

Uncommon pulmonary hamartomas

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Minasian, H. (1977). *Thorax*, 32, 360-364. **Uncommon pulmonary hamartomas.** Endobronchial and multiple pulmonary hamartomas are rare. Three cases are reported—one endobronchial hamartoma, one endobronchial hamartoma accompanied by a similar intrapulmonary lesion, and one multiple hamartoma. These cases typify the clinical presentation and illustrate some of the difficulties in diagnosis and management. The published cases are reviewed.

Although pulmonary hamartomas are not uncommon, those which are intrabronchial or multiple are rare.

Of the 3450 patients subjected to thoracotomy for lung neoplasms at The London Hospital between 1950 and 1976, 33 (0.96%) were found to have pulmonary hamartomas. Two hamartomas were endobronchial, one being accompanied by a similar small peripheral tumour, and one was multiple.

Case 1

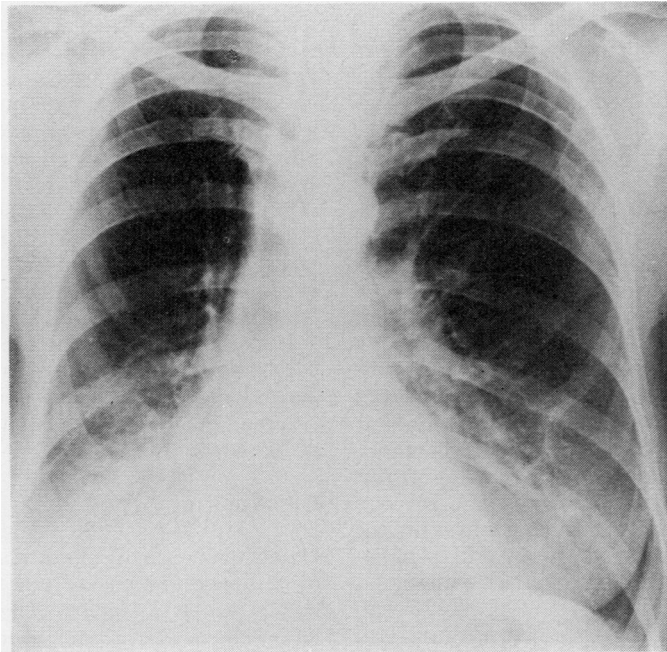
A 62-year-old man was first seen in 1959 with a dry cough, right upper chest pain, and pyrexia. The chest radiograph showed abnormal shadowing in the right upper lobe. Pneumonia was diagnosed, and he was treated with a course of antibiotics which cured his symptoms and pyrexia, but the shadowing on the chest radiograph failed to resolve completely. At bronchoscopy a small nodule was seen on the lower margin of the right upper lobe bronchial orifice. A biopsy showed adipose and myxomatous tissue, suggesting a hamartoma. A few days later a right thoracotomy was performed. There were no visible or palpable masses or enlarged lymph nodes. The posterior aspect of the right main bronchus was exposed and a longitudinal bronchotomy made, displaying a sessile nodule, 4 mm in diameter, which was arising from the wall of the right main bronchus just below the upper lobe bronchial orifice. The nodule was excised and the bronchotomy closed with 3/0 interrupted silk sutures. The patient's recovery was uneventful.

On histological examination the nodule was found to consist mainly of adipose tissue, but it

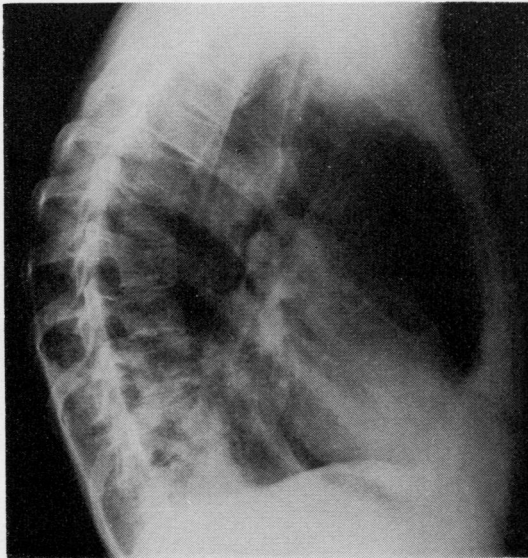
also contained cartilage and bronchial mucous glands.

