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Review Article

Prenatal diagnosis of congenital lobar fluid overload

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ABSTRACT

Prenatal congenital lobar fluid overload (CLFO), which was first described by Ramsay and Byron, is identical to postnatal congenital lobar overinflation. It is characterized by progressive lobar overexpansion that compresses the other adjacent lung lobes. The underlying cause can be an intrinsic cartilaginous abnormality or an extrinsic airway compression. It may be associated with cardiovascular anomalies in 12%–14% of cases and affects males more frequently than females. Most cases are diagnosed postnatally, but early antenatal diagnosis and sequential follow-up are attempted for early treatment, if clinically indicated. This article provided a thorough review of CLFO, including prenatal diagnosis and differential diagnoses, as well as comprehensive illustrations of the perinatal imaging findings of CLFO. Prenatal diagnosis of fetal lung lesions should include CLFO in the differential diagnosis and prompt investigation for associated anomalies.

Introduction

Congenital lobar fluid overload (CLFO) in fetuses, which was first described by Ramsay and Byron and is also referred to as congenital lobar overinflation (CLO) or congenital lobar emphysema (CLE) postnatally, is a rare cystic lung lesion that can often be diagnosed accurately in the neonatal or infantile periods based on presentations of respiratory distress and hyperinflation of pulmonary lobes [1,2]. CLFO is a disease entity of fluid-overloaded and expanded lung tissue, which usually presents on antenatal ultrasonography (US) as a homogeneous hyperechogenicity with or without mass effect to the mediastinum [1,3–19]. On magnetic resonance imaging (MRI), hyperintensity of the affected lung lobe, mass effect to the mediastinum, and intact lung architecture with stretched hilar vessels were frequently observed [1,5,6,11–13,20–27]. From our experience, relative hypointensity of the adjacent and compressed lung lobe is common (Figs. 1–6, cases 1–3), but hyperintensity of the compressed lung tissue can be seen in some cases [11]. The etiology of this malformation remains unclear, but several theories were proposed; these include 1) a bronchial cartilaginous defect that produces a one-way valve effect; 2) endobronchial mucus impaction or proliferative mucosal infolding (polyalveolosis or polyalveolar lobe); 3) extrinsic compression of the bronchi from aberrant vessels or lung parenchymal lesions; and 4) atresia of a lobar bronchus [28–30]. The common locations of CLFO, in the order of frequency, are as follows: left upper lobe, right middle lobe, and right upper lobe [4,31]. Males are more frequently affected than females [32] and about 20% of patients with CLFO have associated congenital cardiovascular anomalies [33,34].

Prenatal diagnosis

Fetal CLFO is usually discovered in the second trimester. Up to the present time, at least 41 cases of fetal CLFO have been

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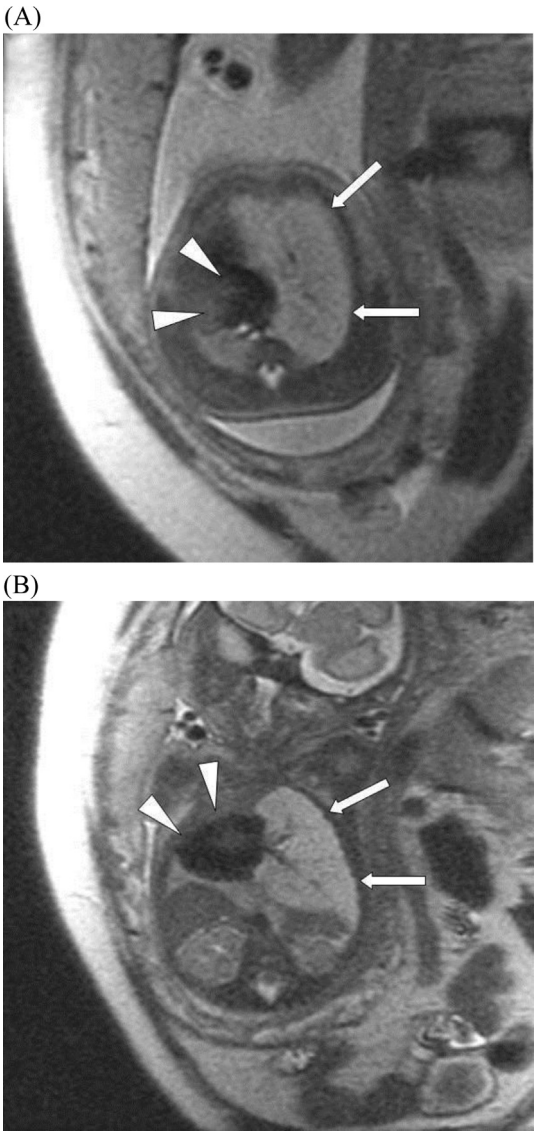

