June 2012 Imaging Case of the Month

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Clinical History: A 46 -year-old man presents to the emergency room with hemoptysis. Frontal and lateral chest radiography (Figures 1A and B) was performed.

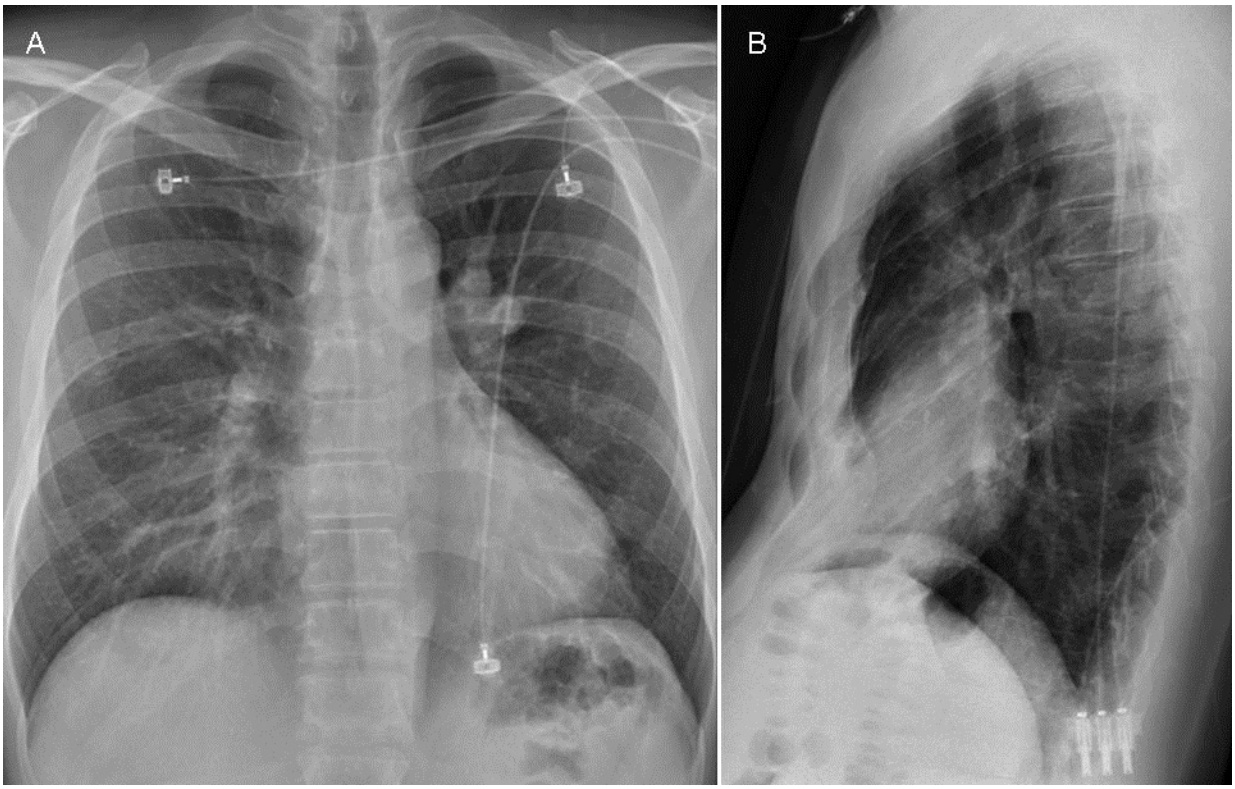

Figure 1. Frontal and lateral chest radiography shows a lobulated, circumscribed lesion within the left hilum. The right hilum appears normal, no lung consolidation is present, and no pleural abnormalities are seen. There is no evidence of mediastinal lymph node enlargement. There is relative lucency involving the left lung, particularly the left upper lobe, compared with the right side.

Which of the differential diagnostic considerations listed below is the ***least likely*** consideration for the appearance of the lesion on the chest radiograph?

