EXTRA LOBAR SEQUESTRATION: DEMONSTRATION OF BLOOD SUPPLY BY DOPPLER ULTRASOUND, CT AND MR ANGIOGRAPHY.

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ABSTRACT

Extralobar sequestration (ELS) is a congenital malformation with anomalous vessel(s) arising from the systemic circulation. ELS may be mimicked by other lesions such as cyst-adenomatoid malformation. Duplex ultrasound, CT and MR angiography suggested the correct diagnosis by demonstrating an anomalous artery arising from the abdominal aorta supplying the ELS.

CASE REPORT

A 25 year -old woman G1P1 at 18th week pregnancy demonstrated a cystic mass in the left hemithorax of the foetus. Follow-up ultrasonography demonstrated a complex mass with cystic and solid components occupying most of the left hemithorax and a diagnosis of Type 2 cyst adenomatoid malformation (CAM) of the lung was made. Early in the third trimester she developed polyhydraminos and the fetal heart was displaced toward the right hemithorax. Delivery was induced at term and a male infant weighing 3,640 gm was born. Apgar scores were 6 at 1 minute and 9 at 5 minutes, but the infant rapidly developed respiratory distress and was intubated and ventilated until surgery.

A chest radiograph revealed a large homogeneous soft tissue mass in the left hemithorax which displaced the heart and mediastinum to the right. Ultrasonography of the chest demonstrated a sharply defined crescentic predominantly echogenic mass with several small vessels. There was a suggestion of a vessel arising directly from the aorta and extending into the mass. Doppler interrogation demonstrated a systemic arterial waveform (Fig 1). Computed tomography of the chest showed a left posterior mediastinal mass extending across the midline with an anomalous artery arising from the abdominal aorta and extending into the mass (Fig 2). The heart was displaced to the right and there was a small left pneumothorax.

An MRI was performed on 1.5T superconducting system (Siemens). Axial T1-weighted TSE(693/12), sagittal TSE(587/12), coronal T2-weighted TSE(3894/112) and postcontrast axial TSE(693/12) and sagittal TSE(587/12) and 3D FISP gadolinium enhanced MR Angiography was performed. The T1-weighted image demonstrated

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a large heterogeneous low signal intensity left posterior mediastinal mass measuring 8.2x4.5x6.4cm that crossed the midline. On T2-weighted sequences the mass was of high signal intensity with several cystic lesions. There was enhancement of the septae and marked peripheral homogeneous enhancement of the mass (Fig 3A). MR Angiography (Fig 3B) demonstrated an anomalous artery arising directly from the abdominal aorta just above the coeliac axis and ascending into the mass.

A provision diagnosis of extralobar sequestration was made and a thoracotomy with ligation of a feeding artery arising from the abdominal aorta at the level of the coeliac axis and resection of the extralobar sequestration was performed.

Histopathological examination confirmed an extralobar pulmonary sequestration. The patient recovered and remains well at eight months.

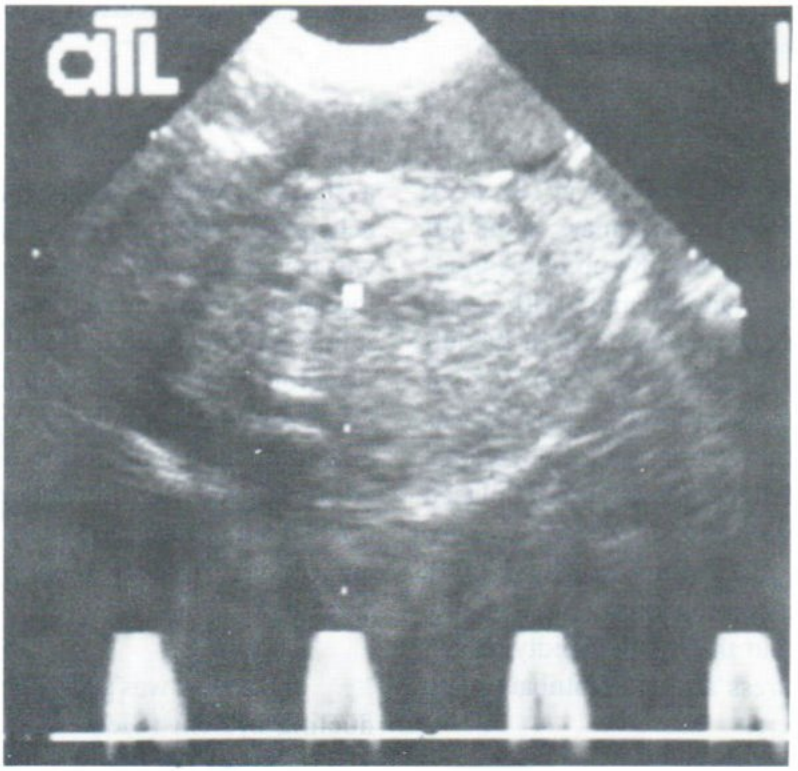


Fig 1 Doppler tracing from the vessel supplying the sequestration demonstrates a systemic arterial waveform.

