

## AND CRITICAL CARE JOURNAL

FOR PULMONOLOGISTS, CARDIOLOGISTS, CARDIOTHORACIC SURGEONS, CRITICAL CARE PHYSICIANS, AND RELATED SPECIALISTS

A Large Thoracic Mass in a 57-Year-Old Patient Rulon H. Sandvliet, Mick Heysteeg and Marinus A. Paul *Chest* 2000;117;897-900 DOI: 10.1378/chest.117.3.897

#### This information is current as of December 4, 2006

The online version of this article, along with updated information and services, is located on the World Wide Web at: http://www.chestjournal.org/cgi/content/full/117/3/897

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### roentgenogram of the month

# A Large Thoracic Mass in a 57-Year-Old Patient\*

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#### (CHEST 2000; 117:897-900)

A 57-year-old woman presented to her general practitioner with progressive shortness of breath that had existed for 1.5 years. The symptoms became obvious while moving to a new home. Because breath sounds of the left lung were found to be almost absent, a chest radiograph was made that showed opacification of almost the entire left hemithorax. Subsequently, the patient was referred to a pulmonologist for further analysis.

Her medical history was unremarkable except for the extraction of a polyp from the uterine cervix. She never smoked and was not on medication.

The patient had no signs of shortness of breath when at rest. Physical examination revealed a BP of 135/75 mm Hg and a regular pulse (64 beats/min). The expansion of the thorax was symmetric. Percussion of the chest was abnormal at the left dorsal lower site, and sounds of breathing were found only at the apex of the lung. The results of routine laboratory studies were normal, as were results of the arterial blood gas analysis.

A chest radiograph (Fig 1) revealed opacification of the left hemithorax, which obscured the lower two thirds of the left lung. A CT scan (Fig 2) showed a large, sharply delineated mass at the site of the left lung. There was some displacement of the mediastinal structures to the right, but there were no signs of infiltration. Only a small part of the left upper lobe appeared to be normal. No abnormalities were seen on the right side.

The following results of spirography showed a clear volume restriction: total lung capacity, 2.85 L (normal, 4.65 L); vital capacity, 1.30 L (normal, 2.74 L). Bronchoscopy revealed a slight narrowing of the left main bronchus, but no endobronchial lesions were seen. The results of cytologic examination of the BAL fluid were negative. A transthoracic puncture revealed RBCs and fibroblasts.

Quantitative perfusion scanning quantified an extensive loss of function on the left side, mainly on the dorsal lower part. The left-to-right ratio was 1:9.

What is the diagnosis?

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Manuscript received May 11, 1999; revision accepted July 6, 1999.

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FIGURE 2. CT scan of the thorax demonstrating a large, welldelineated mass with displacement of the mediastinal structures to the right. No signs of infiltration are seen.

 $\ensuremath{\mathsf{Figure}}\xspace$  1. Chest radiograph showing opacification of almost the entire left hemithorax.

#### Diagnosis: Solitary fibrous tumor of the pleura

When a mass is revealed on a chest radiograph, the first diagnosis to be considered is a malignant disease. Additional information should be obtained to establish the diagnosis prior to surgery and to estimate the chance for a curative thoracotomy. Presumptive diagnoses ranged from a malignant pleural tumor to malignancy occupying the complete lower lobe. Despite the performance of many tests, a conclusive diagnosis was not obtained, and the patient was referred for surgery.

We performed a left thoracotomy, using a standard posterolateral incision. A large tumor was found that originated from the parietal pleura at the dorsal site. There were no attachments of the tumor to the lung, with the exception of a few adhesions at the apex of the lower lobe. A small part of the apex was resected for this reason. The attachment to the pleura had a diameter of 2 cm and was dissected free in continuity with a part of the underlying rib. The stalk contained some large blood vessels, and the ligation of several intercostal arteries was required for complete hemostasis. A smooth tumor measuring  $14 \times 15 \times 7$  cm and weighing 1.5 kg was removed. Histologic examination of the tumor showed spindlelike cells and extensive collagen formation. Mitoses were seen sporadically. The final diagnosis was solitary fibrous tumor (SFT) of the pleura without signs of malignancy. The left lung expanded completely, and recovery was uneventful.

The differential diagnosis of SFT of the pleura includes solitary pleural metastasis, lipoma, pleural fibrosarcoma, intercostal nerve neurilemoma, organized inflammation, and peripheral bronchial carcinoma.<sup>1,2</sup>

The first SFT was described by Wagner<sup>3</sup> in his article "Das Tuberkelähnliche Lymphadenom." In 1931, Klemperer and Rabin<sup>4</sup> discovered a diffuse type of tumor that arose from the mesothelial layer from a localized form that arose from the submesothelial fibrous connective tissue. In 1952, Clagett et al<sup>5</sup> used the term *localized fibrous mesothelioma* to distinguish these usually benign tumors from the more common asbestos-related, malignant mesothelioma. This name also underlines the fact that discussion about its histogenesis is not yet closed.