1. Bronchogenic carcinoma
2. Adenoid cystic carcinoma
3. Carcinoid tumor
4. Sarcoidosis
5. Allergic bronchopulmonary aspergillosis

Correct!
4. Sarcoidosis

Chest radiography shows a unilateral lobulated, circumscribed mass in the left hilum; the right hilum and mediastinum are normal. Lucency in the left lung, particularly left upper lobe, suggests air trapping. These findings indicate a lesion arising from or involving the airway. All the choices above, except sarcoidosis, commonly involve the large airways and may present as a central hilar masses associated with bronchial obstruction. In contrast, while sarcoidosis may present with hilar enlargement due to lymphadenopathy, the hilar nodal involvement is commonly symmetric and frequently accompanied by mediastinal lymph node enlargement; the latter is not present in this case. Furthermore, sarcoidosis is rarely associated with central unilateral endobronchial obstruction.

Clinical Course: The patient subsequently underwent thoracic CT (Figures 2 and 3).

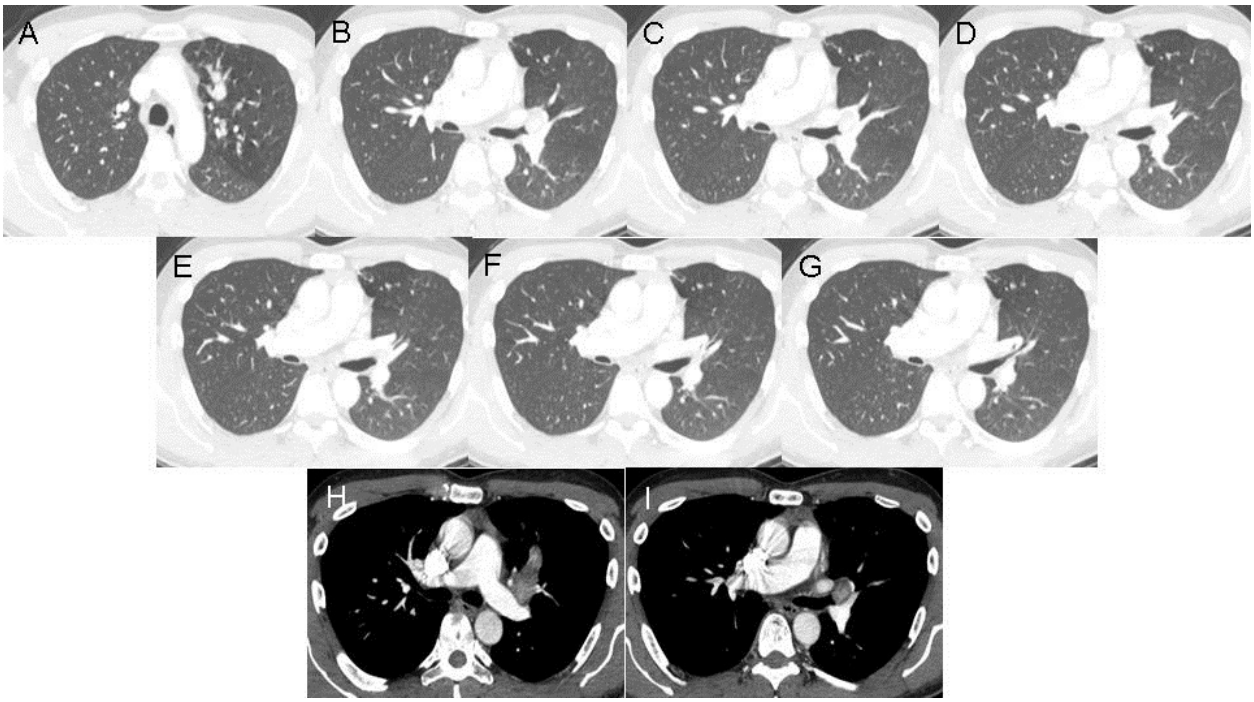

Figure 2. Axial thoracic CT scan (A-G= lung windows, H-I soft tissue windows).