Case 2

A 55-year-old woman was seen in May 1976. A chronic bronchitic for the past six years, she had developed another chest infection which was more severe than usual and failed to clear with antibiotics. There was no chest pain or haemoptysis. The chest radiographs showed collapse of the posterior basal segment of the right lower lobe with no other discrete shadowing (Fig. 1). Screening of the diaphragm showed normal movements. Four examinations of the sputum failed to reveal malignant cells. Pulmonary function tests were suggestive of slight airways obstruction. On bronchoscopy under local anaesthesia the right intermediate bronchus was seen to be obstructed by growth. A biopsy showed bronchial mucosa with areas of squamous metaplasia and underlying cartilage. At a second bronchoscopy a pedunculated growth was observed extending up to the level of the right upper lobe bronchial orifice and occluding the main bronchus. The gross appearance was that of a carcinoma. During an exploratory thoracotomy a hard mass was palpated in the main bronchus. In view of the clinical picture and the findings at bronchoscopy and at operation, a diagnosis of bronchial carcinoma was made despite the histological report. A right pneumonectomy was carried out including removal of the hilar, subcarinal, and paratracheal lymph nodes. The postoperative course was complicated by infection in the remaining left lung and respiratory failure. The patient's condition gradually deteriorated and she died 11 days after the operation.



(a)



(b)

Fig. 1 (a and b) Case 1. Postero-anterior and right lateral chest radiographs showing collapse of the posterior basal segment of the right lower lobe.

The pneumonectomy specimen revealed a benign endobronchial hamartoma containing cartilage, epithelial elements, and blood vessels. A small histologically similar hamartoma, 2 cm in diameter, was also present at the periphery of the lower lobe. The lymph nodes were normal.

Case 3

A woman of 43 was first seen in May 1967 because of an abnormal chest film taken during a mass radiography session. She was asymptomatic at this time and the only past medical history of note was a hysterectomy for fibroids in 1955. The chest radiograph had shown three obvious well-circumscribed opacities in the right lung, the largest of which was in the lower lobe and was 2 cm in diameter. Tomograms did not show any calcification or cavitation. A provisional diagnosis of multiple secondary deposits was made, no further investigations were carried out, and she was kept under regular review. In subsequent years she remained asymptomatic, and serial chest radiographs showed little change in the pulmonary shadows. She was seen again in May 1972, still asymptomatic, but on this occasion there was a suggestion of a slight increase in size of the largest opacity in the right lower lobe. The radiographs also showed a new opacity in the right lung (Fig.

2) and possibly another in the left lung. Her white cell count was 4300 with 43% eosinophils (although this was not borne out in subsequent counts) and the ESR was 4 mm in one hour. The diagnosis considered at this time were atypical eosinophilic granuloma, multiple tuberculoma, and multiple hamartoma, and she was referred to our department. Further investigations were inconclusive and so a thoracotomy was performed. A number of fleshy nodules, one of which was excised, were palpable in the middle and lower lobes. There was no other intrathoracic abnormality and she made an uninterrupted recovery from the operation.

Histology showed that the nodule consisted of bands of smooth muscle, small amounts of collagen, and small cysts and tubules lined by columnar epithelium, consistent with a diagnosis of hamartoma.

Discussion

Hamartoma is a localised malformation made up of cell types which are not foreign to the organ within which it occurs. Those occurring in the lung behave like a benign neoplasm in that they may continue to grow even when body growth ceases.

In 1965, Bateson, reviewing the world literature,

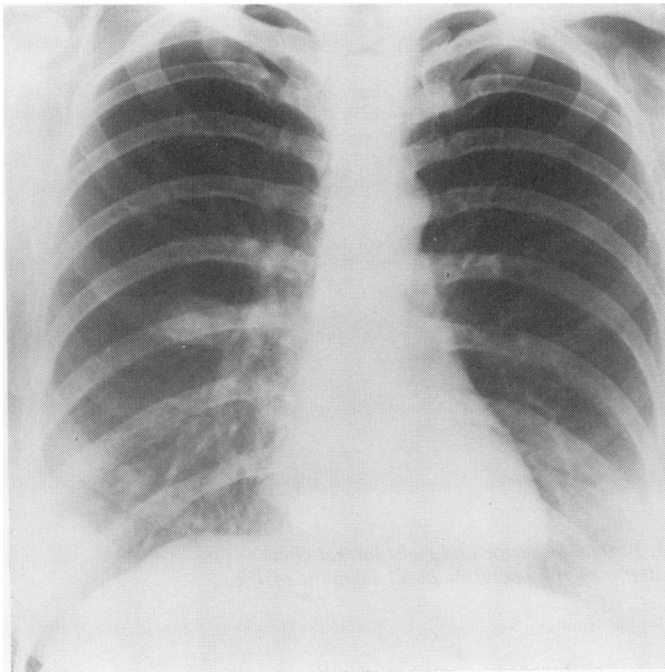


Fig. 2 Case 2. Postero-anterior chest radiograph showing multiple hamartomas in the right lung.

