A localized fibrous tumor of the pleura (LFTP) is a slow growing, primary pleural neoplasm unrelated to asbestos exposure, and accounts for fewer than 5% of pleural tumors (1). It occurs in both sexes and in all age groups, but predominantly affects those in the fifth to seventh decades of life. England et al. (2) found that 60% of these tumors were benign and 40% were malignant, as determined microscopically on the basis of cellularity, pleomorphism, mitotic activity, hemorrhage, and necrosis. Several radiologic descriptions of benign LFTP have been published, but the radiologic findings of malignant LFTP involving invasion of the chest wall have not been reported. We describe a case of malignant LFTP with chest wall invasion, confirmed pathologically, and review the literature.

Index words: Pleura
Pleura, Computed tomography (CT)
Pleura, neoplasms

Case Report

A 60-year-old man with mild chest pain underwent chest radiography, and a large mass was found to occupy the lower two thirds of the left side of the chest, shifting the mediastinum to the right and obliterating the left costophrenic angle (Fig. 1A). CT images depicted a large (14×17 cm), well-defined, heterogeneously enhanced mass, with destruction of the anterior arc of the left fourth rib and invasion of the left anterior chest wall. In the peripheral portion of the mass lesion, the internal contour of the enhanced vessel was serpiginous or tubular, and in the central portion, there was severe necrosis. Due to the presence of the mass, the left lower lobe had collapsed, and the mediastinal structure was displaced to the right. Ipsilateral pleural effusion was substantial (Fig. 1B, C). Transthoracic needle biopsy performed at that time showed that a malignant schwannoma was present.

At thoracotomy, a firm, encapsulated mass, measuring 20×12×10 cm, was found to be attached to the anterior chest wall by a pedicle. The tumor had invaded the adjacent anterior chest wall, and the anterior arc of the left
fourth rib showed mild erosive change without destruction. A cut section of the mass showed that it was yellowish white and had a solid, fibrotic or edematous appearance with central hemorrhage and necrosis (Fig. 1D). Except for the stalk, these findings correlated well with those of CT. Microscopically, the mass was com-

Fig. 1. A. Chest radiograph shows a large mass occupying the lower two thirds of the left side of the chest, shifting mediastinum to the right and obliterating the left costophrenic angle. 
B, C. CT images depicted a large (14×17 cm), well-defined, heterogeneously enhanced mass, with destruction of the anterior arc of the left fourth rib and invasion of the left anterior chest wall (arrow). In the peripheral portion of the mass lesion, the internal contour of the enhanced vessel (arrowhead) was serpiginous or tubular, and in the central portion, there was severe necrosis. Due to the presence of the mass, the left lower lobe had collapsed, and the mediastinal structure was displaced to the right. Ipsilateral pleural effusion was substantial. 
D. The mass is relatively well demarcated, and the cut section of the mass showed that it was yellowish white and had a solid, fibrotic or edematous appearance with central hemorrhage and necrosis. 
E. On microscopic findings, the tumor cells are plump and mitoses are occasionally found (H & E, × 400).
posed of spindle cells in a fascicular pattern; there was a focal myxoid zone and cellularity was high, with frequent mitoses (more than 4/10 high-power field), consistent with a malignant localized fibrous tumor of the pleura (Fig. 1E). The patient was scheduled for radiation therapy.

Discussion

Tumors of the pleura are classified as either diffuse or localized. Diffuse malignant mesothelioma is the most common primary pleural tumor, and is most often related to asbestos exposure. The prognosis is poor. In contrast, a localized fibrous tumor of the pleura (LFTP) is rare, not related to asbestos exposure, and the prognosis after surgical resection is generally good.

The origin of the LFTPs is controversial, with researchers arguing in favor of either mesothelial or submesothelial proliferation (2, 8). A recent study found a common characteristic immunophenotype for localized fibrous tumor of the pleura that confirms a mesenchymal, nonepithelial origin (8).

The tumor has been found in all age groups but has a peak incidence in individuals aged over 50. It is equally common in men and women. Approximately 50% of patients are asymptomatic (2, 9), the lesion being discovered incidentally at routine chest radiography. In symptomatic patients, the most frequent manifestations are chest pain, cough and dyspnea, which occur in 40% of such patients. These tumors are frequently associated with extrathoracic manifestations, including hypertrophic pulmonary osteoarthropathy and hypoglycemia. The former has been noted in 4-35% of patients, while hypoglycemia is usually associated with very large tumors and occurs in less than 5% of patients (2, 9). In our case, neither hypertrophic pulmonary osteoarthropathy nor hypoglycemia occurred.

