

Primary Hemangiopericytoma of the Lung

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SINCE its first description by Stout and Murray¹ in 1942, over 200 cases of hemangiopericytoma have been recorded. Although metastatic pulmonary spread is not uncommon, the tumour occurs only rarely as a primary growth within the lung.

Hemangiopericytoma may be considered a potentially malignant tumour derived from pericytes. These pericytes are modified smooth muscle cells with contractile properties and are capable of regulating the calibre of the capillary lumen. Theoretically the tumour may occur wherever capillaries are found.²

The hemangiopericytoma differs from the glomus tumour (also derived from pericytes) in that it lacks the organoid structure of the glomus and is composed of structurally different cells. The glomus, in addition, is completely encapsulated, often produces pain and is always benign, while the hemangiopericytoma is locally invasive, is painless, and frequently metastasizes.

Silver-staining techniques, in cases of hemangiopericytoma, demonstrate that the neoplastic cell (the pericyte) lies outside the capillary wall; this serves to distinguish the tumour from a hemangioendothelioma.

Although initially thought to occur primarily in superficial structures, especially the subcutaneous tissue and muscle of the extremities, Stout,³ in his review of 197 cases, demonstrated that it occurred with equal frequency in internal structures. Only three of a total of 99 "internal tumours", however, occurred in the "respiratory tract".

A 43-year-old housewife was referred to hospital in April 1959, for surgical correction of an esophageal hiatus hernia.

In addition to symptoms attributable to the hernia, she complained of recurrent episodes of left chest pain occurring every winter over the previous 15 years. The pain was pleuritic in nature and was situated posteriorly below the left scapula. She denied cough or sputum but had noted some dyspnea on exertion over the preceding six months.

On examination, there was percussion dullness and decreased air entry over the left lower chest. No adventitious sounds were heard. Physical examination was otherwise unremarkable.

A radiograph of her chest revealed that the left lower lung field was obscured by a homogeneous density extending up the lateral chest wall (Figs. 1 and 2). It was felt that this represented pleural fluid and/or pleural thickening, but an underlying parenchymal lesion could not be excluded. Radiographs taken six months previously were obtained for comparison and revealed a similar abnormality.

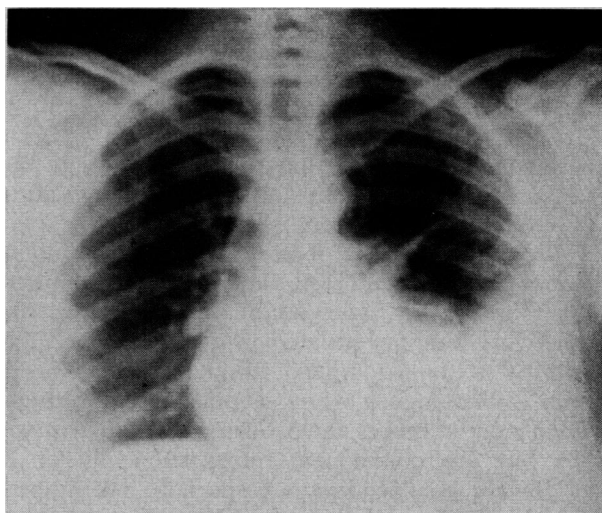


Fig. 1.—Admission chest radiograph in April 1959 showing homogeneous density in the left base suggestive of pleural fluid and/or thickening.

At thoracotomy on April 10, 1959, the left lower lobe was airless and completely replaced by tumour. The tumour was firm and blue-black in colour with multiple small cysts scattered over the surface. It was attached to the region of the hilus of the lung by a pedicle which contained blood vessels but no identifiable bronchus. The tumour was not unduly vascular and was excised without difficulty. The esophageal hiatus hernia was then repaired.

The gross specimen is illustrated in Fig. 3. Microscopically (Figs. 4, 5 and 6) the tumour was composed of multiple vascular spaces lined by flattened endothelium and separated by spindle-shaped cells. The amount of collagen varied from field to field. The cells had no malignant features. In no area was there evidence of any tissue which resembled lung parenchyma, bronchus or cartilage.

