January 2013 Imaging Case of the Month

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Clinical History: A 40-year-old previously healthy man presented with complaints of cough with blood-streaked sputum. Frontal and lateral chest radiography (Figure 1) was performed.

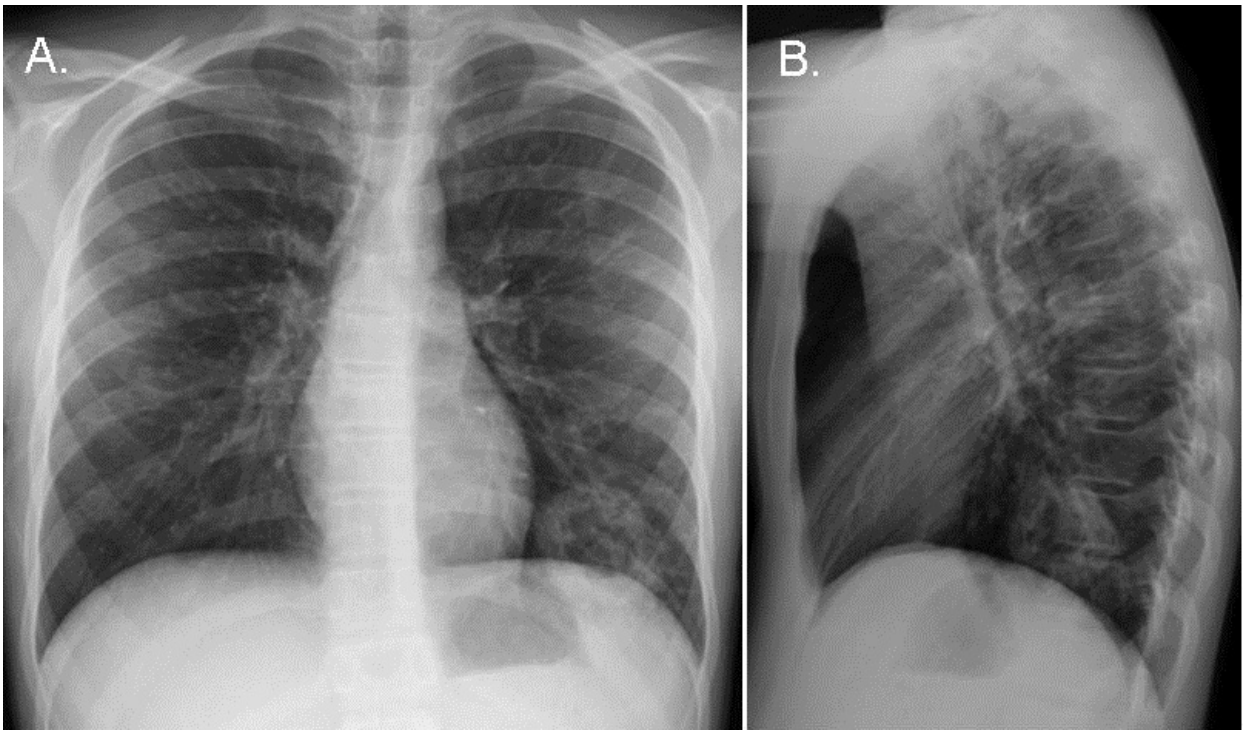

Figure 1. Frontal (A) and lateral (B) chest radiography.

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

1. The chest radiograph shows focal consolidation
2. The chest radiograph shows a loculated left pleural effusion
3. The chest radiograph shows pulmonary cavities
4. The chest radiograph shows tubular opacities suggesting arteriovenous malformations
5. The chest radiograph shows a left diaphragmatic hernia

Correct!
3. The chest radiograph shows pulmonary cavities

A focal opacity is present within the left lower lobe, but it is not consolidation (i.e., homogeneous increased lung attenuation with obscuration of vascular and bronchial wall margins, often associated with air bronchograms); rather, the left lower lobe lesion is lucent centrally, consistent with a gas-containing lesion, or lung cavity. No pleural abnormalities are seen. Streaky perihilar opacities, consistent with airway thickening, are present bilaterally, but no discrete, tubular opacity associated with a nodule- a morphology suggestive of arteriovenous malformation- is seen. The lesion at the left base is near the location that would be expected for a diaphragmatic hernia, but this lesion is gas-containing, representing a pulmonary cavity, and is not solid-appearing, as would be expected for a diaphragmatic hernia. Furthermore, a diaphragmatic hernia will usually obscure the diaphragm contour where the diaphragmatic defect is located, creating an *incomplete border sign* (typical of a lesion outside the lung parenchyma); however, on this image, the entire circumference of the left lower lobe cavity is visible, indicating that it is completely surrounded by air.

