May 2013 Imaging Case of the Month

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Clinical History

A 21-year-old woman presented with complaints of cough. Frontal and lateral chest radiography (Figures 1A & B) was performed. A detail comparison chest radiograph from several years prior (Figure 1C) is presented as well.

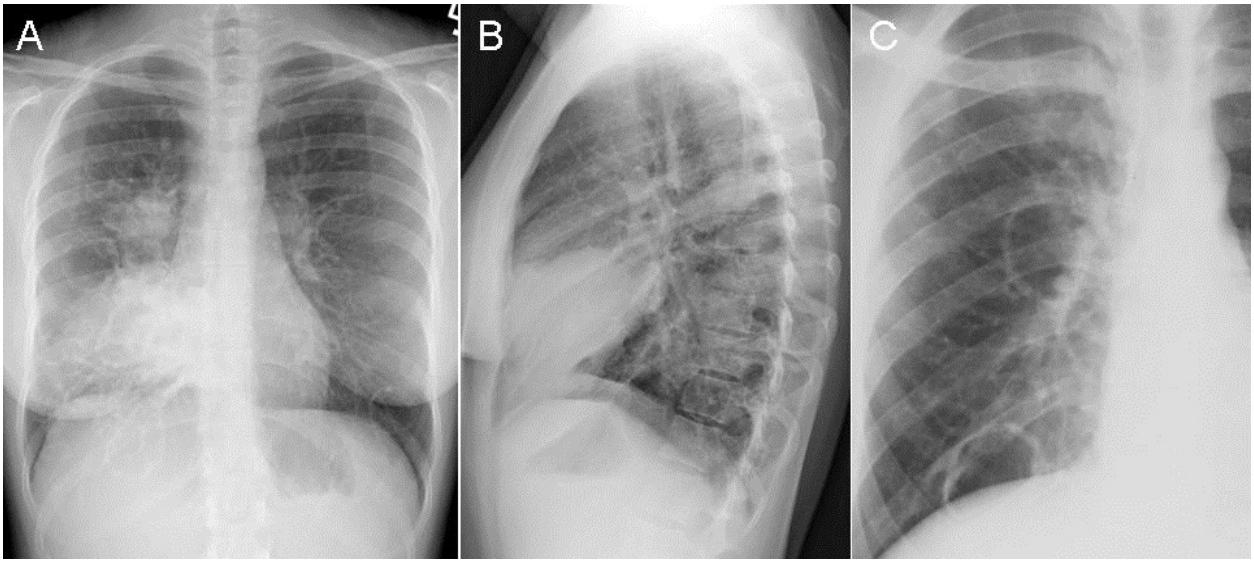

Figure 1. Frontal (A) and lateral (B) chest radiography at presentation and a radiograph from several years earlier (C).

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

1. The chest radiograph predominantly shows bilateral linear and reticular abnormalities
2. The chest radiograph shows a combination of nodules, masses and thin-walled cysts
3. The chest radiograph shows multifocal consolidation with air bronchograms
4. The chest radiograph shows multifocal pleural abnormalities
5. The chest radiograph shows mediastinal widening & hilar lymphadenopathy

Correct!

2. The chest radiograph shows a combination of nodules, masses and thin-walled cysts

The visible mediastinal and hilar contours are within normal limits; no lymphadenopathy is present. The right hilum appears enlarged on the frontal image, but the hilum is shown to be normal on the lateral projection. The appearance of hilar enlargement on the frontal chest radiograph is caused by a mass overlying this region, located slightly posterior to the right hilum, seen to advantage on the lateral radiograph. The interstitium is largely normal; linear or reticular abnormalities, or findings suggesting fibrotic lung disease, are not the dominant abnormalities present on the chest radiograph. The costophrenic angles are sharp and no pleural abnormalities are seen. A mass-like opacity is present in the right middle lobe and in the right suprahilar region, and at least one thin-walled cyst is seen in the left lower lobe, with another in the right mid-lung. No air bronchograms, however, are seen within the focal lung opacities.)

