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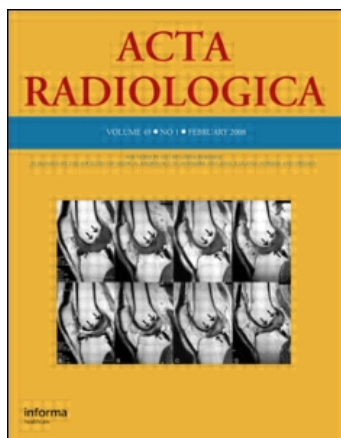
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Solitary Fibrous Tumor of the Lung: Three Rare Cases of Intraparenchymal Nodules

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Solitary fibrous tumor (SFT) of the pleura usually presents as a peripheral mass, in contact with the surface of the pleura. However, on occasion, it can occur separately from the pleura, in the lung parenchyma. We describe the radiological and imaging features of three SFTs of the lung, diagnosed in our department, with relevant clinical data. The diagnosis of SFT of the lung, although rare, should be considered in a slow-growing solitary lung parenchymal nodule.

Key words: Computed tomography; lung neoplasm; solitary fibrous tumor of the pleura (SFTP)

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Solitary fibrous tumor (SFT) of the pleura, first described by Klemperer and Rabin in 1931 (1), is a mesenchymal neoplasm that involves the pleura, unrelated to asbestos exposure or cigarette smoking (2). Usually, SFT of the pleura presents as a peripheral mass, in contact with the surface of the pleura, to which it is joined by a peduncle, which allows mobility within the pleural cavity. Rarely, a fibrous tumor can occur separately from the pleura in the lung parenchyma.

SFT of the lung is a rare mesenchymal tumor entity described in a small number of isolated case reports, mainly in the histologic literature (3, 4). Only in one report were pertaining computed tomography (CT) features described, with particular emphasis on the relative appearance after administration of iodine contrast media (5). We herein describe the radiological and imaging features of three SFTs of the lung, diagnosed in our department, with relevant clinical data.

Case 1

A 44-year-old male textile worker without any evidence of asbestos exposure presented with a

several-week history of chest discomfort on exertion. His past medical history was unremarkable but for a 39-pack-year history of cigarette smoking. Chest radiographs showed a well-defined, homogeneous opacity in the suprahilar region of the left upper lobe (Fig. 1A), which was barely visible on retrospective examination of oblique views of the chest performed 3 years earlier, following a minor blunt thoracic trauma. CT demonstrated a 4-cm left upper lobe mass with well-delineated lobular contour and absence of invasion of the surrounding lung parenchyma. Enlarged left lower paratracheal lymph nodes were seen. The tumor abutted the apical posterior segmental bronchus and several pulmonary vessels (Fig. 1B). After the administration of intravenous contrast material, early scan (25-s delay) showed significant heterogeneous enhancement of the mass, characterized by curvilinear vascular structures encased within the lesion (Fig. 1B). A presumptive diagnosis of a slowly growing tumor of pulmonary origin with rich vascularization was made. At surgery, a segmental resection of the left upper lobe revealed an intraparenchymal, deeply seated, whitish mass, with a bulging cut surface. Pathologic examination revealed an SFT. Neither mediastinal nor pulmonary lymph node

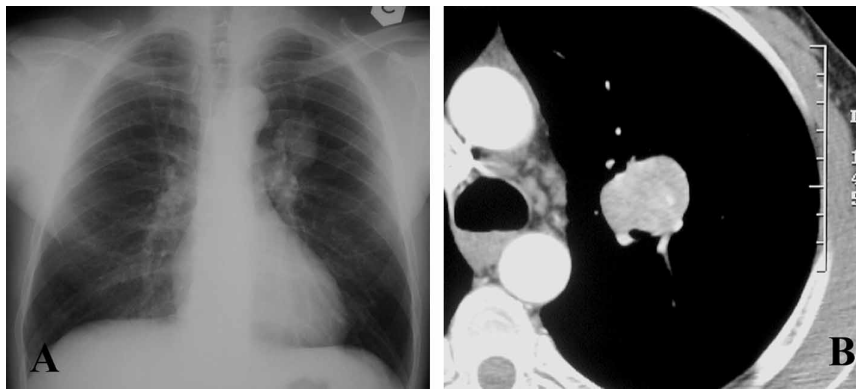


Fig. 1. A. Posteroanterior radiograph shows a lobulated, well-defined opacity of the left upper lobe. B. Corresponding computed tomography scan (mediastinal window) demonstrates strict contiguity of the left upper lobe solid mass with bronchovascular structures.

