Congenital Lobar Emphysema

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23.1 Introduction

Congenital lobar emphysema (CLE) is a pulmonary congenital malformation characterized by hyperinflation. After birth, a dysfunctional valve mechanism (air enters the alveoli thanks to negative pressure but cannot be released because the tracheobronchial tree has collapsed) leads to air trapping. CLE has all the features of obstructive emphysema: compression of lung parenchyma; mediastinal shift, herniation of the emphysematous lobe across the anterior mediastinum; and reduced respiratory reserve due to lowering of the diaphragm [1–2].

CLE is a relatively common congenital malformation of the lung (1 in 20,000 to 1 in 30,000 deliveries). However, it is not always detected during prenatal ultrasonographic evaluation [3]. There does not appear to be a significant predominance for a particular sex [4], but some authors have shown that males are affected three times more often than females, and that it occurs predominantly in Caucasians [5]. Bilateral localization of CLE is not common [2]. The frequency of distribution is [6]:

- superior left lobe (40–50%);
- middle right lobe (30–40%);
- superior right lobe (20%);
- inferior lobes (<5%).

CLE often presents with alveolar destruction, but sometimes alveoli are intact but over-distended. Over-distension of alveoli is associated with polyalveolosis (or "polyalveolar lobe", described for the first time by Hislop and Reid) [7]. The term "polyalveolosis" describes a lobe in which, compared with normal airways and arteries, the total number of alveoli is increased, along with impairment of gas exchange. Clinical and radiological findings cannot be used to differentiate between CLE and polyalveolar lobes. Congenital lobar over-inflation is used to define both entities [2, 6, 8]. Cardiac malformations (12–14% of cases) [9], renal or bones anomalies, pectus excavatum ("funnel chest") and hiatal hernia are possible associated anomalies [3].

23.2 Etiopathogenesis

The origin of CLE is air trapping. The affected lung allows the passage of air during inspiration but not its complete emission during expiration, resulting in pulmonary hyperexpansion. Several theories have been postulated to explain air trapping but the precise etiology is unknown in 50% of cases [3, 9, 10]. The most reliable include [6]:

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- Dysplastic bronchial cartilage or complete bronchial atresia allow the valve effect [11];
- endobronchial obstruction (accumulation of thick mucus [12], excessive mucosal proliferation and subsequent refolding [1]);
- extrinsic bronchial compression by aberrant vascular structures, enlarged heart chambers or thoracic masses [13];
- widespread and atypical bronchial abnormalities that may or may not be related to infections [14, 15].

We could say that CLE is due to intrinsic (bronchomalacia, stenosis) or extrinsic (bronchogenic cyst, vascular structures) bronchial obstruction that led to progressive air trapping and hyperinflation.

23.3 Clinical Features

The spectrum of clinical presentation is broad and is dependent upon: the age of the patient; size and location of the malformation; and associated anomalies. Symptoms can arise at birth (25% of cases), within the first month of life (50% of cases) or sporadically after 6 months of age [6]. Symptoms in the newborn are often associated with rapid progression that requires early resection of the malformation. In general, lesions associated with poorly defined symptoms remain stable or regress and they do not damage the surrounding parenchyma [2, 6, 16].

In the neonatal period, CLE can lead to mild-to-moderate respiratory distress and only occasionally to severe respiratory failure [17]. In the latter, there is severe mediastinal shift, lobar distension, reduced respiratory excursion and pulmonary compression in the affected and controlateral lung. Reduction of breath sounds and hypophonia can be observed at clinical examination. In early childhood, the cardinal symptom is tachypnea that worsens with crying or feeding. Children may also have wheezing, dry cough, stridor, failure to thrive and cyanosis that necessitates urgent treatment. Thoracic infections are typical at older ages [1, 18].

23.4 Diagnosis

The routine use of ultrasonography during pregnancy has changed the natural history of many congenital anomalies and has permitted better comprehension of their pathophysiology [15]. Prenatal diagnosis is fundamental for the planning of birth and the management of congenital malformations as well as for the psychological support of parents. Congenital malformations of the lung are not usually identified before the second trimester. A precise prenatal diagnosis of CLE is difficult [3]. Sometimes the malformation is suspected only by indirect signs (mediastinal shift, polyhydramnios, fetal hydrops). CLE appears as a luminescent unilateral lesion with no systemic vascularization and is sometimes associated with cysts [3, 6, 15-19]. Recently, prenatal diagnosis has become more accurate thanks to the use of magnetic resonance imaging (MRI) of the fetus that allows precise anatomical representation of the thoracic cage and of its contents [6, 20]. Prenatally, a lung-to-thorax transverse area ratio (L/T) of <0.25), polyhydramnios, and hydrops are important prognostic factors [21].

