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Prenatal diagnosis of lobar bronchial atresia

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KEYWORDS: lobar bronchial atresia; lung malformation; prenatal diagnosis

ABSTRACT

We report three cases of fetal lobar bronchial atresia referred to our Fetal Medicine Center during the mid-trimester of pregnancy over the last 15 years. Lobar bronchial atresia can mimic a main stem bronchial atresia on mid-trimester ultrasound examination as it induces extensive lobar enlargement, major mediastinal shift and eversion of the diaphragm. It was associated with severe pulmonary hypoplasia in all three cases, even though polyhydramnios and ascites were absent in two. Termination of pregnancy was performed at parental request after extensive counseling in each of the cases and necropsy confirmed one or two enlarged lung lobes leading to major compression of the remaining lobe(s) of the ipsilateral lung, the contralateral lung and the heart. No other anomalies were observed and the karyotype was normal in all cases. Copyright © 2011 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORT

During the past 15 years, three cases of fetal lobar bronchial atresia have been referred to our Fetal Medicine Center during the mid-trimester of pregnancy (Table 1). The ultrasonographic features in each case consisted of an enlarged pulmonary echogenic mass associated with dilated fluid-filled airways, major mediastinal shift, contralateral lung compression and eversion of the diaphragm (Figure 1). The echogenic mass always appeared to extend to the full extent of the lung and in none of the cases was a normal ipsilateral lung detected. In one case, ascites was present at the time of diagnosis and polyhydramnios occurred a few weeks later. In none of the cases had cystic hygroma or enlarged nuchal translucency been detected at the first-trimester ultrasound scan. Termination of pregnancy was performed at parental request after extensive counseling in each of the cases.

Necropsy in each fetus confirmed the presence of one or two enlarged lung lobes leading to major compression of the remaining lobe(s) of the ipsilateral lung, the contralateral lung and the heart (Figures 2 and 3). No other anomaly was observed and the karyotype was normal in all cases. The lung-to-body weight ratio (LBWR), calculated by dividing the weight of the unaffected lobes by the fetal body weight, was always below 0.015, confirming severe lung hypoplasia. Further dissection demonstrated atresia of one or two lobar bronchi in each case. In two cases involving the left lung, necropsy revealed abnormal lung lobulation, with three left lung lobes.

DISCUSSION

This series confirms that lobar bronchial atresia is a rare entity (with a prevalence of less than 1 per 100 000 births based on our local birth defect registry) associated with severe lung hypoplasia. Bronchial atresia usually occurs as an isolated anomaly and is characterized by a mucocele associated with a blindly terminating bronchus and hyperinflation of the obstructed segment of the lung¹. A mucocele can be detected prenatally by demonstrating dilated anechoic fluid-filled airways during an ultrasound scan. The obstructed lung corresponds to an enlarged hyperechoic lung area, as illustrated in Figure 1. Lobar bronchial atresia induces such an extensive lobar enlargement that it mimics a main stem bronchial atresia. We found that a single plugged lobe can increase in size so much that no ipsilateral normal lung remains detectable at the mid-trimester ultrasound scan.

Severe pulmonary hypoplasia with LBWR below 0.015 before 28 weeks of gestation was always confirmed despite there being no ascites/hydrops or polyhydramnios in two cases. The weight ratio of affected lobes to normal lobes varied from 3 to 25 and ascites/polyhydramnios was

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Figure 1 Ultrasonographic features of fetal bronchial atresia at 25 weeks of gestation (Case 3): enlarged and echogenic left lung associated with dilated fluid-filled airways, major mediastinal shift and contralateral lung compression (a), eversion of the diaphragm (b) and these features shown in the multiplanar view (c).

only present in the most severe case. This corresponded to a double lobe lesion for which the affected lung weight was around double that observed for the cases with a single lobe lesion, with major compression leading to a severely reduced contralateral lung (Table 1 and Figure 3). Ascites and polyhydramnios therefore appear to be very delayed manifestations of lung compression in this context.

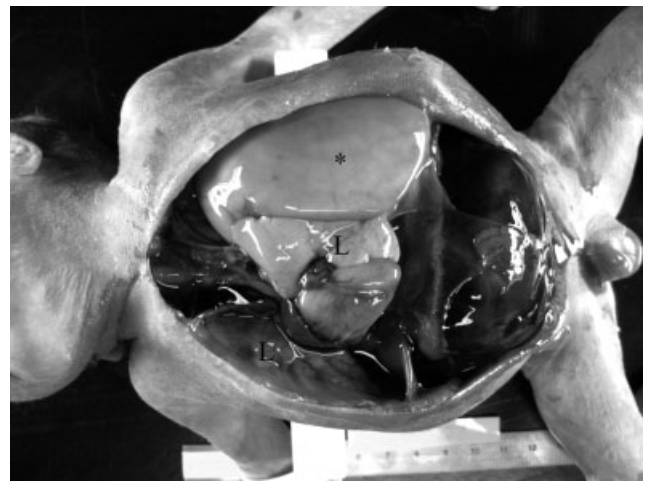


Figure 2 Necropsy findings showing an enlarged (*) accessory middle left lobe caused by bronchial atresia with compression of the normal lung tissue (L) and mediastinal shift (Case 3).