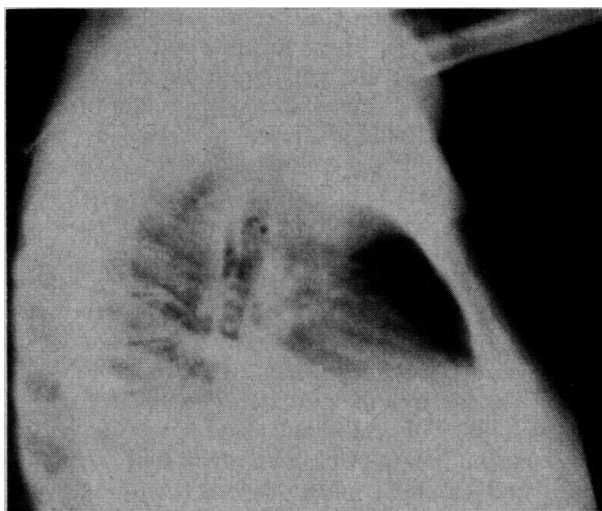


Fig. 2.—Lateral view again shows poorly defined basal lesion.

Her postoperative recovery was uneventful, and she was discharged from hospital two weeks after operation on April 25, 1959.

The patient was seen again three years later. At that time she was completely asymptomatic. Physical examination was unremarkable and failed to reveal any evidence of a peripherally situated primary tumour. A chest radiograph on April 26, 1962 (Fig. 7) revealed minimal pleural thickening at the left base with no evidence of recurrence of the tumour.

DISCUSSION

Hemangiopericytoma may occur at any age but is seen predominantly in the age group from 30 to 50, with an almost equal sex incidence.

According to Stout,³ it occurs with almost equal frequency in superficial and deep structures. Fisher,⁴ however, reviewed 20 cases, only three of which had tumours situated in deeper tissues. The majority of the superficial tumours (25%) occur in the subcutaneous tissue or muscle of the thigh. The deeper tumours have been reported in many sites but are commonest retroperitoneally or in the female genital organs, especially the uterus.

Primary hemangiopericytomas within the lung are rare. Stout³ described three in the "respiratory tract". His review no doubt included the two cases reported by McCormack and Gallivan⁵ in 1954. McCormack *et al.*,⁶ in 1961, reported an additional case of an asymptomatic primary in the right upper lobe discovered on routine chest radiography. No other reference to primary lesions in the lung was found in the literature.

The presenting symptom depends on the location of the tumour. Most superficial tumours present as painless lumps often present for many years. With the exception of the rare congenital variety, there is usually no redness of the overlying skin to suggest the vascular nature of the tumour. The deeply situated hemangiopericytoma may present as an asymptomatic mass or radiographic abnormality or may produce symptoms secondary to pressure on adjacent structures.

Whether the recurrent episodes of chest pain in the case described in this report may be attributed to the tumour is open to question. One might postulate a recurrent inflammatory process in a lobe which is the site of a slowly growing tumour, while the exertional dyspnea of relatively recent onset may well have been due to a reduced pulmonary reserve occurring secondary to a reduction in functioning lung tissue.

The radiological appearance of either primary or secondary hemangiopericytoma of the lung is not characteristic. Of the previously reported cases of lung primaries,^{5,6} one was described as a circumscribed nodule in the right upper lobe and another as an area of increased density in the right upper lobe. The radiological abnormality in the left base in our patient was attributed to pleural fluid and/or thickening. Needless to say, any opacity in the lung parenchyma occurring in a patient with a proven primary hemangiopericytoma

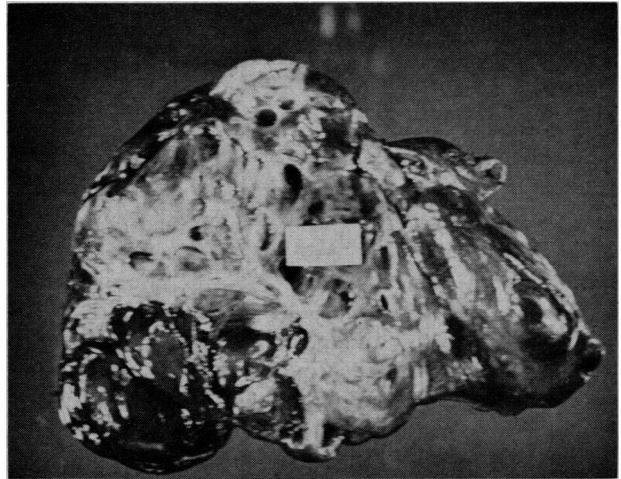


Fig. 3.—Gross appearance of the resected specimen. The tumour was blue-black in colour with multiple small cysts scattered over the surface. No pulmonary or bronchial tissue could be identified in the cut specimen.

