Intrapulmonary thymoma: report of two cases

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Intrapulmonary thymoma is a rarity—only nine cases have been reported. We describe here two cases of intrapulmonary thymoma diagnosed at necropsy and review earlier reported experience.1-4

Case reports

CASE 1
A 46 year old Chinese man presented in July 1983 with intermittent haemoptysis and progressive exertional dyspnoea of six months’ duration. Investigation at another hospital showed consolidation collapse of the left lung. Sputum examination was negative for malignant cells. Fibreoptic bronchoscopy showed narrowing of the left main bronchus due to extrinsic compression; a blind bronchial biopsy showed no evidence of malignancy. An exploratory thoracotomy was planned but no compatible blood was available for transfusion because the patient was found to have positive results in direct and indirect Coombs’ tests. The bronchial obstruction and breathlessness became worse. He was given radiotherapy on the assumption that he had some form of intrathoracic malignancy and was referred to the Queen Mary Hospital for further management.

The patient looked ill. There was no clinical evidence of myasthenia gravis. He had a haemoglobin concentration of 8.2 g/dl with a reticulocyte count of 1.4% and normochromic, normocytic red blood cells. The leucocyte count was 8.2 × 10³/l, and the erythrocyte sedimentation rate was 144 mm in the first hour. The total bilirubin level was 9 μmol/l (0.53 mg/100 ml). There was diffuse gammapathy with an IgG concentration of 4800 mg/dl; antinuclear factor and rheumatoid factor tests gave negative results; serum complement levels were normal. A chest radiograph showed consolidation in the right lung, left pleural effusion, and peculiar curvilinear calcification in the left lung (fig 1a). Transbronchial biopsy of the left lung was carried out through the fibreoptic bronchoscope, but the specimen showed only changes from irradiation.

Despite antimicrobial treatment, the patient died soon after admission to our hospital from bronchopneumonia.

Necropsy findings

In the left lung there were multiple well encapsulated tumour masses. The largest mass was in the upper lobe and measured 12 × 10 cm. It was situated subpleurally and was completely inside the lung. This tumour caused appreciably compression and displacement of the left upper lobe bronchus. The capsule of the tumour was calcified and fibrotic. The tumour was variegated in colour, with soft, fleshy, pinkish areas among haemorrhagic, cystic, and whitish fibrotic areas. Smaller tumour masses, ranging from 2 to 5 cm in diameter, with similar appearances were found in other parts of the lung, mainly in the subpleural region. They were similarly encapsulated (fig 1b).

The right lung showed extensive suppurative consolidation. There was no tumour. Hilar lymph nodes were normal. The involved thymus was seen in its normal anatomical position. It had a normal appearance and was free from tumour. The other organs in the body were unremarkable.

All tumour nodules showed similar histological appearances. They showed many of the typical features of a predominantly epithelial thymoma: branching fibrous septa, hyalinised vessels, perivascular spaces, microcyts, and in many areas a pseudohaemangiopericytoma arrangement of tumour cells (fig 1c). There was no capsular invasion or cytological evidence of malignancy.

CASE 2
A 60 year old Chinese woman was admitted in July 1983 with a history of weakness of neck muscles, dysarthria, and dysphagia of one month’s duration. She also had cough with yellowish sputum and fever. She was ill and febrile on admission.

Examination of the central nervous system showed slurring of speech, drooping of the left eyelid, and weakness of the left lateral rectus muscle and neck muscles. She was mildly dysnpeic with a respiratory rate of 20/min. Crepitations were heard all over the chest on auscultation.

Investigations showed a haemoglobin concentration of 15.1 g/dl and a leucocyte count of 14.5 × 10³/l with 75% polynormorphs. A chest radiograph showed a rounded opacity at the right hilum (fig 2a).

A presumptive clinical diagnosis of carcinoma of the lung with Eaton-Lambert syndrome was made. Treatment with pyridostigmine 60 mg and prednison 10 mg, however, alleviated the muscular weakness. Despite antimicrobial treatment, she succumbed to bronchopneumonia one day after admission, before bronchoscopic examination could be performed.

