Primary Pulmonary Leiomyosarcoma
Review of the Literature and Report of One New Case

H. A. Fadhlí, M.D., A. W. Harrison, M.D., F.C.C.P. and S. H. Shaddock, B.S.
Galveston, Texas

Thirty-eight cases of primary pulmonary leiomyosarcoma have been reported in the literature. The age range has been from 4 to 83 years with 17 of the patients being between 50 and 70 years of age. Twenty were in males and 18 in females. The right lung was involved in 18 cases and the left lung in 16 cases. In one, both lungs and the left atrium were involved. In one only the left parietal pleura was involved. The trachea and the carina were the site of a leiomyosarcoma in one case each. Although metastases to other organs occurred occasionally, there was no metastasis to hilar or mediastinal lymph nodes. The more peripheral tumors were often associated with pleural adhesions.

When they occurred in the lung parenchyma, the leiomyosarcomas usually were ovoid or spherical and often were encapsulated. The cut surface usually was described as white or gray and firm. Microscopically, the tumors typically were composed of large embryonal muscle cells with long nuclei and myofibrils in the cytoplasm.

The most common symptoms, in decreasing order of frequency, were cough, chest pain, dyspnea, hemoptysis, fatigue, and weight loss. Evidence of a lesion was found in all cases in which radiologic examination was performed. The usual findings were the presence of a mass, atelectasis and shift of the mediastinum. In one, a large cavitory lesion with air-fluid level was seen.

Diagnosis was made by bronchoscopy in eight. Twenty-one were diagnosed surgically and of these, 13 had not been evident on bronchoscopy. Nine were found at necropsy.

*From the University of Texas Medical Branch, Department of Surgery, Division of Thoracic Surgery.

FIG. 1: Shows consolidation with pleural reaction and a large cavity with fluid level in the apex of the right lung. FIG. 2: Shows diminution in the amount of fluid in the cavity.
Twenty-six patients received surgical treatment. Pneumonectomy was performed in nine, lobectomy in seven and resection in five. Pneumonectomy or lobectomy with resection of adjacent structures was done in four and in one, four tumors were excised. Two were given radiotherapy. Three refused operation.

In those in whom pneumonectomy was performed, two died during surgery,\(^1\) one died in two months,\(^1\) two died in two years,\(^1,12\) one died in nine years,\(^1\) and three were alive and in good health when they were reported one year,\(^1\) two years,\(^1\) and five years postoperatively. Of those receiving lobectomy, one died on the 13th postoperative day,\(^1\) one died in the sixth postoperative year,\(^1\) two were said to be alive and well four months\(^1\) and one year postoperatively and three were not followed up.\(^1,12,13\) One on whom resection was performed was not followed up,\(^1\) but the other four were in good health three months,\(^1\) one year,\(^1\) six years\(^1\) and nine years postoperatively. The patient in whom four tumors were shelled out was alive six years after surgery without change in the tumors left behind or recurrence at the site of the resected tumors.\(^5\) Among those in whom pneumonectomy or lobectomy with resection of adjacent structures were performed, one died in 18 months\(^1\) and the remaining three were alive and in good health eight months,\(^3\) three years,\(^3\) and 21 years\(^3\) after operation.

One receiving radiotherapy died in eight months\(^5\) and the other in ten months.\(^5\) All who refused surgery died within two years.\(^3,14,15\)

**CASE REPORT**

A 69-year-old man had a chief complaint of blood streaking his sputum for a period of three days. His cough, which had been non-productive for several years, had become productive three months previously of one-half to one cup of thick yellow sputum daily. He had had pain in the right pleural region which was independent of respiration and seemed to diminish with exercise. There had been occasional night sweats and a weight loss of eight pounds during the previous six years. He had smoked one package of cigarettes daily for approximately 40 years.

The blood pressure was 120/60; the pulse was 80 and regular; and respirations were 20. The chest showed an increased anteroposterior diameter with slightly increased resonance. There

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**Fig. 3** Shows further emptying of the cavity after a bout of productive cough. **Fig. 4** Shows enlargement of the recurring tumor in the right upper lung field, with involvement and destruction of the upper five ribs in the right axillary region (7 months postoperatively).
were a few scattered rhonchi. The hemoglobin was 11.7 grams per cent; the hematocrit was 37 per cent, and there were 11,500 white cells per cmm. Sputum culture revealed normal flora and sputum cytology was negative for malignant cells.

An initial chest x-ray film (Fig. 1) on February 8, 1963, showed consolidation with pleural reaction and a large cavity with fluid level in the apex of the right lung. Bronchoscopic examination on February 22, 1963, revealed narrowing of the right main stem bronchus, apparently due to extrinsic pressure, and narrowing of the right upper lobe bronchial openings, due to a severe inflammation. Bronchoscopic and scalene node biopsies were negative for granulomatous disease or malignancy. Progress chest x-ray films showed diminution in the amount of the fluid in the cavity (Fig. 2). On February 27, 1963, a chest x-ray film taken after the patient had a bout of productive coughing revealed further emptying of the cavity (Fig. 3). Because the cavity was thought to be an abscess, it was elected to administer antibiotics, INH and streptomycin for three weeks before surgical exploration.

Right anterior thoracotomy was performed on April 1, 1963. The right upper lobe contained a firm round mass which measured about three inches in diameter and was found to be attached to the posterior chest wall and to the superior segment of the lower lobe by adhesions. Extra-pleural resection of the entire upper lobe was done and a few hilar lymph nodes were excised.

The tumor contained a large cavity 5 cm. in diameter, the walls of which were 1.0 to 2.0 cm. thick and on cut section, the wall had a dirty yellow-gray appearance. The final pathologic diagnosis was that of primary pulmonary leiomyosarcoma with extension to the pleura. The hilar lymph nodes showed no evidence of metastases.

He had a favorable postoperative course and left the hospital on April 22, 1963, to be followed as an outpatient.

On July 25, 1963, he complained of occasional pain in the right axilla and scapular areas. A firm 1 cm. nodule was found in the right axilla. This nodule was fixed to the chest wall. A chest x-ray film showed an area of nodular density in the right upper lateral chest and evidence of destruction of the entire third rib at the axillary area. Roentgenographic examination of the upper gastrointestinal tract and a barium enema showed no abnormal finding.

On November 5, 1963, he complained of pain in the right axilla. The axillary mass at that time measured 8 x 4 cm. A chest x-ray film showed increase in the size of the mass with extension into the right axilla.

He is being followed in the outpatient clinic and receiving two courses of l-sarcolysine without benefit.

References