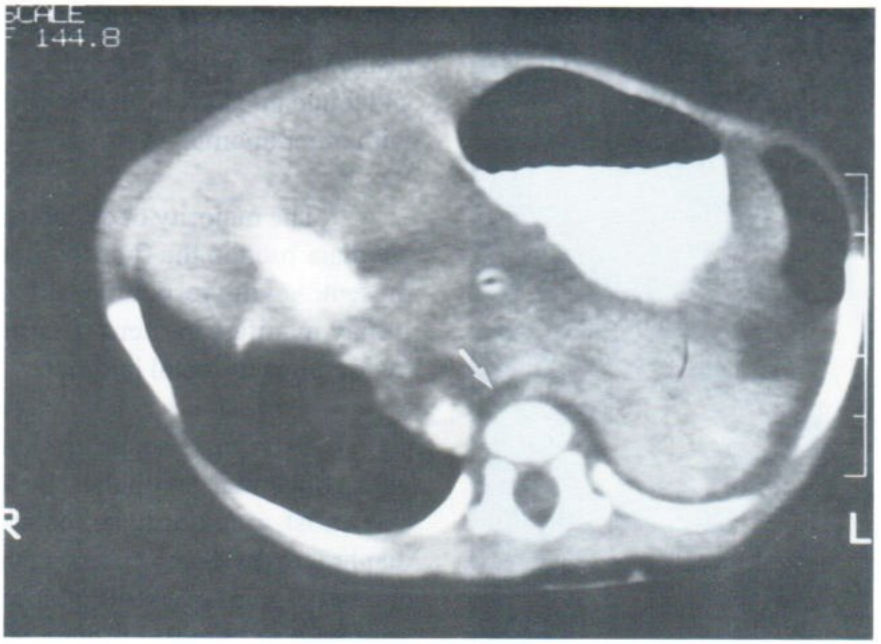
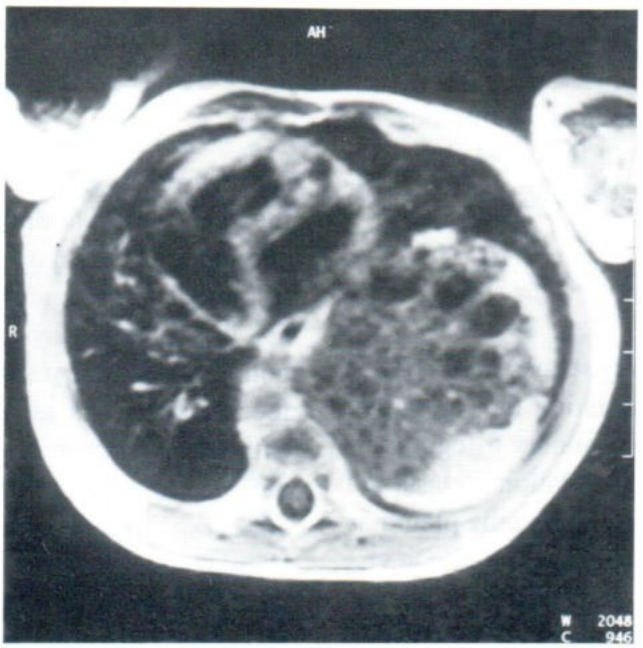
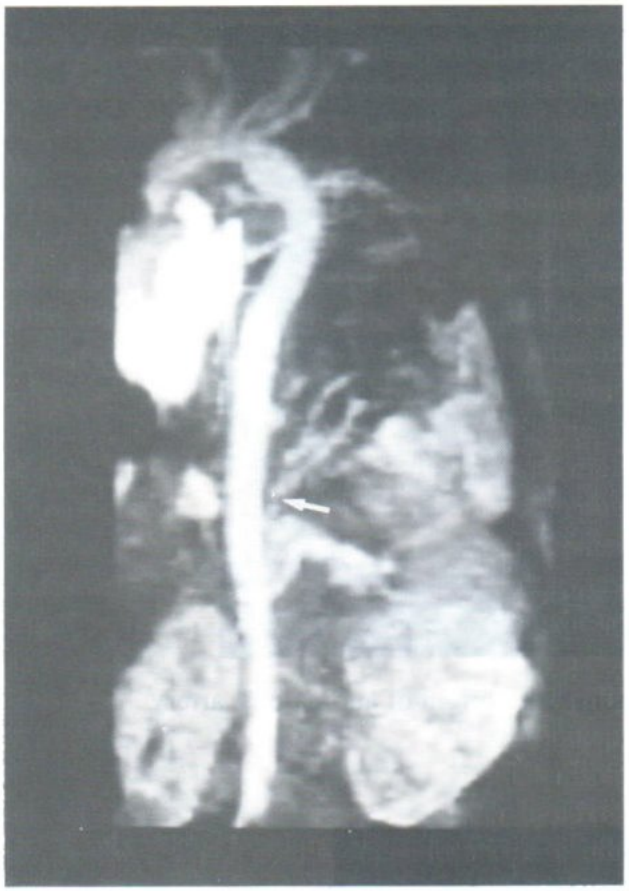


