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SOLITARY FIBROUS TUMOR OF THE PLEURA

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Background: A 62-year-old woman presented to the rheumatologist with generalised joint pain. She was in good health and had no pulmonary complaints. Laboratory findings were normal. Previous medical history showed partial thyroidectomy for a goiter and local resection of a low-grade chondrosarcoma of the right hip, 20 years earlier.

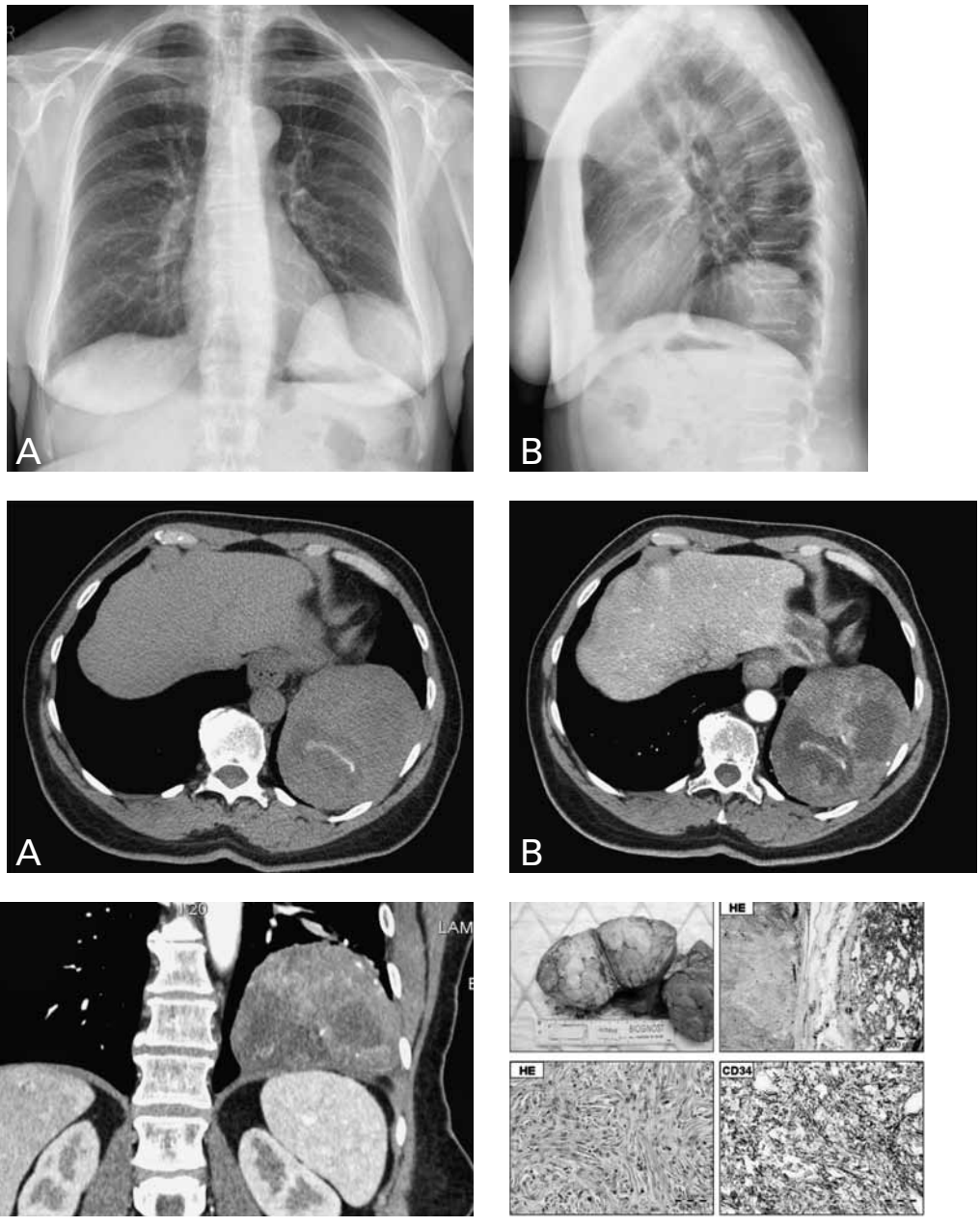

Fig. 1A 1B 2A 2B 2C 3

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Radiological diagnosis

On PA (A) and lateral (B) chest radiographs (Fig. 1), a well-defined rounded mass in the lower part of the left hemithorax is noticed. The mass is contiguous with the left hemidiaphragm and mimics diaphragmatic elevation.

