Haemangioleiomyomatous tumour of the lung

AJAIB S SOORAE¹* AND HOSHANG BHARUCHA²

From the ¹Thoracic Surgical Unit and ²Institute of Pathology, Queen's University of Belfast, Royal Victoria Hospital, Belfast BT12 6BA, Northern Ireland, UK

SUMMARY A case of haemangioleiomyomatous tumour of the lung, occurring as a peripheral, solitary nodule in an asymptomatic 54-year-old man is presented. The tumour was well-demarcated and microscopically it was characterised by the presence of vascular spaces with endothelial, pericytic, and, predominantly, smooth muscle proliferation. Islands of cartilage and slit-like spaces lined by bronchial epithelium make this a hamartomatous lesion of a quite distinctive and unusual variety, which does not fit any of the well-recognised patterns of hamartomas previously described. The long-term prognosis after limited excision is considered to be favourable.

Benign tumours of the lung, in comparison with bronchogenic carcinoma, are relatively rare. We report a case of haemangioleiomyomatous tumour of the lung, which is hamartomatous in nature and, to the best of our knowledge, has not been reported previously.

*Present address: Department of Thoracic Surgery, Fazakerley Hospital, Longmoor Lane, Liverpool 9, UK

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Case report

A 54-year-old man first presented to his doctor in March 1978 complaining of cough and a wheeze. He used to smoke 50 cigarettes a day but had given up smoking five years previously. He was treated with antibiotics and bronchodilators and made a complete symptomatic recovery. Subsequently, a routine chest radiograph showed a rounded opacity in the right upper lobe adjacent to the mediastinum (Fig. 1).

Fig. 1 Posteroanterior chest radiograph showing a rounded opacity in the right upper lobe.

Fig. 2 Anteroposterior tomogram showing an ovoid, homogenous opacity, with no calcification or cavitation, situated in the posterior part of the upper lobe.

At this stage the patient was referred to the thoracic surgical unit for further management.

On examination he was a healthy looking, thick-set man, not anaemic or dyspnoeic, with a pulse rate of 80 per minute and a BP of 120/85 mmHg. There was no lymphadenopathy or hepatosplenomegaly. There was good air entry over both lung

fields with normal breath sounds. The haematocrit and the rest of the blood chemistry were normal with an ESR of 5 mm in the first hour. Sputum cytology was negative for malignant cells. On bronchoscopy the bronchial tree was normal on both sides. Anteroposterior tomograms showed a well-defined, homogenous, ovoid opacity situated in the posterior part of the upper lobe. No calcification or cavitation was noted (Fig. 2).

At right thoracotomy in July 1978, a soft mass, 3 × 3 cm, was found in the posterior segment of the upper lobe. It was considered to be a benign lesion, and hence a wedge excision, with a good margin of healthy lung tissue around it, was carried out. Recovery was uneventful, and the patient was discharged on the 11th postoperative day. Five months later the patient remains well and asymptomatic.

PATHOLOGY

Gross appearance

The surgical specimen showed a well-circumscribed, fleshy, tan-coloured nodule, about 3 cm in diameter, with a central area of haemorrhage and surrounded by normal lung parenchyma. No obvious attachment to a recognisable bronchus was visualised, and the vasculature to this tumour was quite unremarkable.

Microscopic appearance

Routine histological preparations at 6 micron

Fig. 3 Foot's reticulin preparation shows endothelial and pericyte proliferation (Foot's reticulin × 400).

Fig. 4 *The vascular component is now reduced to slit-like spaces with a predominant spindle-cell component (Masson trichrome $\times 400$).*

thickness were stained with haematoxylin/eosin, Masson trichrome stain, Foot's reticulum method, Congo red, elastic Van Gieson, and the Picro-Sirius red technique.

The lesion consisted of vascular spaces devoid of elastin and lined by variable amounts of endothelial cells. Some of the vascular spaces were lined by a single layer of endothelial cells, while in other areas

Fig. 5 *Interweaving bundles of spindle-shaped cells with the characteristics of smooth muscle (Masson trichrome $\times 400$).*

Fig. 6 An epithelial-lined space resembling a bronchiole (Haematoxylin and eosin $\times 250$). Spaces like these were situated within the tumour in addition to several compressed normal bronchi found at the periphery of the tumour.

there was pericyte proliferation (Fig. 3). In addition, there were areas where the vascular component was reduced to very thin, slit-like channels, which were surrounded by a proliferation of spindle-shaped cells with tapered nuclei resembling smooth muscle cells (Fig. 4). The reticulum pattern was quite distinctive and revealed areas that resembled a haemangio-endothelioma. In other areas, the reticulum merely outlined thin, slit-like blood vessels. The Masson trichrome stain confirmed the presence of smooth muscle as the most predominant component in most areas of this neoplasm (Fig. 5). The picro-Sirius method revealed very little collagenous stroma within the lesion. Another interesting feature was the presence of epithelial-lined spaces, which were undoubtedly bronchial in origin (Fig. 6).

Discussion

Hamartoma is the most common benign pulmonary tumour.^{1,2} It has generally been considered a tumour-like malformation of developmental origin since the original description by Albrecht in 1904.³ However, there are others who believe it to be a true, benign neoplasm of the connective tissue of the bronchial walls.^{4,5}

Several types of pulmonary hamartomas have been described, the commonest being a small, fibrocartilaginous, solid mass discovered on a routine chest radiograph, usually in an asymptomatic adult patient.⁶ Fibroleiomyomatous hamartoma is much less common, is often multifocal in origin, and is seen in asymptomatic middle-aged women.^{7,8}

Pulmonary haemangiomas, which are rare, are also considered by many authors to be hamartomas.⁹ This tumour has several features that categorise it into a hamartoma but it does not fit into any of the reported descriptions.

There were no characteristic clinical, radiological, or gross features of this tumour. Like other benign tumours situated in the lung parenchyma, it was discovered on a routine chest radiograph as a well-circumscribed peripheral nodule. The symptoms of cough and a wheeze, with which the patient presented initially, were probably unrelated to the tumour.

Histologically, the tumour was characterised by the presence of endothelial, pericyte, and smooth muscle proliferation. There were focal areas where one or the other element predominated, and the presence of slit-like spaces covered by bronchial epithelium completed the picture of a hamartomatous lesion of bronchial origin. The presence of vascular spaces with proliferating smooth muscle makes this lesion quite distinctive, and we believe it has not previously been documented. The prognosis, as in other types of hamartoma, is considered to be favourable, and hence limited surgical excision is believed to be adequate.

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Requests for reprints to: Dr H Bharucha, Institute of Pathology, The Queen's University of Belfast, Grosvenor Road, Belfast BT12 6BL, Northern Ireland, UK.