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CASE REPORT

# Primary Haemangiopericytoma of The Lung

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*A 37-year-old woman underwent a left lower lobectomy for an undiagnosed tumour, which was reported on histopathology as a primary haemangiopericytoma. She subsequently succumbed to secondaries in the spine. A brief review of the literature shows that this potentially malignant tumour, arising from pericytes, rarely involves the lung and does not satisfy the usual microscopic criteria for malignancy.*

Haemangiopericytoma is a tumour thought to arise from the pericytes,[1] cells with protoplasmic processes contained within the alveolar capillary and the pulmonary vascular basement membrane. It occurs in many parts of the body, but the lung is one of the least common sites.[2] In the lung it may present as a small asymptomatic nodule, usually discovered on a routine chest radiograph or as a large symptomatic lesion. Its behaviour is often unpredictable, since distinction between benign and malignant forms is difficult or even impossible. This report describes a rare case of haemangiopericytoma of the lung.

A 37-year-old woman was admitted with the history of cough and haemoptysis of three months' duration. The results of physical examination and laboratory investigations were essentially within normal limits. Chest radiography revealed a well-defined shadow in the left lower lobe (Fig 1). The rest of the lung fields was clear without any suggestion of hilar or mediastinal adenopathy. These find-

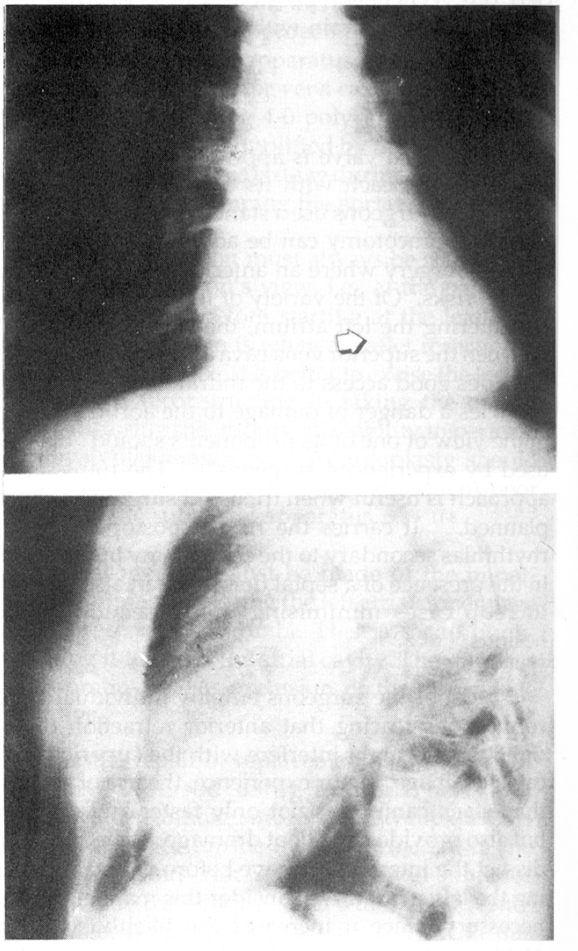

Fig 1. *Posteroanterior and lateral chest radiographs showing the round homogenous shadow in the left lower lobe.*

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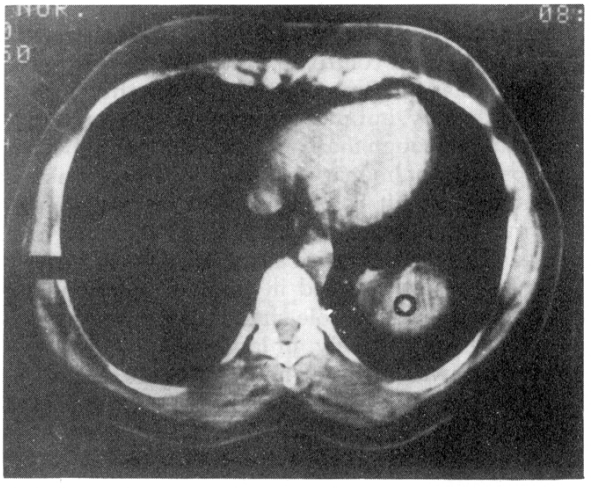

Fig 2. *Computed tomographic scan showing the tumour.*

ings were corroborated by a computed tomographic scan of the chest (fig 2) which also ruled out a cyst. Bronchoscopy was noncontributory. Fine needle aspiration for cytology was not attempted.

A left posterolateral thoracotomy revealed a tumour involving the basal segments of the left lower lobe. There was no involvement of the chest wall or significant lymphadenopathy. A left lower lobectomy was performed. On gross section, it was a basally located, 6 cm x 4.5 cm, well circumscribed, firm, greyish white tumour not arising from the bronchus.