CT scan of the thorax (Fig. 2), axial non-enhanced CT scan (mediastinal window setting) confirms a solitary, large, well-defined soft tissue mass. The tumor forms acute angles with the adjacent pleural surface and contains central areas of low-density and linear calcifications. There is no erosion nor scalloping of the adjacent ribs and no chest wall invasion. On axial (B) and coronal (C) contrast-enhanced CT scan (mediastinal window setting), a heterogeneous contrast-enhancement of the lesion is seen. The central low-density areas are better delineated on these images and correspond to central necrosis. The coronal reformatted image shows the broad base on the diaphragm.

Figure 3 shows the pathology findings after surgical resection and includes the gross specimen (macrophotograph) and microphotographs (hematoxylin-eosin and CD34 stain).

Radiological diagnosis

Based on imaging findings, the diagnosis of *solitary fibrous tumor of the pleura* was made, which was histopathologically confirmed after surgery, by showing a fibrous tumor of the pleura composed of spindle cells with collagen stroma, with diffuse strong positivity for CD34.

Discussion

Solitary or localized fibrous tumors of the pleura (SFTP) are rare mesenchymal neoplasms mostly affecting the pleura. Lesions with the same histological characteristics can be found in the mediastinum, lung, pericardium and heart as well as in extrathoracic locations such as breast and abdomen. Primary pleural neoplasms are rare, representing less than 5 to 10% of all pleural neoplasms. The prevalence of SFTP is about 2.8 cases per 100.000 registered hospital patients. SFTP equally affects both sexes and occurs most often in the 6th to 7th decade. No relationship has been found between SFTP and exposure to cigarette smoke, asbestos or other environmental pollutants.

Symptoms occur in about 50% of patients and may include cough, dyspnea, hemoptysis, chest pain and heaviness or sensation of a mass moving within the chest. Paraneoplastic syndromes such as hypertrophic osteoarthropathy are seen in 10% of cases and hypoglycaemia in 5% of cases.

SFTP appear on plain radiographs as solitary, homogeneous, well-defined, lobular, solitary mass lesions, typically in contact with the pleural surface or within a fissure. They are mostly found in the middle and inferior hemithorax, and when in contiguity with the diaphragm, they may mimic diaphragmatic elevation on plain chest X-rays. When large, SFTP may demonstrate mass effect on the mediastinum and adjacent lung structures.

On CT a small SFTP shows a homogeneous, well-defined non-invasive, lobular soft-tissue mass with – in most cases – typical acute angles in contact with the pleura. Larger lesions are more heterogeneous and show areas of necrosis, myxoid changes, hemorrhage or cystic degeneration. Calcifications are associated with central necrosis and seen in 7% of the cases. Enhancement after intravenous contrast administration is usually heterogeneous. A pedicle is often present, but rarely visible on imaging studies.

If pedunculated, changes in position of the tumor follow changes in respiration or body position.

On MRI SFTP are of low or intermediate signal intensity on both T1- and T2-weighted images, which is thought to be related to high content of fibrous collagenous tissue and hypocellularity. High SI on T2-weighted images may also occur and may relate to necrosis, cystic or myxoid degeneration and hypercellular areas. MRI is superior to CT in assessment of tumor extension and in excluding chest wall and diaphragm invasion.

SFTP must be differentiated from malignant mesothelioma, which in contrast to SFTP, often presents with chest pain, is smaller in size and frequently invades the thoracic wall. Moreover, in case of asbestos exposure, pleural plaques (calcified or non-calcified) can often be seen. Pleural metastasis of chondrosarcoma is extremely rare and only occurs with high-grade malignancies.

Although atelectasis, pleural effusion and significant mass effect are signs in favour of malignancy, differentiation between benign or malignant SFTP cannot reliably be made on imaging studies. Since its malignant potential, aggressive surgical resection is the treatment of choice. The prognosis for patients with SFTP is generally favourable. Paraneoplastic symptoms – when present – usually disappear after surgery.

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