Several images from a thoracic CT scan (Figure 2) performed 1 year prior to the presentation chest radiograph were subsequently located.

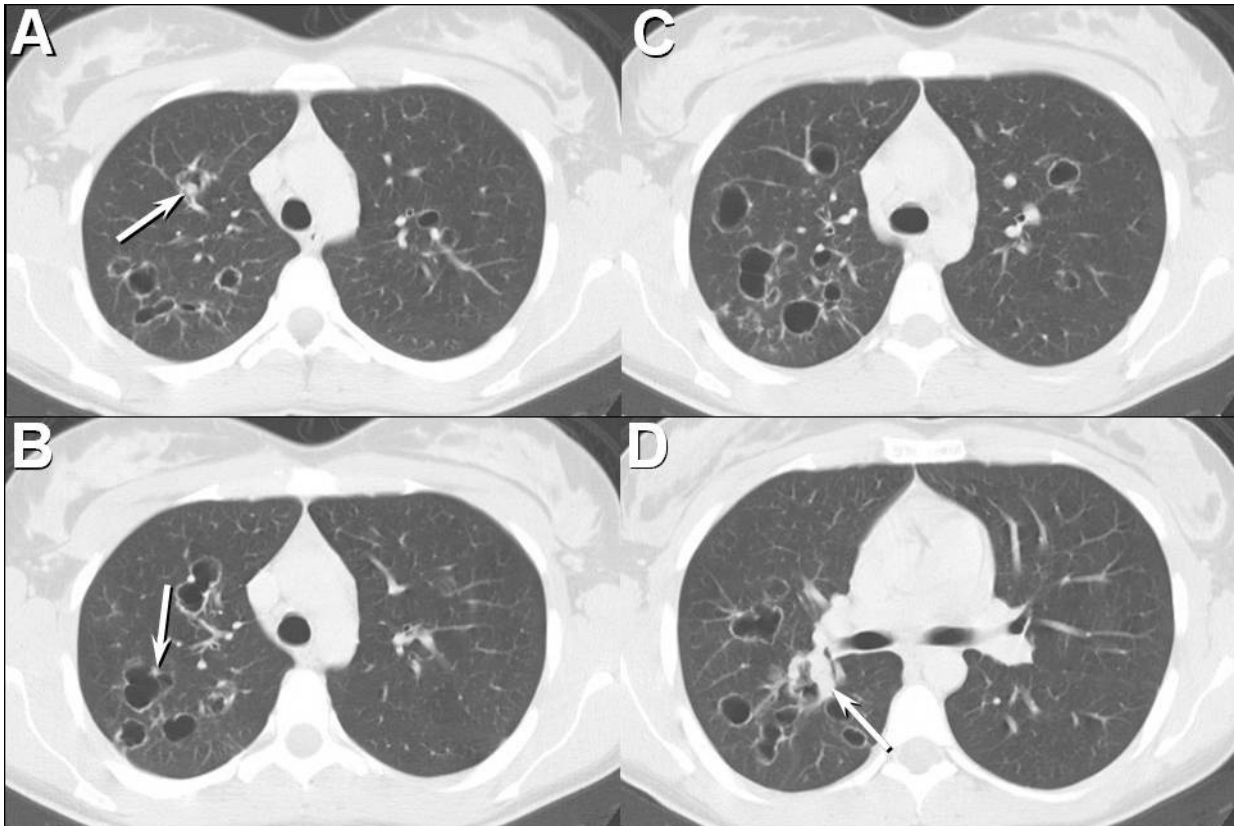

Figure 2. Thoracic CT performed 1 year prior to presentation.

Which of the following statements regarding this CT examination is ***most accurate***?

