

MALIGNANT TERATOID TUMOUR OF THE LUNG: ? MALIGNANT HAMARTOMA

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In a previous paper (Jensen and Schiødt, 1958) describing the conditions of growth of 22 lung hamartomas we mentioned that in one case a malignant tumour had been ruled out, because the evidence of it having been developed from a hamartoma was doubtful. This case will now be reported and discussed.

CASE REPORT

A man, aged 66, had attended regularly for radiographic control of the lungs since 1944. No abnormality had been detected. About the middle of November, 1950, a walnut-sized, well-defined shadow was observed in the left third intercostal space (Fig. 1). The patient neither coughed nor expectorated. Weight 104.5 kg., height 187 cm., B.S.R. 5 mm./hour. Bronchoscopy and bronchography showed no abnormality in the bronchial tree. A radiograph taken just over one month later (Fig. 2) showed that the infiltration had grown about 1 cm. both across and lengthwise. The patient's general condition had remained good. There was no loss of weight.

Cancer being suspected, thoracotomy was done on January 19, 1951. The left lung surface was found to be free. A somewhat oblong, hard tumour was palpated centrally in the upper lobe. The superior segments of the upper lobe were resected, and a soft lymph node in the hilum, as well as a similar node at the origin of the segmental bronchus, were also removed.

On out-patient control two months later the patient was feeling well. Radiological examination revealed a fluid level above the diaphragm, but no other abnormality. Radiographs taken on May 18 showed unchanged conditions. On August 10 the patient looked sallow and was feverish. He had lost weight, and x-ray examination disclosed a massive blurring of the lower field of the left lung. On August 22 the outlines of more than one tumour mass were apparent. There were no signs of metastases.

On August 30 explorative thoracotomy was done. A pleural exudate and a closely adhering lung were found. In the paravertebral sulcus, on a level with the third and fourth ribs, necrosing tumour tissue was

encountered; this extended so far into the thoracic wall and towards the vertebral column that radical operation was out of the question.

On November 3, 1951, one year after the discovery of the tumour, the patient died. Necropsy was prohibited by his relatives.

Pathological examination of a preparation from the first operation showed a $3 \times 10 \times 15$ cm. large piece of lung tissue containing, close to the line of resection, a $1 \times 1.5 \times 3$ cm. large, well-defined mass. The anterior segmental bronchus was occluded and seemed to disappear into the tumour; but it was easily shelled out from the lung tissue, having no demonstrable relation to the bronchus. The surface and the cut surface were lobulated, the consistency was firm, elastic, and cartilaginous. The remaining lung and bronchi presented no changes.

On microscopy the tumour tissue presented a polymorphous histological picture. Most of the tissue had a mesenchymal character, with gradual transitions between loose connective tissue, myxoedematous tissue, and rounded areas of a character resembling foetal cartilage, showing in several places pronounced nuclear polymorphism (Fig. 3). "Mature" cartilage was not demonstrated. Here and there the connective tissue had proliferated, having a sarcomatous character. There was lymphocyte infiltration. In addition islands of squamous epithelium, here and there with marked corneous pearl formation, were found. The transition from epithelial tissue to stroma was in several places indistinct. Narrow clefts lined with a low stratified squamous epithelium were also found (Fig. 4); but not the ramified clefts lined with columnar epithelium characteristic of hamartomas. Parts of the tumour were rich in vessels, some of which were large and cavernous. There was no fat. The whole tumour was well defined, surrounded by compressed but otherwise normal lung. The tumour was nowhere seen to be in relation to bronchial structures. Lymph nodes displayed sinusoid reticulosis, but no signs of malignancy.