The patient subsequently underwent thoracic CT (Figure 2) for further characterization of the pulmonary abnormalities seen at chest radiography.

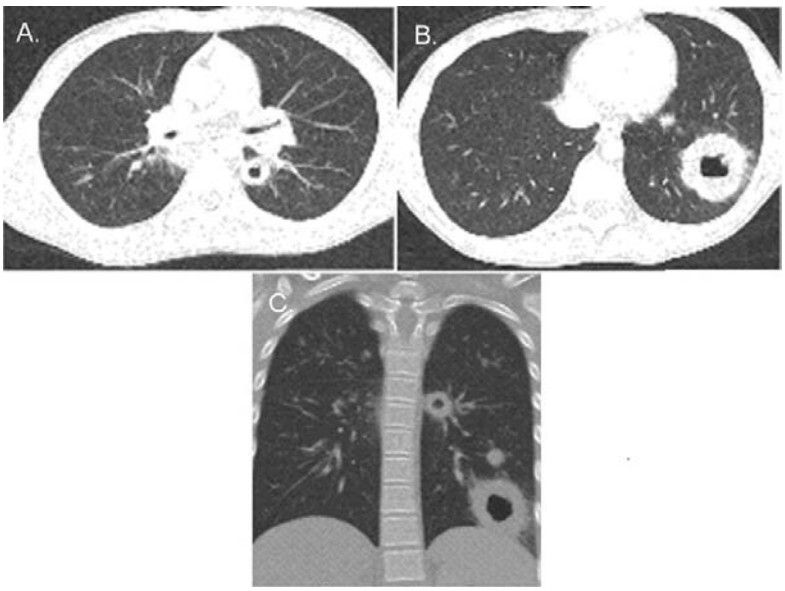

Figure 2: Thoracic CT displayed in lung windows (axial A and B, coronal C).

Regarding the thoracic CT, which of the following statements is ***most accurate***?

1. The thoracic CT shows multiple nodules and cavities
2. The thoracic CT shows small perilymphatic nodules
3. The thoracic CT shows a random dissemination, or “miliary,” pattern
4. The thoracic CT shows multifocal ground-glass opacity
5. The thoracic CT shows multifocal high attenuation mucous impaction

Correct!
1. The thoracic CT shows multiple nodules and cavities

Multiple variably sized nodules and cavities are present. The dominant lesion, as seen on the chest radiograph, is present in the left lower lobe, but another cavity is present in the medial aspect of the superior segment of the left lower lobe, and small nodules are also seen in the upper and lower lobes bilaterally. The term “perilymphatic” is usually applied to small nodules, typically ≤1 cm, that are primarily seen along bronchovascular structures, interlobular septae, and fissural surfaces. Perilymphatic nodules are commonly encountered in diseases affecting the lymphatic system, such as sarcoidosis and caricinomatosis. While some of the nodules on this thoracic CT are seen along bronchovascular bundles, the nodules are not related to the fissures, making a perilymphatic distribution unlikely. Similarly, a random, or “miliary,” pattern is typically said to be present when small nodules are seen diffusely distributed throughout the lung, relatively equally in relation to fissural surfaces, along bronchovascular structures, and in the centrilobular regions. This pattern is typical of processes spreading hematogenously to the lung, but is not present in this case. Multifocal ground-glass opacity is not present in this case. A few foci of ground-glass attenuation are seen around the dominant left lower lobe cavity and in a peribronchial location, but these findings are minor in comparison with other abnormalities on the scan. High attenuation mucous impaction, typical of allergic bronchopulmonary aspergillosis, cannot be diagnosed when presented with only lung window images; soft tissue windows are needed for such an assessment. The slightly wider window widths and levels used for the coronal image set (Figure 2C) do not show high attenuation bronchial impaction. Finally, while some peribronchial opacity is present, clear evidence of bronchiectasis and mucous impaction is not seen.

Regarding the assessment of lung cavities at chest radiography, which of the following is ***correct***?

1. The wall thickness of a lung cavity should be assessed at its thickest portion
2. The presence of an air-fluid level within a lung cavity is diagnostic of lung abscess
3. Malignant causes of lung cavities usually produce cavities with a smooth inner lining
4. When the thinnest portion of a lung cavity is <2 mm, the cavity is unequivocally benign

Correct!