found 457 reported cases of hamartoma, 89 of which were endobronchial. Up to 1964 only 51 cases of endobronchial hamartoma had been reported in the English literature (Stengel *et al.*, 1964). More recent reports of cases include those by Zeidler and Vogt-Moykopf (1971), Sibala (1972), Pirozynski and Schwarz (1973), and Shah *et al.* (1973). There appear to be only 17 previously reported cases of multiple pulmonary hamartoma (Sargent *et al.*, 1970; Sulser and Bühler, 1975; Becker *et al.*, 1976).

The incidence of pulmonary hamartoma among the adult population is given by McDonald *et al.* (1945) as 0.25%, representing 23 cases in nearly 8000 postmortem examinations. le Roux (1964) reported that in Edinburgh during the period 1950–59 only 27 patients with pulmonary chondromatous hamartomas were seen. Two of these were endobronchial. During the same period 3000 cases of lung carcinoma were seen. These figures are comparable with those in our department.

Bateson (1965) showed that the median age for presentation of an endobronchial hamartoma is between 50 and 60. The youngest case on record is that of a 16-year-old boy (Perry, 1959).

The male to female ratio is given by Bateson (1965) as 5:1 and by McDonald *et al.* (1945) as 3:1. The multiple variety, however, seems to occur exclusively in females, and at a mean age of 46 (range 20–74) years.

In published cases of endobronchial hamartoma, the most frequent clinical features were cough, dyspnoea, haemoptysis, and pyrexia. Almost every case appears to have some degree of bronchial obstruction with distal pulmonary sepsis and/or atelectasis. The chest radiograph often shows the latter changes but fails to reveal the primary lesion. The clinical and radiological features may be indistinguishable from those of a carcinoma. Bronchoscopy invariably reveals the abnormality, but it is difficult or impossible to distinguish it from a carcinoma from its gross appearances. Histology of the biopsy material shows no evidence of malignancy, but, in view of the macroscopic appearances and the relative rarity of benign lung neoplasms, the surgeon may easily assume a technical biopsy error and regard the condition as malignant. Preoperative diagnosis may therefore present a difficult problem.

The multiple pulmonary hamartoma does not usually cause symptoms or signs, the only positive findings being on the chest radiograph. The case reported here shows most of the characteristics of this condition and demonstrates the diagnostic dilemmas and difficulties.

Microscopically the commonest constituent of

endobronchial hamartoma is cartilage, followed in frequency by loose connective tissue, fat, bone, bone marrow, smooth muscle, lymphoid cells and epithelial cells, and epithelial cell clefts respectively (Bateson, 1965). The multiple variety characteristically lacks cartilage and consists mainly of smooth muscle—hence the name 'leiomyomatous hamartoma'. Benign uterine hamartomas often coexist. One previous case had an associated cutaneous leiomyoma (Massachusetts General Hospital, 1963) and another had a benign gastric leiomyoma (Muller *et al.*, 1974).

Zeidler and Vogt-Moykopf (1971) demonstrated that endobronchial hamartomas are fairly evenly distributed between the two sides, 54.5% on the left and 45.5% on the right. The commonest site is the left lower lobe bronchus. The multiple type is usually bilateral.

If a firm diagnosis of endobronchial hamartoma can be reached before operation, then, provided the lesion is favourably sited, bronchotomy and excision is the treatment of choice. In the absence of a preoperative diagnosis and in a lesion sited distally, lung resection is necessary. Often the lung distal to the obstructing hamartoma is atelectatic and irreversibly damaged. The multiple hamartoma requires no treatment.

The second case reported here is particularly interesting because there was a second intrapulmonary peripheral hamartoma, histologically similar to the endobronchial hamartoma. This would seem to support the view of Bateson (1970) that the intrapulmonary and endobronchial chondromatous hamartomas are similar, the only difference being the site of their origin and the direction of their growth.

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