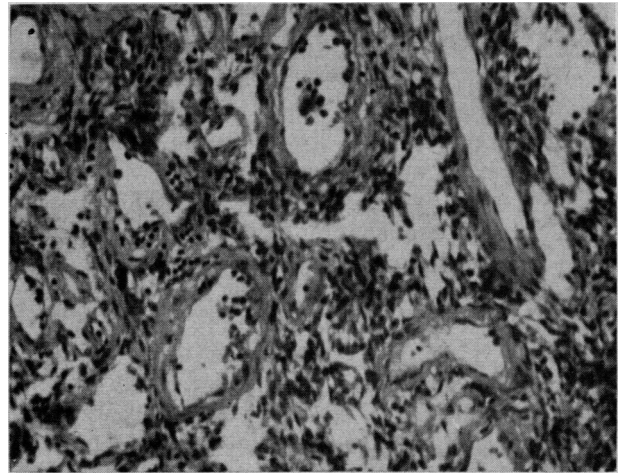


Fig. 4.—Low-power microscopic view of the resected specimen. The multiple vascular spaces are separated by sheets of spindle-shaped cells ($\times 200$).

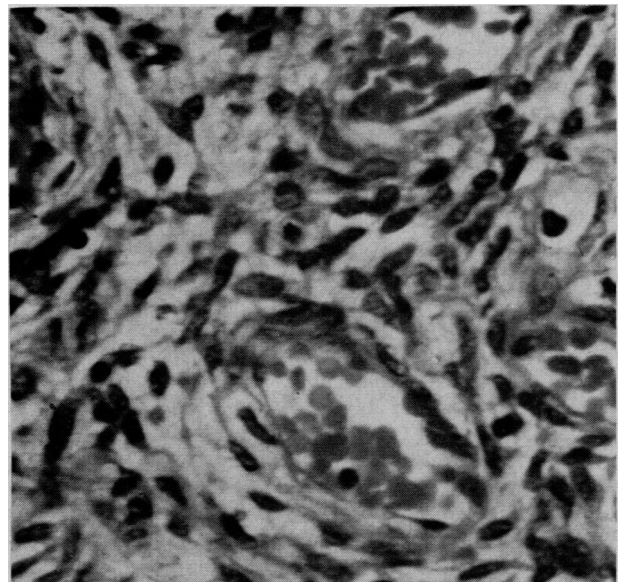


Fig. 5.—High-power microscopic view shows a capillary lined by normal endothelium and containing red blood cells, at the bottom of the photograph. The spindle-shaped pericytes may be recognized lying outside the capillary wall. These cells had no histologically malignant features ($\times 500$).

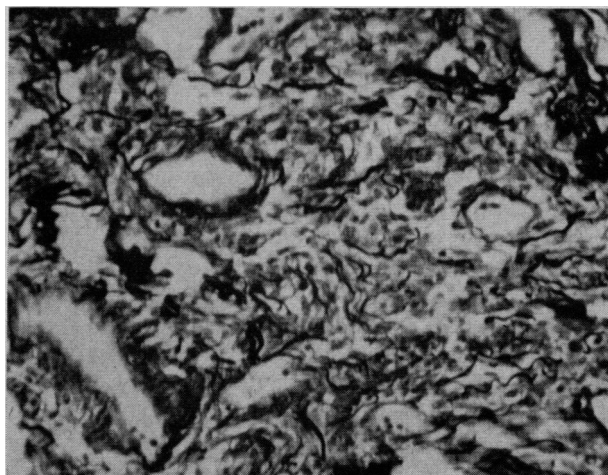


Fig. 6.—Silver-staining technique demonstrates that the tumour cells lie outside the capillary walls which are outlined in black. This serves to distinguish the tumour from a hemangioendothelioma ($\times 200$).

elsewhere should raise the suspicion of metastatic spread.