1. The thoracic CT shows worsened multifocal, migratory consolidation compared with the presentation chest radiograph (Figures 1A & B)
2. The thoracic CT shows more prominent bilateral nodular opacities compared with the presentation chest radiograph
3. The thoracic CT shows the presence of bilateral thin-walled cysts, some with nodular components
4. The thoracic CT shows features suggesting fibrotic lung disease
5. The thoracic CT shows numerous bilateral nodules that later became cystic and cavitary on the presentation chest radiographs

Correct!

3. The thoracic CT shows the presence of bilateral thin-walled cysts, some with nodular components

The thoracic CT shows multiple, somewhat clustered, largely thin-walled cysts, although some of the cysts show peripheral nodular components (arrows). Some confluent opacity is present within the right perihilar region, but no overt consolidation is present. While cysts and nodules are present in combination on both the thoracic CT and the presentation chest radiographs, the thoracic CT is dominated by cystic change, whereas the follow up chest radiograph obtained at presentation is dominated by masses; therefore, the progression between these two studies is more suggestive of cysts developing into nodules and masses, and not the reverse, as suggested by choice "5". Similarly, the nodular abnormalities are increasingly prominent on the follow up chest radiograph obtained at presentation compared with the thoracic CT, so choice "2" is not the best among the available choices. No features to indicate fibrotic lung disease are present.).

Several focused images from a thoracic CT scan (Figure 3) performed 4 years prior to the presentation chest radiograph (Figures 1A & B) were obtained.

