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Case Report

Congenital lobar emphysema case report: A frequently misdiagnosed disease

Rahaf Ibrahim [a,*], Sawssan Ali [b], Bassam Darwish [c]

[a] Pediatric Department, Damascus University Pediatric Hospital, Damascus, Syria
[b] Pulmonary Pediatric Department, Damascus University Pediatric Hospital, Damascus, Syria
[c] Thoracic Surgery Department, Damascus University Al Mowasat Hospital, Damascus, Syria

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ABSTRACT

Introduction and importance: Congenital Lobar Emphysema is a rare cystic lesion of the lung which may be misdiagnosed and managed as pneumonia or pneumothorax.
Case presentation: We presented a case of a congenital cystic lung malformation, the case demonstrated a 29-day-old boy who presented with respiratory distress. He was initially diagnosed as pneumonia and was given unnecessary antibiotic treatment in another hospital without improvement. Subsequently, the diagnosis of congenital lobar emphysema of the left upper lobe was made on the basis of the clinical and radiologic features. The condition wasn't detected before birth. He underwent a successful left thoracotomy with left upper lobe lobectomy.
Clinical discussion: Congenital Lobar Emphysema poses a challenge in diagnosis. It may mimic other causes of respiratory distress.
Conclusion: Congenital Lobar Emphysema requires a high index of clinical and radiological suspicion to make an early diagnosis so that timely treatment will be offered.

1. Introduction

Congenital cystic lung lesions include congenital cystic adenomatoid malformations (CCAM), pulmonary sequestrations, congenital lobar emphysema (CLE), and bronchogenic cysts [1]. CLE is the overinflation of one or more lung lobes. When the lobe increases in size, it compresses and displaces mediastinal components and the rest of the lung. Clinical presentation depends on the degree of hyperinflation of the affected region [2].

Diagnosis requires a strong clinical suspicion. We describe a case of a 29-day-old boy with CLE who was initially diagnosed with pneumonia. We report this case because of its rarity. CLE is generally misdiagnosed and managed as pneumothorax or pneumonia. This study aims to contribute toward a greater understanding of CLE. This anomaly should be kept in mind in differential diagnoses of infants presenting with respiratory distress and must be diagnosed as early as possible to give timely and appropriate treatment.

All our cases has been reported in line with THE CARE 2017 guidelines [3] and THE SCARE 2020 criteria [4].

2. Case report

A 29-day-old, term, male child, weighing 3.5 kg, presented to our pediatrics emergency ward with progressively increasing respiratory distress for seven days and had worsened two days ago. The child was treated in another hospital and was referred to us. The mother gave a history of fatigability and diaphoresis while feeding. There was no history of cough or fever. The child was admitted in another hospital and was treated for pneumonia. Chest X-ray at that time showed collapse consolidation of the apical segment of the right lung which was misread as lobar consolidations. He was given intravenous antibiotics for seven days without improvement and hence referred to our hospital for further management and examination. During this course, her blood investigations were essentially normal. Past history was unremarkable except mother noticed occasional episodes of fast breathing and cyanosis. Antenatal history of mother was uneventful. Post natal period was unremarkable. Mother had several antenatally ultrasound scans but no lung pathology was detected or reported. The initial physical examination revealed respiratory distress with rapid shallow breathing using the accessory breathing muscles. Inter-costal and sub-costal

* Corresponding author. Damascus University Pediatric Hospital, Damascus, Syria.
E-mail addresses: Dr.r.e345@gmail.com (R. Ibrahim), Dr.Sawssanali@gmail.com (S. Ali), bdarwish1956@gmail.com (B. Darwish).

