

Bilateral intralobar pulmonary sequestration

A case report

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Abstract

Bronchopulmonary sequestration is a congenital lung malformation consisting of a non-functioning lung segment. Arterial supply to such a segment is found to be systemic rather than pulmonary, and by definition there is no communication with the tracheobronchial tree. It accounts for about 6% of all congenital pulmonary malformations. Bilateral bronchopulmonary sequestration is yet more uncommon. These malformations can be classified as either intralobar sequestration (the commoner type), or extralobar sequestration (in 14- 25%).

Keywords

Sequestration; bronchopulmonary; intralobar

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History

A 58 year old lady was referred to Accident and Emergency with a sudden onset of severe chest pain radiating to the back.

Clinical features

The pain was described as tearing in nature. The patient, who had no previous history of hypertension, was found to have a blood pressure of 180/105mmHg. No significant difference was recorded between right and left arm blood pressure measurements. The patient admitted to a chronic cough, but otherwise she had no family or medical history of note. Physical examination was unremarkable.

Investigations

Initial evaluation included a sinus rhythm electrocardiogram and a non elevated creatine kinase MB isoenzyme. Other parameters including temperature, full blood count, urea and electrolytes, activated partial thromboplastin time, international normalized ratio, erythrocyte sedimentation rate and c-reactive protein were all found to be within the normal range. A postero-anterior chest radiograph was reported as normal (Figure 1).

Treatment and outcome

The constant severe pain persisted despite the administration of NSAIDs. The suspicion of a dissecting aortic aneurysm was raised, and an urgent CT angiogram of the aorta was performed.

Progress

The CT angiography was performed on a 16 slice helical CT scanner (GE Brightspeed) with a pitch of 0.9:1, 260 mA, and 120 kV. One hundred millilitres of iodinated contrast medium (350 mgI ml⁻¹ Omnipaque™) was injected intravenously using an automatic infusing pump at a flow rate of 4 ml sec⁻¹. There was no evidence of a dissecting aneurysm on CT angiography, but a dilated aortic bulb was evident (Figure 2), however this is not typically of clinical relevance with this patient's presenting complaint.

An incidental anomalous finding was also detected. Both lower lung lobes were supplied directly by systemic branches (around 5mm in diameter) arising from the ventral part of the thoracic aorta, at the level of the 10th thoracic vertebra. (Figures 3-5)



Figure 1: PA erect chest radiograph taken at the A&E department

Particularities noted in the lung parenchyma included multiple thin walled cysts, some of which were found to be fluid filled, while others contained air. These together with emphysematous changes in the surrounding region (figures 6,7), are due to post-obstructive hyperinflation of the sequestered lung. The characteristic finding of mucoid impaction of the nearby bronchus surrounded by hyperinflated lung was also identified in this case (Figure 8).



Figure 3: Axial CT image showing the origin of the anomalous arteries from the ventral part of the aorta

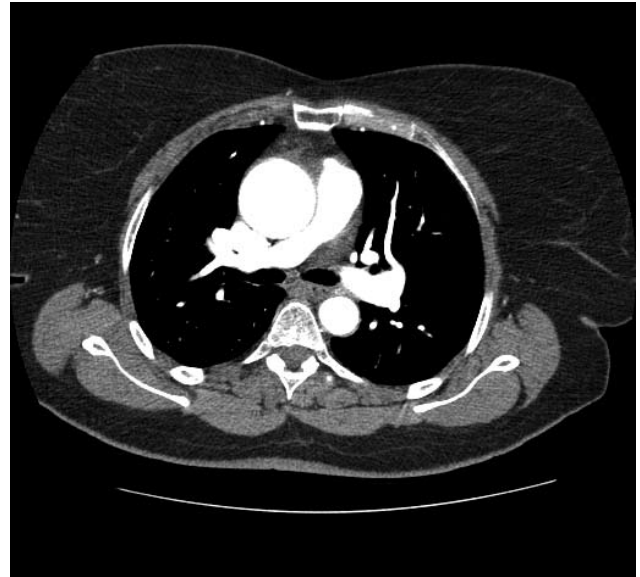


Figure 2: CTA (Aorta) – showing dilatation of the aortic bulb

Discussion

Definition and classification of bronchopulmonary sequestration

Classically a triad is used to describe bronchopulmonary sequestration: (1) a non functioning lung segment, (2) with no communication with the tracheobronchial tree, (3) and having a systemic arterial supply.^{1,2}

In intralobar sequestration, though the affected lobe is separated from the bronchial tree (as described by the triad), it is enclosed by visceral pleura.