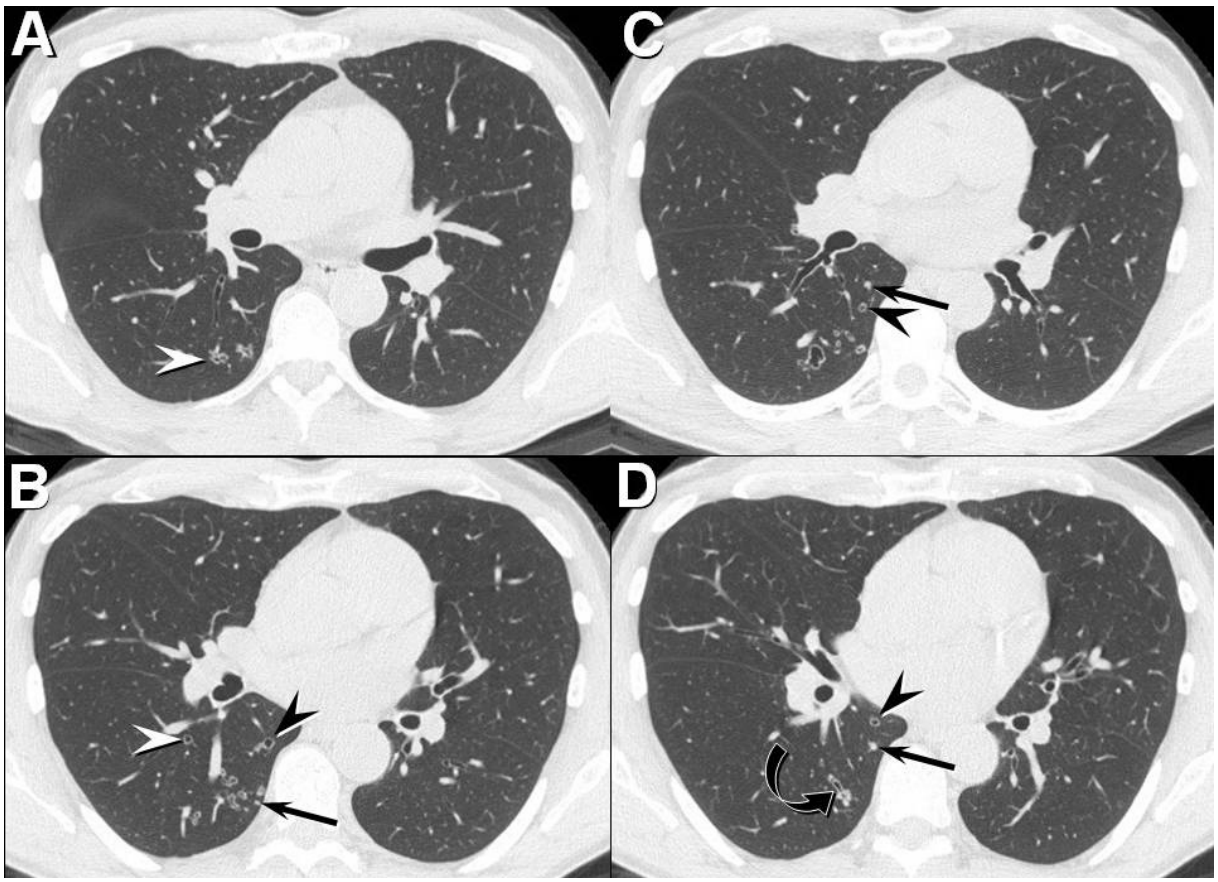

Figure 3. Thoracic CT 4 years prior to presentation

Which of the following statements regarding this CT examination is ***most accurate***?

1. The thoracic CT shows numerous thin-walled cysts
2. The distribution of the abnormalities seen on CT is dependent in nature
3. The thoracic CT shows cystic change consistent with honeycombing
4. The thoracic CT shows numerous small nodules consistent with a “miliary” pattern
5. 1 and 2
6. 2 and 3

Correct!
5. 1 and 2

The focused thoracic CT images show numerous small cysts (arrowheads), some irregularly shaped and some tubular-shaped (curved arrow), the latter suggesting an airway etiology. The cysts are not thick-walled, stacked, and subpleural in morphology, as would be expected with honeycombing. A few centrilobular nodules are present (arrows), but the nodular pattern is not a miliary one. There is a conspicuous clustering of the abnormalities- both cysts and nodules- in the superior and posterior basal segments of the right lower lobe, consistent with a dependent distribution.

The patient subsequently underwent thoracic CT (Figure 4) for further characterization of the pulmonary abnormalities seen at presentation chest radiography (Figure 1A & B).

