PRIMARY PULMONARY SARCOMA

A Case Report

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It is generally agreed that the incidence of primary malignant pulmonary neoplasms has increased markedly in recent years, although the extent of the increase varies in different countries. American observers (1, 2) believe that primary malignant tumors of the lung form about 1 or 2 per cent of all cancers. According to Hunt (3) and Davidson (4), European statistics show a higher incidence, this type of tumor being found in about 7 per cent of all autopsies on patients dying with malignant disease, and ranking next in frequency to carcinoma of stomach, breast, uterus, and esophagus. Recent reports (5, 6, 7) also indicate that intrathoracic tumors are apparently increasing much more rapidly than the general increase in cancer would warrant, due probably to better diagnostic methods and increased human longevity. It is evident, however, that the greater incidence of these tumors is due to the increased recognition of pulmonary carcinoma and that primary sarcoma of the lungs still remains an obscure and comparatively rare condition.

Adler (8) in 1911 collected from the literature 90 cases of pulmonary sarcoma, but the evidence presented in many cases is insufficient to warrant their inclusion in this group. Among 9246 autopsies at the Breslau Pathological Institute only four primary sarcomas of the lung were found (9). Stout (10) states that true mesoblastic malignant tumors of the lung are very rare, and Ewing (11) that mesoblastic tumors only occasionally arise in that organ. Ball (12), in a review of the German, French, and English literature since 1900, was able to collect only 14 authentic cases of primary pulmonary sarcoma. The following case, therefore, because of its rarity and its unusual extensions into neighboring structures, was deemed worthy of reporting in detail.

Case Report

S. A. J., a mechanic, aged eighteen, was referred to the Queen Alexandra Sanatorium, London, Ontario, in March 1931, with complaints of persistent cough, hemoptysis, and slight loss of weight.

His family history was negative. He had had the usual childhood diseases, and typhoid fever at twelve years of age. Otherwise he had always enjoyed good health and had participated considerably in amateur sports.

Present Illness: The onset of the present illness occurred about the middle of January 1931, with symptoms of rhinitis, moderate cough, and expectoration. Some days later,
following a strenuous game of hockey, the patient coughed up about half an ounce of blood clot. The next day he consulted his physician, who found his temperature slightly elevated. The cough persisted, with moderate expectoration containing an occasional blood clot. However, the patient felt quite well and worked steadily until the end of February 1931. He had no dyspnea or chills and noticed no fatigue or change in strength, although he lost some five pounds in weight in about six weeks.

On admission to the sanatorium on March 7, 1931, the temperature was 99.8° F., pulse 96, respiration 22, weight 120 pounds.

The physical examination revealed a moderately well nourished youth, slightly flushed, who did not appear acutely ill. The physical findings were negative with the exception of those within the thorax. The chest was rather poorly developed. Impaired resonance was noted over the upper right chest, with widening of mediastinal dullness in the first

![Figure 1: Roentgenogram showing a circular, discrete area of moderately dense homogeneous opacity occupying the inner halves of the first and second intercostal spaces on the right](image)

and second right interspaces anteriorly and in the interscapular region on the right side posteriorly. The breath sounds were bronchovesicular in type over the first and second interspaces anteriorly. The heart appeared slightly enlarged to the left, with a systolic murmur heard about half way between the pulmonic and mitral areas.

**Laboratory Findings:** A blood count revealed 4,700,000 red blood cells, with 70 per cent hemoglobin and 8,600 white blood cells per cubic millimeter, with 79 per cent polymorphonuclears. Urinalysis showed a faint trace of albumin. The Wassermann and Kahn tests were negative. Repeated sputum tests were negative for acid-fast organisms. Stereo-roentgenograms of the chest revealed a moderately dense, homogeneous, circular, discrete deposit 2.5 inches in diameter occupying the inner half of the right first and second interspaces.