1. The wall thickness of a lung cavity should be assessed at its thickest portion

If the thickest portion of the cavity wall is ≥16 mm, the cavity is very likely malignant in nature; in contrast, if the thickest part of the cavity wall is ≤4 mm, the lesion is very likely benign. When the thickest portion of the cavity measures between 5-15 mm, the cavity is indeterminate- malignant or benign etiologies are nearly equally likely. Characterizing a cavity detected at chest radiography by measuring the cavity wall at its thinnest portion does not reliably discriminate between benign and malignant etiologies. Malignant cavities, often, but not always, produces cavities with lobulated or irregular internal linings, whereas benign causes of lung cavities may produce smooth internal linings. The exception to the latter is pulmonary abscesses, which often produce irregular internal cavity linings. An air-fluid level is commonly present in pulmonary abscesses and less commonly present in cavitary pulmonary malignancies, but the presence or absence of an air-fluid level within a pulmonary cavity does not reliably establish the etiology of the cavity.

The differential diagnosis of the imaging findings for this patient ***includes*** which of the following?

1. Metastatic disease
2. Amyloidosis
3. Rheumatoid lung disease
4. Granulomatosis with polyangiitis (aka Wegener's Granulomatosis)
5. Septic pulmonary emboli
6. All of the above

Correct!
6. All of the above

All of the possibilities listed may produce multiple pulmonary nodules and cavities. Distinguishing among these possibilities requires clinical information (i.e., if the patient has a known primary malignancy, usually a squamous cell malignancy), or whether or not the patient has a clinical presentation suggestive of septic embolization (such as fever, a history of an infected indwelling catheter, endocarditis, intravenous drug abuse, etc). Cavity pulmonary amyloidosis is extremely uncommon, and may or may not be accompanied by other systemic features of amyloidosis, but can be diagnosed with biopsy of affected tissue. Similarly, cavitary rheumatoid nodules are rare, and features of rheumatoid arthritis may, or may not, be evident simultaneously. Granulomatosis with polyangiitis (aka Wegener's Granulomatosis) may be associated with features of renal involvement and / or sinus disease, and elevated c-anti-neutrophil antibodies against protease 3 in cytoplasmic granules (c-ANCA), and sometimes myeloperoxidase, may be present in 90% or more of affected patients.

Further investigation found that the patient had red cell casts in his urine, but no evidence of sinonasal disease.

What is the ***appropriate next step*** for the evaluation/management of this patient?

1. Bronchoscopy with transbronchial biopsy
2. Serial thoracic CT to assess for growth or change in the cavities and nodules
3. ^{18F}FDG-PET scanning
4. c-ANCA antibody assessment and renal biopsy
5. Percutaneous transthoracic fine needle aspiration biopsy

Correct!
4. c-ANCA antibody assessment and renal biopsy

Positive c-ANCA testing is suggestive of the diagnosis of granulomatosis with polyangiitis, but is not specific for that diagnosis and the absence of c-ANCA positivity does not exclude the diagnosis of granulomatosis with polyangiitis. Therefore, a tissue diagnosis from an area of active inflammation is preferred to establish the diagnosis of granulomatosis with polyangiitis conclusively. Given the clinical evidence of renal involvement in this patient, renal biopsy could provide tissue to confirm the diagnosis of granulomatosis with polyangiitis. Bronchoscopy with transbronchial biopsy may be able to provide a tissue for diagnosis also, but the amount of tissue obtained with this procedure is relatively small and may be insufficient to establish the diagnosis of granulomatosis with polyangiitis, and the larger amount of tissue that may be procured with renal biopsy may be preferable. Percutaneous transthoracic fine needle aspiration biopsy often provides a larger amount of tissue for diagnosis compared with bronchoscopy and transbronchial biopsy, but is associated with a higher complication rate. Neither serial thoracic CT to assess for growth or change in the cavities and nodules nor ^{18F}FDG-PET scanning would provide management-altering information in this patient. The patient clearly has an active, undiagnosed pulmonary parenchymal process, and therefore follow up imaging is not appropriate, and the presence of absence of glucose utilization within the pulmonary lesions would not mitigate the need to establish a diagnosis immediately.

The patient subsequently underwent c-ANCA testing, which was positive, and percutaneous renal biopsy showed segmental necrotizing glomerulonephritis with crescent formation.

The ***most likely*** diagnosis for the lesions in this patient is which of the following?

1. Septic emboli
2. Amyloidosis
3. Metastatic disease
4. Granulomatosis with polyangiitis
5. None of the above

Correct!
4. Granulomatosis with polyangiitis

Positive c-ANCA testing is not associated with the other diagnoses listed. The results of the renal biopsy- segmental necrotizing glomerulonephritis- are typical of granulomatosis with polyangiitis).

Diagnosis: Granulomatosis with polyangiitis (aka ANCA-associated granulomatous vasculitis or Wegener's Granulomatosis)

References

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