CLINICAL CASE

Congenital lobar emphysema in right upper and middle lobes in a 2-month-old female patient

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ABSTRACT

Background. Bronchopulmonary malformations demonstrate a low frequency in the population. Their presentation is sporadic and may be associated with malformations in other systems. Within this group is congenital lobar emphysema, which is a developmental anomaly of the lower respiratory tract manifesting as hyperinflation of one or more lung lobes. Among its causes, entrapment of air through a valve mechanism at the bronchus or gigantism by overdevelopment of lobular alveolar lung parenchyma has been proposed.
Case report. We report the case of a 2-month-old female patient who from birth presented with respiratory disease classified as pneumonic process. The patient was treated with multiple antibiotic schemes as well as placement of pleural seals complicated by pneumothorax and pleural effusion. Subsequently, diagnosis of congenital lobar emphysema was made and supported by laboratory studies. Surgery was carried out where involvement of the middle and upper lobes of the right lung was demonstrated.
Conclusions. The patient presented a favorable evolution with a good expansion of residual right lung and correction of mediastinal positions.

Key words: congenital lobar emphysema, congenital pulmonary malformation, respiratory difficulty, posterolateral thoracotomy.

INTRODUCTION

Congenital lobar emphysema (CLE) is a congenital lung malformation characterized by partial obstruction of the airways. The bronchi allow entry of the inspired air but, during exhalation, the bronchi collapse with subsequent hyperinflation of one or more lobules of the normal pulmonary structure. This overinflation causes a focal retention of air upon exhalation, with the consequent distention of the affected lobe and compression of adjacent structures. According to the theory suggested by Hislop and Reid, the defect would result in a poor development of cartilage (bronchomalacia) that sustains the bronchus of the affected lobe or in an intraluminal obstruction.[1-3]

In half of the cases, no cause is found and in the other half of the cases there is intraluminal obstruction, either by folds of the bronchial mucosa, by rotation, by bronchial stenosis or granulation tissue being the most common cause. In 2% of cases the cause is extrinsic, due to vascular rings or mediastinal lymphadenopathy. A second theory proposes as a mechanism of CLE the overgrowth of the alveoli in the affected area. This theory is based on pathological findings of pulmonary acini with a large number of alveoli of normal size without the presence of airway abnormalities (polyalveolar form).[2]

CLE comprises 1.4 to 2.2% of all congenital malformations. The incidence is reported as 1 case per 20,000 to 30,000 live births and predominates in males (ratio 3:1).[4-6] The most-often affected lobes are the left upper lobe (47-50%), right middle lobe (28-24%), right upper lobe (18-20%) and lower lobes (5%).[4,5] CLE is associated with other congenital malformations in 14-40% of cases, the most common being patent ductus arteriosus, interventricular septal defects, diaphragmatic hernia and renal malformations.

Symptoms usually appear at birth in 33% of cases, 50% during the first month and most before 1 year of age. The

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clinical picture is that of any lesion that occupies the chest and, depending on the severity of the malformation, may present as progressive respiratory distress rapidly evolving in the first months of life or demonstrate a more gradual and insidious onset and may present no symptoms for years. It usually affects children who present with progressive dyspnea or respiratory insufficiency. The severity depends on the size of the overdistended lobe, compression of lung tissue around it and the degree of displacement of the mediastinum.

CLE is one of the causes of unilateral hyperlucid lung; therefore, AP and lateral chest x-rays are essential in the diagnosis and monitoring of these entities. On chest x-ray we can observe an overdistention of the lobe or affected lobes and bronchovascular ill-defined lines in its interior, atelectasis of the contralateral lung, flattening of the ipsilateral hemidiaphragm and contralateral mediastinal shift. High-resolution chest computed axial tomography (CAT) demonstrates emphysematous area and compression of the affected parenchyma.[7,8]

Lobectomy has been the traditional treatment in patients with severe symptoms of CLE or of neonatal onset. At present, conservative treatment is more acceptable in asymptomatic children of any age or who demonstrate mild or moderate respiratory symptoms, especially with a normal bronchoscopic examination. In these cases, treatment is based on observation and close monitoring becauses many involute. Conservative management in children can be carried out under strict monitoring, providing information about the disease and its complications to the parents and training them to identify alarm data. In case of persistent respiratory symptoms, elective lobectomy should be performed, thus avoiding increased risks for the patient.[9]

Clinical Case

We present the case of a 2-month-old female who, since her birth in the ISSSTE Regional Hospital in La Paz, Baja California, Mexico, presented a picture characterized by severe shortness of breath accompanied by cough, expiratory stridor, central cyanosis during feeding, as well as the presence of intercostal and thoracoabdominal dissociation, which was classified as a pneumonic process. The process was managed with multiple schemes of antibiotics and with the placement of a pleural seal device twice to solve an alleged pneumothorax and a pleural effusion as a complication of the infectious process.

