Medical Imaging Quiz – Case 47

An asymptomatic 32-year-old woman presented to the pathologist for clinical evaluation in order to take a health certificate. Medical history and physical examination revealed none abnormality. Thus, she referred to our department for a chest x ray which revealed a solid mass at the lower lobe of the left lung. Chest computed tomography (CT) was then performed showing a large solid mass with great contrast medium enhancement. CT guided fine needle biopsy was decided in order to confirm the diagnosis (fig. 1).

Comment

Carcinoid tumors of the lung are a fascinating but uncommon group of pulmonary neoplasms. Typical carcinoid tumors of the lung represent the most well-differentiated and least biologically aggressive type of pulmonary NET. These tumors characteristically grow slowly and tend to metastasize infrequently. Atypical carcinoid tumors have a more aggressive histologic and clinical picture. They metastasize at a considerably higher rate than do typical carcinoid tumors and, therefore, carry a worse prognosis.

Bronchopulmonary carcinoid tumors are reported to represent about 10% of all carcinoid tumors. Between 1% and 6% of all lung tumors are carcinoid tumors. Some 80–90% of tumors develop within a bronchus of subsegmental size or greater. About 10–15% of tumors arise in a mainstem bronchus; however, they rarely appear in the trachea. About 10–20% of tumors are located in the pulmonary periphery. Peripheral pulmonary carcinoid tumors most often are asymptomatic and usually are discovered incidentally.

They are one of the differential diagnoses considered in evaluation of a solitary pulmonary nodule.

Atypical carcinoid tumors account for about 10% of all pulmonary carcinoid tumors. They can present in the same locations as typical carcinoids, but they occur more commonly as peripheral lesions. At least 50% of pulmonary atypical carcinoid tumors present in the periphery of the lung. The average age of people at occurrence of typical carcinoid tumors is 40–50 years, but typical carcinoid tumors have been reported in virtually every age group. Carcinoid tumors of the lung generally have a better prognosis than other forms of pulmonary malignancy. They have an overall 5-year survival rate of 78–95% and a 10-year survival rate of 75–90%. Typical carcinoid tumors have been found to have a much better prognosis than do the atypical variety. Atypical have a more aggressive nature and a greater tendency to metastasize.

Between 25% and 39% of patients with a carcinoid pulmonary tumor are asymptomatic. The vast majority of symptomatic patients have symptoms directly involving the bronchopulmonary tree. Carcinoids developing within large airway structures grow slowly and can become quite large, causing bronchial obstruction and symptoms such as persistent cough, hemoptysis, and recurrent or
obstructive pneumonitis. Wheezing, chest pain, and dyspnea may also be noted. Carcinoid syndrome occurs in about 2% of cases of pulmonary carcinoid tumors, much less frequently than it does in cases associated with gastrointestinal (GI) carcinoid tumors.

An abnormal finding on chest radiography is present in about 75% of patients with a pulmonary carcinoid tumor. Findings include either the presence of the tumor mass itself or indirect evidence of its presence observed as parenchymal changes associated with bronchial obstruction from the mass. Changes associated with bronchial obstruction include persistent atelectasis, consolidation secondary to pneumonia, and changes of bronchiectasis and hyper-inflation. CT can demonstrate more detail about nodules, masses, or suspicious parenchymal changes, such as persistent atelectasis or obstructive pneumonia found on plain chest radiography. It may reveal nodules or masses that are not well-visualized on plain chest radiography by virtue of their small size or their position, such as those located in a retrocardiac position. Intravenous contrast in CT also can be useful in differentiating malignant from benign lesions. Malignant lesions generally have increased vascularity and show greater enhancement than benign lesions on contrast CT. Because carcinoid tumors are highly vascular, they also possess this feature.

About 75% of pulmonary carcinoids are visible on bronchoscopy. In most cases, the physician makes the diagnosis of pulmonary carcinoid tumor on the basis of the findings from bronchoscopy plus a combination of radiologic studies. At present, most endoscopists perform bronchoscopic biopsy of these lesions for histologic diagnosis. Percutaneous needle biopsy may be useful for tissue sampling of peripheral pulmonary nodules. As with transbronchial biopsy, the amount of tissue sampled may be quite limited, making exact histologic determination difficult. The diagnostic yield for a specific benign diagnosis in solitary pulmonary nodules is 12–68%. A negative finding on biopsy should not produce a false sense of confidence in the examining physician. A combination of clinical findings, patient risk factors, and data from all completed diagnostic studies should enter into the decision whether to proceed with surgical removal of a pulmonary nodule or to observe it for a longer period.

All pulmonary carcinoid tumors should be treated as malignancies. Because surgical resection is the only treatment known to achieve cure, all pulmonary carcinoid tumors without evidence of distant metastatic disease should be resected completely as long as no contraindication to surgery exists. Chemotherapy and radiation therapy have been used in the treatment of metastatic disease, but have met with virtually no success. After surgery, patients are followed clinically and with plain chest radiography every 2–3 months for the first year. If no evidence of recurrence is discovered within this period, surveillance intervals are extended to every 6 months. Additional studies, such as CT, are performed only if suspicion of recurrence arises.

References

4. TRAVIS WD, BRAMBILLA E, MÜLLER-HERMELINK HK, HARRIS CC. World Health Organization classification of tumours: Pathology and genetics of tumours of the lung, pleura, thymus and heart. IARC Press, Lyon, 2004

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