involvement was detected. The patient had an uncomplicated postoperative recovery, and remains well 4 years after surgery.

Case 2

A 64-year-old man came to our hospital for routine health screening examinations. He was completely asymptomatic and denied any specific medical history or family history. He was an active smoker with a 48-pack-year smoking history and had worked his whole life in the textile industry, with no occupational exposures. Chest radiographs revealed a left lower lobe opacity (Fig. 2A). CT scans demonstrated a well-margined round mass, approximately 6 cm in size. The mass had hetero-

geneous soft-tissue attenuation, and showed intense, heterogeneous enhancement after administration of intravenous contrast material (Fig. 2B). Physical examination was unremarkable, and routine laboratory investigations, spirometry, and arterial blood gases were normal. Bronchoscopy showed no endobronchial lesions. A fine-needle aspiration biopsy was interpreted as showing a low-grade spindle-cell lesion, possibly a hemangiopericytoma. At thoracotomy, a left lower lobectomy revealed a completely intraparenchymal mass. Systematic sampling of pulmonary and mediastinal lymph node stations was performed. Histologic examination showed SFT with no lymph node involvement. Postoperative recovery was uneventful, and the patient remains well 3 years after the operation.



Fig. 2. Chest radiograph (A) and contrast-enhanced computed tomography scan (mediastinal window) (B) show a well-defined mass in the left lower lobe, with some intralesional vessels.

Case 3

A 47-year-old asymptomatic man—an active smoker with a 20-pack-year smoking history—was referred for evaluation of a solitary pulmonary nodule. He had no medical or surgical history, and no significant occupational exposure. Physical examination and laboratory studies were normal. Chest radiograph showed a peripheral opacity in the left lower lung field. On CT, the nodule appeared as a well-defined, intraparenchymal 3-cm mass in the lower lobe. The tumor abutted a pulmonary vessel, without sign of aggressiveness, with no sign of encasement, and showed slightly heterogeneous enhancement after administration of intravenous contrast material (Fig. 3A and B). Bronchoscopy was normal. Although the radiographic findings suggested a benign nature, the lesion remained indeterminate.

Incidentally, an anterior mediastinal mass was found, which was thought most likely to represent a thymoma, and a resection was recommended. During the operation, a nonencapsulated tumor, measuring $9 \times 5 \times 4$ cm, was found to be adherent to the anterior mediastinal fat tissue and the pericardium. En bloc removal of all visible tumor, surrounding thymus gland and adipose tissue, and adherent pericardium was carried out. Microscopic confirmation of negative margins was achieved by frozen section. The left pleural cavity was then opened, and a well-circumscribed, intraparenchymal 3.5-cm lesion was resected from the lower lobe. Final pathology showed both Masaoka stage II thymic carcinoma (World Health Organization type C) and an intrapulmonary SFT of the lower left lobe. The patient was subsequently treated with adjuvant chemotherapy, and remains clinically well 1 year after the operation.

Discussion

Solitary fibrous tumor of the pleura is a rare neoplasm that accounts for less than 5% of all pleural tumors (6). Such mass lesions have also occasionally been reported in other thoracic areas (mediastinum, pericardium, and pulmonary parenchyma) as well as in extrathoracic areas such as the meninges, epiglottis, salivary glands, thyroid, kidneys, and breast (7). Although half of these lesions are pleural, it is not necessary to construct a pleural origin for the intraparenchymal lesions. Several explanations for this intrapulmonary location have been proposed (3, 4): 1) direct continuity of subpleural mesenchyma with interlobular septa connective tissue; 2) invagination of visceral pleura of origin, with mechanical pressure causing slow intrapulmonary growth; and 3) origin from pulmonary parenchyma fibroblasts, with possible entrapment of alveolar pneumocytes and small bronchioles, not to be regarded as a sign of aggressiveness.