**Progress Notes:** The patient remained in the sanatorium for some five months. The evening temperature was usually around 99° F., with an occasional elevation as high as 100° F., with a pulse of 80 to 112. There continued to be a slight hacking cough with as much as an ounce of mucopurulent sputum, frequently blood-stained. Moreover, free blood was coughed up on frequent occasions, sometimes amounting to four ounces. The
lesion in the right lung extended steadily and there was an effusion into the right pleural space. On March 30, 1931, 100 c.c. of a dark bloody fluid, which failed to show evidence of clotting, was aspirated from the right pleural cavity. Four months later 10 c.c. of a thick, bright red fluid was aspirated from the center of the mass in the right lung. This fluid clotted readily, and sections of the clot showed typical areas of spindle-cell sarcoma interspersed throughout normal blood clot. The patient was discharged from the sanatorium two weeks later. At this time the physical findings were impairment of resonance over the upper two-thirds of the right lung field, bronchial breathing, and marked pectoriloquy with numerous medium moist râles over the upper half of this side anteriorly. The patient was considerably weaker and had lost about 22 pounds.

At home the patient's condition became progressively worse. He rapidly became emaciated, marked dyspnea developed, and cyanosis was frequently present. He con-

Fig. 2. Roentgenogram of later date than Fig. 1, showing considerable increase in extent of the mass, with sharply defined convex lower border

continued to cough up small amounts of blood. Death occurred on March 3, 1932, fifteen months after the onset of symptoms.

Summary of Autopsy: The majority of the post-mortem findings concerned the thorax. The right pleural cavity was practically obliterated by firm fibrous adhesions, and it was impossible to remove the right lung intact. On removal the lung was seen to be replaced almost entirely by a crumbly, grayish-white, hemorrhagic tumor mass, only a narrow rim of atelectatic lung tissue being seen about the margin of the new growth. The tumor was very friable and broke off readily into large, soft, translucent, grayish-white masses. There was extensive hemorrhage throughout, and the central portion of the tumor appeared to be cystic in character, filled with blood clot and necrotic tumor tissue. Medially the tumor could be traced into and along the right main bronchus to about 1.5 cm. above the bifurcation of the trachea. The right bronchus was entirely occluded and the growth enroached on the opening of the left main bronchus. The tumor was not attached to the bronchial mucoa but appeared to be growing along the lumen of the bronchus. It also occupied the lumen of the right pulmonary vein and extended along this structure into the left auricle, which was practically filled by a firm, brownish-red, oval mass of tumor about 4 × 2 cm. The tumor lay free in the auricle but
appeared to have invaded the intimal lining of the vein. The cavity of the auricle was
filled fairly completely by the tumor mass and there was definite obstruction of the
auriculo-ventricular valve.

The left lung revealed a number of small, firm, discrete nodules, the largest about
4 mm. in diameter, radiating from the hilus into the lower portion of the upper lobe and
the upper portion of the lower lobe. On section these nodules were firm, grayish-white,
and somewhat translucent.

The liver externally appeared normal but section revealed several small nodules, the
largest about 5 mm. in diameter, similar to those seen in the left lung. No other tumor
nodules could be found throughout the remainder of the body, although a careful search
was made for any other possible primary tumor.

Microscopically the tumor is extremely cellular and is composed of two types of cells,
round and spindle cells. The spindle cells appear to predominate and are seen throughout
the entire tumor. The firmer peripheral portions of the tumor and the extensions into

**DISCUSSION**

Excluding lymphosarcoma, which is not regarded as primary in the
lung, pulmonary sarcoma is usually classified on a morphological basis
into two groups: (1) spindle-cell sarcoma, the more common type, oc-

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**FIG. 3**

**FIG. 4**

**FIGS. 3 AND 4. GROSS APPEARANCES OF MEDIAL ASPECT OF RIGHT LUNG (FIG. 3), SHOWING
THE RIGHT BRONCHUS OCCLUDED BY TUMOR GROWTH, AND OF THE HEART (FIG. 4) OPENED
TO SHOW THE TUMOR IN THE RIGHT PULMONARY VEIN, EXTENDING INTO AND OCCUPYING
THE RIGHT ATRIAL CAVITY**

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the bronchus and pulmonary vein are composed almost exclusively of spindle cells, while
the round cells are more numerous in the soft, friable central portion. There is no line of
demarcation between the two types of cells. The spindle cells in some areas have a
tendency to be arranged in bundles, but the round cells show no attempt at any orderly
formation. Mitotic figures are common and the tumor is apparently growing quite
rapidly. Sections stained by silver impregnation show delicate reticulum fibers between
the cells. These fibers are most numerous in the areas of spindle cells. The spindle cells
are silver positive. Sections stained by Masson's trichrome method show that the cells
do not possess fibroglia fibers. These findings are consistent with a diagnosis of sarcoma.
occurring as a circumscribed tumor in elderly patients, usually of slow progression and without metastases; (2) round-cell sarcoma, a rapidly progressing tumor in youthful subjects, very cellular, composed of large or small round cells, prone to hemorrhage and necrosis but showing little tendency to metastasize.