Histological examination of a biopsy specimen from the second operation on August 30 showed tumour tissue consisting of closely packed, dark, polymorphous, here and there spindle-shaped nuclei and

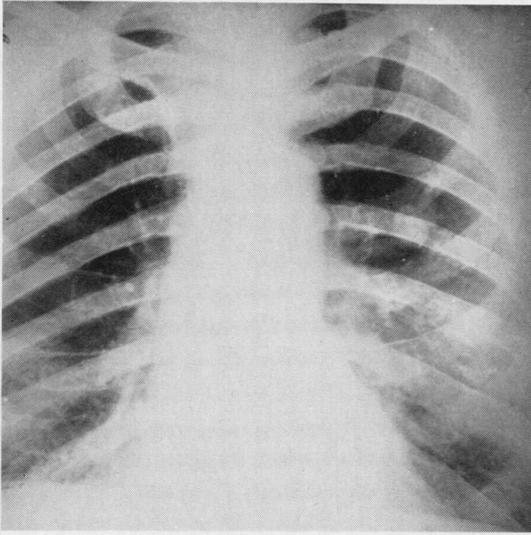


FIG. 1.—Radiograph of the thorax taken in the middle of November, 1950. A well-defined shadow is seen in the left third intercostal space.

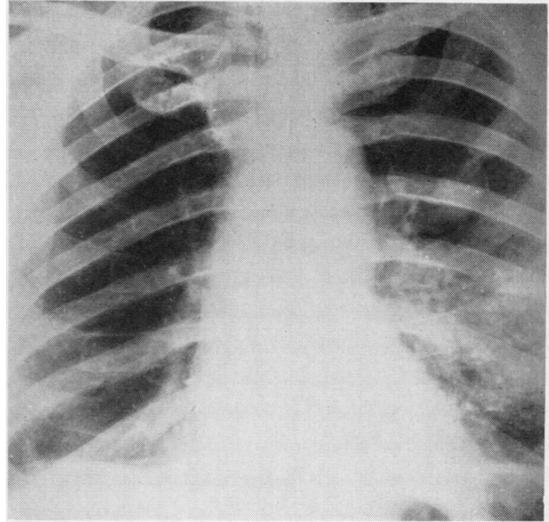


FIG. 2.—Radiograph of the thorax taken at the beginning of January, 1951. The shadow in the left third intercostal space is seen to have grown.

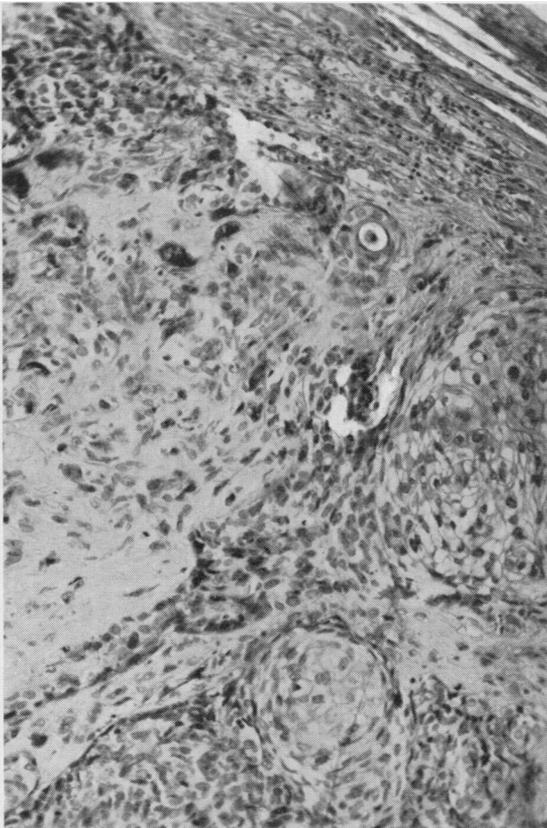


FIG. 3.—Photomicrograph (\times about 145) of a preparation from the first operation, showing solid tissue with nuclear polymorphism and indistinct transition to connective tissue, here and there reminiscent of foetal cartilage and in other places of epithelial formation. At the top right compressed pulmonary tissue is seen.