In the first 24 h of life, CLE has the radiological appearance of a radiopaque mass due to fluid accumulation in the affected lobe that is dependent upon delayed clearance [2]. Progressive over-inflation occurs together with fluid absorption that leads to acinar shadowing, a reticular interstitial pattern and, finally, lobe hyperlucency [1, 2, 9, 22] (Fig. 23.1).

The emphysematous lobe herniates and crosses the mediastinum, compressing the normal parenchyma and causing mediastinal shift. Intercostal spaces are expanded and the diaphragm is flattened [1, 9]. The lesion does not change during breathing but has a reduced blood supply. Computed tomography (CT) is useful for the diagnosis and scintigraphy can be undertaken the diagnosis is unclear. CT

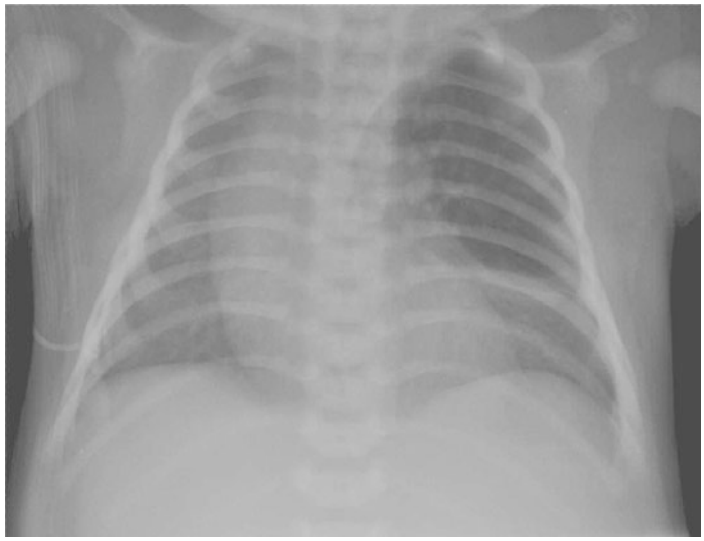

Fig. 23.1 Chest radiograph showing hyperluciency of the affected left lobe and mediastinal shift

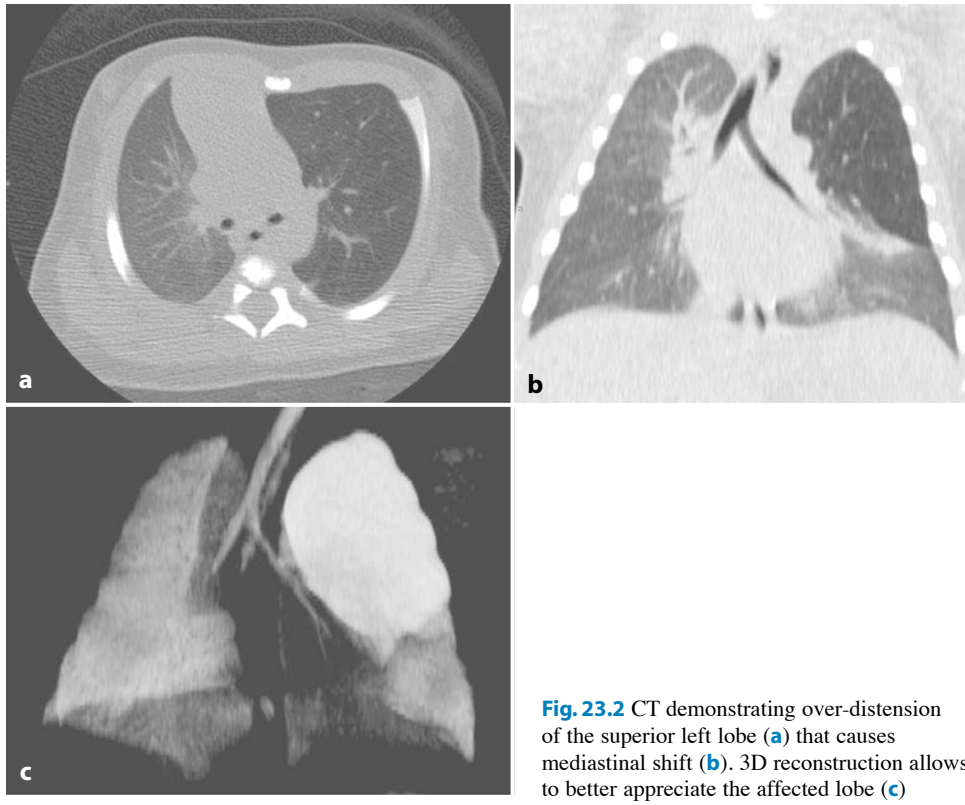

Fig. 23.2 CT demonstrating over-distension of the superior left lobe (**a**) that causes mediastinal shift (**b**). 3D reconstruction allows to better appreciate the affected lobe (**c**)

