Primary Pulmonary Leiomyosarcoma

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A review of the literature1-21 reflects the rarity of primary, smooth-muscle-cell tumors of the lung. Of a total of 24 such tumors reported since the turn of the century, 16 were leiomyosarcomas (Table I) and eight benign leiomyomas (Table II).

Case Report

J. R., a 19-year-old white male farmer, was first seen on June 7, 1954, for complaints including weakness, anorexia, and loss of 35 lbs. weight since March, 1954. Although a recent posteroanterior chest film had been reported as normal, chest x-ray film was repeated, revealing a large round tumor mass with small nodular extensions (Fig. 1), all of which had been previously hidden by the cardiac shadow. A left lateral x-ray film of the chest localized this mass in the left lower lobe behind the heart.

The patient suddenly became dyspneic and developed signs and symptoms of severe internal hemorrhage. X-ray films now showed the left hemithorax to be opaquely blurred with some shift of the mediastinum to the right (Fig. 2). Closed (water-sealed) drainage through catheters introduced intercostally resulted in escape of considerable bloody fluid. X-ray film showed opacity of the lower two-thirds of the left lung field with some improvement in the position of the mediastinum, but dyspnea, requiring the administration of oxygen through an intranasal catheter continued. He was given whole blood transfusions and his blood pressure maintained at 120/80. He was started on penicillin, and his oral temperature of 104° fell over a period of a few days to 101°. His pulse rate fell from 150 to 120 per minute. He was unable to void voluntarily (necessitating an indwelling Foley catheter), sweated profusely and complained of dyspnea, weakness and chest and back pain. Bronchoscopy and studies of the bronchial secretions for malignancy were negative.

On June 29, 1954, operation was performed through a standard left thoracotomy incision. The pleural space was filled with an extensive blood clot and some free bloody fluid. Almost as soon as removal of this blood clot was undertaken, the anesthetist reported that free bloody fluid was filling and escaping out of the endotracheal tube. The operator (A.M.T.) introduced his hand blindly through the extensive blood clot to the upper hilar region and compressed the left main stem bronchus there, later applying a Satinsky clamp to this region. The anesthetist was then able to clear the tracheobronchial tree and maintain a satisfactory anesthetic for the remainder of the operation.

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The large, round, nodular tumor mass occupying the lower posterior and medial portion of the left lower lobe was removed in toto by left lower lobectomy. An enlarged hilar lymph node was also removed. Frozen section reports indicated that the lymph node showed no evidence of malignancy, but it appeared that the tumor mass itself was probably malignant, although the exact type of malignancy could not be definitely determined at the time. In view of this report, the remainder of the lung was removed. Several small areas of the pericardium, diaphragmatic surface and parietal pleura were selected for biopsy.

The 300 gm. tumor in the left lower lobe measured 11 cms. in its maxi-

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<th>Author</th>
<th>Year Reported</th>
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<tr>
<td>Neumann¹</td>
<td>1938</td>
<td>Two cases pulmonary leiomyosarcoma, females, aged 63 and 83. (Autopsies)</td>
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<tr>
<td>Brunn et al²</td>
<td>1940</td>
<td>Leiomyosarcoma, left lower-lobe bronchus, 55 year old male. (Patient refused operation; died of the disease)</td>
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<tr>
<td>Randall et al³</td>
<td>1946</td>
<td>Leiomyosarcoma, right lung, 34 year old Negro. (Died during operation for removal of tumor)</td>
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<tr>
<td>Holinger et al⁴</td>
<td>1950</td>
<td>Myosarcoma, anterior wall of trachea, five year old white female. (Successfully removed)</td>
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<tr>
<td>Mishkin⁵</td>
<td>1951</td>
<td>Myosarcoma, right main bronchus, six-year-old white female (Died after roentgen-ray therapy)</td>
</tr>
<tr>
<td>Johnson et al⁶</td>
<td>1952</td>
<td>Leiomyosarcoma, right upper lobe, 52 year old white male. (Successfully treated by pneumonectomy)</td>
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<tr>
<td>Killingsworth et al⁷</td>
<td>1953</td>
<td>Leiomyosarcoma, left lower lobe, seven year old white female. (Successfully treated by pneumonectomy with partial excision of left atrium)</td>
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<tr>
<td>Watson et al⁸</td>
<td>1954</td>
<td>Six cases, pulmonary leiomyosarcomas. (3 successfully treated by surgical excision)</td>
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<tr>
<td>Rosenberg et al⁹</td>
<td>1955</td>
<td>Concurrent leiomyosarcoma and carcinoma, right bronchus, 62 year old white male, (Treated by pneumonectomy)</td>
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mum diameter (Fig. 3). It was non-encapsulated but separated easily from the surrounding lung leaving a shallow ragged depression. The cut surface of the tumor was homogenous and of mottled yellow-brown color.

