

PULMONARY INTRALOBAR SEQUESTRATION IN A PATIENT WITH CYSTIC FIBROSIS

A case report

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Abstract

We report a case of a 3 years old girl in whom pulmonary intralobar sequestration (ILS) was suggested but surgery, extended clinical and laboratory examinations led to the correct, final diagnosis of Cystic Fibrosis. (CF)

ILS has been variably considered as either a congenital anomaly or an acquired postinflammatory process. The association of ILS and CF is rare. This unique case raises questions regarding the pathogenesis of ILS.

To our knowledge such a condition has previously not been reported in children.

Key words: intralobar sequestration, Cystic fibrosis in children

Case report

A 3-year old girl, who weighed 3200 g at birth was born at normal gestation after normal pregnancy, she presented shortly after birth with tachypnoea and grunting. The general practitioner thought it to be adaptational disturbances, but at 2 month of age the girl was referred to the hospital. Physical examination revealed tachypnea, dyspnea, cough, nasal flaring, hypoxia and somatic retardation. Arterial blood gases showed marked hypoxemia, with high CO₂ tension.

Decreased breath sound with crepitation above the lungs suggested the diagnosis of pneumonia.

The heart rate was 160, and the breath frequency was 50-60. There was no history of fever and hemoptyses.

Several clinical and radiological examinations, including chest X-ray, US, ruled out cardiac or central origin, diaphragmatic or hiatus hernia. Chest X-ray showed thorax asymmetry and the left lung seemed to be smaller than the right. (Fig. 1.)

Perfusion lung scintigraphy (Tc 99) revealed decreased perfusion activity at the left lower lobe. Chest CT scans showed a sharply demarcated triangular atelectasis in this region and MRI suggested the diagnosis of sequestration. (Fig. 2-3.).

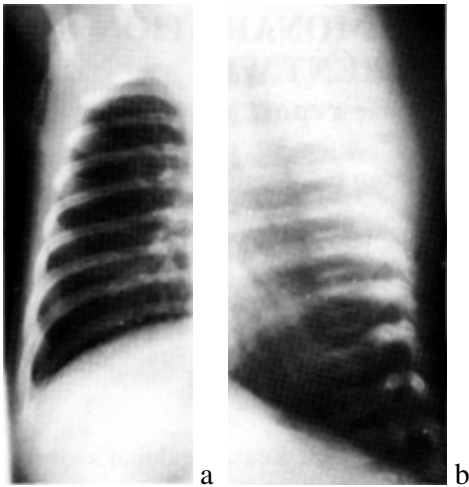


Fig. 1: Chesi X-Ray, AP view: thorax (lung) assymetry, mild overinflation on the right side (a); lateral view: hyperinflation in the basal segment hut there was no sign of any other anomaly (h).

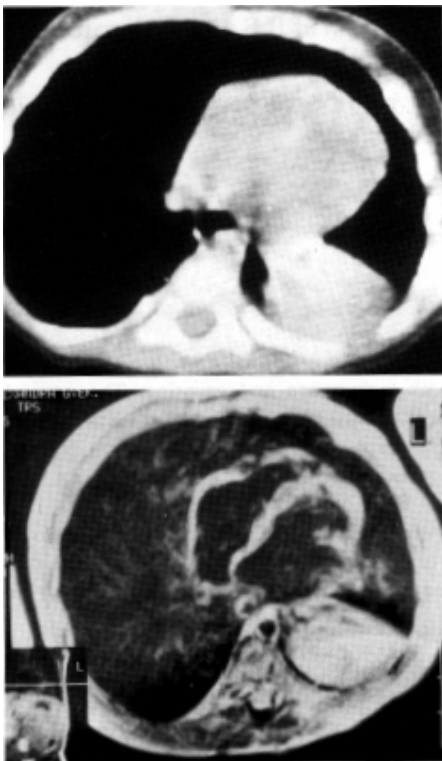


Fig. 2., 3.a

Fig. 2: CT scans showed supraphrenic triangular atelectasis behind the heart.

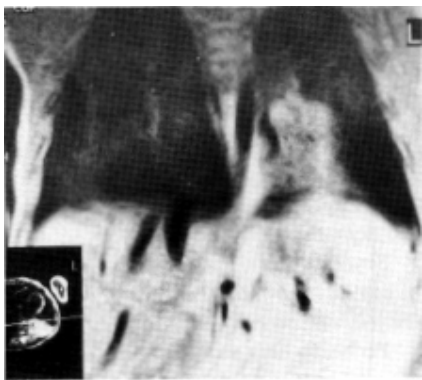


Fig. 3.b.

Fig. 3: MR T2 weighted axial (a) and coronal (b) scans showed inhomogenous, hyper intense "mass" with an abnormal vessel from the descending aorta (arrow).

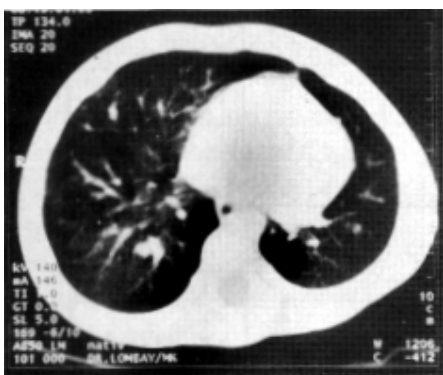


Fig. 4: Repeated CT scan showed hyperinflated areas of the lungs and some widened bronchus shadow with thinned wall.

At 4,5 months of age surgery was done and histology proved the sequestered lung tissue with acute and chronic inflammatory changes and fibrosis.

After surgery the girl was in good clinical condition, but shortly after recurrent obstructive bronchitis began with tachy- dyspnea, coughing, purulent sputum. Repeated clinical and laboratory and CT examinations at 17 months of age revealed the diagnosis of cystic fibrosis. (Fig. 4.).

Discussion

Bronchopulmonary sequestration is a relatively rare anomaly, which has been known for over 100 years (2., 3). Despite this fact there are still some unanswered questions about the pathogenesis. Sequestration is defined as a mass of nonfunctioning pulmonary tissue that does not have any connection with the normal tracheobronchial tree and is often supplied by an anomalous systemic artery. It is classified into two types: intralobar sequestration, which is located within the normal lung and extralobar which is located outside the normal lung [1, 4]. Intralobar sequestration is much

less often associated with other anomalies (14%), than it is with the extralobar form (about 50%). (Table 3). It is more likely that intralobar sequestration is a direct result of postinflammatory degenerative changes, as frequently diagnosed in adults. Treatment of choice is surgery as early as possible. [4]. Diagnostic imaging modalities have an important role for the diagnosis (table 4). In our case the CT scans showed the pulmonary anomaly but MRI revealed the characteristic signs of the sequestration. Although the surgery was successful there was no significant improvement clinically which suggested any other diseases. Repeated laboratory examinations revealed the correct diagnosis finally.

We could find a case of a 38 years old patient with CF in the literature [5] to whom at autopsy intralobar sequestration was found and which was thought to be a coincidental association but we have no knowledge of any similar pediatric case in the relevant literature.

We may repeat Tomashefski et al' [5] statement: "this unique case raises questions regarding the pathogenesis of ILS and allows comparison between the morphology of ILS and that advanced CF-associated disease.

References

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