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Atypical pulmonary sequestration (bronchoarterial pulmonary malinosculation) in a child

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Introduction

The pulmonary sequestration spectrum and related congenital lung anomalies present an extremely complex and varied group of bronchopulmonary vascular malformations (1). Previous attempts at nomenclature and classification have proved inadequate in all cases. We report a case with extralobar pulmonary sequestration (EPS) presented by accessory lung with own pleural investment, systemic blood supply and pedicle communication with the tracheobronchial tree. The findings differed from the classical description of ERS and corresponded to bronchoarterial pulmonary malinosculation (BAPM) reported by Clements and Warner (1).

Case Report

The patient was a 10-year-old girl, whose disease clinically started 1 month prior to hospital admission, with cough and fever. A chest roentgenogram at that time showed left side dorsobasal opacification suggesting pleuropneumonia or incapsulated pleuritis. The child was treated with a course of antibiotics which cured its pyrexia but still remained continuing the abnormal shadowing in the left dorsobasal side of the chest. Due to computer tomographic data suspicious for paraesophageal hernia, the child was referred to our department.

On physical examination she was in a good general condition with moderate scoliosis with convexity to the left side. Her chest was clear to ausculation with decreased breath sounds at the left base. Results of laboratory studies were within normal limits. There was moderate hypoxaemia (PaO_2 66·4 mmHg, $PaCO_2$ 41·0 mmHg). The spirography revealed a low degree of restriction VC 79%, minute ventilation 69%, FEV/FVC 83%. Further radiologic studies and computed tomographic scanning illustrated an ovoid shaped cystic and partly solid appearing mass in the left dorsobasal part of the chest (Plate 1).

The aortography showed the presence of four arteries in this area, originating from the intercostal arteries and one branch coming from the descending aorta.

Through a left-sided thoracotomy approach left upper and lower lobes were found of normal appearance and an isolated dark blue pyramid mass located in the dorsobasal part of the thorax, connected with the pulmonary hilus. The mass had its own pleural investment separate from the adjacent normal lobes. Dissection was carried out. No branches of the pulmonary artery feeding the mass were discovered. There were four arteries originating from the intercostal arteries and two veins draining to the hemiazygous. In the upper part of the lesion a pedicle was identified leading to the hilus, located under the lower lobar bronchus and containing a narrow bronchus with two ramifications entering the accessory lung parenchyma.

After ligation of the blood vessels and the bronchus communicating with the tracheobronchial tree, the mass was removed. The pathology specimen consisted of a pyramidal mass weighing 86 g and dimensions 5·5 × 10 × 15 cm covered by a smooth to finely wrinkled pleura. On the cut surface dilated cyst-like bronchial ramifications blended with atelectatic lung parenchyma. Microscopically the lesion was presented by inflammated saccular bronchiectasiae, uniformly dilated bronchioles, and cleft-like spaces lined by cubiodal epithelium (Plate 2). There were atelectatic areas and signs of pneumonia in stage of organization. These gross and microscopic findings supported the surgical diagnosis of EPS. The postoperative course was unremarkable and the child was discharged on the eighth day.

Comments

EPS is a rare congenital anomaly of the lung (2,3,4). Out of 1347 pulmonary malformations, Vogt

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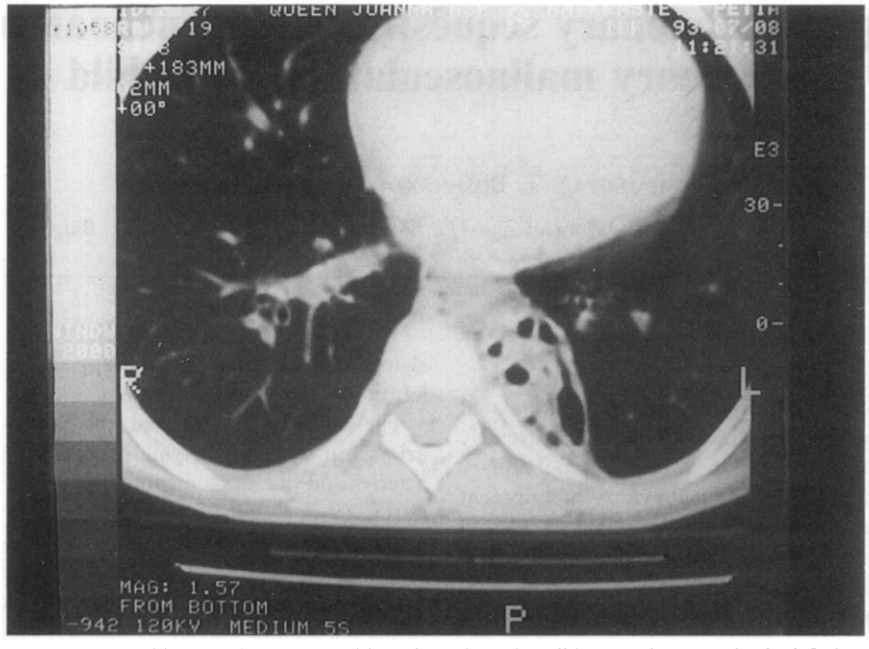

Plate 1 Computer tomographic scan shows an ovoid cystic and partly solid appearing mass in the left dorsobasal part of the chest.