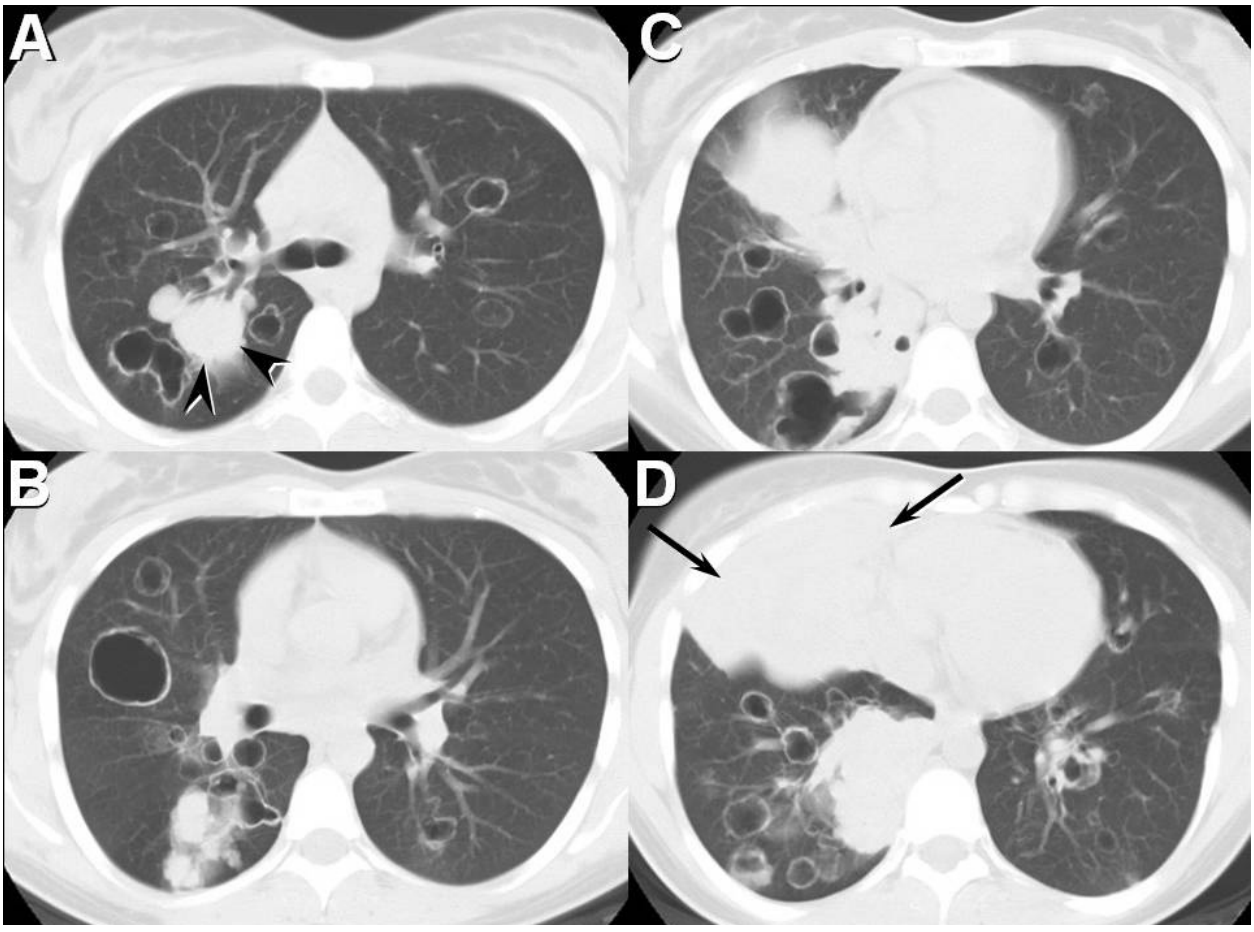

Figure 4. Thoracic CT obtained at presentation, shortly after presentation chest radiography

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

1. Comparison among the thoracic CTs shows multifocal, recurrent, and migratory nodules and cystic lesions
2. The thoracic CT shows worsening of interstitial thickening suggesting a progressive fibrotic abnormality
3. Comparison of the thoracic CTs shows serial progression of the cystic and cavitary process
4. The presentation thoracic CT shows findings suggesting development of a bronchopleural fistula

Correct!

3. Comparison of the thoracic CTs shows serial progression of the cystic and cavitary process

The serial thoracic imaging studies suggest a progressive abnormality characterized initially by small cysts, likely an airway-related process, which subsequently enlarge and develop nodular components, eventually progressing into masses. The thoracic CT confirms the right middle lobe mass (arrows) seen at presentation chest radiography and shows the right suprahilar mass (arrowheads) located within the posterior segment of the right upper lobe. These abnormalities are not waxing and waning / recurrent, or migratory; rather, they are relentlessly progressive. No pleural abnormalities are present, and the marked abnormalities on the thoracic CT studies are not interstitial in nature. No features to suggest fibrotic lung disease- such as architectural distortion, reticulation, traction bronchiectasis, and honeycombing- are evident.

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

1. Presumptive broad spectrum antibiotic treatment
2. ^{18F}FDG-PET scanning
3. Bronchoscopy with bronchoalveolar lavage and biopsy
4. Percutaneous transthoracic fine needle aspiration biopsy
5. Surgical lung biopsy
6. 3 or 4

Correct!
3. Bronchoscopy with bronchoalveolar lavage and biopsy
4. Percutaneous transthoracic fine needle aspiration biopsy
6. 3 or 4

