PEDIATRIC PATHOLOGY CASES

Pulmonary Sequestration Presenting as Fetal Hydrops

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Pulmonary sequestration is a rare developmental anomaly of the lung characterized by nonfunctional pulmonary tissue without communication with the tracheobronchial tree and receiving an aberrant systemic arterial blood supply. Few cases of non-immune hydrops fetalis associated with this entity have been reported. A 2 day old male baby born by cesarean section at 31 weeks gestational age due to fetal hydrops is presented. Autopsy revealed a hydroptic baby with extralobar pulmonary sequestration and bilateral pulmonary hypoplasia. The clinicopathologic presentation of this unusual pulmonary developmental anomaly is discussed.

Key words: Extralobar pulmonary sequestration, Fetal hydrops, Pulmonary anomaly

Pulmonary sequestration is a segment of nonfunctional lung that lacks a normal connection to the tracheobronchial tree and possesses an aberrant systemic blood supply, usually arising from the descending aorta. Pulmonary sequestration can be: intralobar, located within the visceral pleura of the normal lung tissue or extralobar, invested with its own visceral pleura. These lesions are rare pulmonary malformations accounting for 0.15 to 6.4% of all congenital pulmonary anomalies (1). They are almost always unilateral, the left side of the chest affected twice as often as the right and there is a male to female ratio of 3:1 in extralobar lesions (2).

Clinical presentation depends on the type of sequestration; recurrent infection in infants and children is the rule in intralobar cases while asymptomatic lesions found incidentally in fetuses and neonates characterize the extralobar type (1).

Prognosis is variable, ranging from lethal outcome to even spontaneous regression of some lesions (3,4). Survival has been reported after successful medical management (5,6,7) even in those cases with the worst clinical presentations.

Case Report

A 2 day old male baby was born by cesarean section due to fetal hydrops at 31 weeks gestational age to a 41 year old G5P4A0 mother. Hydrops was detected the day before his birth. A previous obstetric sonogram done at 20 weeks was normal. At birth, Apgar scores were 3-2-3 at one, five and ten minutes respectively and body weight was 2105g. The neonate was floppy, with no respiratory effort, generalized edema, and acrocyanosis. He was endotracheally intubated and given positive pressure ventilation. The right hemithorax was decompressed placing a catheter as well the abdomen placing catheters on both lower quadrants, obtaining 160 ml of serous fluid from the abdomen and approximately 10ml from the thorax. The patient was admitted to the Neonatal Intensive Care Unit for mechanical ventilation and management of his critical status. During his short hospital stay, Cardiology and Genetics Services evaluated the patient and there were no cardiac findings on echocardiogram to explain fetal hydrops. Metabolic screening tests and chromosomal studies were recommended by the geneticist. A portable chest/abdomen roentgenogram revealed complete left lung opacification and a mediastinum shifted to the right considered to be due in part to patient’s rotation, versus a left pleural effusion, diaphragmatic hernia or other space occupying lesion. Despite all medical efforts, he had a cardiac arrest requiring prolonged cardiopulmonary resuscitation. The patient expired two days after birth. The blood cultures were negative. There was no evidence of hemolysis and the direct Coomb’s test was negative.

Pathology Findings

Postmortem examination revealed a moderately hydroptic baby with generalized edema, more noticeable in the dorsum of the feet, hands and face. Body weight was 2280g, when the expected is 1543 ± 519g. Body measurements were within the expected for his gestational age.

Upon opening the cavities, there were 35 ml of serous...
ascitic fluid, bilateral pleural effusions of serosanguinolent fluid, 25 and 30 ml on the right and left side respectively, and a pericardial effusion of 15ml of serous fluid. The subcutaneous tissues were edematous. When the pleural cavities were inspected, a 3.5 X 3.0 X 1.0 cm mass was noted in the left supradiaphragmatic area with a pedicle connected to the thoracic aorta (Figures 1, 2 and 3). The mediastinum appeared in the midline. Externally, this mass was similar in appearance to the native lungs. On section, it was dark pink, and rubbery. The lungs revealed normal lobation and a congested rubbery parenchyma. Both lungs appeared smaller than the expected, the left much more than the right. (Figure 1). The rest of the macroscopic exam revealed no other structural abnormalities.

![Figure 1. Note supradiaphragmatic mass (*) and hypoplastic lungs (arrows).](image1)

Microscopic examination of this supradiaphragmatic mass revealed a pulmonary sequestration characterized by uniformly dilated bronchioles, alveolar ducts, and alveoli (Figure 4). This histology contrasted markedly to the lungs which presented formation of hyaline membranes, focal hemorrhages and early pneumonia.