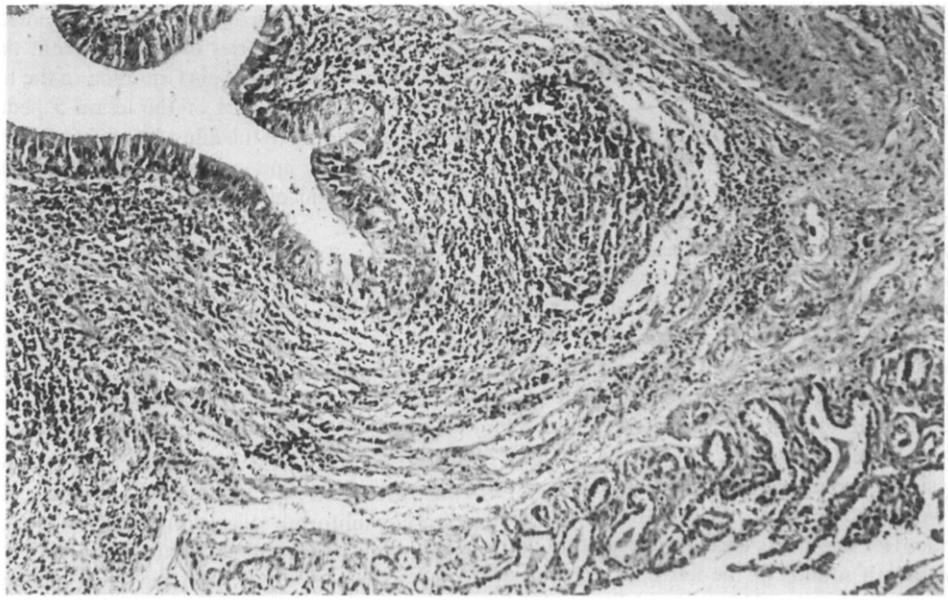

Plate 2 Dilated bronchiole with abundant lymphoid infiltration in the wall and adjacent lung parenchyma, fibrosis and cleft-like spaces, lined by cubiodal epithelium. Haematoxylin-eosin stain, × 100.

Moykopf, Rau and Branscheid (1991) found accessory lung tissue in 127 patients (9·9%) and only in four cases differentiated into EPS (5).

EPS is believed to arise from an outpouching of the foregut separate from the normally developing lung, with its own blood supply (6).

In our case there was a narrow accessory bronchus located in a pedicle which was not separated from its foregut source and connected the trancheobronchial tree with the accessory lung. The theory of a separate foregut source in the embryology of EPS seemed to be improbable in our case. The pulmonary artery was

not developed and the systemic arterial blood supply from the aorta was persisting in this accessory lung parenchyma. The case presented an atypical EPS, an entity which did not find place in the used classifications of pulmonary malformations (2,4–6). Our findings were consistent with BAPM described by Clements and Warner (1987) as an abnormality of both bronchopulmonary airway and the arterial blood supply to an area of lung. The abnormality of bronchopulmonary communication may range from absent connection with a blind-ending bronchus (agenesis) or atretic membrane to apparently normal patency with a gradation of abnormality in between (1).

The classification of the newly-named pulmonary malinosculation spectrum proposed by Clements and Warner (1987) by means of a simple descriptive approach, included all congenital anomalies and improved our understanding and management of these complicated lesions.

Our patient did not have specific clinical symptoms. The scoliosis with convexity to the left side had some impact on the clinical signs of decreased breath sounds.

Due to the presence of communication between the sequestrated tissue and the tracheobronchial tree the child had suffered from recurrent inflammation in the sequestrated lung with development of saccular bronchiectasiae, giving a cystic appearance of the lesion.

The finding of bronchiectatic changes together with uniformly dilated bronchioles and alveoli lined by cubiodal epithelium required differential diagnosis with cystic adenomatoid malformation type 2. The clinical diagnosis in our case relied upon X-ray, computer tomography and mainly upon aortography. A diagnosis of lung sequestration by colour coded Doppler sonography was reported in 1992 by Deeg, Holbeck and Singer (7).

Extirpation was the only reasonable approach after the diagnosis had been established. The low partial pressure of oxygen suggested that there was a significant shunt through the EPS which was an additional reason for surgical removal. The ligature of the blood vessels should be done very carefully since there was a danger from bleeding due to the high blood pressure in the aorta and its branches supplying the sequestrated tissue.

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