Given the progressive nature of the abnormalities, ^{18}FDG-PET scanning would likely contribute little management-altering information. Relatively little tracer accumulation in the lesions at ^{18}FDG-PET would not allow a conservative, observatory approach. In fact, such an approach has already occurred and progression of the abnormalities has clearly been established. Elevated tracer uptake within the pulmonary abnormalities would not distinguish among the numerous infectious, inflammatory, and neoplastic etiologies that must be considered in this case. Percutaneous transthoracic fine needle aspiration biopsy would be a reasonable choice, particularly if directed at the mass within the right middle lobe abutting the chest wall. Percutaneous transthoracic fine needle aspiration biopsy of cysts and cavities is also possible, but laceration of the cavity wall may occur because the needle is frequently directed towards the cavity wall to maximize the possibility of obtaining sufficient tissue for diagnosis, and, if the internal wall of the cavity is vascularized, this can result in significant bleeding. Additionally, while not confirmed in all reports on the subject, some feel that percutaneous transthoracic fine needle aspiration biopsy of cavitary lesions is associated with an increased rate of biopsy-related complications, including cough, hemoptysis, and air embolism. Bronchoscopy with bronchoalveolar lavage and biopsy would be a reasonable choice for this patient given the close association of a mass with the posterior segment right upper lobe bronchus. In this patient, surgical lung biopsy may be considered in the event that either percutaneous transthoracic fine needle aspiration biopsy or bronchoscopy with bronchoalveolar lavage and biopsy are unable to obtain a diagnosis.).

The patient subsequently underwent two additional thoracic CT studies just over 1 year (Figure 5) following presentation.