A detailed description of the pathological features and variants of this tumour is beyond the scope of this report. The main cell in each instance is the pericyte which lies outside the capillary wall. The amount of vascularity and fibrosis is variable. It should be borne in mind, as pointed out by Stout,³ that it is impossible to predict on purely histological grounds which tumours are malignant and may metastasize. Fisher⁴ described one patient with a primary tumour in the neck which, after initial excision, was interpreted as being histologically benign. This patient subsequently had three local recurrences and later died from widespread metastases.

Metastases occurred in 11.7% of Stout's cases³ and in 45% of Fisher's patients.⁴ Primary lesions occurring in the lower extremity, especially the thigh, are prone to metastatic spread. Fifty per cent of all metastases occurring in Stout's 197 patients arose from primary lesions in the leg. Of

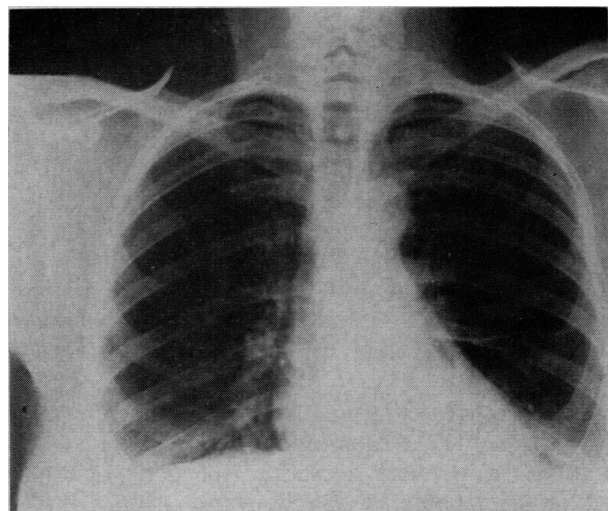


Fig. 7.—Follow-up chest radiograph three years after resection shows only minimal residual pleural thickening at the left base.

Fisher's nine patients with metastases, the tumour originated in the leg in five. The most frequent metastatic sites are lungs, bones, and regional lymph nodes.

The treatment of choice of peripheral tumours would seem to be wide surgical excision, regardless of the histological degree of malignancy. This does not apply to the superficially occurring congenital variety, which is invariably benign. The choice of wide excision is supported by Wise⁷ and Fisher.⁴ All of Fisher's cases that were histologically malignant and treated by simple excision had local recurrence. All of the cases that were histologically malignant had no metastases and were treated by amputation were cured.

The number of cases of primary lung hemangiopericytomas treated surgically is limited to three. All three were treated by lobectomy. Follow-up data are not available in one case while of the remaining two McCormack's patient⁵ was well one year after operation while our patient is alive and well three years after resection.

In spite of the great vascularity of the tumour in many instances, most workers have found that it is highly radioresistant. Mujahed, Vasilas and Evans,⁸ however, irradiated 10 patients, either after biopsy or after incomplete removal; they felt that nine of these were improved on the basis of reduced pain, decrease in tumour size or arrested growth.

CONCLUSIONS

Certain facts concerning this interesting tumour are worthy of note: The tumour occurs with equal frequency in superficial and deep structures. It is often malignant, and metastases have been reported in as many as 45% of patients. Primary hemangiopericytomas in the lower extremity should be managed with caution, as these are most likely to metastasize. The degree of histological malignancy is not a good criterion on which to base treatment. Wide excision seems the best form of treatment for peripherally situated tumours with the exception of the benign congenital form. The value of irradiation therapy has yet to be established but might be of benefit in cases of local recurrence or metastases. Lobectomy would seem to be a suitable procedure in cases of primary hemangiopericytomas within the lung.

SUMMARY

A successfully treated case of primary hemangiopericytoma of the lung is discussed. A brief review of the English literature on this disease is presented.

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