Radiological studies were conducted where we observed on the plain chest x-ray a hyperlucent image and air trapping in the right lung, atelectasis in the contralateral lung and mediastinal shift to the left side (Figures 1 and 2). The study was complemented with chest CT image.

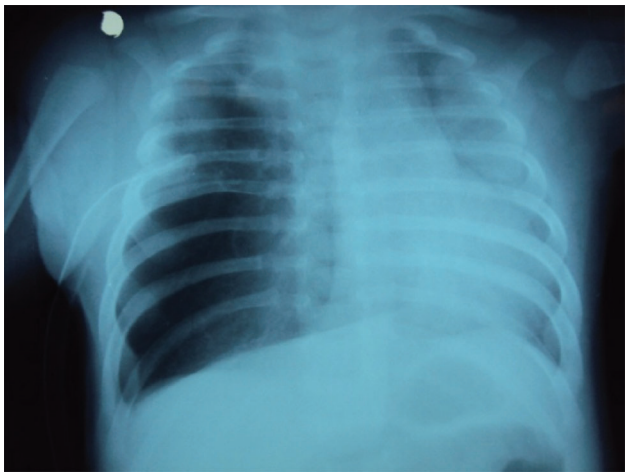

Figure 1. Right hyperinflated hemithorax with hyperlucency and deviation of the mediastinum toward the contralateral side; presence of left apical atelectasis. Horizontalization of the costal arches and abatement of both hemidiaphragms.

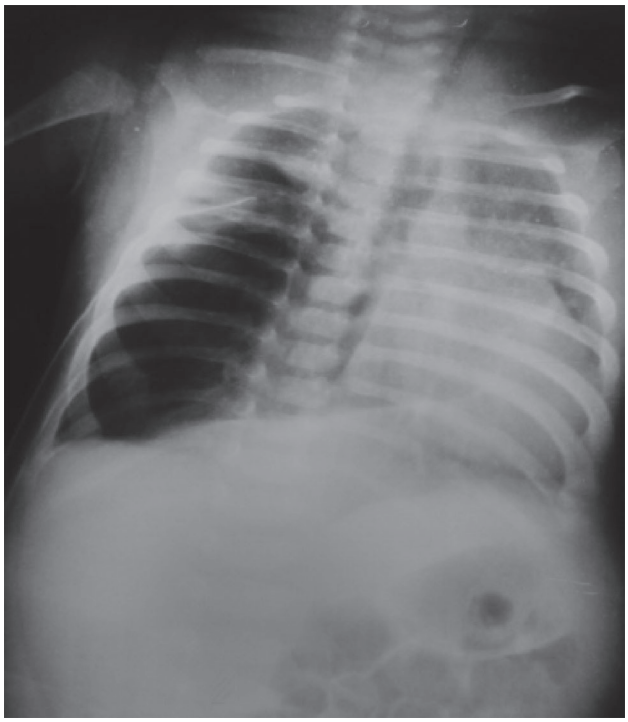

Figure 2. Hyperdistention of the right lung and deviation of the mediastinum towards the left side, which persisted after placement of a pleural catheter.

On the superior cuts an apparent emphysematous zone was demonstrated in the right upper lobe (Figure 3) along with normal lung parenchyma in the lower cuts, images compatible with right CLE. The decision was to transfer the patient from the Regional Hospital of Baja California to a highly specialized hospital in Mexico City for surgical correction of the condition. However, during the airlift, the patient had a cyanotic event and respiratory difficulty increased; therefore, the patient was urgently sent to Guadalajara, Jalisco, Mexico and transferred to Hospital Regional Dr. Valentín Gómez Farías (HRVGF).

At the HRVGF, initial physical examination revealed a patient with mild respiratory distress, tachypnea, intercostal retractions and nasal flaring, with a healing chest tube opening at the right rib cage (placed during initial management at the referral hospital), with supplemental oxygen via facial mask with 90% saturation. Paraclinical tests were carried out with the following results: laboratory blood results: hemoglobin (Hg) 11.6 g/dL, hematocrit (Hct) 34%, white blood count (WBC) 9.770, 56.1% neutrophils, lymphocytes 35.8%, monocytes 5.4%, and platelets 376,000. Blood chemistry reported glucose 96 mg/dL, BUN 12 mg/dL, urea 25 mg/dL, and creatinine 0.41 mg/dL. Serum electrolytes reported Na 139.4 mmol/L, K 5.1 mmol/L, and Cl 102.9 mmol/L. Clotting times reported prothrombin time (PT)12 sec, partial thromboplastin time (PTT) 25.9 sec, and fibrinogen 333.

