INTRALOBULAR PULMONARY SEQUESTRATION

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Background: A 59-year-old non-smoking woman, with a history of gall stones, type 2 diabetes mellitus and hypertension, was referred to our hospital because of persistent coughing, night sweats, headaches and weight loss. Clinical examination revealed no abnormalities. The laboratory findings on admission were as follows: erythrocyte sedimentation rate was 39 mm/h, C-reactive protein level was 62 mg/mL and leukocyte count was 8.0 x 10^9/L. Bronchoscopic examination showed a normal bronchial tree.

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Work-up

Conventional radiograph of the thorax (A: postero-anterior view, B: lateral view) shows a sharply circumscribed oval density of about 4 cm in diameter is visible in the posterior right lower lobe of the lung (arrows).

Contrast-enhanced CT scan of the thorax (Fig. 2) (A: reformatted image in the coronal plane, B: reformatted image in the sagittal plane) shows a lesion located in the paravertebral and basal region of the right lung (arrows). A prominent vessel is seen within the lesion. On axial section (C), there are several vessels inside the lesion, of which the main one is originating from the aorta (arrow). None of the vessels seem to be compromised. Venous drainage is via the pulmonary vein. Furthermore, in the liver multiple well-circumscribed low density areas, matching cysts, are visible. On axial image, lung window setting (D), the lesion is embedded in the lung tissue, sharing with it a common pleural investment.

Radiological diagnosis

Based on the radiological findings, the diagnosis of pulmonary sequestration was made.

Since, on CT scan the lesion was embedded in the lung tissue, sharing with it a common pleural investment, and had an abnormal vascular supply, the diagnosis was further refined to pulmonary sequestration of the ILS (intralobar) type.

Discussion

The term sequestration, which is derived from the Latin verb ‘sequestrare’, meaning “to set apart”, was first used by Pryce in 1946. Pulmonary sequestration is a development lung disease, defined as nonfunctioning pulmonary tissue lacking normal communication with the bronchial tree and supplied by a systemic artery. Two forms of sequestration are recognized: extralobar (ELS), which is separated from the lung tissue by a separate lining of pleura, and intralobar (ILS), which is embedded in the normal lung, sharing with it a common pleural investment.

ILS accounts for about 75% of pulmonary sequestration findings. It is considered an acquired disease, with recurrent episodes of pulmonary infection affecting approximately 50% of the patients < 20 years of age. In most cases, the lower lobes are involved, typically the left lobes (in 55-60%), always above the diaphragm. The sequestration is usually supplied by an artery arising from the thoracic aorta, whereas the venous drainage is typically performed by the pulmonary veins. In rare cases, ILS is associated with congenital malformations. There is no gender predominance in the occurrence of ILS.

ELS is considered a congenital disease. It results from an accessory lung bud that in some cases maintains the original connection with the intestine, allowing communication between the sequestration and the gastrointestinal tract. Typically, it is a single, asymptomatic lesion, ovoid or pyramidal shaped and measuring from 3 to 6 cm. The diagnosis of ELS is usually a result of incidental findings on routine chest radiographs or at the occasion of laparotomy or thoracotomy.

Traditionally, angiography was required for a definite diagnosis of pulmonary sequestration. Recent advances in diagnostic accuracy of CT-angiography have resulted in the use of this imaging modality as a non-invasive alternative diagnostic method for pulmonary sequestration. In particular, 3-D CT provides accurate information on the aberrant vascular supply and can replace angiography.

The treatment for pulmonary sequestration is surgery. The identification and control of the aberrant artery branch, above or below the diaphragm, are essential for preventing hemorrhage. Postoperative results have been reported to be excellent.

Bibliography