Extrapleural solitary fibrous tumors, with the exception of mediastinal tumors, typically follow a benign course, in contrast to SFT of the pleura, which may demonstrate features of local invasion, local recurrence, or intrathoracic spread (8). Radiologically, SFTs are well-defined ovoid or round pulmonary nodules or masses on plain chest radiographs. To our knowledge, the CT features of intraparenchymal SFTs have been described in only one case report (5), in which Patsios et al. reported significant enhancement in early scans (30-s delay). All tumors in our series demonstrated analogous intense and heterogeneous enhancement as well, ranging in dimensions between 3.5 and 6 cm, with sharp margins and round or oval shape. Morphologically, alternating hypercellular and hypocellular sclerotic foci have been observed together

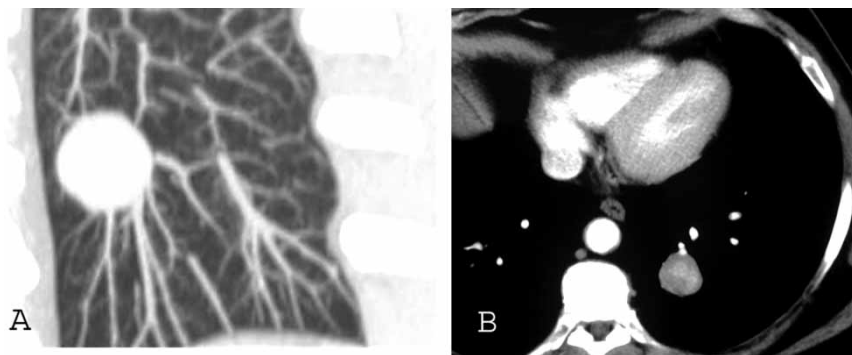


Fig. 3. Coronal reformatted maximum intensity projection (A) and axial computed tomography (mediastinal window) (B) images show a well-marginated nodule with central enhancement abutting vascular structures.

with haphazard proliferation of cytological bland spindle cells separated by variable amounts of hyalinized collagen, often with entrapped bronchiolar and alveolar epithelium. In two cases (1 and 3), the observation of a broad area of contact with bronchovascular structures suggested a presumptive diagnosis of carcinoid, which in about 20% of cases presents as solitary pulmonary nodules in the lung periphery distal to the segmental bronchi, typically as slow-growing nodules (9, 10). CT contrast enhancement and calcifications are common. Atypical carcinoids are also usually peripheral, large, and well-circumscribed masses, similar to that in case 3. Another differential diagnosis is hamartoma, which represents the third most common cause of a solitary pulmonary nodule, following granuloma and carcinoma in frequency (11). Calcification and a fat component are specific findings in hamartomas. However, as many as two-thirds have no foci of calcification or fat density evident on CT, and are difficult to differentiate from malignant solitary pulmonary nodules. Dynamic magnetic resonance imaging (MRI) has been used to assess tumor vascularity (microvessel counts) and interstitium (degree of elastic and collagen fibers), and to predict outcome among patients with peripheral small lung carcinoma (12). Results of several MRI studies of pulmonary lesions have suggested that the kinetic indexes and morphologic parameters of dynamic MRI allow accurate differentiation of malignant and benign lesions (12).

The diagnosis of SFT, although rare, should be considered in a slow-growing, well-defined solitary lung parenchymal nodule. In our opinion, no diagnostic features allow a reliable differentiation from other benign lung tumors.

In conclusion, when an intraparenchymal mass not showing signs of aggressive behavior is found and examined with CT, especially in nonsmokers with no risk factors for lung carcinoma, the diagnosis of intrapulmonary SFT of the pleura should be taken into consideration.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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