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Congenital Lobar Emphysema in an Infant: A Case Report from the University Hospital of Mali

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Abstract

Congenital lobar emphysema is characterized by distress due to hyperinflation of the affected lung lobe(s). We report the case of a 2-month-old infant with congenital left lower lobe emphysema. A 2-month-old female infant, born at term without incident, was hospitalized for respiratory distress. On a postnatal day 15, respiratory distress occurred. On admission, he weighed 3400 g and was apyretic. He had a polypnea of 58 cycles/min and a oxygen saturation of hemoglobin of 90% on room air. A chest radiograph revealed hyperclarity of the left lung. Chest CT revealed left lower lobe emphysema. He underwent a left lower lobectomy. The postoperative course was uneventful. He was discharged from the hospital 7 days after surgery without sequelae. Physicians should be aware that congenital lobar emphysema can present with respiratory distress in infants. A chest CT scan confirms the diagnosis. Surgical treatment is effective.

Keywords

Congenital Lobar Emphysema, Infant, Pediatrics, Mali Hospital

1. Introduction

Congenital lobar emphysema is a pulmonary malformation characterized by progressive distension of one, sometimes several, lobes of the lung, compression, and collapse of the remaining lung parenchyma, and mediastinal shift to the

opposite side [1] [2] [3]. About 50% of cases have no identified etiology. Partial or complete obstruction of the lobar bronchus is likely to be the cause. Many extrinsic or intrinsic phenomena have been described: bronchomalacia, flange, stenosis, vascular compression, and bronchogenic cyst. The lung parenchyma is normal [3]. It is diagnosed in most cases after birth but prenatal diagnosis is possible. Cardiovascular abnormalities are associated in 10 - 15% of cases [1] [3]. Congenital lobar emphysema is a rare bronchopulmonary malformation with a prevalence of 1 in 20,000 to 30,000 [1]. It is an important cause of respiratory distress in the neonatal period [2] [4]. It remains a diagnostic and therapeutic challenge in developing countries because of the low level of technical facilities [5]. The left upper lobe is most often affected followed by the middle and right upper lobe. Involvement of the left lower lobe is rare (1%) [4]. We report a clinical case of congenital lobar emphysema of the left lower lobe in a 2-month-old infant diagnosed and treated in the pediatric department of the University Hospital of Mali.

2. Observation

This was a 2-month-old female infant whose parents had no known medical or surgical history. There was no notion of consanguinity in their marriage. His mother had 2 pregnancies and has 2 living children. Our infant is the 2nd child of the couple from a pregnancy well followed until term without major incidents. She was born in natural way without any notion of resuscitation at birth. Her birth weight was 2500 g.

According to her mother, the onset of symptoms was due to an influenza syndrome treated with oral amoxicillin/clavulanic acid without success at 45 days of age. The parents were concerned about the progressive onset of respiratory discomfort and whining. They decided to bring her to the pediatric department of the Gabriel Touré University Hospital, where an emergency chest X-ray showed hyperclarity of the left hemithorax (**Figure 1**). She was referred to us for management.

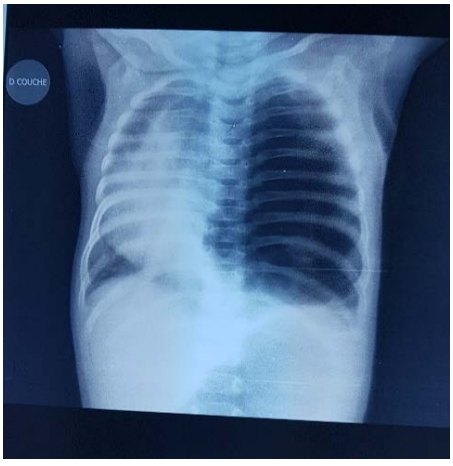

Figure 1. Supine chest X-ray showing left avascular hyperclarity with right mediastinal deviation.

On admission, she had a temperature of 36˚C. She weighed 3400 g with a height of 52 cm and a head circumference of 38 cm. There was no cyanosis or pallor.

Respiratory: the thorax was symmetrical. She had polypnea with a respiratory rate of 62 cycles/min and hemoglobin oxygen saturation (oxygen saturation of hemoglobin) of 78% under air, intercostal pulling, thoraco-abdominal rocking. On pulmonary auscultation, the vesicular murmur was abolished on the left.

Cardiovascular: there was a regular tachycardia at 180 beats per minute without murmur. The rest of the clinical examination was normal. She was admitted to the hospital and put on fasting and oxygen (sufficient amount for oxygen saturation of hemoglobin ≥ 95%) after disobstruction of the nasal cavity. She was infused with 10% glucose serum at a rate of 100 ml/Kg/d.

To further explore the hyperclarity revealed by standard radiography, a chest CT scan was ordered. It showed a bulky bullous partitioned image occupying almost the entire left lung field associated with a displacement of the mediastinum to the right by mass effect in favor of a left congenital lobar emphysema (**Figure 2**).