Fig. 1. (A) Axial and (B) coronal MRI of CLFO in case 1 at 20 weeks gestational age shows hyperintense and expanded left upper lobe (arrows), suggesting fluid overload and causing compression of the heart toward the right (arrowheads).

reported in English literature (Table 1). Quinton et al. have reported the earliest diagnosis of CLFO at 18 gestational weeks [4]. Prenatal US remains the reference standard of imaging modalities to evaluate the lesion. The most common manifestation on US is a unilateral, bright, hyperechogenic lesion in the affected lung [5], but Alamo et al. [13] and Babu et al. [17] reported that some CLFO cases can present as cystic mass lesions (Table 1). On fetal MRI, CLFO appears as a fluid-overloaded segment or lobe with high signal intensity and stretching of the hilar vessels without architectural distortion of the adjacent pulmonary lobes [1]. Some cases show partial or complete regression on serial follow-up US in the third trimester, depending of the etiology of the blockage. However, it is sometimes difficult to distinguish CLFO from microcystic congenital pulmonary airway malformation (CPAM) and bronchopulmonary sequestration (BPS) on prenatal US [1]. Nowadays, fetal MRI can be a helpful complementary modality to US to differentiate among CLFO and other fetal cystic lung lesions owing to its better tissue contrast, larger field of view, enhanced anatomic evaluation, more detailed

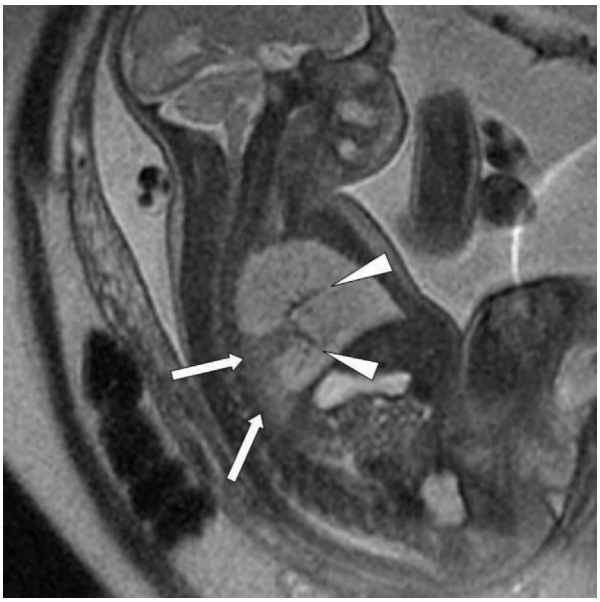

Fig. 2. Sagittal MRI of CLFO in case 1 at 20 weeks gestational age shows a hyperintense and expanded left upper lobe and a small, hypointense, compressed adjacent left lower lobe (arrows). Intact lung architecture with stretched hilar vessels (arrowheads) is observed. The lesion disappeared at 37 weeks gestational age and postnatal chest radiograph of the infant was normal.

demonstration of the lesion extent, as well as the capability of detecting other associated congenital anomalies.

Differential diagnosis

Fetal cystic lung lesions are rare but significant in the spectrum of congenital lung malformations (CLMs). These lesions include CLFO, which is synonymous with postnatal CLE, BPS, CPAM, and bronchial atresia (BA) [1]. Antenatal differential diagnoses of CLFO should be done for appropriate treatment recommendations.

Congenital pulmonary airway malformation

CPAM, previously known as congenital cystic adenomatoid malformation (CCAM), is characterized by adenomatoid proliferation of bronchiole-like cysts in the lung and lack of normal alveolar development [35]. It has been the most commonly diagnosed lung malformation prenatally, accounting for 30%–40% of all congenital lung diseases [20]. The outcome of CPAM is unpredictable. Some postulate that CPAM with <57% of the total lung volume would resolve completely, whereas CPAM with >84% of total lung volume will not [1]. One important prognostic factor is the presence of hydrops; if not treated, more than 90% of these fetuses die before birth [36]. CPAM is thought to be a predisposing condition to lung neoplasms, such as pleuropulmonary blastoma or rhabdomyosarcoma [5]. Malignant transformation to bronchoalveolar carcinoma is another issue that was reported to develop on a preexisting CPAM in young adulthood [5].

Stocker et al. distinguished three types of CPAM based on cyst size and histopathology. Type I consisted of large cysts (at least one dominant cyst >1 cm in diameter); type II comprised numerous small cysts <1 cm; and type III has microcystic lesions <0.2 cm in diameter [37]. Nowadays, this classification had been expanded to include two additional subtypes: type 0 displays acinar dysgenesis or dysplasia, whereas type IV exhibits distal acinar origin [37].