In the literature, there is a great diversity in nomenclature. Some of these terms are *localized mesothelioma*, *benign fibrous mesothelioma*, *benign localized fibroma*, and *submesothelial fibroma*.<sup>6</sup> To evade the discussion about the histogenesis, the name *solitary fibrous tumor* is advised.<sup>6,7</sup> Such tumors also are described in other sites with mesenchymal tissue such as the lung, pericardium, mediastinum, upper respiratory tract, peritoneum, lever, thyroid, and orbit.<sup>8–10</sup> The incidence of SFT of the pleura is approximately 2.8 cases per 100,000 registered hospital patients.<sup>7,11</sup> SFTs represent < 5% of all neoplasms involving the pleura.<sup>1,12</sup> Two thirds of these tumors arise from the visceral pleura, and one third arise from the parietal pleura. Thirteen percent of the reported cases had an aggressive clinical behavior with local infiltration and local recurrence. Even distant metastases have been described. The remaining 87% had a benign clinical behavior, and radical resection is generally believed to be sufficient treatment.<sup>1,4,11,13</sup>

About 54% of the patients are symptomatic. The most frequent symptoms, dyspnea, coughing, and chest pain, are present in 40% of the patients. Extrathoracic manifestations include arthritic pain in and clubbing of the fingers, called *hypertrophic pulmonary osteoarthropathy*, and hypoglycemia.<sup>11,13–15</sup>

The most important radiologic tool is the chest radiograph. The image of the tumor depends on its size and varies from a sharply delineated round or lobulated mass, with or without pleural effusion, to opacification of the complete hemithorax. In pedunculated tumors, the position of the tumor may change with the position of the patient. CT scanning and MRI are important to evaluate the relationship of the tumor to neighboring structures and to evaluate the resectability of the tumor. In 8 to 17% of the patients, a pleural effusion can be seen, and in 7%, calcifications are present in the tumor. Angiography plays only a minor role.<sup>1,14</sup>

On CT scanning, a sharply delineated, sometimes lobulated mass with the same density as the musculature can be seen. Contrast enhancement is usually intense and homogeneous as a result of the rich vascularization of the tumor. However, a CT scan also may show nonenhancing areas that are due to necrosis, myxoid degeneration, or hemorrhage within the tumor. Displacement rather than invasion of adjacent structures is characteristic. MRI is the most sensitive instrument to exclude invasion of neighboring structures. It defines the extent of the tumor more clearly and provides better tissue characterization. Angiography may be helpful in suggesting the diagnosis in SFTs arising from the parietal pleura.<sup>1,14</sup>

Fine-needle aspiration cytology is mostly inconclusive because the tumor is composed of acellular and hypercellular portions. This makes it difficult to obtain representative material for cytologic analysis. Moreover, the aspiration material mostly consists of fibroblasts, which provide no specific diagnosis.<sup>2</sup> However, a promising report of two cases in which the diagnosis was obtained by fine-needle aspiration biopsy (cytology) recently has been published.<sup>12</sup>

The definitive diagnosis is obtained by histology, usually after surgery.<sup>6,11</sup> Macroscopically, these tu-

mors confirm the image seen on the chest radiograph. Half of the tumors are pedunculated, and half are sessile. On sectioning, they are gray-white and harbor areas with necrosis or hemorrhage. In reported cases, the average diameter is 5 to 10 cm, and the average weight is 100 to 400 g.

The microscopic picture is dominated by a "patternless pattern," with fibroblasts, collagen, and reticule fibers haphazardly arranged or in a hemangiopericytoma-like pattern. Sometimes both patterns coexist.<sup>6,7,11</sup> Acellular and hypercellular areas usually can be seen.

Immunohistochemical analysis shows these tumors to be negative for keratin, S-100, carcinoembryonic antigen, and factor VIII, and positive for vimentin and CD- $34.^{2,12}$ 

The prognosis depends, first, on the resectability of the tumor, second, on its size, and then, in decreasing order of importance, on the mitotic count, polymorphism, and necrosis within the tumor.<sup>8,14,15</sup> Surgical resection is curative in most patients.

#### References

- 1 Desser TS, Stark P. Pictorial essay: solitary fibrous tumor of the pleura. J Thorac Imaging 1998; 13:27–35
- 2 Ali Ž, Hoon V, Syed H, et al. Solitary fibrous tumor. Cancer 1997; 81:116–121

- 3 Wagner E. Das tuberkelähnliche lymphadenom (Der cytogene oder reticulirte Tuberkel). Arch Heilk (Leipzig) 1870; 11:497
- 4 Klemperer P, Rabin CB. Pulmonary neoplasms of the pleura: a report of five cases. Arch Pathol 1931; 11:385–412
- 5 Clagett OT, McDonald JR, Schmidt HW. Localized fibrous mesothelioma of the pleura. J Thorac Surg 1952; 24:213– 230
- 6 Scharifker D, Kaneko M. Localized fibrous "mesothelioma" of pleura (submesothelial fibroma). Cancer 1979; 43:627–635
- 7 Dalton WT, Zolliker AS, McCaughey WTE, et al. Localized primary tumors of the pleura. Cancer 1979; 44:1465–1475
- 8 Batsakis JG, Hybels RD, El-Naggar AK. Solitary fibrous tumor. Ann Otol Rhinol Laryngol 1993; 102:74–76
- 9 Dorfman DM, To K, Dickersin GR, et al. Solitary fibrous tumor of the orbit. Am J Surg Pathol 1994; 18:281–287
- 10 Vallat-Decouvelaere A, Dry SM, Fletcher CDM. Atypical and malignant solitary fibrous tumors in extrathoracic locations. Am J Surg Pathol 1998; 22:1501–1511
- 11 Okike N, Bernatz E, Woolner B. Localized mesothelioma of the pleura. J Thorac Cardiovasc Surg 1978; 75:363–372
- 12 Drachenberg CB, Bouquin PM, Cochran LM, et al. Fine needle aspiration biopsy of solitary fibrous tumors. Acta Cytol 1998; 42:1003–1010
- 13 Nelson R, Burman SO, Kiani R, et al. Hypoglycemic coma associated with benign pleural mesothelioma. J Thorac Cardiovasc Surg 1975; 69:306–314
- 14 Ferretti GR, Chiles C, Choplin RH, et al. Localized benign fibrous tumors of the pleura. AJR Am J Roentgenol 1997; 169:683–686
- 15 Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura. Cancer 1981; 47:2678–2689

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