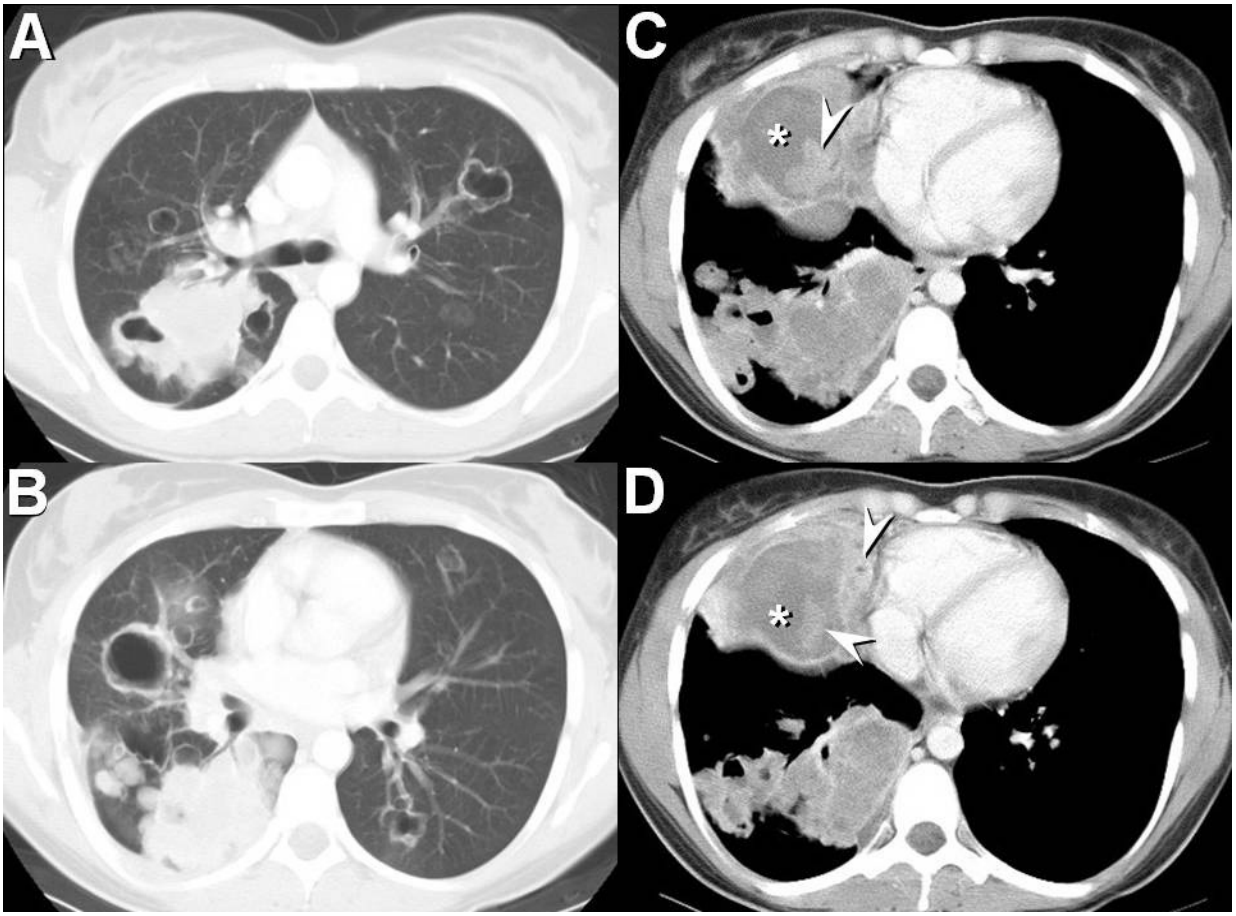

Figure 5. Thoracic CT obtained just over 1 year following presentation, shown in lung and soft tissue windows.

The imaging findings of the various studies presented ***suggest*** which of the following ***diagnoses***?

1. Septic embolization
2. Progressive metastatic malignancy
3. Tracheobronchial papillomatosis complicated by malignant degeneration
4. Progressive lymphangioleiomyomatosis
5. Progressive pulmonary benign metastasizing leiomyomas

Correct!

3. Tracheobronchial papillomatosis complicated by malignant degeneration

Thoracic CT obtained just over 1 year following presentation, shown in lung and soft tissue windows, shows progression of the thin-walled, nodular cysts into frankly cavitary lesions with grossly nodular walls. The images displayed soft tissue windows show that the cavitary areas contain both internal low attenuation, consistent with necrosis (*) as well as nodular, enhancing foci (arrowheads) related to the internal cavity walls.

While septic embolization certainly can produce nodules that subsequently cavitate, in this patient, cysts and cavities are present initially, subsequently complicated by the development of nodules and masses. Furthermore, the progression of the abnormalities has occurred over years and in the absence of clinical features of infection, making septic embolization very unlikely.

Pulmonary benign metastasizing leiomyomas may cause multiple pulmonary nodules and masses, and even cavitation of the pulmonary abnormalities has been reported in this condition. Typically patients with pulmonary benign metastasizing leiomyomas have a history of uterine leiomyomas who have undergone hysterectomy, and that history was not provided in this case. Additionally, while the pulmonary nodules and masses in patients with benign metastasizing leiomyomas may show growth over time, typically the growth is much slower than that seen in this patient, and the pulmonary abnormalities in such patients are frequently asymptomatic. Finally, the dependent distribution of the pulmonary abnormalities in this patient would not be expected with benign metastasizing leiomyomas.

The extensive nodular and mass-like components of the pulmonary abnormalities in this patient are not consistent with pulmonary lymphangioleiomyomatosis; that condition is dominated by thin-walled cysts, with only small nodules, often reflecting multifocal micronodular pneumocyte hyperplasia, occasionally present. Progressive metastatic malignancy is a prominent consideration for the abnormalities in this patient, and the rate of progression and development of nodular and mass-like components related to the cystic lesions is consistent with that diagnosis. However, no history of extrathoracic malignancy was provided and nor was such apparent clinically. Finally, the small clustered nature of the cysts, in a dependent distribution, as noted on the earliest thoracic CT (Figure 3), would be quite atypical for pulmonary metastatic malignancy. On the other hand, the presence of small pulmonary cysts and nodules located in a dependent distribution, with subsequent enlargement of the cystic lesions, in a young patient, is very suggestive of tracheobronchial papillomatosis. In that setting, the development of nodular components within the cyst walls, progressing to frank masses, is highly suggestive of the development of squamous cell malignancy, which is a known complication of this condition.

