Prenatal Sonographic Features of Intralobal Bronchopulmonary Sequestration

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Congenital lung lesions that can be diagnosed on the basis of prenatal sonography include cystic adenomatoid malformations and extrapulmonary bronchopulmonary sequestrations. Intralobal bronchopulmonary sequestration is a rare congenital malformation of the lung. The prenatal sonographic features of this condition are reported here.

Case Report

A 20-year-old woman, gravida 2, para 1, was referred for sonography to rule out fetal anomalies. She was 25 weeks by menstrual age. Her first child was a girl and had no abnormalities. There was no history of consanguinity.

Sonography was performed with an HDI 5000 scanner (Philips Medical Systems, Bothell, WA) and broadband convex probes of 2 to 5 and 4 to 7 MHz and a linear probe of 5 to 12 MHz. Biometric measurements corresponded to 23 weeks. There was a large, brightly echogenic mass filling the left hemithorax (Fig. 1). The fetal heart was pushed to the right. A color Doppler study showed a large artery arising from the descending thoracic aorta and supplying the mass (Fig. 2). The venous drainage was by a pulmonary vein into the left atrium (Fig. 3). These features were indicative of intralobal bronchopulmonary sequestration. There was no other malformation. The parents opted for termination of the pregnancy.

On fetal autopsy, the right lung and upper lobe of the left lung were found to be hypoplastic. The left lower lobe was markedly enlarged, filling the left thoracic cavity and displacing the heart to the right of the midline. The right branch of the pulmonary artery was supplying the right
lung. The left pulmonary artery showed 2 divisions. One was found supplying the upper lobe of the left lung, and the other branch was found to be fibrosed. A large muscular arterial branch was found arising from the descending thoracic aorta and supplying the left lower lobe of the lung (Fig. 4A). The vein from this lobe was connected to the left atrium (Fig. 4B). Microscopically, the enlarged left lower lobe showed cut sections of terminal bronchi, respiratory bronchioles, and alveoli. The interstitium was well developed and showed occasional congested blood vessels.

Discussion
Bronchopulmonary sequestrations are rare abnormalities that constitute 0.15% to 6.4% of all congenital pulmonary malformations.1 Bronchopulmonary sequestration is generally defined as nonfunctioning lung tissue that is not in normal continuity with the tracheobronchial tree and that derives its blood supply from systemic vessels.2 Pryce3 further classified the lesion as intralobar or extralobar on the basis of the morphologic patterns. Intralobar bronchopulmonary sequestration consists of an abnormal
segment of lung tissue that shares the visceral pleural covering of an otherwise normal pulmonary lobe and that lacks normal communication with the tracheobronchial tree, although variable amounts of air may be contained within the anomalous tissue. Intralobar sequestrations occur almost always within the lower lobe and slightly more often on the left side. The artery supplying the intralobar bronchopulmonary sequestration usually arises from the descending thoracic aorta. Most intralobar bronchopulmonary sequestrations are drained by the normal pulmonary vein into the left atrium. Extralobar bronchopulmonary sequestration consists of a discrete accessory mass of nonaerated lung tissue that is invested by its own pleura. It histologically resembles normal lung tissue, although all structures are dilated. Typically, an anomalous artery arising directly from the thoracic or abdominal aorta supplies it. The venous drainage of extralobar bronchopulmonary sequestration is usually systemic, through the azygos, hemiazygos, or superior vena cava into the right atrium. Therefore, the major differentiating feature between intralobar and extralobar bronchopulmonary sequestrations is the venous drainage.

Prenatal sonographic features of extralobar bronchopulmonary sequestration have been reported. To our knowledge, there are only 2 reports of prenatal diagnosis of intralobar bronchopulmonary sequestration. In the first, an antenatal diagnosis of a cystic adenomatoid malformation was made. Postnatally the child underwent surgery at the age of 2.5 years, and the histopathologic diagnosis was intralobar bronchopulmonary sequestration. In the second report, the antenatal diagnoses considered were cystic adenomatoid malformation and pulmonary sequestration. Postnatally, the child underwent surgery on day 5, and a final diagnosis of intralobar sequestration was made. In both reports, the diagnosis of intralobar pulmonary sequestration was made postnatally.

On prenatal sonography, bronchopulmonary sequestrations, both the intralobar and extralobar types, appear as brightly echogenic masses in the chest. Depending on the size, the sequestration can compress or displace the heart and the rest of the lungs. The microcystic form of a cystic adenomatoid malformation also has a similar appearance. The hallmark of bronchopulmonary sequestration is the anomalous artery supplying it, which arises, in most instances, from the aorta. In the case presented here, the anomalous artery supplying the mass was seen arising from the descending thoracic aorta. The only differentiating feature between intralobar and extralobar bronchopulmonary sequestrations, which can be seen prenatally, is the pattern of venous drainage. The improved resolution of gray scale images and the improved sensitivity of color Doppler images available in recent equipment have made consistent visualization of the pulmonary veins possible. In this case, the venous drainage of the mass was via the pulmonary vein to the left atrium. This feature is characteristic of intralobar bronchopulmonary sequestration.
Although extralobar bronchopulmonary sequestration is accepted universally as a congenital anomaly, some authors have indicated an acquired origin for intralobar bronchopulmonary sequestration. The present case supports the theory that intralobar bronchopulmonary sequestration is also congenital in origin. It can manifest in infancy as congestive heart failure\textsuperscript{9–11} because the lesion acts as a communication between the systemic and pulmonary circulations as it receives blood supply by an anomalous artery from the thoracic aorta and drains by a pulmonary vein to the left atrium. The postnatal prognosis of intralobar bronchopulmonary sequestration is related to the size of the mass. When it is very large, the prognosis is poor because of hypoplasia of the rest of the lungs and more severe systemic-pulmonary communication, leading to earlier onset of cardiac failure.

References


![Figure 4. A](image1.png) Autopsy photograph showing the descending aorta (AO) and the anomalous artery (arrowhead) from the aorta supplying the mass (IL). B, The pulmonary vein (arrowhead) is shown draining the mass (IL) into the left atrium.