Other complementary examinations were ordered, including:

- A cardiac ultrasound to look for an associated heart defect came back normal.
- A blood count without abnormalities: hemoglobin (14.3 g/dl), hematocrit (42.1%), white blood cells (7.2000/mm^3), platelets (69,000/mm^3 with platelet aggregates).
- An undisturbed blood ionogram: sodium (133 mmol/l), potassium (5.1 mmol/l), chlorine (95 mmol/l), magnesium (1.06 mmol/l) and phosphorus (18.5 mmol/l).

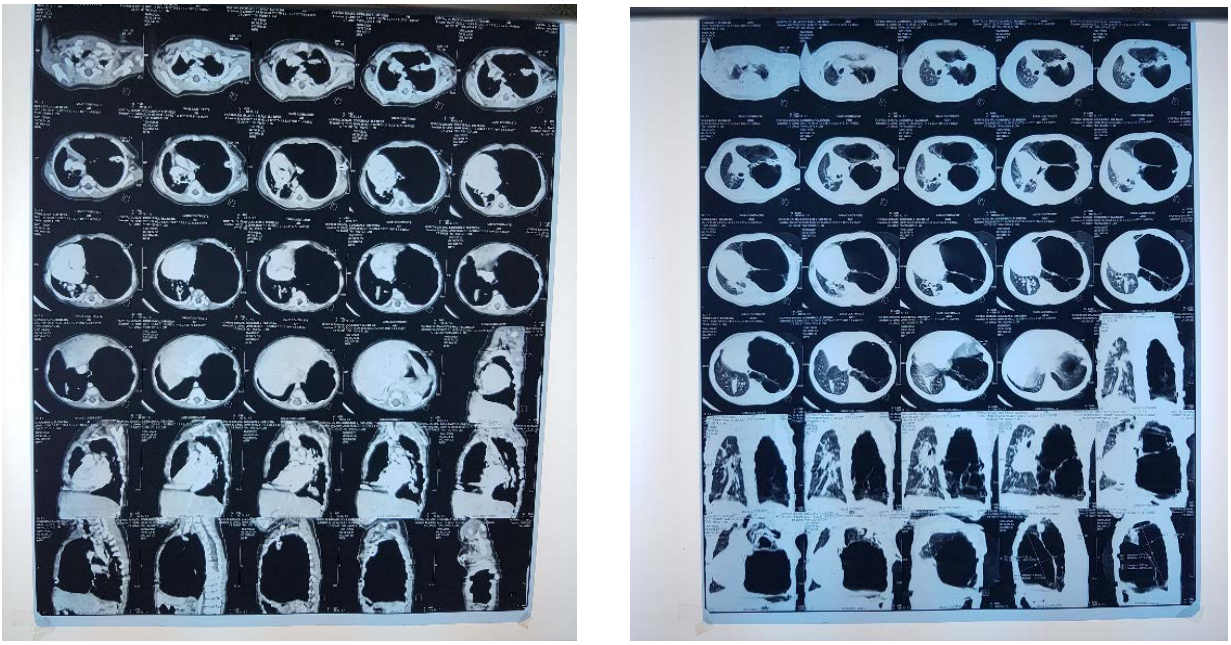

Figure 2. These sections show a voluminous compartmentalized bullous image occupying almost the entire left lung field (with a stump of parenchyma at the apical level) associated with a displacement of the mediastinum to the right by mass effect and a compensatory hypervascularization of the right lung parenchyma evoking a compressive left lower bullous dysplasia.

- C-reactive protein was equal to 0.23 mg/ln, blood glucose: 6.9 mmol/l.
- Prothrombin time: 100%, activated partial thromboplastin time: 33 seconds, and Rh group: O positive.

She was operated on in the 3rd week of hospitalization by the thoracic surgery department. She underwent a left lower lobectomy.

The immediate postoperative course was relatively calm with the good general condition, respiratory rate at 40 cycles/minute, hemoglobin oxygen saturation at 88% on air, and heart rate at 146 beats/minute. She was fasted and on oxygen for a sufficient amount of hemoglobin oxygen saturation ≥ 95%. Medical treatment including 10% glucose serum at 100 ml/Kg/d, paracetamol injection 60 mg/Kg/d slow IV, cefotaxime 50 mg/Kg/8h direct IV for 7 days was initiated.

After one week of treatment, she was in good general condition, hemoglobin oxygen saturation 99% on air, respiratory rate 36 cycles/minute. Feeding was resumed.

On the 25th day of hospitalization, she was discharged with a weight of 4000 g. She was on -air and apyretic. Her respiratory and heart rates were normal. The control frontal chest radiograph was normal (Figure 3).

On the tenth-day post-discharge visit, she had no complaints. Her nutritional status was good, the temperature was 37˚C. Her chest was symmetrical with no signs of respiratory struggle. Her respiratory rate was 55 cycles/minute. She was on the air. The surgical wound was clean and healing. Cardiorespiratory auscultation was normal.