Transthoracic fine needle aspiration biopsy of the subpleural right middle lobe mass was performed, and malignant squamous cells were identified at cytopathology. The

patient subsequently underwent right pneumonectomy (Figure 6), which provided additional confirmation of the diagnosis.

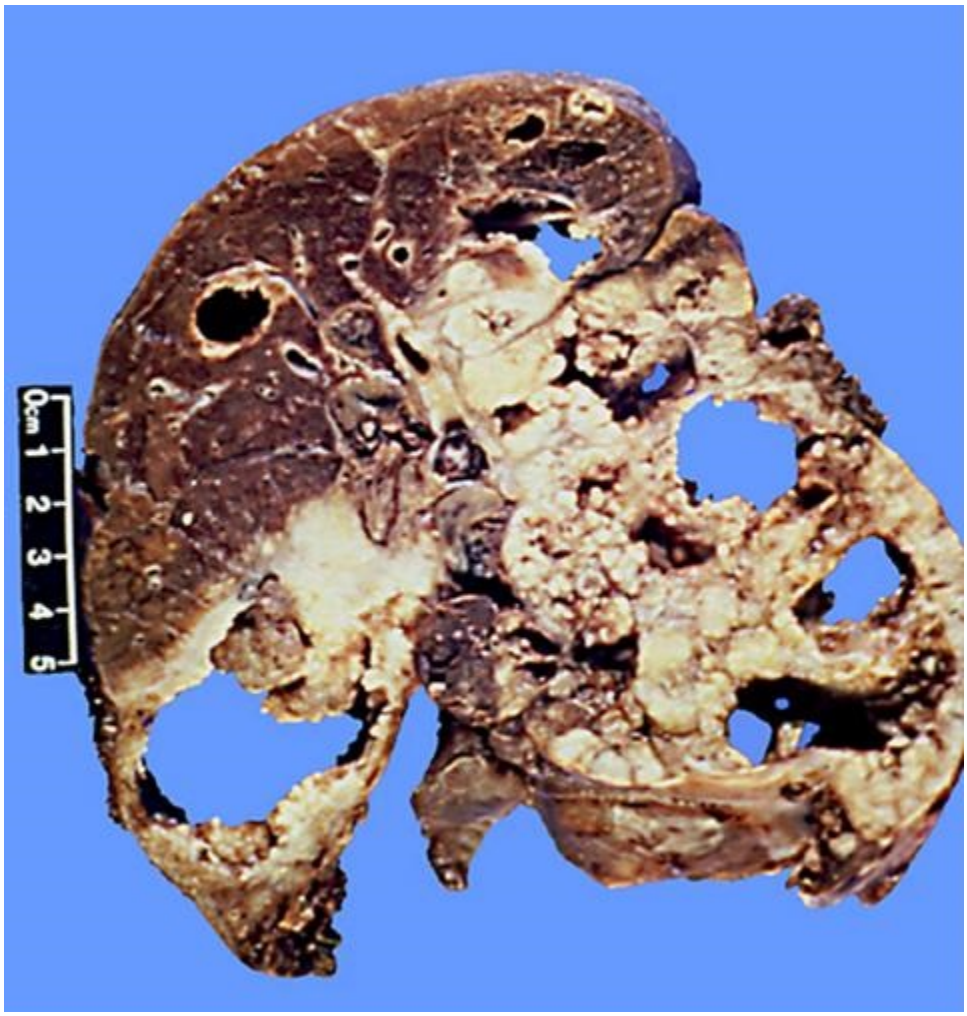

Figure 6. Gross pathological specimen following right pneumonectomy shows multiple, upper and lower lobe cavities with extensive nodularity of the inner walls.

Diagnosis: Tracheobronchial Papillomatosis complicated by malignant degeneration, with development of multiple squamous cell carcinomas

References

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