Microscopic examination demonstrated round, oval and fusiform cells (Fig 3) with vesicular pleomorphic nuclei, some showing prominent nucleoli. Normal and abnormal mitotic activity could be seen. The cells were arranged in groups with focal trabecular pattern, intervening cleft like spaces and capillaries, mostly collapsed, some containing blood. Adjacent lung tissue showed oedema, collapse and chronic inflammatory cells. The overall histopathological appearance was suggestive of a malignant haemangiopericytoma of the lung.

The patient's postoperative recovery was uneventful. However, three months later, she complained of severe back pain and was found to have developed secondaries in the thoracic spine. She died soon thereafter.

## Discussion

Haemangiopericytoma has been described as a mesenchymal neoplasm of vascular origin characterised by numerous capillaries surrounded by tumour cells and a rich reticulum network. The entity was first described by Stout and Murray[1] in 1942. In their original description the authors postulated that the tumour is composed mainly of pericytes, a cell type first identified by Zimmerman[3] in 1923.

Although haemangiopericytoma may arise in the lung and mediastinum, it is usually extrathoracic with a definite musculoskeletal predilection.[3-6] The

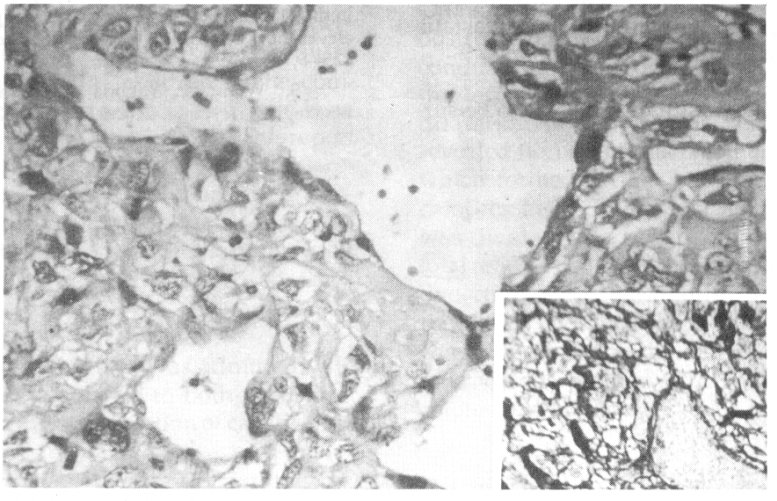

Fig 3. *Photomicrograph of the tumour showing multiple vascular spaces lined by flattened cells and intervening areas with plump, oval or spindle shaped cells and moderate pleomorphism (H&E x 400). Inset shows reticulin preparation (x 100) showing reticulin fibres radiating away from the vascular channel.*

tumour may appear at any site with capillaries, but most often affects soft tissues of the extremities and trunk. The most commonly affected areas are, in order of frequency, the extremities, retroperitoneum, head, neck and trunk. Pulmonary haemangiopericytoma is rare, but the clinical features do not differ from those of other lung tumours. It may present as a small asymptomatic nodule incidentally discovered on a routine chest radiography or as a large symptomatic lesion. Our patient presented with cough and haemoptysis.

We feel that the chances of making an intraoperative diagnosis based on frozen section may be remote in such cases unless the possibility of existence of haemangiopericytoma is kept in mind.

The tumour may appear at any age, but is usually seen in the fifth and sixth decades without any sex prediliction. It should be differentiated from other soft tissue tumours with abundant vascular channels such as extraskeletal mesenchymal chondrosarcoma, infantile fibrosarcoma, synovial sarcoma and malignant histiocytoma.[4] Most of these cases are probably aggressive and infiltrative with a strong tendency to metastasise.[5,6] Their symptomatology and prognosis depend on the duration of presentation. Conventional histological criteria for malignancy, such as mitotic activity, pleomorphism and cellularity are considered unreliable in haemangiopericytoma and often malignancy can be defined only from the tendency to recur or metastasise.[2] Its overall recurrence rate is about 50 per cent but that of pulmonary haemangio pericytomas is around 30-35 per cent with about 80 per cent appearing within the first year.[7] Metastatic spread can occur to the lung, liver and bone.

The only effective treatment appears to be surgical excision, although radiotherapy has been useful for local tumour control in some cases of recurrence or metastasis.[8] Opinions differ on the role of chemotherapy, but most attempts have been disappointing.[9] Though the experience with radiotherapy for primary pulmonary haemangiopericytoma has been minimal, it has been useful in palliation of pulmonary metastases and relief of superior vena caval obstruction. Its use as an adjuvant to surgery has been recommended in cases of recurrence; if symptoms in inoperable situations demand palliation; or preoperatively, in larger lesions involving the chest wall, where the diagnosis of haemangiopericytoma has been established.[5]

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