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Bronchial mucoepidermoid tumour in a child presenting with organomegaly due to secondary amyloidosis: case report and review of the literature

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Abstract Childhood bronchial mucoepidermoid tumours (BMET) are rare. A 12-year-old boy with hepatosplenomegaly underwent liver biopsy which diagnosed amyloidosis. Chest radiograph and CT, performed for recurrent respiratory symptoms, identified a left lower lobe tumour, which was subsequently excised. Histology showed a BMET. A literature review reveals 51 reported cases of BMET in children. Common presenting symptoms include fever, cough and recurrent pneumonia. Diagnosis is often delayed and patients with recurrent respiratory symptoms should undergo CT or bronchoscopy. The association between amyloidosis and BMET in this case is unique and has not been previously described, but may be coincidental.

Introduction

Primary pulmonary neoplasms are rare in children and are usually reported as isolated cases. The most commonly encountered entities include bronchogenic carcinoma, plasma cell granuloma and bronchial adenoma. In the latter group, bronchial mucoepidermoid tumour (BMET) is a rare variant, occurring in 2.5–7.3 % of adenomas and comprises 0.1–0.2 % of primary lung cancers [1, 2]. Usually occurring as a polypoid endobronchial mass within the proximal airways, it presents with features related to bronchial irritation and obstruction such as fever, cough, haemoptysis or recurrent pneumonia [1, 2]. The low malignant potential of childhood BMET renders them amenable to conservative surgical resection. We report a unique case of a BMET in a child previously diagnosed with secondary amyloidosis. While there may be a causative link between these entities, this association has not been previously described and may be coincidental.

Case report

A 12-year-old boy was referred with haematemesis/haemoptysis, epistaxis and a 4-year history of hepatosplenomegaly. Review of the medical records revealed that the patient presented 4 years previously with cough, weight loss and abdominal swelling. There had been no significant respiratory symptoms, but he had generalised lymphadenopathy, finger clubbing and hepatosplenomegaly for which he was extensively investigated. Chest X-ray demonstrated left hilar opacification. Laboratory investigations showed a leucocytosis, microcytic anaemia, raised ESR and positive antibodies to core and surface antigens of hepatitis B. Liver biopsy diagnosed amyloidosis and the patient was treated for presumed unrelated left lower lobe pneumonia.

In the ensuing 4 years repeated respiratory symptoms, in particular, cough and occasional haemoptysis, precipitated numerous hospital admissions. On this admission, the patient had digital clubbing, generalised lymphadenopathy and hepatosplenomegaly. There were clinical stigmata of hepatic decompensation. Respiratory examination revealed a pleural rub with dullness to percussion, coarse crackles and decreased breath sounds over the left lower lobe. Liver biopsy confirmed amyloidosis. The CXR showed an ill-defined, left lower lobe retrocardiac mass (Fig. 1). A CT scan, performed because of the nonspecific roentgenographic features in a patient with recurrent respiratory symptoms with digital clubbing suspicious of bronchiectasis, revealed a partly calcified mass with peripheral pockets of air, suggesting that the mass was within a cavity (Fig. 2). A bronchus was seen leading to the mass, but there was no bronchiectasis (Fig. 3). Surrounding passive atelectasis was present and hilar nodes were enlarged. The mass was excised. Macroscopically it was endobronchial and could be easily peeled away from the smooth wall of a dilated bronchus. Histology diagnosed BMET without nodal metastases. The patient recovered well post-operatively.