Spindle cells predominated an interlacing pattern, with variations in-
cluding ovoid, ribbon and tumor giant cells (Fig. 4). With von Gieson's stain the tumor cells took a yellow color. When stained with phosphotungstic acid hematoxylin, some of the elongated cells exhibited blue myogial fibrils.

The remaining lung tissue showed hyperemia. The bronchi showed no significant pathologic change. The additional specimens (hilar lymph nodes, a segment of pericardium, pleura and the remainder of the left lung) showed no involvement by the tumor.

Discussion

Opinion among the pathologists varied as to the malignant potential of this type of tumor. It was pointed out that some of the more benign types of uterine myomata have been proved to metastasize while the more pleomorphic varieties apparently can remain localized for long periods. Expert opinion on this tumor's microscopic picture favored the probability that this was a benign tumor.22

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<tr>
<td>Forkel&quot;</td>
<td>1910</td>
<td>Leiomyoma, left upper lobe, 63 year old female. (Autopsy)</td>
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<tr>
<td>Deussing&quot;</td>
<td>1912</td>
<td>Multiple primary pulmonary myomas, 57 year old female. (Autopsy)</td>
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<tr>
<td>Franco&quot;</td>
<td>1929</td>
<td>Voluminous leiomyoma, right upper lobe, male. (Autopsy)</td>
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<tr>
<td>Brahy&quot;</td>
<td>1941</td>
<td>Fibroleiomyoma, 22 year old female, right lower lobe. (Successfully treated by resection of lower two-thirds right lower lobe)</td>
</tr>
<tr>
<td>Williams et al&quot;</td>
<td>1950</td>
<td>Leiomyoma, left lower lobe, eight year old female. Youngest patient in series of benign tumors. (Successfully treated by pneumonectomy)</td>
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<tr>
<td>Turkington et al&quot;</td>
<td>1950</td>
<td>Leiomyoma, left lower lobe, 57 year old female. Only case diagnosed preoperatively by bronchoscopic biopsy. (Successfully treated by pneumonectomy)</td>
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<td>Freireich et al&quot;</td>
<td>1951</td>
<td>Leiomyoma, right upper lobe, 61 year old male. (Successfully treated by pneumonectomy)</td>
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<tr>
<td>Pierce et al&quot;</td>
<td>1954</td>
<td>Leiomyoma, 24 year old white male, right middle lobe. (Successfully treated by lobectomy)</td>
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The immediate and early post-operative course was uneventful and the patient was discharged from the hospital on the 12th post-operative day. He appeared to be in excellent health when, four months after operation, two hard, freely movable, painless subcutaneous nodules approximately 2 cms. in diameter were noted—one in the left anterior abdominal wall and the other on the right thigh. These lesions increased in size, one extending to the skin surface where an ulceration 2 cm. in diameter developed. In December, 1954, this ulcerated lesion was excised, revealing a 3.5 cm. tumor mass, light red and soft to rubbery. Microscopic examination (Fig. 5) showed a pleomorphic tumor, more highly undifferentiated in most areas than the resected pulmonary lesion, composed of solid sheets of fairly large round to ovoid cells, with wide areas of necrosis present throughout the tumor tissue. A few areas showed spindle-shaped cells resembling those seen in the tumor removed from the lung.