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recession was evident. He was irritable and restless but not febrile. The heart rate was 160 beats/min, respiratory rate was 70/min and maintaining oxygen saturation of 88% on room air. He was kept under observation with supplemental oxygen using a hood. Examination of respiratory system revealed decreased air entry on auscultation, but more on the left than right, and heart sounds were shifted to right side. Other systems appeared normal. Chest X-ray showed marked hyperinflation of left upper lobe with contralateral mediastinal shift and collapse of the ipsilateral remaining lung field, and a portion of the left lung is herniated across the midline (Fig. 1 [a]). For further evaluation Chest CT was advised which gave findings of hyperinflation and poor vascularization of left upper lobe with compressive atelectasis of the lower lobes and subsequent contralateral mediastinal displacement to the right side (Fig. 2). Echocardiography and abdominal ultrasound revealed normal findings. A diagnosis of CLE of the left upper lobe was made on the basis of the typical clinical and radiologic features. The child was scheduled for left upper lobectomy. The patient was taken to the operating room. He underwent left thoracotomy with left upper lobe lobectomy. The operation was performed uneventfully, showing a grossly overdistended left upper lobe. Intra-operatively the vital parameters were stable. Post operatively patient remained vitally stable. He fed well, and his tachypnea and recession improved significantly. Post-operative X-ray of chest revealed expanded right lung and left lower lobe with sings of left upper lobectomy, no mediastinal shift was noted (Fig. 1 [b]). The child was discharged on the third post-operative day. Macroscopically, the specimen composed of pulmonary lobe measuring (9 × 5 × 2) cm. Microscopic findings demonstrated large alveoli lined by flattened epithelium, focal presence of intra-alveolar hemorrhage, with focal small alveoli, fibrotic walls, presence of chronic inflammatory infiltrate. No evidence of granulomatous formation, proliferated histiocytes or tumoral proliferation (Fig. 3). With these findings, the diagnosis of congenital lobar emphysema was confirmed. On follow-up, he continues to feed well and is thriving nicely with appropriate growth for age.

3. Discussion

CLE is a rare malformation of lung development, its estimated incidence is approximately 1/20,000–1/30,000. It often occurs in the neonatal period with hyperinflation of one or more pulmonary lobes. The left upper lobe is the most frequently involved lobe [5]. In our case, the affected lobe was the upper left lobe. The etiologies of CLE are either intrinsic included bronchomalacia and mucosal plugging or they are extrinsic included vascular malformation, tumors and cysts, the factors that leading to a subtotal closure of the bronchus and valve effect with air trapping in the affected lobe [6]. The spectrum of clinical symptoms of CLE is broad ranging from very mild symptoms to acute respiratory distress syndrome and depending upon: the age of the patient; size and location of the anomaly; and associated malformations. Symptoms can start at birth in 25% of cases, within the first month of life in 50% of cases or sporadically after 6 months of age [7]. In our case the patient presented with respiratory distress, the symptoms started within the first month of life. CLE may be misdiagnosed as pneumothorax and in such cases insertion of an intercostal drainage tube may be done. On the other hand, many children may be misdiagnosed as pneumonia leading to unnecessary antibiotic treatment and loss of valuable time in diagnosis. A retrospective analysis was done on forty patients with cystic lung lesions. The final diagnosis was CCAM in 19 patients, CLE in 11 patients, and bronchogenic cysts and pulmonary sequestration in 5 patients each. Of these, 20% had received a course of prior antitubercular therapy and 30% had an intercostal drain inserted [8]. Prabhu SM et al. reviewed 54 children operated for congenital cystic lung lesion, 15 children (27%) with an inadvertent chest tube insertion preoperatively based on plain radiograph findings. The patients comprising 10 cases of CCAM and 5 cases of CLE. CCAM was misdiagnosed as complicated pneumatocele and pneumothorax, while CLE was misdiagnosed as tension pneumothorax. They concluded that chest tube insertion in congenital cystic malformations increases the rate of associated complications, and chest CT has a definite role in early diagnosis and deciding appropriate management in these cases [9]. In a 30-year case series in two university hospitals, 20 children with CLE underwent surgery. One patient with severe respiratory distress was misdiagnosed with hypertensive pneumothorax and underwent chest tube drainage [10]. Ten children with CLE were reviewed in order to emphasize the importance of differential diagnosis with pneumothorax. In this series, three patients were mistakenly diagnosed as pneumothorax and intercostal drains were inserted in the emergency department [11]. In our case, the child was being treated for pneumonia without improvement until the right diagnosis was established, and he received the appropriate treatment. The prenatal diagnosis of CLE is established through the ultrasound scan or MRI. With these investigations CLE can be differentiated from other congenital lung lesions. Postnatal diagnoses are made directly after birth in approximately 25–50% of all cases. Where the diagnostic tools are chest X-ray and CT scan. Echocardiography is essential to exclude cardiac anomalies [12]. In our case, the condition wasn't detected before birth. The diagnosis of CLE was made after birth on the basis of the clinical symptoms and radiologic features, and was confirmed with histologic examination. Echocardiography was normal. Cardiac anomalies occurring in 14–20% of the patients with CLE. Other systems anomalies (renal anomalies, musculoskeletal anomalies, gastrointestinal tract, and others) and syndromes (Williams–Beuren syndrome, Miller–Dieker syndrome, Niemann–Pick disease, Fanconi aplastic anemia)