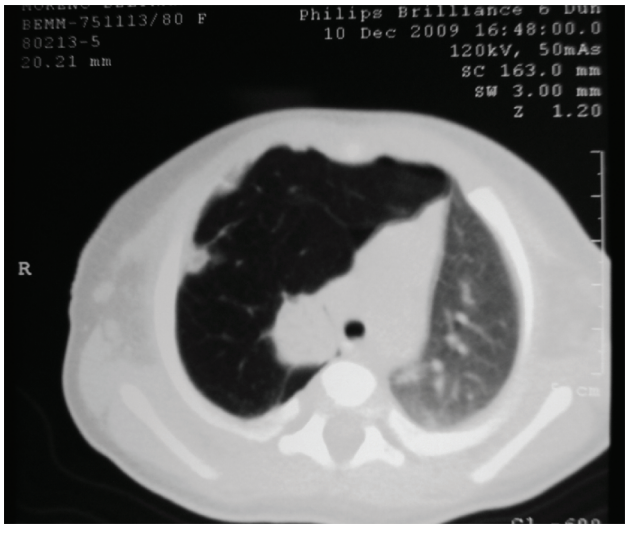

Figure 3. Apical cut tomography. Increase in volume of the right lung and emphysematous parenchyma with deviation of the contralateral lung.

The patient was referred for consultation to the Pediatric Surgery Service who confirmed the referring diagnosis. The patient was then scheduled for an elective surgical procedure. Right posterolateral exploratory thoracotomy was performed with findings of hyperinflated right upper and middle lobes protruding from the surgical wound despite the loss of negative pressure in the chest cavity. The lower lobe demonstrated normal characteristics and was collapsed. Middle and superior lobectomy was performed. The lower lobe was expanded with positive pressure, occupying 60% of the capacity of the chest cavity. The procedure was completed by placing a tube thoracostomy Pleur-Evac system.

In the Pediatric Intensive Care Unit (PICU) during the immediate postoperative period, the patient presented blood loss through the thoracotomy (~200 mL). Samples were taken and a PT of 18.3 sec and PTT that did not clot were reported. A second surgical exploration was required and bleeding from intercostal vessels was found. Hemostasis was performed to control bleeding.

During the immediate postoperative period the patient presented with coagulopathy and a sepsis process that required intensive management with mechanical ventilatory support, antibiotics (cefepime, amikacin) and correction of hematological abnormalities with vitamin K, fresh frozen plasma and packed red blood cells. The patient had high blood pressure for age requiring management with captopril and spironolactone. Pediatric Cardiology Service performed an echocardiogram with findings of a normal-appearing heart with mild pulmonary hypertension.

The patient progressed satisfactorily. The residual right lung had good postsurgical expansion and the mediastinal structures were centralized (Figure 4). Thoractomy catheter was removed on the sixth postoperative day. Sepsis and blood abnormalities were also corrected. Subsequently, the patient was discharged from the PICU to the pediatric floor where she completed antimicrobial treatment and eventually was discharged to home.

Surgical specimen demonstrated pink-colored lobules and purple areas of emphysematous appearance (Figure 5). Histopathology of the surgical specimen reported multiple dilations of the airspace corresponding to bronchioles, alveolar ducts and alveoli with some lymphoid nodular hyperplasia bronchi as well as areas of atelectasis (Figure 6), interstitial pneumonitis (Figure 7), xanthogranulomatous reaction to foreign body (Figure 8), presence of emphysema (Figure 9) and hemorrhagic areas (Figure 10).