![Figure 2. Note hypoplastic lung (arrow) and extralobar sequestration (*).](image2)

![Figure 3. Note extralobar sequestration ( ) and pedicle connected to the thoracic aorta (arrow head). Hypoplastic lung (arrow).](image3)

![Figure 4. Note uniformly dilated bronchioles, alveolar ducts and alveoli.](image4)
Discussion

Pulmonary sequestration is a segment of nonfunctional lung lacking a normal connection to the tracheobronchial tree, and receiving an aberrant systemic blood supply, usually arising from the descending aorta. Two forms exist: intralobar sequestration, located within the visceral pleura of the normal lung tissue, and extralobar sequestration, invested with its own visceral pleura (1). These lesions are rare pulmonary malformations accounting for 0.15 to 0.4% of all congenital pulmonary anomalies (1). They are almost always unilateral, the left side of the chest affected twice as often as the right, and there is a male to female ratio of 3:1 with extralobar lesions (2).

The clinical presentation depends on the type of sequestration; recurrent infection in infants and children is the rule in intralobar cases while asymptomatic lesions found incidentally in fetuses and neonates characterize the extralobar type (1). In 25% of the extralobar cases, the diagnosis is done prenatally, about 60% of the patients present by three months of age, and approximately 10% of patients are asymptomatic. Extralobar pulmonary sequestration may be seen in older children and they have been reported in patients as old as 81 years old [8]. Presenting symptoms are often noted on the first day of life and include cyanosis, dyspnea, and difficulty in feeding. Fetal non-immune hydrops, anasarca, pleural effusions, or localized edema may present along with maternal polyhydramnios. Postnatal detection is suspected when hydrops and neonatal respiratory distress persist in the absence of other organic disease.

Many theories have been proposed to explain the embryology of pulmonary sequestration. A widely accepted theory has explained it as a supernumerary bud that arises caudal to the normal lung and migrates caudally with the esophagus. If the lung bud develops after the pleura has formed, it develops separate from the lung and becomes surrounded by its own pleura, forming an extralobar pulmonary sequestration (2). A high incidence (65%) of associated anomalies have been noted of which 50% are congenital pulmonary airway malformation (CPAM). Other associated anomalies include bronchogenic cysts, cardiovascular malformations, pectus excavatum, congenital diaphragmatic hernias and abnormal foregut communications (8).

Although not a common presentation, extralobar pulmonary sequestration has been associated with the development of non-immune fetal hydrops and this association has been reported in the literature to carry a very poor prognosis for survival. It is known that the overall prognosis depends on the size of the lung mass and the secondary effects of hypoplasia and hydrops fetalis. Hydrops is a harbinger of fetal or neonatal death. The hydrops has been postulated to be secondary to a twist in the vascular pedicle of the sequestrated segment causing venous and lymphatic obstruction resulting in a pressure gradient producing ultimately an effusion (3,5). This space occupying lesion may cause pulmonary hypoplasia, mediastinal shift, and tension hydrothorax from fluid or lymph secretion by the mass, with effusions into the body cavities leading to fetal hydrops. Large intrathoracic masses may also cause vena cava obstruction and cardiac compression leading to cardiac failure and hydrops. Esophageal compression by the thoracic mass causes interference with fetal swallowing of amniotic fluid resulting in polyhydramnios.

The natural history of these lesions is variable, with a significant percentage regressing during the antenatal period. The presence of hydrops is the single most important prognostic factor in the prenatal assessment of these pregnancies; the overall prognosis for these fetuses is extremely poor without intervention. Lesions not associated with hydrops can be expected to have a good outcome, and may be managed conservatively to term. If hydrops is detected before 32 weeks gestation, definitive antenatal intervention is indicated by either serial thoracentesis, placement of a pleuroamniotic shunt or in utero surgical resection of the lesion (5,9). When diagnosis is made after the 32 weeks of pregnancy, steroids lung maturation induction and counseling for early delivery with postnatal surgery is the standard of care (5). All fetuses with a pulmonary sequestration should be delivered at a tertiary care center where immediate resuscitation and surgery can be completed if required. Alcoholicization of the pedicle supplying the sequestrated segment with definitive resolution by the time of birth has been also described by Niccoli et al as a new therapeutic approach (10). Alcohol causes perivascular tissue toxicity and necrosis without remote untoward effects from the target organ (10).

With the advent of routine antenatal ultrasonography, pulmonary mass lesions are being identified prenatally with increasing frequency (6). The combination of an aberrant systemic blood supply (identified by color-flow Doppler sonography) and a well-defined echodense, homogenous lung mass is pathognomonic for the diagnosis of pulmonary sequestration (11,12).

The patient's early death precluded further diagnostic studies. The patient's autopsy disclosed an extralobar pulmonary sequestration as the cause of the hydrops and pulmonary hypoplasia, in addition to a superimposed hyaline membrane disease, early pneumonia, and focal pulmonary hemorrhages telling patient's death despite all medical efforts.
Resumen

El secuestro pulmonar es una anomalía del pulmón caracterizada por tejido pulmonar no funcional sin comunicación con las vías traqueobronquiales, supliéndola por una circulación sistémica aberrante. Pocos casos de hidropesia fetal no-immune asociados a esta entidad han sido publicados. Presentamos a un niño de dos días de edad que nace luego de una cesárea por hidropesia fetal. La autopsia reveló un bebé hidrópico con secuestro pulmonar extralobular e hiperplasia pulmonar. Se discute la presentación clinico-patológica de esta anomalía.

References