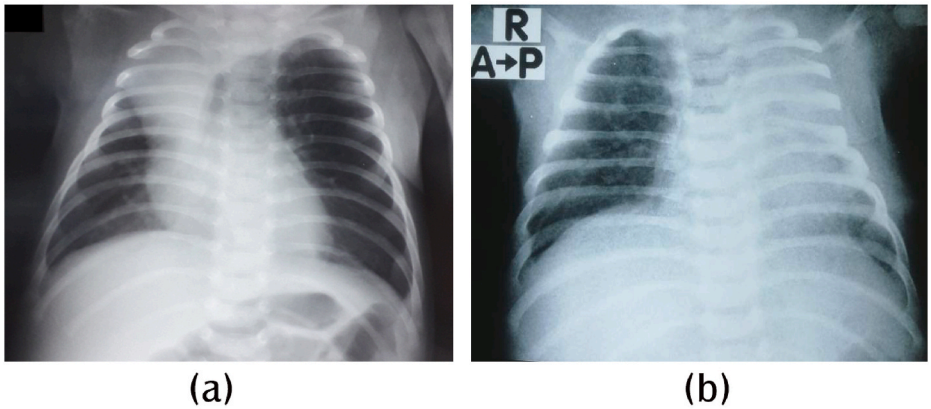

Fig. 1. (a) Chest X-ray showed marked hyperinflation of left upper lobe with contralateral mediastinal shift and collapse of the ipsilateral remaining lung field, and a portion of the left lung is herniated across the midline. (b) Post-operative X-ray of chest revealed expanded right lung and left lower lobe with sings of left upper lobectomy, no mediastinal shift was noted.

