Neonatal respiratory distress due to congenital lobar emphysema

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Section: Paediatric radiology
Area of Interest: Paediatric
Procedure: Diagnostic procedure
Technique: CT
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 27 days, male

Clinical History:

A 27-day-old male newborn was admitted for progressive dyspnoea evolving from the 7th day of life and becoming oxygen-dependent. He was a full term infant born of a twin pregnancy. An antenatal ultrasound was performed but did not show any abnormality. Frontal chest X-ray and CT revealed pathognomonic findings.

Imaging Findings:

Fig. 1: Anteroposterior chest X-ray in a 27-day-old newborn shows a hyperlucent left upper lobe with attenuated vascularity. There is an overdistension of the left upper lobe with widened intercostal spaces and mediastinal shift to the right. Left lower and right upper lobar atelectasis are also noted.

Fig. 2: Thoracic CT scan confirms chest X-ray findings.

Fig. 2a: Axial thoracic CT shows a hyperlucent left upper lobe with attenuated vascularity. There is left upper lobe overdistension with a midline herniation and mediastinal shift to the right.

Fig. 2b: Coronal CT scan shows bilateral atelectasis.

Discussion:

Congenital lobar emphysema (CLE) is a rare lung malformation characterised by overinflation of one or more lobes leading to compression of the lung parenchyma and displacement of mediastinal structures. [1] Several pathological mechanisms are suggested, including bronchial wall cartilage deficiency, and intrinsic or extrinsic bronchial obstruction resulting in a check-valve mechanism with progressive lobar overinflation. There is no apparent cause in about 50% of cases. [2] The left upper lobe is the most often affected (in about 42% of cases) and bilateral CLE is extremely rare. [2, 3]

Patients typically present with respiratory distress occurring most often in the neonatal period, with hyperresonance of the hemithorax involved and decreased or absent breath sounds.

Chest X-ray which is often performed first-line in case of respiratory distress will show a hyperlucent and hyperinflated lung segment or lobe with attenuated vascularity, partial collapse of adjacent lung segments, and mediastinal shift to the contralateral side.

CT is performed to confirm the diagnosis, to rule out other abnormalities, and explore mediastinal vascular...
structures. [4]
A prenatal ultrasound can reveal an echogenic and homogeneous lung lesion with a tubular cystic structure at the hilum. [5]
On MRI these findings appear as T2 hyperintense lung areas and a T2 hyperintense tubular structure corresponding to a dilated bronchus with trapped fluid. [5]
Nuclear imaging will show the absence of perfusion of the affected lobe.

Bronchial atresia is another congenital malformation leading to lobar inflation. It is characterised by bronchial obliteration that often affects a segmental bronchus resulting in mucus impaction and segmental hyperinflation.
Chest plain film findings include a finger-like perihilar opacity. Thoracic CT will more accurately show mucus impaction with a distal hyperlucent lung.
Swyer-James syndrome is an acquired form of lobar or segmental emphysema as a result of an infectious bronchiolitis.

Patients with mild symptoms should be followed up unlike patients with respiratory distress who require surgical treatment (lobectomy). [6]

Congenital lobar emphysema is an uncommon surgical cause of neonatal respiratory distress that can be life-threatening. [7] The diagnosis is based on the clinical examination and imaging findings. The treatment consists of lobar resection (lobectomy) in symptomatic patients.

**Differential Diagnosis List:** Congenital lobar emphysema, Bronchial atresia, Pulmonary hypoplasia, Swyer-James syndrome, Pneumothorax

**Final Diagnosis:** Congenital lobar emphysema.

**References:**
Description: Frontal chest radiograph shows an overdistended and hyperlucent left upper lobe with widening intercostal spaces and mediastinal shift to the right. Left lower and lower right upper lobar atelectasis are also noted. Origin: Pediatric Radiology Department CHU IBN ROCHD / Casablanca MOROCCO
Figure 2

Description: Axial thoracic CT (lung windows) confirms CXR findings. Origin: Pediatric Radiology Department CHU IBN ROCHD / CASABLANCA MOROCCO
Description: Coronal thoracic CT also confirms chest X-ray findings. Origin: Pediatric Radiology Department CHU IBN ROCHD / CASABLANCA MOROCCO