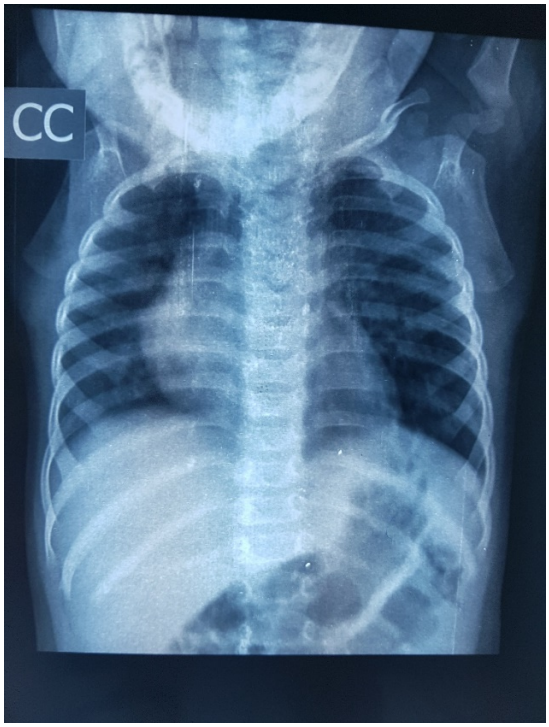

Figure 3. Frontal chest X-ray, supine, 1st control after left lower lobectomy on the 25th day of hospitalization: showing expansion of the left lung parenchyma (upper lobe) and return of the mediastinum to its place.

On the fourth-month post-discharge visit, she had no complaints. She was apyretic and weighed 5000 g. The control chest X-ray was normal.

3. Discussion

Congenital lobar emphysema is characterized by progressive distension of one lobe, sometimes two lobes [2]. It is a rare disease with an annual incidence of 1/20,000 to 1/30,000 births [1]. Most cases occur during the neonatal period or early infancy. Less severely affected patients may present in early childhood, or more rarely, in adulthood [1]. The clinical picture varies from asymptomatic or mildly symptomatic disease to severe respiratory failure requiring immediate intervention. Symptomatic patients present with respiratory distress, dyspnea, tachycardia, cyanosis, and failure to thrive [1]. The left upper lobe is most frequently affected (41% of cases), followed by the right middle lobe (34% of cases) and the right upper lobe (21% of cases), bilateral forms: 3%, and the right or left lower lobes: 1%. Congenital lobar emphysema is associated with cardiac defects in 15% - 20% of cases [1] [4] [6].

About 50% of cases have no identified etiology. However, the disease could be explained by malformation or absence of cartilage rings, intrinsic obstruction by excess mucus, extrinsic obstruction (vascular or bronchial), or hyperinflation alone. A polyalveolar lobe could also be involved [1] [4] [7].

Chest X-ray and chest CT scan are the primary diagnostic imaging methods. Radiography reveals hyperclar-affected lobes, mediastinal collapse, and collapse of unaffected ipsilateral segments. CT scan reveals affected lobes and vascular involvement [1]. Differential diagnoses also include congenital pulmonary airway defect, pneumonia, bronchiolitis, and foreign body inhalation. A prenatal diagnosis can be made by ultrasound by identifying hyperechoic areas of the fetal lung that is not always specific for congenital lobar emphysema [1]. Lobectomy of the affected lobes is the most widely accepted treatment with satisfactory efficacy. Asymptomatic or mildly symptomatic cases can be managed conservatively, but follow-up is required. Thoracoscopic resection appears to be more effective postoperatively [1] [4]. Asymptomatic cases may regress spontaneously. The long-term prognosis is usually good for asymptomatic cases, if diagnosed early and operated on. However, the disease is sometimes lethal [1] [4].

We report a case of left congenital lobar emphysema in a 2-month-old female infant. The age of onset of clinical signs of congenital lobar emphysema ranges from 2 weeks to six months [8]. Our infant presented with his first symptoms at 45 days of life. He was a female infant as reported by Souleymane Diatta *et al.* [9] in their series and contrary to most series where the male/female ratio was 3/1 [8]. Respiratory distress was the mode of the revelation of the disease in our infants as described in the literature and by most authors [1] [5] [6]. The frontal chest X-ray showed a left hyperclarity with a deviation of the mediastinum to the right orienting towards left lobar emphysema confirmed by the thoracic scanner specified the involvement of the left lower lobe. This location represents 1% of

congenital lobar emphysema [4]. Our clinical form is slightly different from that of Badiu *et al.* [10], Latif *et al.* [11], and Felipe *et al.* [12] who reported left upper lobe involvement. It is also different from that reported by Tibana *et al.* [6] who found emphysema in the right middle lobe. We performed a successful left lower lobectomy. This technique is the one performed by most authors for symptomatic neonates or infants [4] [13].

4. Conclusion

Congenital lobar emphysema is an uncommon condition. Left lower lobe involvement is a rare clinical form. The diagnosis of congenital lobar emphysema is difficult in pediatric departments. Therefore, it should be considered in neonates or infants in respiratory distress. Chest X-ray and, preferably, chest CT scan are essential tools for its diagnosis. Surgical treatment is effective and does not carry any risk. The younger the child, the more often it is performed.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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