Fig 2 Axial contrast-enhanced CT demonstrates origin of anomalous artery from the abdominal aorta. (arrow)



Fig(3A) Axial postcontrast MR image. Extralobar sequestration with areas of low signal intensity corresponding to cystic areas with enhancement of the wall. The heart is markedly displaced into the right hemithorax.



Fig(3B) MR angiogram depicts anomalous artery (arrow) arising from just above the celiac axis and ascending into the sequestration.

DISCUSSION

Extralobar pulmonary sequestration (ELS) is a rare congenital malformation that is part of the spectrum of broncho-pulmonary foregut malformations. Pulmonary sequestration is classified as intralobar being encased in the same pleura as the adjacent lung or extralobar with a separate visceral pleural investment. Pulmonary sequestration consists of lung tissue usually cystic which has no normal communication with the tracheobronchial tree or pulmonary artery. Extralobar sequestration make up 25% of pulmonary sequestration and 90% occur at the left base. They can also occur in the mediastinum, within the diaphragm, and in retroperitoneal sites. Retroperitoneally pulmonary sequestration are asymptomatic and found incidentally or at autopsy if unassociated with other anomalies.

The majority of extralobar sequestration's are diagnosed in the first six months of life, often presenting with respiratory distress and cyanosis. It may manifest in-utero if associated with polyhydramnios or foetal hydrops.

Extralobar sequestrations are associated with congenital anomalies in 50-65% of cases. The most commonly associated is congenital diaphragmatic hernia, diaphragmatic eventration or paralysis. In large sequestrations, or those found with a large congenital diaphragmatic hernia or pleural effusion, pulmonary hypoplasia may occur. Other anomalies found include bronchogenic cysts, pericardial defects, cyst-adenomatoid malformation, foregut duplication-with communication to the oesophagus or stomach, ectopic pancreas and vertebral anomalies.

Pulmonary sequestration drives its blood supply from a systemic artery arising directly from the thoracic or abdominal aorta in 80%. The anomalous artery may arise from the splenic, gastric, subclavian or intercostal artery in 15% and in 5% it is supplied by the pulmonary artery or by both the pulmonary and systemic circulation. The feeding artery is generally single but 20% of extralobar sequestrations are supplied by multiple arteries. Venous return is typically systemic in 80%, but drainage can be through the Azygos, Hemiazygos or venacava to the right atrium. The venous return is partly through the pulmonary vein in 20% of cases. Systemic arterial supply is not pathognomonic of sequestration. It can occur in other congenital conditions such as arteriovenous fistulae, pulmonary aplasia and systemic arteries can supply the normal lung parenchyma. In ELS the usual radiographic finding is a cystic or solid mass in the left lung base. Other findings include

normal appearing lung, hyperlucent areas, a combination of solid and cystic mass, or pneumonia.[1]

Typical sonographic findings are of a uniformly echogenic mass surrounded by a thin echogenic rim which distinguishes it from pulmonary consolidation and atelectasis.[2] Diagnosis is confirmed if a systemic feeding vessel is demonstrated and Doppler demonstrates a systemic arterial waveform. Antenatal sonography can demonstrate nonimmune hydrops foetalis, pleural effusions or maternal hydramnios. Pulmonary sequestration is identified as an echogenic mass separated from the lung.

The CT appearances show multicystic abnormalities with pulmonary cysts and emphysema which is probably due to air trapping from collateral ventilation. Dynamic, ultrafast high resolution CT demonstrate the anomalous artery supplying the sequestration.[3] The systemic arterial supply to sequestation is prone to partial or complete thrombosis. Calcification within the anomalous artery and sequestration has been demonstrated.[1]

MR angiography can similarly demonstrate systemic arterial supply to sequestration. MRI can demonstrate solid and mucous containing components of sequestration. However, emphysematous changes and calcification are not demonstrated.[4,5]

The definitive treatment for sequestration is surgical resection, and preoperative assessment of the presence and location of an anomalous systemic artery supply is the primary objective of imaging. Classification into intralobar and extralobar sequestration by the absence or presence of a separate pleural investment are not of primary importance for surgery. Therefore, all imaging techniques capable of demonstrating anomalous vessel are utilised to diagnose pulmonary sequestration. Non-invasive imaging technique include sonography, helical CT and MR angiography.

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