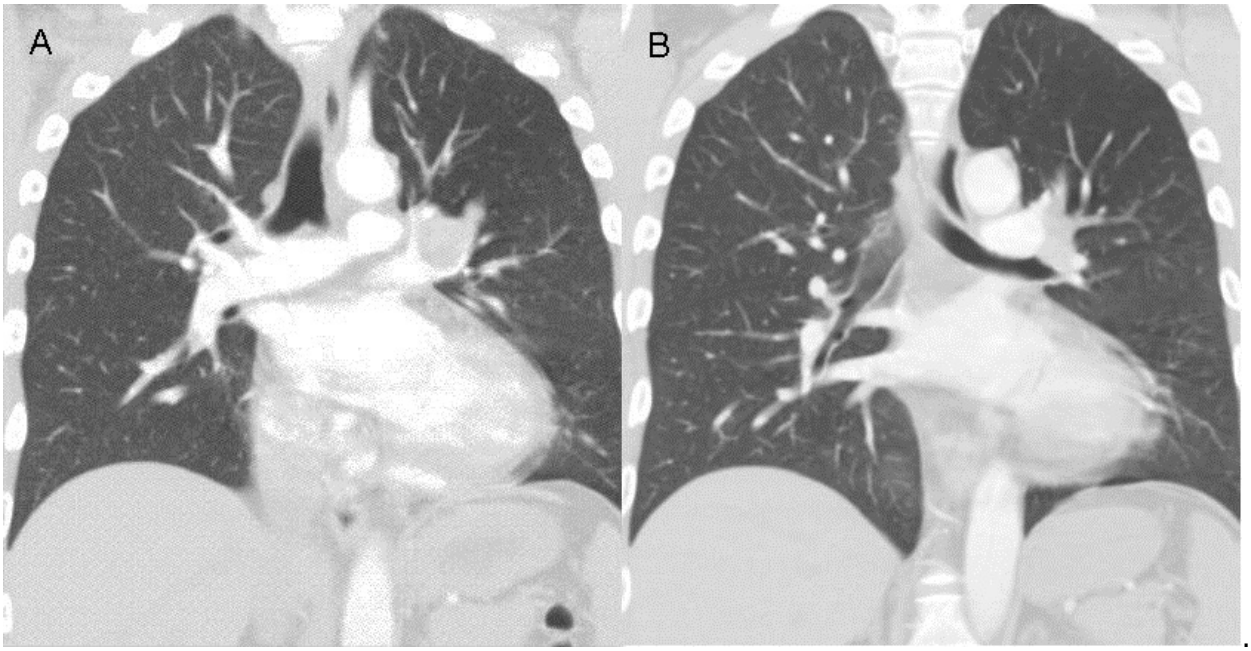

Figure 3. Coronal thoracic CT images (lung windows).

Regarding the CT appearance of the lesion, which of the following is ***most*** accurate?

1. The lesion shows internal necrosis
2. The lesion has an aggressive, infiltrating morphology
3. The lesion is associated with post-obstructive consolidation
4. The lesion is circumscribed, arising from the left upper lobe airway
5. The lesion shows no evidence of enhancement

Correct!

4. The lesion is circumscribed, arising from the left upper lobe airway

The images show a fairly well-defined, or circumscribed, lesion arising from the left upper lobe bronchus, filling and expanding the anterior segment left upper lobe bronchus. While the lesion, and associated mucus impaction, shows minimally lobulation, it is not infiltrative-appearing, nor is it overtly invading adjacent structures; rather, it appears somewhat indolent. The lesion is associated with air trapping in the left upper lobe, rather than post-obstructive consolidation- note the left upper lobe hyperlucency best seen on Figure 2B, outlined by the left major fissure posteriorly. The internal portions of the lesion, best appreciated on the soft tissue window images Figures 2H and I, show the internal aspects of the lesion to be hyperattenuating, consistent with enhancement, rather than displaying the low attenuation typical of necrosis.

Which of the following represents the ***most appropriate*** next step in management at this point?

