Congenital Lobar Emphysema in Infants

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ABSTRACT
Congenital lobar emphysema in infants is a disorder that is detected most often in newborns or young infants. We report here the case of a 4-month-old infant who at two months suddenly presented upper respiratory infections, treated symptomatically, but evolution showed shortness of breath, wheezing, weight deficit. Thoracic ultrasound revealed left upper lobe hyperinflation causing mediastinal displacement to the right, a slightly reduced blood supply at this level, and a lobar emphysema appearance. Bronchoscopy evidenced a thickening in the left bronchial tree, due to left upper lobe emphysema.

Keywords: congenital lobar emphysema, congenital lobar emphysema in infants, infant

INTRODUCTION
Congenital lobar emphysema (CLE) is a congenital anomaly of the lung, with a prevalence of 1 in 20,000 to 1 in 30,000 (1). Most of the cases present in the neonatal period, with a male to female ratio of 3:1 (2, 3). CLE has also been reported with other associated anomalies with double superior vena cava and horse shoe kidney (4). One case is reported with polysplenia, a syndrome characterized by bilateral bi-lobed lungs and bilateral pulmonary atria along with liver, which is symmetrically placed in the midline and multiple nodules of spleen (5).

CASE REPORT
A 4-month-old male infant was admitted for respiratory distress, wheezing and failure to thrive. Personal physiological history showed that he was the first child from a physiological pregnancy, born at term by C-section, with a birth weight of 2850 g, an Apgar score 8/9, and a normal psychosomatic development. Family history was not significant, and personal pathological history included a congenital infection of undetermined etiology.

The clinical onset of the disease was two months before admission, on the occasion of an acute upper airway infection episode, which was...
treated by the family doctor with symptomatic drugs, but evolution showed persistent breathing difficulty, wheezing, and weight deficit. Objective examination at admission evidenced a patient with a weight of 4650 g (< 5th percentile), a length of 63 cm (75th percentile), weight index = 0.77, afebrile, with pale skin, reduced subcutaneous cellular tissue, laryngeal stridor, pulmonary hypersonority in the left hemithorax, vesicular murmur with prolonged expiration, bilateral sibilant rales, more frequent in the right hemithorax, mild intercostal and substernal retractions, respiratory rate 42 breaths/min, heart rate 166/min, SaO₂ = 95-96%.

Laboratory investigations detected no pathological changes, and echocardiography excluded congenital cardiac malformations. Chest X-ray showed left upper lobe hyperinflation causing mediastinal displacement to the right, with a slightly reduced blood supply at this level (Figure 1).

Chest computed tomography detected left upper lobe hyperinflation causing mediastinal displacement to the right, with a slightly reduced blood supply at this level and a lobar emphysema appearance (Figure 2).

Bronchoscopy evidenced omega-shaped epiglottis, short paramedian aryepiglottic folds, anteriorly displaced arytenoids; during inspiration, aryepiglottic folds and arytenoids made contact on the median line, starting to vibrate. The trachea and the right bronchial tree were completely permeable, without pathological changes, and the left bronchial tree was permeable at the level of the tertiary carinae of the lingular bronchus. In the superior branch of the left upper lobe bronchus, a thickening due to left upper lobe emphysema, without pathological secretions, was found.

Regarding differential diagnosis, cystic fibrosis was excluded based on two negative sweat tests, and cystic adenomatoid malformation was also excluded by chest computed tomography.

DISCUSSION

Congenital lobar emphysema (CLE) is a rare congenital malformation, characterized by alveolar distension and pulmonary hyperinflation, with contralateral pulmonary atelectasis (3). CLE is most frequently diagnosed in the neonatal period, 5% of patients being diagnosed around the age of six months. Diagnosis can also be established prenatally by ultrasound, as well as at school age (6, 7). Familial CLE cases have also been reported (8). The incidence of left lung lobe involvement is 43% of all cases, our infant belonging to this group; the middle right lobe is affected in 32% of cases, the upper right lobe in 28% of cases, while bilateral involvement occurs in 20% of cases (9).

The cause of congenital lobar emphysema can be identified in 50% of patients and can be attributed to a congenital bronchial cartilage defect, extrinsic compression of aberrant vessels, bronchial stenosis, viscous bronchial mucus, and mediastinal displacement to the opposite side of bronchial obstruction (10). In 10% of cases, congenital lobar emphysema can be associated with congenital cardiac malformations, which are excluded by echocardiography (11).

Patients frequently present tachycardia, tachypnea, costal retraction, with progressive accumulation of carbon dioxide in the affected lobe, and evolution towards respiratory failure. The pulmonary expansion of the two hemithoraces is asymmetrical, with the presence of bronchi, pulmonary hypersonority in the affected...
lobe, and diminished respiratory and cardiac sounds. The infant had polyneum, tachycardia, pulmonary sonority was detected by percussion; auscultation revealed vesicular murmur with prolonged expiration and bilateral sibilant rales, intercostal and subcostal retractions; respiratory failure was not associated, SaO₂ values being higher than 92%.

Chest X-ray describes hyperinflation of the affected lobe, with pulmonary atelectasis and contralateral mediastinal displacement, and bronchoscopy can be normal or changed. The suspicion of congenital lobar emphysema requires additional imaging investigations such as CT/MRI, which allow diagnosis and initiation of early treatment for a favorable prognosis (11-14). In order to diagnose the infant, chest X-ray and CT were performed, which evidenced left upper lobe hyperinflation causing mediastinal displacement to the right, with a slightly reduced blood supply at this level, and bronchoscopy detected a minimal thickening in the upper branch of the left lobe bronchus.

Conservative treatment is used in asymptomatic patients with minimal respiratory manifestations. Children over the age of two years with moderate respiratory symptoms and normal bronchoscopy can also be treated conservatively. In patients with severe respiratory manifestations, resection of the affected lobe segment is performed as an alternative to lobectomy (14). The case was interpreted as congenital lobar emphysema with mild/moderate respiratory manifestations, and was treated by conservative therapy, with a good clinical evolution of symptomatology.

**TAKE-HOME MESSAGE**

Unilateral clinical findings in a bronchiolitis-like young infant should point to a congenital lung malformation.

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**REFERENCES**