Although he continued to feel well for several months, he lost weight and eventually complained of chest pain, weakness and some shortness of breath. On December 6, 1954, a chest x-ray film (Fig. 6) revealed several metastatic nodules in the remaining lung. Early in March, 1955, he was again hospitalized when a large mass was visible and palpable in his right thigh. He died on March 22, 1955.

Autopsy Findings: A soft, white, well-circumscribed, extremely friable tumor extended through the intercostal spaces at the level of the stump.
of the sixth rib (which had been partially removed when the left pneumonectomy was performed). This tumor, palpable externally as a small mass beneath the intact skin, was an extension of similar tumor tissue filling the left pleural space, firmly adherent to the left chest wall, to the diaphragm and to the apex of the chest cavity. Two well circumscribed large nodules, each approximately 8 cm. in diameter and showing areas of necrosis, were present within this intrapleural mass of malignant tissue. Some blood was present in the left pleural space. The fatty tissue surrounding the pericardial sac showed infiltration by the firm pale white tumor tissue, which appeared well-circumscribed. The heart, somewhat dilated and enlarged, with cloudy swelling of the myocardium, was shifted to the left. The enlarged right lung contained two pedunculated tumors on its surface, and five other tumor masses within the lung substance varying from 2 to 8 cms. in size. These soft white well-circumscribed tumors, the larger of which showed central necrosis, could be freely shelled out of the surrounding hemorrhagic lung tissue. The enlarged tracheobronchial lymph nodes on the right side showed evidence of tumor metastases. In the abdomen, the lymph nodes at the head of the pancreas and at the hilus of the liver were enlarged and contained metastatic tumor nodules. The liver weighed 2300 gms., the cut surface showing the mottled red appearance of chronic passive congestion with fatty infiltration. The spleen was also enlarged, the splenic pulp being soft and dark red and the follicles being extremely prominent. The other abdominal organs were essentially normal. Under the fascia of the right thigh was a tumor mass measuring 6 by 9 cms. which could easily be shelled out and appeared to displace rather than infiltrate the muscles.

Discussion

A total of 17 primary pulmonary leiomyosarcomas, including our own, has been reported since the turn of the century,1-9 nine in females and eight in males, the patients' ages ranging from four to 83 years. The tumors were successfully excised in seven, the surgical procedures varying from local excision of the neoplasm from the trachea4 or lung,6 to lobectomies8 and pneumonectomies.6,7,9 In one, radical pneumonectomy with excision of a portion of the left atrium was necessary to remove the neoplasm.7 It appears, therefore, that primary leiomyosarcoma of the lung, when localized, may be eradicated surgically. Roentgen ray therapy is also effective as a palliative procedure.

Primary pulmonary leiomyoma has been reported in a total of eight cases,10-17 five in females and three in males, the ages ranging from eight to 63 years.

The low incidence of benign bronchial tumors in general as compared to malignant tumors can be appreciated by the report by Adams21 covering a 16 year series totaling 164 cases in which the ratio of malignant tumors was 22 to one. In a review of the literature, Randall and Blades9 noted the scarcity of reports of mesenchymal tumors of bronchial origin, and particularly of benign tumors arising from smooth muscle.
Acknowledgment: Autopsy was performed on our case and report kindly furnished by Lorin E. Dickemann, M.D., Pathologist, St. Elizabeth’s Mercy and Grace Hospitals, Hutchinson, Kansas.

SUMMARY

A case of a patient with leiomyosarcoma of the lung treated by pneumonectomy is reported. A review of the literature disclosed that 16 such primary pulmonary malignant lesions have been reported, six of which were apparently treated successfully by surgical excision. Leiomyoma, according to the reports in the literature, occurs less frequently, having been reported in only eight cases.

REFERENCES

22 Brines, O. A., Chief of Department of Pathology, Wayne University Medical School, Detroit, Mich.: Personal communication.