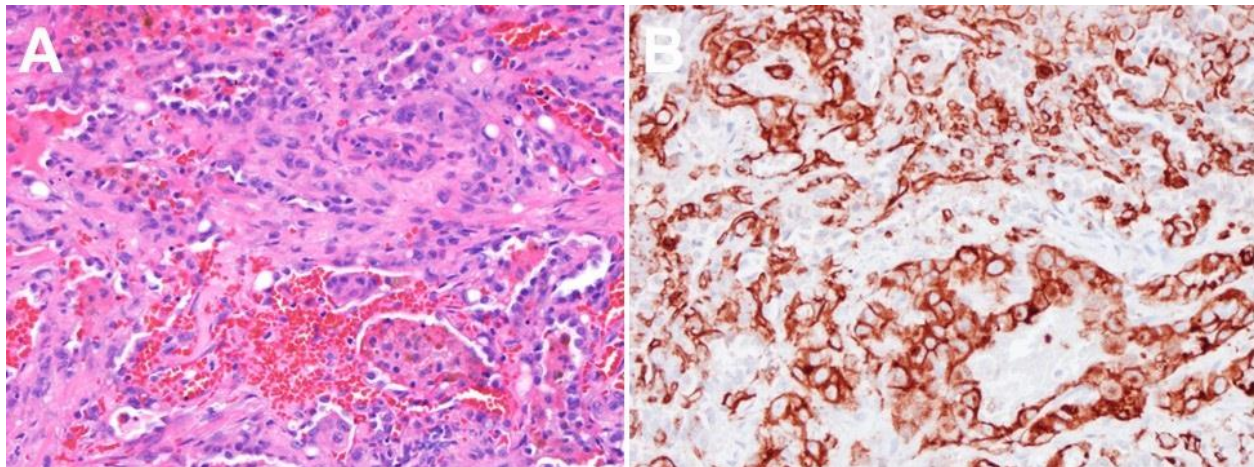


Figure 3. A: High power view showing malignant cells and hemorrhage. B: Immunohistochemistry for ERG.

A diagnosis of angiosarcoma was made.

Which of the following are **true** regarding angiosarcoma?

1. Administration of high doses of recombinant interleukin 1 (rIL-1) may be effective
2. Angiosarcomas respond well to chemotherapy
3. It is the most common cancer seen in the 20-30 age group
4. Most angiosarcomas originate in the lung
5. Prognosis with angiosarcoma is poor



**Correct!**

### **5. Prognosis with angiosarcoma is poor**

Angiosarcoma is an uncommon tumor but can affect young adults as well as the old (3). It usually presents with multiple solid lung nodules or masses but can present with cavitating lesions or even ground glass opacities or consolidation (4). Most cases in the lung are thought to represent metastatic disease, however, primary pulmonary angiosarcoma has been reported although it is very rare (5). There is no standard therapy (3,5). Surgical resection, radiation, and chemotherapy have all been attempted. Surgery has been the mainstay for locally confined disease. Chemotherapy and radiotherapy often produce an initial response. Systemic administration of high doses of recombinant interleukin 2 (rIL-2) also has been reported to be effective in primary angiosarcoma of the lung (6). However, regardless of the treatment prognosis usually remains poor. Median survival has been reported at 7 months (3).

### **References**

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