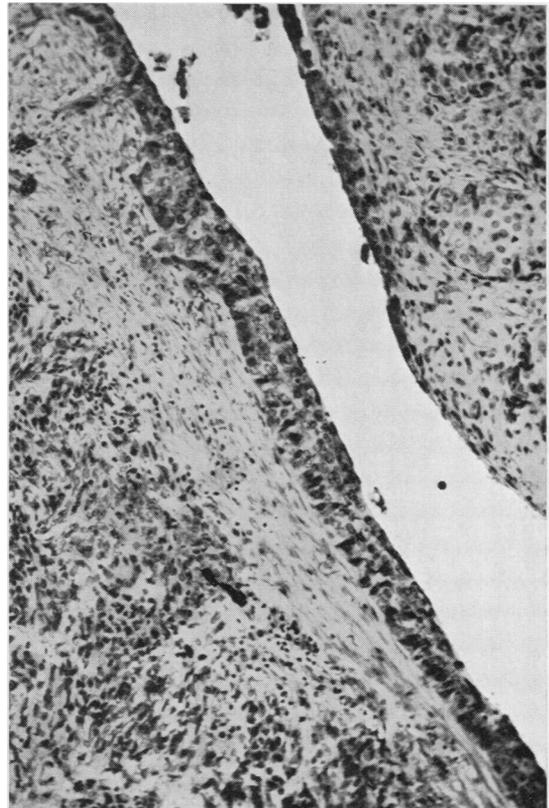


FIG. 4.—Photomicrograph (\times about 72) of a preparation from the first operation showing a long cleft bounded by epithelial-like tissue.

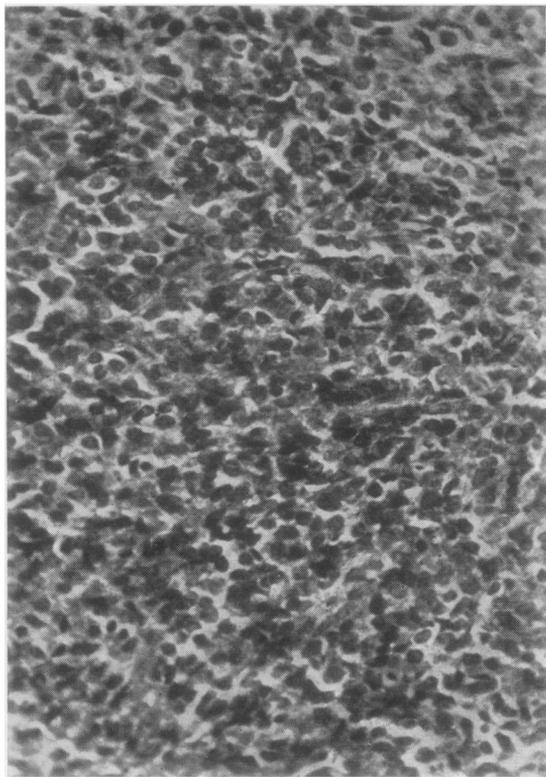


FIG. 5.—Photomicrograph (\times about 280) of a biopsy specimen from the second operation showing a polymorphous, sarcoma-like tumour rich in cells.

scant cytoplasm. There were no epithelial cells, nor was the stroma present in the tumour tissue (Fig. 5). The lesion was diagnosed as a malignant tumour, rather of the type of pure sarcoma.

DISCUSSION

A developmental malformation, including a hamartoma, may be the site of a malignant tumour. A well-known instance of this is the malignant teratoma of the testis. As for hamartoma of the lung, it is suggested in the literature that the chance of malignant transformation is extremely small. Few cases of malignant lung hamartoma have been reported, and none of these is quite certain.