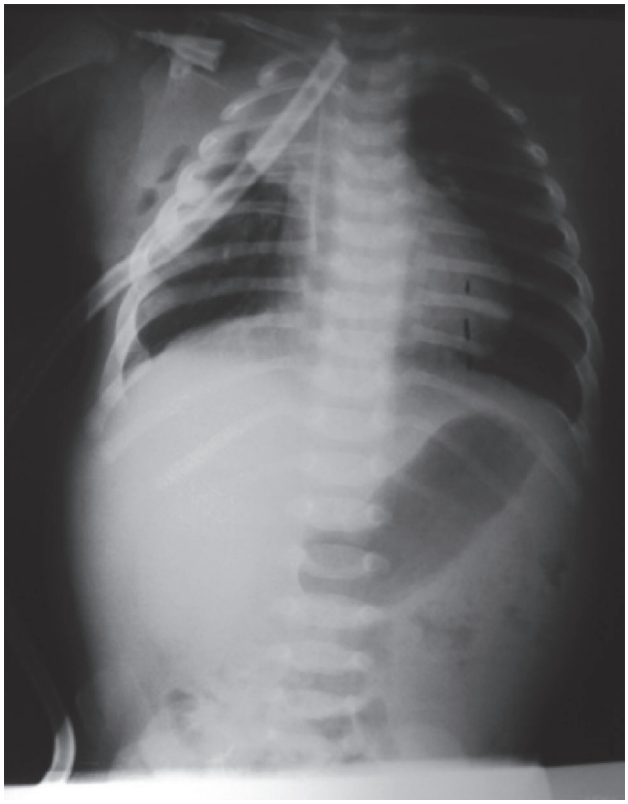

Figure 4. Expansion of the residual right lung and centralization of mediastinal structures after surgery.

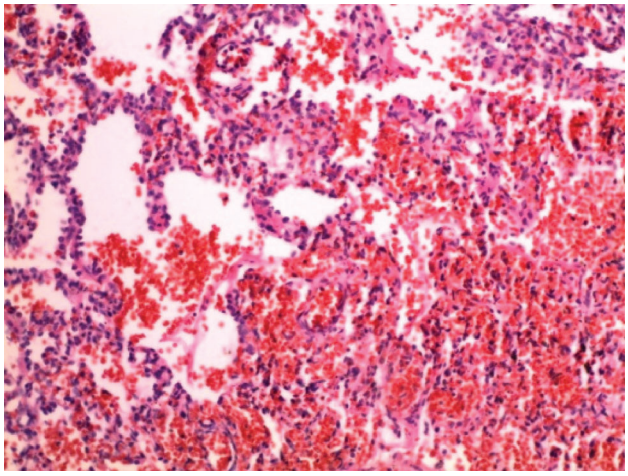

Figure 5. Macroscopic images of right superior and middle pulmonary lobes. Increase in volume of the lobes as well as an emphysematous appearance is demonstrated.

DISCUSSION

Treatment of CLE varies depending on the clinical condition of the patient.[10-12] An alternative is the expectant management in asymptomatic patients without cardiovascular decompensation or respiratory compromise, for whom a strict clinical and radiological follow-up must be maintained. Follow-up time varies in different studies reported with the longest being 17 years, demonstrating a decrease in hyperexpansion of the affected lobe.[8]

Another alternative is surgical management consisting of resection of the affected lung segment through exploratory thoracotomy and/or thoracoscopy. This is indicated

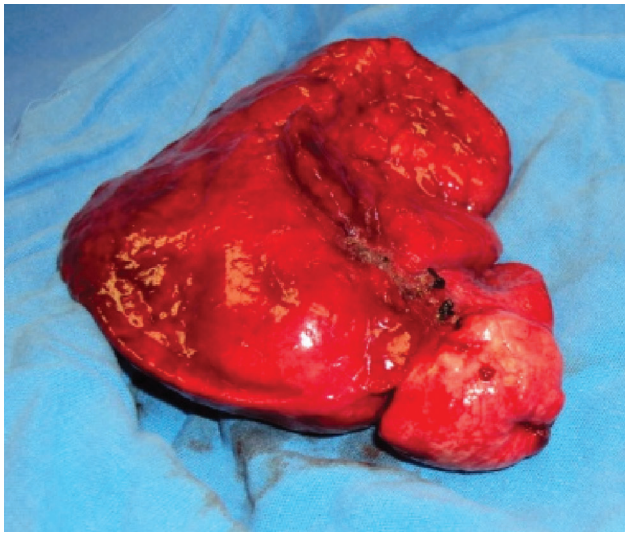

Figure 6. Microscopic histopathological image where zone of atelectasis is observed along with deformity and compression of the alveolar spaces and walls with appearance of intraluminal pneumocytes.

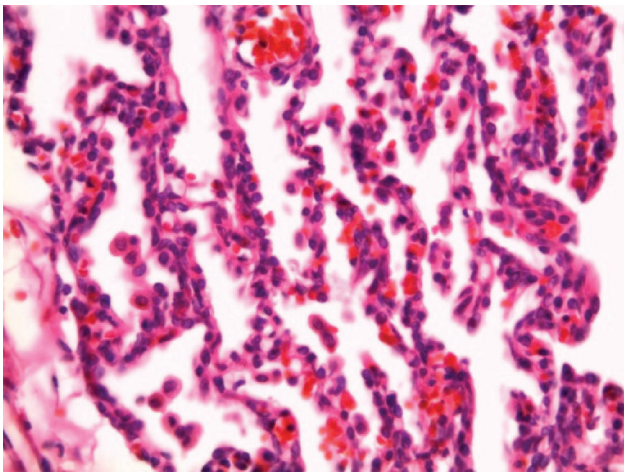

Figure 7. Microscopic histopathological image where interstitial pneumonitis is seen. Alveolar walls show proliferation of fibroblasts, fibrocytes and collagen as well as blood vessels with lymphocytic infiltration.

