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CASE REPORT

Congenital lobar emphysema: conservative treatment and 13-year follow-up

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Abstract

Congenital lobar emphysema is a congenital malformation characterized by overdistension of the affected lobe without lung parenchyma abnormality. It can manifest in the neonatal period or later, depending on the number of lobes involved. Bilateral involvement is rare; in general, hyperinflation occurs more commonly in the left upper lobe. The diagnosis is clinical, confirmed by chest X-ray and computed tomography. Bronchoscopy can help especially in the differential diagnosis of foreign body and mucous plugs. This procedure also helps define the pathogenesis of congenital lobar emphysema identifying bronchial anomalies. In most of the cases the treatment is surgical, based on the excision of the affected lobe. In some cases, when there are mild or no symptoms, the management can be conservative. The aim of this study was to report a case of nonsurgical congenital lobar emphysema and its 13-year-follow up.

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INTRODUCTION

Congenital lobar emphysema (CLE) is a congenital disorder characterized by hyperinflation of one or more lung lobes with A histologically normal parenchyma. In most cases, only one lobe is involved, with the upper left (LSE) being the most commonly affected[1].

It is a rare disease, with a prevalence of 1 case in 20,000 to 30,000 live births. The male/female ratio is 2:1.[1-5].

Other congenital malformations such as congenital heart malformations, pectus excavatum, anterior mediastinal defects, diaphragmatic hernia and hiatal hernia may be associated. The diagnosis is based on clinical and radiological findings[3,6].

Clinical manifestations usually occur in the neonatal period, although, in rare cases, some newborns are asymptomatic and the diagnosis is incidental in late childhood or adulthood[2,3]. Symptoms are variable, from mild respiratory signs to severe respiratory failure. The most frequently observed signs and symptoms are dyspnea, tachypnea, intercostal retraction, wheezing, cough, cyanosis, asymmetrical breath sounds. older children may present with recurrent respiratory infections[1,2].

The most common rEadiological changes are hyperinflation of the affected lobe, contralateral mediastinal deviation and compression of the adjacent lung tissue, generating atelectasis. High-resolution computed tomography (HRCT) of the chest confirms the radiological diagnosis and outlines the anatomically affected lobe[1-3,6,7].

Bronchoscopy allows the identification of anatomical alterations and can also exclude endobronchial obstruction caused by bronchial mucous plug, foreign body or even extrinsic compression[6,7].

In severe cases, the treatment is surgical with excision of the affected lobe. In patients with mild to moderate symptoms, conservative treatment has been recommended. This type of treatment may be dubious, especially in those who develop recurrent pulmonary infection at the same time.[8-10]

The objective is to report a case of non-surgical CLE and the satisfactory result in the 13-year follow-up.

CLINICAL CASE

A 3-month-old boy was referred to the University Hospital because of tachypnea. His mother reported this symptom since birth with worsening after the diagnosis of bronchiolitis at 2 months of life. Pregnancy and delivery were uneventful. He was on exclusive breastfeeding. His mother had asthma and his father had allergic rhinitis.

Physical examination revealed an active and reactive infant, tachypneic on room air (respiratory rate of 60 breaths per minute), without cyanosis. Breathing sounds were reduced in the left hemithorax, with no adventitious sounds. There were no other abnormalities on physical examination. Chest radiography showed a hyperinflated left hemithorax with an area of hypertransparency in the left upper lobe and contralateral mediastinal shift, without atelectasis. HRCT confirmed the suspicion of ELC (Figure 1A). Flexible bronchoscopy was performed to rule out mucous plug due to bronchiolitis. This examination showed a congenital defect in the LSL bronchus cartilage (Figure 2).