shows an enlarged affected lobe that is hypodense with atelectasis of the surrounding parenchyma. Contrast-enhanced CT along with magnetic resonance angiography is particularly useful for the diagnosis of congenital lobar over-inflation due to extrinsic compression (vascular rings, cystic lesions) (Fig. 23.2). Scintigraphic perfusion/ventilation

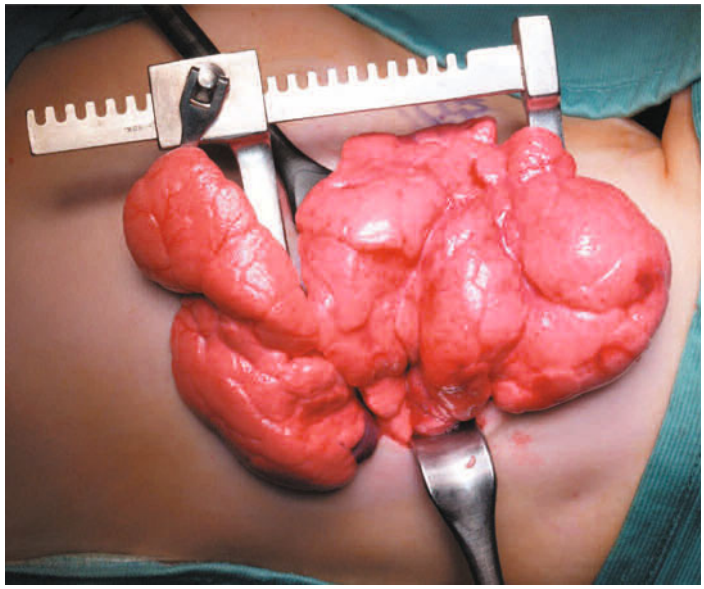

Fig. 23.3 After thoracotomy, the affected lobes "pop out" from the wound

scans demonstrate delayed uptake and washout of the radioisotope and little blood flow [6]. Bronchoscopy may show intrinsic or extrinsic obstructions but is not so widespread because it may exaggerate the distress [1, 23].

The differential diagnosis includes acquired emphysema (usually associated with high-pressure ventilation), pneumothorax, aspiration of foreign bodies, cystic congenital lesions, diaphragmatic hernia, and bronchogenic cysts. The position of the collapsed lung (which is not around the hilum in CLE) and lung markings (that are not present in pneumothorax and only attenuated in CLE) help the differentiation from pneumothorax [2].

23.5 Management

The treatment of congenital malformations of the lung is planned using a multidisciplinary approach. A limited number of cases with severe fetal hydrops profit from prenatal intervention, otherwise most cases are treated only after birth [17]. Symptomatic lesions should be resected by lobectomy [24-26] to allow compensatory pulmonary growth and to improve respiratory function. Acute postnatal respiratory distress requires emergency surgery. Respiratory function should be supported with high-frequency ventilation, administration of nitric oxide or extracorporeal membrane oxygenation to stabilize the patient. Positive-pressure ventilation may increase the amount of air trapped, thereby worsening the emphysema. Likewise, placement of a drain tube for presumed pneumothorax impairs clinical status [24]. The management of asymptomatic lesions is controversial, especially with regard to the need and timing of surgery. Stable CLE can reduce over time and have a good prognosis, justifying a "wait and see" approach [3, 4, 17, 18, 24]. These patients should undergo CT at around 6 months of age. If necessary, elective surgery can be arranged after radiological evaluation.

Lobectomy with traditional thoracotomic access is undertaken. The affected lobe characteristically "pop outs" through the wound [6] (Fig. 23.3). Thoracoscopy has been advocated by some authors as an alternative method [27] but has some limitations. That is, the expanded lobe prevents the creation of a work space and the procedure should be carried out only in stable patients by expert surgeons. A minimally invasive approach can be used if the over-inflation is mild or after effective endoscopic decompression of the affected lobe.

23.6 Outcome

CLE carries a good prognosis. A high percentage of cases are treated successful with conservative management, but symptomatic patients require surgery. The prevalence of long-term pulmonary growth after lobectomy is excellent [6].

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