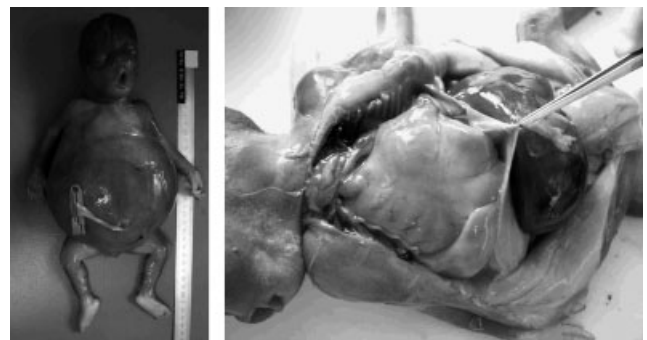


Figure 3 Postmortem images showing ascites associated with extremely enlarged middle and lower right lung lobes at 25 weeks of gestation (Case 2).

The clinical presentation of congenital bronchial atresia in children or infants consists of respiratory distress and/or recurrent pneumonia². Young adult cases are sometimes identified. However, the cases reported in children or adults are described as having segmental or subsegmental atresia, leading to less extensive lesions. Some surgeons have observed gaps in the segmental bronchi as in a true atresia, whereas other cases have been described as having a membranous web bulging into the lumen³. This suggests that there is probably not only one unequivocal type of atresia on histology. The pathogenesis of the condition has not yet been elucidated.

Unlike laryngeal or tracheal obstruction (Congenital High Airway Obstruction Syndrome), which can occur in syndromes such as Fraser syndrome, no syndromes including lobar bronchial atresia have yet been described. Bronchial atresia probably arises as a result of a developmental interruption. Considering the embryological development of the lungs, two theories are proposed. The first theory suggests that a nest of proliferating cells separates the developing bronchial bud during weeks 5–6 of gestation and continues to branch independently. As a consequence, normal branching distal to the atresia is maintained without actual connection to the central

Table 1 Prenatal and postmortem characteristics of three cases of lobar bronchial atresia

Characteristic	Case 1	Case 2	Case 3
GA at diagnosis (weeks)	23	19	24
Side of the lesion	Left	Right	Left
Apparent size of the lesion	Whole lung	Whole lung	Whole lung
Mediastinal shift	Major	Major	Major
Eversion of diaphragm	Present	Present	Present
Ascites	Absent	Present	Absent
Amniotic fluid volume	Normal	Polyhydramnios (> 22 weeks)	Normal
Evolution	TOP (24 weeks)	TOP (25 weeks)	TOP (25 weeks)
Fetal body weight (g)	1010	1510	1030
Lesion weight (g)	48	97	34
Weight of unaffected lobes (g)	10	4	13
LBWR*	0.0099	0.0026	0.0126
Level of atresia	Upper lobe	Middle and lower lobe	Middle lobe
Abnormal lobulation	Present (three left lobes)	Absent	Present (three left lobes)

*Lung to body weight ratio (LBWR) calculated by dividing the weight of the unaffected lobes by the fetal body weight. GA, gestational age; TOP, termination of pregnancy.

airways⁴. This early theory could also favor the abnormal lung lobulation that we observed in two cases and that has been described in up to 11% of cases in Fraser syndrome⁵. One can speculate that the absence of ultrasonographic signs on the first-trimester scan (no enlarged hyperechoic lung and no mediastinal compression or enlarged nuchal translucency), despite early embryological insult, could be caused by reduced lung fluid secretion during the early pseudo-glandular stage of lung development. The second theory assumes that a vascular accident with ischemia and scarring causes secondary bronchial atresia at around the 16th week of gestation when all airway branches are fully developed^{6,7}. However, one report has revealed that the blind end of the proximal or distal bronchus was lined with bronchial epithelium without scar formation or granuloma on pathological examination in most cases of congenital bronchial atresia². This indicates that bronchial atresia does not seem to be induced by acquired inflammatory change.

It has been suggested that many, if not all, lung malformations, may represent a continuum of anomalies associated with unrecognized airway obstruction^{8,9}. Langston *et al.* proposed a 'malformation sequence' to explain the spectrum of pulmonary anomalies, depending on the level, timing and degree of bronchial obstruction⁹. An atretic bronchus early in gestation might favor the formation of a cystic adenomatoid malformation or a bronchogenic cyst, whereas obstruction later in gestation (at 16–18 weeks) might predispose the lung to the development of bronchopulmonary sequestration or lobar emphysema^{3,9}. Our case series and its distinct ultrasonographic features could correspond to a total and early obstruction of a lobar bronchus.

In conclusion, the prenatal diagnosis of lobar bronchial atresia is based on ultrasonographic features usually detected during a mid-trimester scan. Major mediastinal shift, contralateral lung compression and eversion of the diaphragm are associated with severe pulmonary

hypoplasia. Ascites or polyhydramnios may be present in the most severe cases. A nest of proliferating cells that separates the developing bronchial bud during weeks 5–6 of gestation and continues to branch independently could represent a better explanation for the pathogenesis of the condition in comparison to the theory that it results from a secondary vascular accident with ischemia and scarring.

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