Evaluation by the pediatric surgery team indicated lobectomy, which was not performed due to some recurrent respiratory viral infections. The child evolved with wheezing triggered by respiratory viruses, but remained asymptomatic during the period between these events, and the 1-year follow-up HRCT showed improvement in hyperinflation, with a slight increase in the left upper lobe, without mediastinal shift (Figure 1B) . Expectant treatment was maintained with regular clinical follow-up, with normalization of respiratory rate with increasing age. At 8 years of age, he was asymptomatic, with a normal physical examination and a gradual improvement in the HRCT scan (Figure 1C). Currently 13 years old, he remains asymptomatic, only being treated for asthma attacks.

DISCUSSION

Currently, technological developments in ultrasound imaging have allowed the prenatal diagnosis of ELC that shows a hyperechogenic lung[1,2]. However, the definitive diagnosis must be performed in the postnatal follow-up, as these abnormalities in the fetal lung parenchyma may spontaneously regress in the third trimester of pregnancy[1,2,8,11]. Postnatal diagnosis must be careful, as it can be easily confused with pneumothorax or foreign body[1,3,7,9]. It is important to emphasize that the chest X-ray of the newborn in the first days of life may show an opaque lung that later becomes radiolucent[2,5]. The diagnosis of our patient was suggested only after birth, in an investigation of persistent tachypnea after an episode of acute viral bronchiolitis.

The cause of ELC is identified in 50% of cases. Intraluminal bronchial obstruction such as redundant bronchial mucosa, bronchial stenosis, bronchial torsion or rotation, polyalveolar lobe, and congenital deficiency of bronchial cartilage are intrinsic factors reported as the etiology of ELC. Likewise, the presence of vascular rings, hilar adenopathy, esophageal or bronchogenic cysts and congenital heart malformations, are considered extrinsic factors that lead to bronchial obstruction and subsequent ELC. In the remaining cases, the etiology is unknown; no bronchial abnormalities can be demonstrated[1,2,5-7,9]. The reported case had abnormal cartilage of the LSL bronchus detected by flexible bronchoscopy (Figure 2).

Symptoms may vary depending on the degree of involvement and the location of the affected lobe. The most severe cases are described in LSE[9]. Generally, children are symptomatic in the first days of life[3,7]. About 5% of cases may show symptoms by 4-6 months of age. Tachypnea, dyspnea, and eventually cyanosis may be present. Sometimes, respiratory symptoms progressively worsen associated with acute respiratory infection of the upper airways[6,7]. Contralateral mediastinal displacement can cause displacement of heart sounds, decreased venous return, abnormal ventilation/perfusion ratio and hypoxia[12]. In the case studied, there was persistence and worsening of tachypnea after an episode of acute viral bronchiolitis.

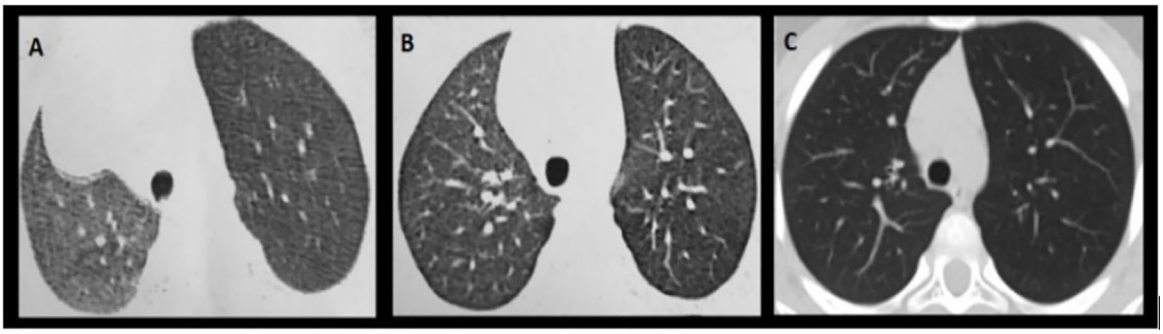


Figure 1. Chest HRCT shows hyperinflated left upper lobe with contralateral mediastinum shift on 02/12/2007 (A), gradual improvement of hyperinflation and shift on 11/24/2008 (B) and 5/15/2015 (C).