Tapie (quoted by Hickey and Simpson, 1926) is stated to have published in 1892 a case of osteoma in the left lower lobe; the tumour was lying in close relation to an epithelial neoplasm. The patient, aged 28, also had multiple adenocarcinomatous infiltrations in both lungs. There seems to be no definite evidence of development from a hamartoma. Greenspan (1933) described a case of primary osteochondrosarcoma of the

lung, which is often termed "malignant hamartoma" in the literature. However, the author himself found the chondroblastic tissue in the bronchus itself to be the most likely site of origin; epithelial structures were not demonstrated. Lowell and Tuhy (1949) described a chondrosarcoma filling both branches of the pulmonary artery and continuing into lung. The authors found that the structure and the site were against a hamartoma as the point of origin. One of the most convincing cases of malignant hamartoma is that published by Simon and Ballon (1947). This was a histologically verified hamartoma of the lung, which here and there showed pronounced polymorphism of the mesenchymal tissue and invasion of mediastinal fatty tissue. The authors concluded that, "although, from a clinical point of view and in the absence of metastases, this tumour may be regarded as benign, it is apparent from an objective point of view that malignancy cannot be excluded." Kuyjer (1955), in a society report, described a case where a man, aged 18, had a benign, typical hamartoma of the lung removed. Six months later the patient was reoperated upon. The histological diagnosis was then spindle-cell sarcoma. The patient died three months later, and necropsy revealed round-cell sarcoma. The case seems convincing according to the description, but has not been substantiated by illustrations. Adams (1957) prefers the name of "chondro-adenoma" to that of hamartoma and designates Simon and Ballon's case as one of "ossifying adenoma." Adams himself reports a similar example, which is not convincingly malignant. Cavin, Masters, and Moody (1958) report a case of hamartoma with development of a leiomyosarcoma in a girl, aged 22 months. These authors cite a fatal case of "chondroma of the lung" described by Sherwood and Sherwood (1932). We have been unable to procure the original article. The difficulties of substantiation in the stated cases from the literature may be classified into two categories: (1) either an obvious hamartoma of the lung was present, the malignancy of which may be called in doubt (Simon and Ballon), or (2) the patient had an unquestionable malignant teratoid tumour, of which the development from a hamartoma was doubtful (Tapie, Greenspan, Lowell and Tuhy).

In the case described above our problem is in the latter category. At first operation a solid macroscopically cartilaginous, well-defined tumour was found in the parenchyma of the lung unrelated to the bronchus. Histologically the tumour consisted chiefly of mesenchymal structures, among which were large areas having the

appearance of foetal cartilage (Fig. 3). In addition, there were definite epithelial cells, here and there lining narrow clefts (Fig. 4). It was, in other words, a teratoid tumour, but composed exclusively of germ layers and constituents relating to pulmonary tissue. This accords with Albrecht's (1904) definition of a hamartoma, but the overall picture was not typical of such, columnar epithelium, fat, and mature cartilage being absent. On the other hand, cavernous vessels and, as is characteristic of hamartomas, an abrupt transition peripherally to compressed but otherwise normal pulmonary tissue was found. This somewhat unusual picture was, perhaps, due to the presence of unequivocally malignant features in the mesenchymal tissue already present in the first specimen. During the further course the tumour developed into a definite sarcoma (Fig. 5) as in Kuyjer's case.

In the case under review it was not proved, but seemed highly probable, that the tumour had developed from a hamartoma. Metastases from a malignant teratoma—in practice from a testis—can be left out of account. The testes were normal on clinical examination, and on the first operation the tumour was considered to be benign. Teratoid tumours in free lung parenchyma are rare (Liebow, 1952; Collier, Dowling, Plott, and Schneider, 1959), unlike in the mediastinum, where benign teratomas are relatively frequent. In our case the tumour had no connexion with apparently normal mediastinum. The fact that a

previous routine radiological examination of the patient's lungs had not revealed infiltration does not preclude a diagnosis of hamartoma (Jensen and Schiødt, 1958). The growth rate of the tumour reported here was considerably faster than has been observed for any one of 22 benign hamartomas of the lung.

SUMMARY

A case is reported of a teratoid, well-defined tumour with epithelial and in places malignant mesenchymal components, localized in free lung parenchyma. The patient was a man, aged 66. The possibility of a diagnosis of malignant hamartoma is discussed. The tumour recurred as a pure sarcoma after operation, and the patient died after one year.

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