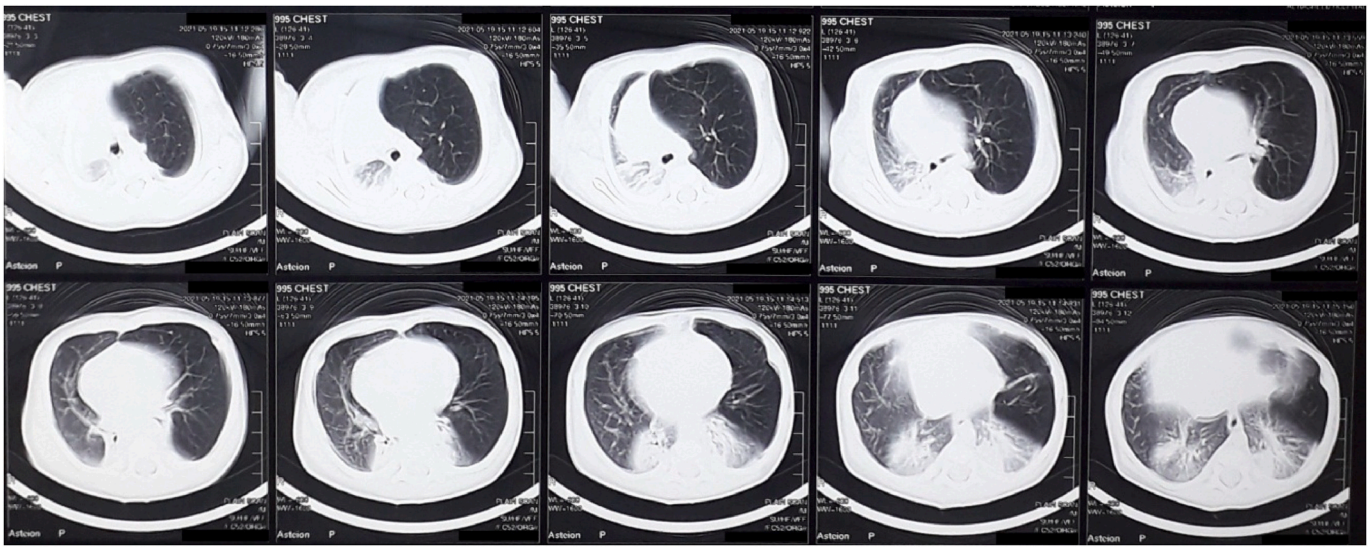

Fig. 2. Chest CT was advised which gave findings of hyperinflation and poor vascularization of left upper lobe with compressive atelectasis of the lower lobes and subsequent contralateral mediastinal displacement to the right side.

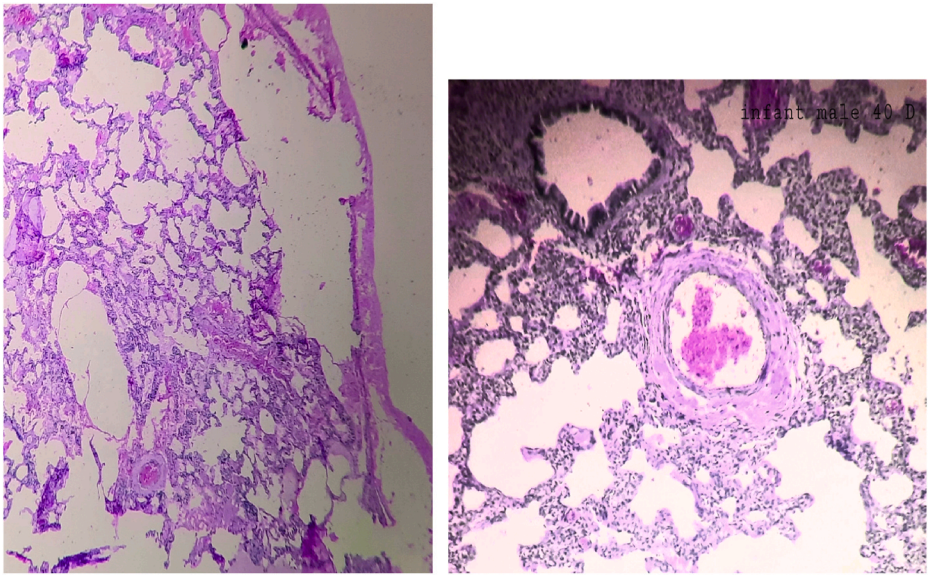

Fig. 3. Microscopic findings of congenital lobar emphysema.

may accompany CLE rarely [13]. In our case, there wasn't any other systems malformations or accompanied syndromes. Management of patients with CLE consists of dealing with the underlying mechanism. Those with minor symptoms may be treated expectantly whereas those who more severely affected will require lobectomy, which is often curative and it allows the surrounding compressed lung to expand [14]. In our case the decision of surgical treatment was made after a case discussion with pediatric pulmonologist and thoracic surgeon.

4. Conclusion

A high index of clinical and radiological suspicion is required to diagnose this rare anomaly which may mimic other causes of respiratory distress. Radiologic investigations are helpful adjunct in making the diagnosis. Early identification and proper management can lead to dramatic improvement in the clinical condition of the patient.

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Ethical approval

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

Consent

The patient's mother consented to the publication of this deidentified case report.

Author contributions

All authors contributed to the development of the manuscript and the care of the patient presented. All authors approved the final manuscript.

Trial registry number

None.

Guarantor

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Declaration of competing interest

None.

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