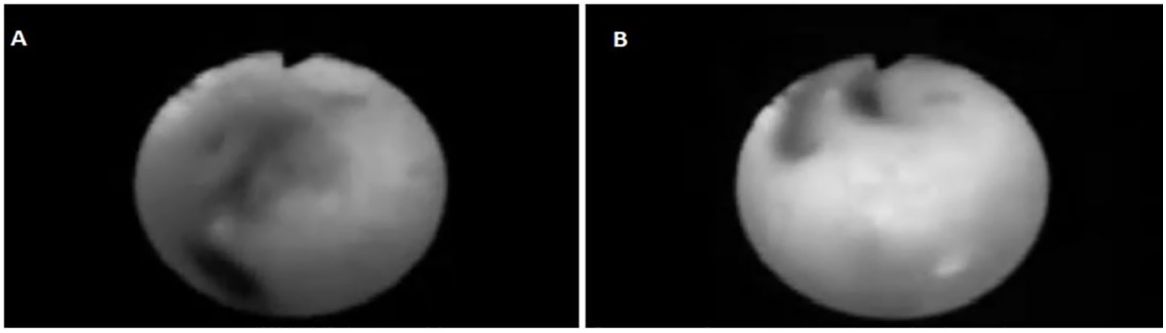


Figure 2. Flexible bronchoscopy shows the normal bronchus orifice of the left lower lobe and, in the arrow, the slit orifice of the upper bronchus (A) which, when crossed, shows the normal distribution of the orifices of the superior lobes and lingula (B).

Chest radiography often reveals hyperlucency in the affected lobe. The mediastinum is contralaterally deviated and may present with progressive ipsilateral atelectasis of the remaining normal lung[1,3,6,7]. In some cases, the diagnosis is confused with pneumothorax. However, a characteristic of ELC is the pulmonary marks throughout the affected lobe that distinguish it from pneumothorax, cystic lesion or diaphragmatic hernia[1,4,9]. HRCT is able to demonstrate an area of localized bronchial abnormality or focal bronchial obstruction, in addition to ruling out other causes of extrinsic compression such as cardiomegaly, vascular ring, cyst lesion or mediastinal mass[1,3,6,11]. Figure 1A shows the HRCT of the chest at the time of diagnosis of the patient in question.

Previous studies have suggested flexible bronchoscopies for all patients eligible for conservative treatment[1,7]. This procedure has the advantage of showing dynamic changes in the airways that cause endobronchial collapse and airway patency distal to the anatomical abnormality. Rigid bronchoscopy can distort the appearance of the airways by physical traction and is not useful[13,14]. In the present case, flexible bronchoscopy demonstrated bronchomalacia, due to alteration in the cartilage at the entrance to the LSE.

In all symptomatic children, surgical resection of the affected lobe was, for a long time, the only indicated and universally accepted treatment modality[5]. However, patients with mild symptoms were treated conservatively, with regular clinical follow-up[1,5-8]. Eigen et al[15] reported that the lung function of non-lobectomized children was similar to those whose approaches had been surgical.

So far, despite some wheezing episodes related to climate change and respiratory viruses, the clinical evolution of this case has been satisfactory. HRCT performed after four years showed improvement in the hyperinflation of the affected lobe, probably related to the growth of the child, with consequent development of the bronchus gauge, reducing the degree of obstruction caused by the cartilaginous defect, in addition to the growth of unaffected lobes (lingula and inferior) (Figure 1).

The demonstration of the cartilaginous defect of the LSL bronchus by bronchoscopy and the satisfactory result reinforced the decision to maintain watchful waiting for our patient.

However, the choice of conservative treatment must be careful, the patient must be closely monitored, as progressive respiratory distress and recurrent respiratory infections may result in the decision for surgery to treat the ELC[1,5-8].

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