1. Surgical resection
2. Percutaneous fine needle aspiration biopsy
3. Bronchoscopy
4. Follow up imaging with ^{18}FDG-PET
5. Follow up thoracic CT in 3-6 months to assess for growth

Correct!
3. Bronchoscopy

Choosing surgical resection at this point is somewhat premature- the exact nature and precise location of the lesion have not yet been characterized, and non-surgical causes for the left upper lobe lesion- such as focal allergic bronchopulmonary aspergillosis- while unlikely, have not been excluded. The lesion is too centrally located to attempt percutaneous fine needle aspiration biopsy- the likelihood of complication increases when such central lesions undergo percutaneous approaches for biopsy. Although imaging with ^{18}FDG-PET could be attempted, while tracer utilization would prompt a tissue diagnosis, lack of tracer utilization would not provide information useful for further management- an endobronchial lesion in a patient with hemoptysis would still remain and require evaluation. Follow up thoracic CT is also not appropriate. Follow up thoracic CT is typically reserved for lesions that are indeterminate as regards malignant potential- the intention is to show growth to indicate aggressive potential, versus stability to indicate a non-aggressive lesion, avoiding intervention in the latter case. This situation most commonly occurs in patients with asymptomatically detected lung nodules, or for whom the nodule detection is likely incidental to the indication for the thoracic CT. In this circumstance, the left upper lobe lesion is certainly the cause of the patient's hemoptysis, and therefore intervention is required. Bronchoscopy is the best method for further investigation of this lesion.

Bronchoscopy was performed and showed a mass protruding into the left upper lobe airway, thought to be arising from the lingular bronchus (Figure 4).

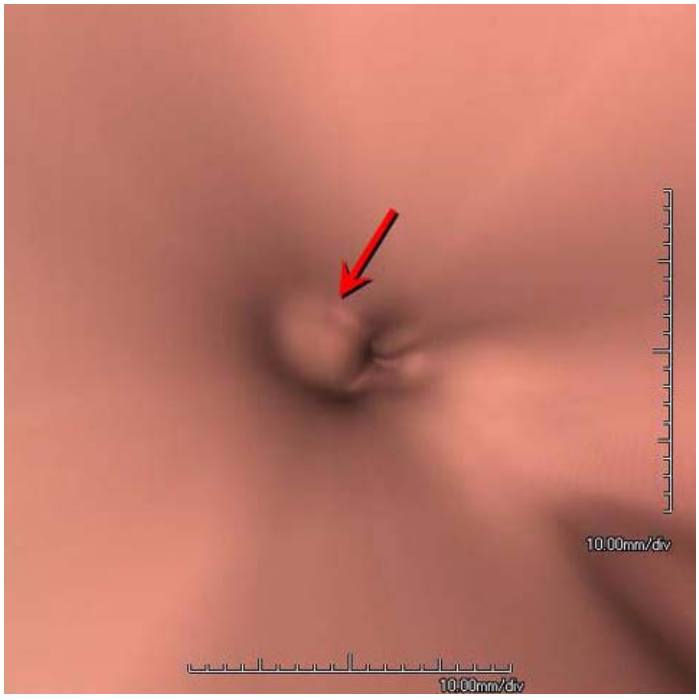

Figure 4. Virtual bronchoscopy showing the internal perspective of the trachea and left upper airways- the mass protruding from the bronchus is visible (arrow).

The lesion was highly vascular, and therefore biopsy was not attempted. The right-side airways, left lower lobe bronchus, and trachea appeared normal. The patient subsequently underwent surgical resection of the lesion, which established the diagnosis of a well-differentiated neuroendocrine neoplasm consistent with “typical” carcinoid tumor).

Diagnosis: Left upper lobe well-differentiated neuroendocrine neoplasm (“typical” carcinoid tumor).

Which of the following regarding carcinoid tumor is ***false***?

1. Carcinoid tumors are true malignancies
2. Carcinoid tumors frequently present as hilar or perihilar masses on chest radiography
3. Thoracic CT features of carcinoid tumor include prominent enhancement, post-obstructive air trapping and / or consolidation, and calcification
4. Pulmonary carcinoid tumors frequently result in the carcinoid syndrome
5. Typical carcinoid tumors often show relatively little tracer uptake at ^{18}FDG-PET

Correct!