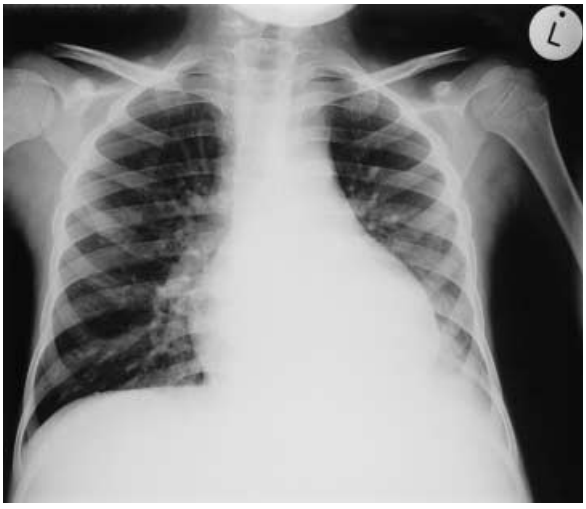

Fig. 1 CXR shows an ill-defined, left lower lobe, retrocardiac mass

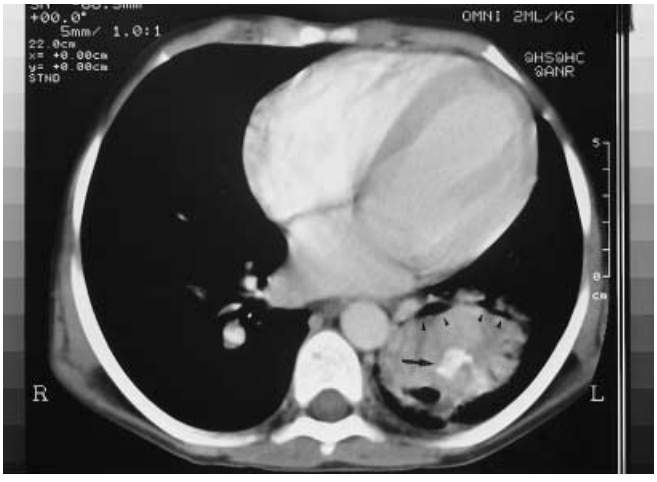

Fig. 2 Axial contrast-enhanced CT shows a soft-tissue mass with central calcification (*large arrow*) and peripheral gas pockets (*small arrows*)

Discussion

Endobronchial tumours are rare in childhood. They are benign in less than 10 % of cases and include hamartomas, leiomyomas and mucus gland adenomas. The malignant group includes the traditionally labelled bronchial adenomas, a class of primary endobronchial tumours which includes adenoid cystic carcinomas (cylindroma), carcinoid and mucoepidermoid tumours (MET) [3]. The term 'adenoma' implies a group of benign conditions and is a misnomer – all three entities can, on occasion, display malignant characteristics. This is recognised in the current WHO classification of lung tumours in which the category of adenomas includes benign tumours only, e. g. serous cell and mucus gland adenomas. The latter group is subdivided into MET when squamous differentiation is present and monomorphic adenomas when absent. Certain authors regard MET as intrinsically malignant, but cognisance should be taken of the fact that the WHO classification has a category of mucoepidermoid carcinoma for malignant types [4].

BMET, first described by Smetana in 1952, represents the rarest variant of bronchial adenomas, accounting for 2.5–7.3 % of cases. Until 1997, only 51 cases had been reported in patients under 16 years of age [2]. Recent analysis demonstrates a male preponderance of 3:2 and a ratio of right- to left-sided lesions of 22:21 [2]. Childhood MET typically arise in a mainstem bronchus or in the proximal portion of lobar bronchi as a polypoid endobronchial growth which compresses adjacent lung [1, 3, 4]. Histologically, the tumour arises from bronchial mucus glands, mimics its architecture and is covered by normal respiratory epithelium. MET can be classified as low or high grade. Childhood tumours are usually low grade. Two high-grade variants have been described with lymph-node metastases reported in one of these cases [2, 5]. Only one low-grade example with lymph-node metastases has been documented [2].

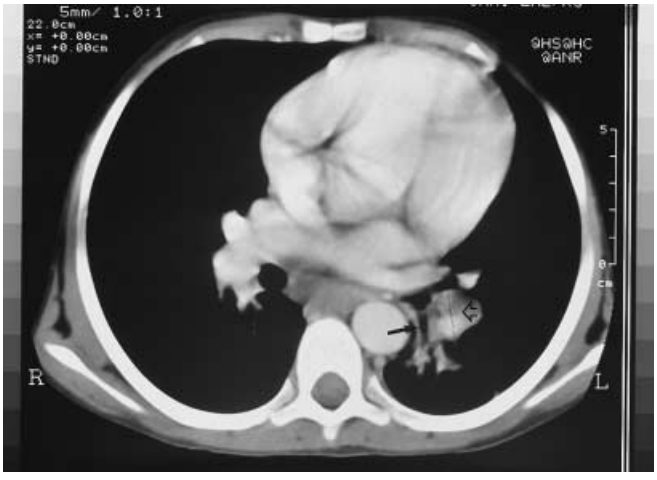

Fig. 3 Axial contrast-enhanced CT shows a left lower lobe bronchus (*closed arrow*) leading to the mass (*open arrow*)

The clinical features relate to bronchial irritation or compression and include recurrent pneumonia (48 %), persistent cough (35 %), haemoptysis (12 %), fever (12 %) and wheezing (10 %) [2]. Most of these symptoms were reported in the index patient, but the current case is unique because of the coexistence with secondary amyloidosis. It is unclear whether the amyloidosis is due to the BMET or a coincidental finding. Although amyloidosis is a well-recognised complication of many primary pulmonary processes, such as tuberculosis or cystic fibrosis, to our knowledge there have been no reports of an association between bronchial adenomas and amyloidosis.

The chest radiograph is usually abnormal. In a study by Kim et al. [6], the CXR demonstrated a central mass with post obstructive atelectasis in 33 % and solitary pulmonary or endobronchial nodules in 66 % of cases. At CT, the tumours were oval or lobulated, conformed to the bronchial branching pattern and contained punctate calcification in 50 % of cases.

Bronchoscopy with biopsy is the diagnostic modality of choice. The low malignant potential makes conservative surgical resection the preferred therapeutic option and carries an excellent long-term prognosis.

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