Necropsy findings

At necropsy a lobulated fleshy tumour, 4 cm in diameter, was found at the hilum of the right lung. It was well encapsulated and was almost completely within the lung parenchyma. It was not attached to any large bronchus, and its medial surface was flush with the surface of the lung. The cut surface was tan coloured with areas of haemorrhage (fig 2b). The rest of the lung was consolidated. No tumour was found in the left lung, which also showed pneumonic...
consolidation. Hilar lymph nodes were not enlarged. There was no tumour in the thymus, which was identified in its normal position in the mediastinum. The other organs were normal.

Microscopically, the tumour was a lymphocyte predominant thymoma (fig 2c). The fibrous capsule completely separated the tumour from the lung parenchyma, and was continuous with the visceral pleura.

Discussion

Primary intrapulmonary thymoma is a rare tumour. Worldwide only nine cases have been published. We have reviewed the features of the seven cases reported in English and German\(^1\)–\(^5\) (two further cases have been reported, in Polish and Russian,\(^6\) but are not considered here). Three patients were male and four female, ranging in age from 14 to 71 years.

Kalish conveniently grouped intrapulmonary thymomas into peripheral and hilar types according to their proximity to the hilar region.\(^5\) All five peripheral intrapulmonary thymomas reported were found in the right lung.\(^4\)–\(^6\) The tumours were relatively small, solitary, and resectable, the largest being 6.5 cm in the greatest diameter.\(^*\) Only one of the five patients had symptoms, presenting with myasthenia gravis and finally dying from this even though the

Fig 1  Case 1: (a) Chest radiograph after radiotherapy showing curvilinear calcification (arrow) in the left lung and left pleural effusion and consolidation in the right lung. (b) Necropsy specimen of the left lung showing multiple tumour masses encapsulated by calcified fibrous tissue. The largest is seen in the upper lobe; smaller ones are seen subpleurally in the lower lobe and the apex of the upper lobe. (c) Photomicrographic appearances of a typical predominantly epithelial thymoma displaying a pseudohaemangio-pericytoma pattern, microcysts, and perivascular spaces. (Haematoxylin and eosin, × 300.)
tumour had been excised.\textsuperscript{1} The other patients remained well after surgery\textsuperscript{4-6} or died from unrelated causes.\textsuperscript{4}

Case 1 in the present report belongs to the peripheral group. The left rather than the right lung was affected, and this is the first reported case of multiple intrapulmonary thymoma. The largest mass, measuring 12 cm in its greatest diameter, was also the largest tumour of its kind reported so far. We consider the smaller tumour nodules in the present case to be multiple primaries rather than metastases because they were well encapsulated, there was no evidence of lymphatic spread or local infiltration, and the microscopic appearance of the tumour was that of a benign thymoma rather than a thymic carcinoma.\textsuperscript{5} Another peculiar feature of the case is the haematological picture. It is unlike the red cell aplasia occasionally associated with thymoma.\textsuperscript{10} Unfortunately a full haematological investigation was not performed before the death of the patient. Possibly the positive Coombs test results are related to the thymoma.

The tumour in case 2 belongs to the hilar group. It differs from the two previously reported in that it was situated in the hilum of the right lung rather than the left\textsuperscript{2,3} and it was associated with myasthenia gravis, which caused the death of the patient. The previous two patients survived after surgical excision of the tumour. Tumour size is similar in all three cases.

It is now evident, perhaps not surprisingly, that both the peripheral and the hilar types of intrapulmonary thymoma may occur in either the left or the right lung. The tumour may be asymptomatic or associated with myasthenia gravis; it may be multiple or solitary; and it affects both sexes in a wide age range.

All cases were diagnosed after pathological examination following surgical excision or at necropsy. Although few cases have been studied so far, helpful clinical diagnostic
clues are: (a) longstanding intrapulmonary chest radiographic shadows, particularly when there is a curvilinear shadow suggesting the calcified capsule of a tumour; (b) the presence of myasthenia gravis; and (c) absence of histological or cytological evidence of carcinoma. The prognosis so far appears good in symptom free patients, but poor in those with myasthenia gravis or with large tumours.

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Reference