Figure 4: Sagittal CT reconstruction showing the origin of the right anomalous artery, which is seen turning dorsally to the affected lobe

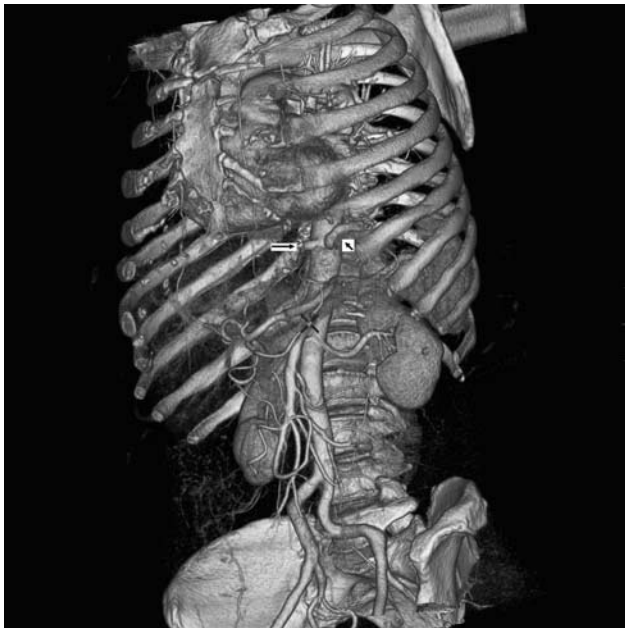


Figure 5: 3D reconstruction: arrows point to the anomalous arterial supply

Embryology and pathophysiology

The most frequently supported embryological theory of sequestration formation involves an accessory lung bud that develops from the ventral aspect of the primitive foregut.³ The pluripotential tissue from this additional lung bud migrates in a caudal direction with the normally developing lung. It receives its blood supply from vessels connecting the aorta and covering the primitive foregut. These attachments to the aorta remain



Figure 6: A second axial CT image showing the course of the right anomalous artery, and also the emphysematous changes of the affected lobe

to form the systemic arterial supply of the sequestration. Early embryologic development of the accessory lung bud results in formation of the sequestration within normal lung tissue. The sequestration is encased within the same pleural covering. This is the intrapulmonary variant. In contrast, later development of the accessory lung bud results in the extrapulmonary type that may give rise to communication with the gastrointestinal tract.⁴



Figure 7: Image showing the emphysematous changes of the affected lobe



Figure 8: Mucus plugging of the affected bronchus

About 10% of cases are associated with congenital anomalies: cardiac (including Tetralogy of Fallot), renal (failure of ascent and rotation), cerebral anomalies and skeletal deformities.

Clinical findings

More than one half of intrapulmonary sequestrations are diagnosed in later childhood or even in adulthood.³ Neonates and infants are usually asymptomatic. In contrast, more than one half of extrapulmonary sequestrations are diagnosed in patients younger than 1 year. Often, this is because other congenital anomalies are present, including congenital diaphragmatic hernia, cardiac malformations, and gastrointestinal malformations.

In the extrapulmonary form, males are affected approximately 4 times more often than females. Incidence is equal in males and females in the intrapulmonary type. Fifteen percent of patients remain completely asymptomatic.⁵

Clinical manifestations may include: pain, repeated chest infections⁶ in the same location, high output congestive heart failure, cough and sputum production and haemoptysis.⁴ The condition most commonly affects the posterobasal segments, and is slightly more common on the left. The morbidity and mortality rates are exceedingly low if resection of the mass precedes repeated infection. Postoperative results are uniformly good.

Complications of sequestration include massive spontaneous non-traumatic pulmonary haemorrhage, chronic inflammation, and fibrosis of the affected lung.⁵

Radiological findings

Several imaging modalities can be used to confirm a diagnosis of sequestration.^{6,7} Chest radiography is not very specific, though it might show recurrent pneumonias localized to the lower lobe. Sometimes, cavitation and cysts may also present. Bronchography will show the absence of communication of the rudimentary bronchial system. The arterial supply can be demonstrated using conventional angiography. Usually, a single large artery (though multiple smaller arteries can be found in about 16% of cases) arises from the distal thoracic aorta, or the proximal abdominal aorta. Rarer origins may be from the celiac trunk, the splenic artery, intercostals or coronary arteries. The venous supply is usually via normal pulmonary veins to the left atrium. In contrast to the extralobar type of sequestration, only

5 % of cases drain into the systemic veins (azygos, hemiazygos, intercostals, and the superior vena cava).¹ However, the latter two modalities of investigation are now considered as obsolete and have been largely replaced by CT^{8,9} and MRA.¹⁰

CT scan aids not only in diagnosis, but also in the differentiation between intra- and extralobar sequestration. As opposed to the former, in extralobar sequestration, a homogenous well-circumscribed soft tissue density mass is usually seen. Intralobar sequestration consists of single/multiple thin-walled cysts containing fluid, air-fluid levels or air alone. Typical features include mucoid impaction of bronchus surrounded by hyperinflated emphysematous lung. CT angiography allows the visualisation of one/two anomalous systemic arteries arising from the aorta. Characteristically, there is enhancement of the sequestered lung at the same time as the thoracic aorta on rapid sequential CT scans.^{5,9} When the sequestration communicates with the gastrointestinal tract, it is termed bronchopulmonary foregut malformation.

Anatomical diagnosis

Bilateral Intralobar pulmonary sequestration with systemic supply derived from lower thoracic aorta.

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