The tumor described here is rather difficult to place in either of these classes, but presents points of resemblance to both types. The central portion of the tumor, composed mainly of round cells, showing marked necrosis and hemorrhage and apparently growing quite rapidly, resembles the malignant round-cell variety. The peripheral portions, however, composed of spindle cells with definite connective-tissue fibrils and a tendency to be arranged in bundles, definitely suggest the spindle or fibrosarcoma type. Histologically the two types of cells are quite similar and have undoubtedly taken their origin from a common progenitor. The variation in the cells is due to stages in the differentiation of the original cell type, the spindle cell being the more mature form. This interpretation suggests that the morphological classification represents only a superficial difference in the various sarcomas of the lung, and that primary pulmonary sarcoma, whether composed of round or spindle cells, arises from a common cell type, the variation in cells being due to stages in the differentiation of the primitive mesenchymal cell.

The differentiation between pulmonary carcinoma and pulmonary sarcoma is admittedly at times a difficult procedure. The presence of spindle cells in a tumor is not sufficient evidence to justify a diagnosis of sarcoma. Barnard (13) regards the so-called "oat-cell sarcoma" as a medullary carcinoma of the bronchus, and the type cell of this class
of tumor is the so-called oat-cell or a large or small spindle cell. Duguid (14), also, in an analysis of 78 cases, finds that the majority are of this type of bronchial carcinoma. Shennan (15) believes that spindle cells are present in some areas in most carcinomas and discusses the differential diagnosis. The gross and microscopic features in our case are so typical that no question should arise regarding the diagnosis of the tumor.

It is impossible in this case to trace the primary origin of the tumor, but the recent work on the origin of the alveolar epithelium appears to afford a plausible source for this type of malignancy. It was formerly considered that the pulmonary alveoli were lined by a definite epithelial covering, derived from the bronchial mucosa, which is entodermal in origin. Fried (16, 17, 18) in 1927, in a study of the origin of histiocytes in the lung, concluded that these cells arise from the alveolar lining which is not epithelial but mesenchymal in origin and belongs to the reticulo-endothelial system. Rose (19) concludes that the lung is of dualistic origin, the bronchi originating from entoderm invading a layer of mesoderm which forms the alveoli. He finds no evidence of an extension of bronchial mucosa into the alveoli and regards the septal cells as mesodermal in origin. Gardner and Smith (20) and Foot (21) also regard the septal cells as belonging to the reticulo-endothelial system. This conception of the mesodermal origin of the septal cells makes it easier to understand the production of sarcoma from the pulmonary alveoli and suggests a possible source for the tumor presented.

The extension of the tumor along established channels, bronchus and pulmonary vein, without gross involvement of these structures, is a striking feature in this case. However, invasion of the pulmonary vein and growth along this structure into the cardiac chambers have been described for primary carcinoma of the lung. Adler (8), in an analysis of 374 primary carcinomas of the lung, records this type of invasion in one case. Mead (1) recently reported a similar case of cardiac invasion by way of the pulmonary vein and calculated that this type of extension occurs in 0.26 per cent of cases of primary carcinoma of the lung. The present case appears to be the first instance in which this type of extension has been recorded for a primary sarcoma of the lung. It is interesting to note, however, that Müller (22) describes a somewhat similar condition in a fibrillated sarcoma of the left auricle, one portion of which extended along the pulmonary vein, and was directly continuous with a sharply circumscribed tumor nodule in the middle lobe of the right lung. The comparative scarcity and insignificance of other metastases in the present case agrees with the majority of recorded cases, in which metastases are not prominent, although Fishberg (23) states that metastasis in pulmonary sarcoma usually occurs early.

**Summary**

A case of primary sarcoma of the lung with unusual extension to the bronchus and to the heart by way of the pulmonary vein has been recorded.
It is suggested that sarcomas of the lung arise from a common source and that later differentiation of the cells determines the classification. A possible source of pulmonary sarcoma is discussed.

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