4. Pulmonary carcinoid tumors frequently result in the carcinoid syndrome

Both typical and atypical carcinoid tumors are true malignancies, although typical carcinoid tumors are often relatively indolent neoplasms that are associated with a generally good prognosis following complete surgical resection. Most pulmonary carcinoid tumors (60-70%) present as centrally located masses, arising from the main, lobar, or segmental bronchi. Due to their central endobronchial location, pulmonary carcinoid tumors often present as hilar or perihilar masses on chest radiography, often associated with post-obstructive changes, such as atelectasis, consolidation, or air trapping. At thoracic CT, the endobronchial nature of carcinoid tumor is often readily apparent, and the lesions often show calcification. Intense enhancement following intravenous contrast administration at thoracic CT is common with carcinoid tumors, owing to their relatively vascular nature. However, pulmonary carcinoid tumors often do not show significant tracer accumulation at ^{18}FDG-PET scanning- this is one of the well-known pitfalls in tumor detection and staging using ^{18}FDG-PET. Pulmonary carcinoid tumors are uncommonly associated with the carcinoid syndrome- abdominal cramps, flushing of the face, neck or upper chest, diarrhea, palpitations, hypotension, and wheezing. The carcinoid syndrome is mediated by release of vasoactive substances, such serotonin release, and occurs most commonly in patients with gastrointestinal carcinoid tumors, frequently win the setting of liver metastases.

Neuroendocrine cell neoplasms arise from Kulchitzky cells in the bronchial mucosa and include “typical” carcinoid tumors, “atypical” carcinoid tumors, large cell neuroendocrine neoplasms, and small cell carcinoma, in increasing order of aggressiveness. Pulmonary carcinoid tumors account for more than 25% of all carcinoid tumors and 1-2% of pulmonary neoplasms. Most (80-90%) carcinoid tumors are “typical” carcinoid tumors, with the remaining 10-20% “atypical” carcinoid tumors. Men are about as equally affected as women, with some reports suggesting a female predominance, and others indicating a male predominance; men are more commonly affected with atypical carcinoid tumors. The age of presentation is wide, averaging about 46 years old, although carcinoid tumors are among the commonest pulmonary neoplasms in children and adolescents. Typical carcinoid tumors are unassociated with smoking, whereas some association with smoking with atypical carcinoid tumors has been recognized.

Carcinoid tumors tend to present with symptoms of central airway obstruction, such as cough, wheezing, and hemoptysis. Carcinoid tumors are occasionally detected asymptomatically. Atypical carcinoid tumors may be more peripherally located than typical carcinoid tumors, and therefore present at a slightly later age than typical carcinoid tumors.

Pulmonary carcinoid tumors commonly present as hilar or perihilar masses at chest radiography, associated with post-obstructive atelectasis, pneumonitis, consolidation, or air trapping. Thoracic CT commonly shows a round or oval endobronchial lesion, associated with post-obstructive atelectasis or air trapping, arising in the main, lobar, or segmental bronchi. Mucus plugging distal to the lesion is common. Carcinoid tumors

may be located distal to segmental airways, and then are classified as peripheral, in 16-40% of patients. The relationship of the tumor to the airway is often not evident for peripheral lesions, and these tumors often present as solitary pulmonary nodules. Calcification may be seen in up to 30% of lesions at CT but is often not appreciable at chest radiography, and is more commonly encountered in central lesions. Typical and atypical carcinoid tumors closely resemble one another at imaging, although atypical carcinoid tumors tend to be larger at diagnosis and more commonly associated with lymphadenopathy, reflecting their more aggressive nature. Atypical carcinoid tumors are more likely to be found in the lung peripheral than typical carcinoid tumors.

Contrast-enhanced thoracic CT in patients with carcinoid tumors often shows intense contrast enhancement of the lesion. However, lack of significant tracer utilization at ^{18}FDG-PET is well-recognized with carcinoid tumors, but is not a universal finding, and occasionally typical carcinoid tumors are associated with increased tracer utilization at ^{18}FDG-PET.

Pulmonary carcinoid tumors are generally treated surgically when possible, and outcomes are usually excellent. Post-operative survival is diminished with atypical carcinoid tumors compared with typical lesions, although still improved compared with bronchogenic carcinomas. The histopathological lesion classification and status of nodal involvement are the two most important factors influencing patient prognosis.

References

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