Case reports

Intralobar bronchopulmonary sequestration: antenatal diagnosis

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Abstract

A child with the antenatal diagnosis of pulmonary cystic adenomatoid malformation underwent thoracotomy and an intralobar bronchopulmonary sequestration was found. Histological examination of the resected specimen showed cystic adenomatoid malformation within the sequestered segment.

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Congenital lung lesions which may now be diagnosed antenatally by ultrasound examination include cystic adenomatoid malformation[1] which is a rare lesion with multiple cysts of differing sizes sometimes causing mediastinal shift. Cystic adenomatoid malformation may be responsible for acute neonatal respiratory distress, and in the longer term may cause repeated respiratory infections and undergo malignant change. Paediatric surgeons should be made aware of the possible diagnosis and plans made for the further obstetric, neonatal, and surgical management.

We describe a case in which the antenatal diagnosis of pulmonary cystic adenomatoid malformation was made, and at subsequent elective lobectomy an intralobar bronchopulmonary sequestration was found. The resected specimen showed cystic adenomatoid malformation within the sequestered segment.

Case report

A fetus of 34 weeks gestation was initially thought to have a diaphragmatic hernia on routine antenatal ultrasound scanning. Subsequent prenatal scanning suggested the diagnosis should be revised to cystic adenomatoid malformation of the lung affecting the left lower lobe. At birth a baby girl was delivered uneventfully and without respiratory distress. Postnatal computed tomographic scans performed at two weeks and 20 months both reported a multicystic lesion (fig 1) compatible with the features of cystic adenomatoid malformation.

An elective left lower lobectomy was performed when the child was aged two and a half to preclude malignant transformation. At thoracotomy an obvious transdiaphragmatic vessel was noted passing to the abnormal segment of

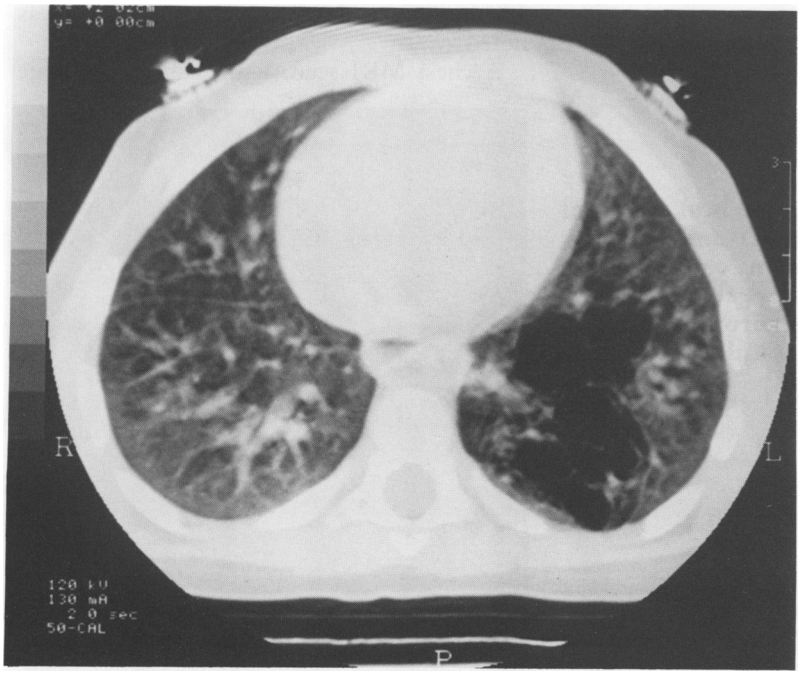

Figure 1 Computed tomographic scan of chest showing the cystic lesion in the posterior basal segment of the left lower lobe.