Macroscopically, LFTP appears as a firm, round to oblong, gray-white tumor, usually circumscribed by a thin capsule. Approximately 65-80% of LFTPs arise from the visceral pleura, and the rest from the parietal pleura (2, 9). Forty percent are attached to the pleura by a pedicle, in which case they are usually benign; most are attached to the visceral pleura, but some malignant LFTPs also have a pedicle (2). Most unresectable tumors originate from the parietal pleura. Tumors range in diameter from 1 to 39 cm; those larger than 10 cm are more likely to be malignant (2). The tumor in this case was connected to the pleura by a pedicle, but due to severe invasion of the chest wall, we were unable to identify its origin.

Microscopically, an LFTP is composed of haphazardly arranged fascicles of elongated spindle cells separated by varying amounts of collagen (2, 9). Tumor cellularity varies from one area to another; tumors may be predominantly fibrous and acellular, cellular with less collagen, or mixed. Myxoid change or hyalinization occurs in areas of dense collagen tissue, and in large tumors, areas of necrosis and hemorrhage may occur. The fibrous and acellular nature of these tumors makes their diagnosis at transthoracic needle biopsy difficult (9); the lesion in this present case was initially identified as a malignant peripheral nerve sheath tumor after transthoracic needle biopsy, but confirmed as a malignant LFTP after removal, and it appears that for correct diagnosis, surgery is required. England et al. (2) found that 60% of these tumors were benign and 40% were malignant, as determined microscopically on the basis of cellularity, pleomorphism, mitotic activity, hemorrhage, and necrosis. They have a relatively good prognosis: all benign tumors and 45% of malignant tumors respond to surgical excision (2). The majority of unresectable tumors have more aggressive histologic findings and associated pleural effusion.

Radiologically, an LFTP appears as a solitary, sharply defined, and sometimes lobulated mass, varying in diameter from 1 to 39 cm, and may be located in an interlobar fissure or adjacent to the diaphragm, mediastinum, or chest wall (1, 3-6). The mass is connected to the pleura and classically forms obtuse angles with the chest wall. In case involving large masses or these attached to the pleura by a pedicle, the angles with the chest wall may be acute, mimicking a mass of pulmonary origin (1, 4). Calcification or pleural effusion was evident in four (7%) and ten (17%) of 58 cases reviewed in the AFIP series (1). Compared with those which are benign, malignant tumors commonly give rise to complications (6), and large tumors may compress adjacent lung parenchyma and mediastinal structures. The tumor under review was a large mass that compressed the adjacent lung and mediastinum, with ipsilateral pleural effusion, but correct evaluation of its origin was precluded by its large size. Pedunculated tumors may be mobile, changing in position according to respiration or posture, or from image to image during serial chest radiography (4). In this case, the tumor had a stalk, but the mass was not mobile due to its large size and invasion of the adjacent chest wall.

At CT, a well-delineated, often lobulated mass in con-
tact with the pleura is typically observed, and on unenhanced CT scans, soft-tissue attenuation is apparent (3). Calcification is principally noted in large tumors and is related to areas of necrosis (2), and as a result of the rich vascularization of the tumor, contrast enhancement is usually intense and homogeneous. CT, however, may reveal non-enhancing areas that correspond to necrosis, myxoid degeneration, or intra-tumoral hemorrhage (3). As on chest radiographs, the angle between the tumor and the chest wall may cause confusion in locating the tumor; this sign is more likely to be helpful in localizing a small lesion, which forms obtuse angles with the chest wall, than large masses, the angles of which are most often acute. With regard to the identification of a pedicle, this was possible in four of 16 patients in a study by Mendelson et al. (10). CT findings that suggest a malignant fibrous tumor include a diameter greater than 10cm, central necrosis, and ipsilateral pleural effusion (6). In this present case, accurate determination of the origin of the tumor was not possible due to its large size, but since there was destruction of the adjacent rib and invasion of the chest wall though to a relatively minor extent compared with the tumor’s bulk, it is assumed that the tumor originated from the pleura. In addition, the tumor’s large size and its central necrosis, as well as the presence of ipsilateral pleural effusion and the destruction and invasion which occurred, suggest the its potential malignancy.

The differential diagnosis of a localized mass abutting or invading the pleura include lipoma, peripheral bronchogenic carcinoma, and other pulmonary neoplasms (7). Accurate evaluation of the origin of the tumor is not always possible, but when a tumor is large and there is severe necrosis, invasion of the chest wall, and ipsilateral pleural effusion, the possibility of a malignant pleural tumor should be considered. Diagnosis based only on transthoracic needle biopsy is inaccurate; for accurate diagnosis and treatment, surgical resection is required.

References