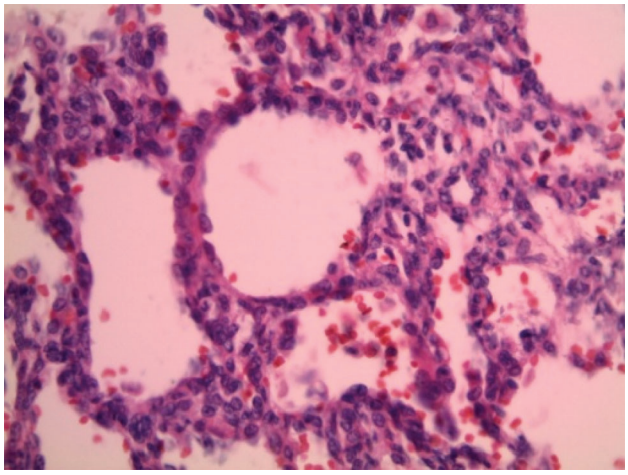

Figure 8. Microscopic histopathological image where xanthogranulomatous reaction to foreign body is noted. Anticoagulation tissue necrosis, multinucleated giant cells and lipid vacuoles.

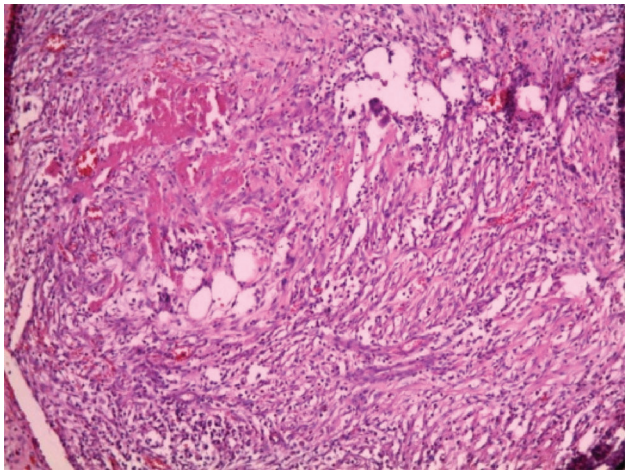

Figure 9. Microscopic histopathological image where the presence of emphysema is appreciated. Panoramic view of alveoli with rupture of walls and formation of sacular structures of variable size and shape.

in symptomatic patients with respiratory compromise and/or cardiovascular decompensation. Therefore, in this case surgical treatment was decided upon, which was carried out successfully with a favorable outcome, presenting good residual right lung expansion and correction in the position of the mediastinal structures. The only occurring sequel was mild pulmonary hypertension.

It should be noted that the importance of this case is twofold: 1) the low incidence of ipsilateral bilobular CLE because no other case was found to be previously reported in the Mexican literature, and 2) the importance of timely diagnosis, avoiding confusion with the presence

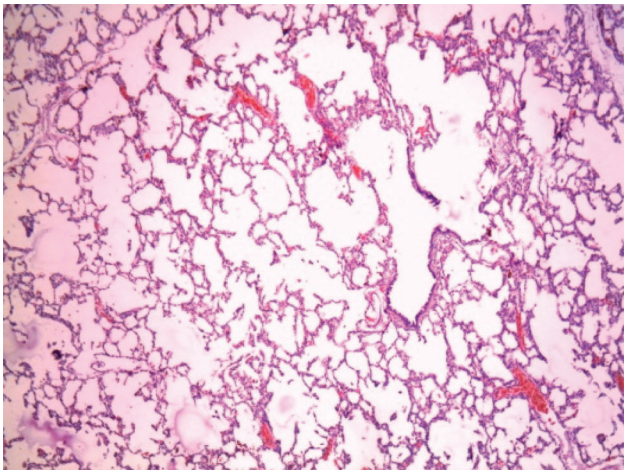

Figure 10. Hemorrhagic zone. Alveoli sac shows distention at the expense of extravasated erythrocytes and intraluminal proteinaceous material with loss of architecture, with functional residual alveolar sacs in the left upper quadrant.

of pneumothorax, pneumomediastinum or pleural effusion, thereby avoiding unnecessary placement of chest tubes, which may cause many complications.

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