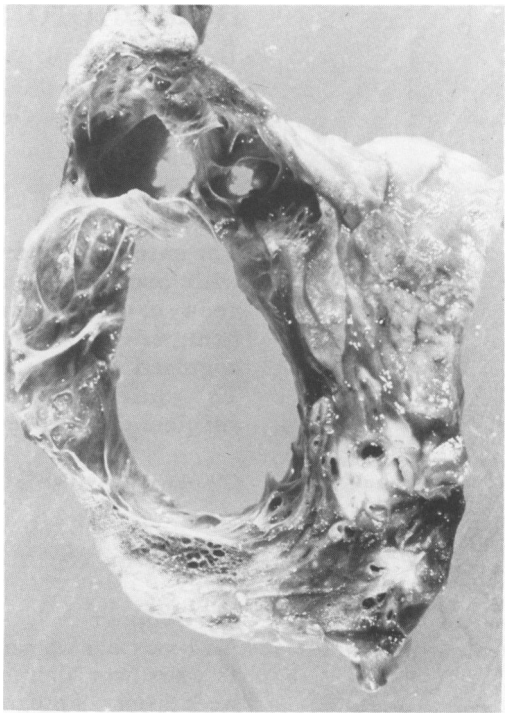

Figure 2 Cut section of resected specimen showing cystic change.

the lower lobe. An operative diagnosis of bronchopulmonary sequestration was made and lobectomy completed.

Macroscopically the lobe measured 9 × 7 × 7 cm and on section showed a consolidated posterobasal segment with overlying pleural thickening. Multiple cysts measuring up to 2·5 cm in diameter were present in the consolidated segment (fig 2) and an aberrant vessel passed through the diaphragmatic surface of the abnormal area. Histological examination revealed cystic structures lined by cuboidal to pseudostratified columnar epithelium with smooth muscle fibres in the walls, and amorphous eosinophilic debris with foamy macrophages in their lumina. The interstitium was infiltrated by chronic inflammatory cells with lymphoid follicle formation. The pathological appearances were those of type II cystic adenomatoid malformation with numerous cysts arising in an intralobar bronchopulmonary sequestration.[2-4]

On retrospective review of the antenatal ultrasound examination no evidence was seen of the abnormal systemic arterial supply to this sequestered segment of lung.

Discussion

To our knowledge intralobar bronchopulmonary sequestration in which an unexpanded portion of lung with no tracheobronchial connection and a systemic arterial supply has not hitherto been reported to be diagnosed prenatally.[5,6] This condition should now be included in the differential diagnosis of abnormal intrathoracic lesions detectable on antenatal ultrasound scan when a careful search should be made for an aberrant systemic blood supply.

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Progressive tracheal and superior vena caval compression caused by benign neurofibromatosis

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Abstract

The case history presented is of a patient with progressive tracheal and superior vena caval compression caused by a benign neurofibroma, a previously unrecognised feature of neurofibromatosis. The patient was successfully treated by surgical decompression.

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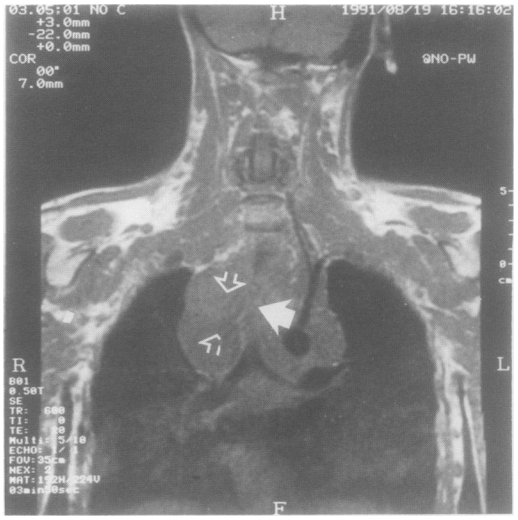
Case report

A 21 year old woman, a known asthmatic for 11 years, was admitted to hospital with a four week history of progressive shortness of breath associated with dry cough and considerable stridor. She was initially treated with increasing doses of bronchodilators and oral steroids, but continued to deteriorate. The patient's father was known to suffer from Von Recklinghausen's disease and no other member of the family was known to be affected.

On examination she looked unwell and was short of breath at rest. She had multiple cafe au lait spots and subcutaneous nodules, her face was puffy, and she had prominent subcutaneous veins on the top half of her body. Inspiratory stridor of moderate degree was noted.

A chest MRI scan (figure) showed a large



MRI scan of the chest showing a lobulated mediastinal mass (empty arrow) causing lower tracheal compression (solid arrow). © *University Department of Medical Illustration.*