A Case of Intra-Abdominal Extralobar Pulmonary Sequestration: Prenatal Detection, Postnatal Diagnostic Approach and Outcome

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ABSTRACT

This paper describes the perinatal and postnatal outcome of a case considered to have a fetal intra-abdominal extralobar pulmonary sequestration.

Routine midtrimester fetal ultrasonographic scan detected an abdominal echogenic mass between aorta and stomach. It was considered primarily as an intra-abdominal extralobar pulmonary sequestration. At postnatal 2 months, computed tomography scan with intravenous contrast described it as an extralobar pulmonary sequestration at the level of diaphragmatic hiatus by showing the aberrant blood supply to the sequestration with venous drainage into the portal confluence. The patient is now 4 years old and asymptomatic; she is being followed-up by a pediatric surgeon.

Intra-abdominal extralobar pulmonary sequestrations usually remain asymptomatic throughout life; they may be safely observed without surgery unless they become symptomatic or show any change in the characteristics of the radiologic appearance.

Keywords: Abdominal cavity, Bronchopulmonary sequestration, Childhood, Pregnancy, Second trimester, Computed tomography, X-Ray

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Introduction

Pulmonary sequestration is an uncommon congenital anomaly with an estimated incidence of 0.15% to 1.7% in the general population (1). Approximately 25% of sequestrations are extralobar and 10% to 15% of the cases are intraabdominal (2). In this report, we describe the perinatal and postnatal outcome of a case considered to have a fetal intra-abdominal extralobar pulmonary sequestration.

Case Report

A 29 year-old primigravid woman applied for antenatal care in the first trimester. First trimester ultrasonographic evaluation revealed normal nuchal translucency thickness and fetal anatomy. Routine mid trimester fetal ultrasonographic scan detected an abdominal echogenic mass between aorta and stomach with a diameter of 13x15 mm in axial section (Figure 1) and 18x15 mm in longitudinal section in a 20-week fetus.

A cystic hypoechoic area was present in the center of the mass. Color Doppler ultrasound could not determine the blood supply of the mass. Ipsilateral kidney and adrenal gland could be visualized separately. It was considered primarily as an intraabdominal extralobar pulmonary sequestration (ELS). No other fetal anomaly was detected. The mass size was 25x25 mm in axial section and 26x21 mm in longitudinal section (Figure 2) at 25 weeks of gestation and did not change until birth.

Figure 1: Transverse image of the fetus at 20 weeks of gestation shows echogenic abdominal mass with a hypoechoic area
At 39 weeks of gestation, she gave birth to a female weighing 3860 g with an Apgar score of 8 and 9 at 1 and 5 minutes, respectively. The infant had no respiratory and feeding problems post-delivery, and physical examination was completely normal. In the early postnatal life, the findings of neonatal abdominal sonographic examination and magnetic resonance imaging (MRI) were compatible with an ELS at the level of diaphragmatic hiatus. An ipsilateral normal adrenal gland identified on postnatal MRI effectively excluded adrenal hemorrhage and an adrenal tumor. At postnatal 2 months, computed tomography (CT) scan with intravenous contrast described it as an ELS with a diameter of 28 mm in longitudinal section and 17x12 mm in axial section at the level of diaphragmatic hiatus by showing the aberrant feeding arteries of the sequestration (Figure 3) arising from the left gastric artery and celiac trunk with venous drainage into the portal confluence.

Computed tomography scan with intravenous contrast done in November 2014 did not show significant changes in the size of solid mass that had been measured 30x15x15 mm in diameter. The patient is now 4 years old and asymptomatic; she has been followed-up by a pediatric surgeon. The patient’s family has not decided for surgery yet.

**Discussion**

Pulmonary sequestration is a rare congenital anomaly in which a mass of lung tissue lacks normal communication with the tracheobronchial tree. Pulmonary sequestration has classically been divided into intralobar and extralobar forms. Intralobar sequestrations lie within the pulmonary visceral pleura and typically have a systemic blood supply from the aorta with drainage into a pulmonary vein. ELS have their own pleural investment, maintaining complete anatomic separation from the normal lung. They typically have a systemic blood supply with systemic venous drainage, although blood supply may be from a pulmonary artery (3). Most ELSs are found postero-medially in the left lower chest but can occur within the diaphragm, below it or rarely in other locations (4).

The detection of an echogenic mass in the fetal abdomen gives a diagnostic challenge to the ultrasonographer. The major differential diagnosis of a prenatal/neonatal suprarenal mass includes adrenal hemorrhage and neuroblastoma (5). Adrenal hemorrhage may be echogenic when first detected but becomes hypoechoic or anechoic within several days or shrinks in the first month of life (6). On prenatal sonography subdiaphragmatic ELS is usually echogenic, is left-sided, and can be identified in the second trimester. Neuroblastoma is most often cystic, right-sided, and identified in the third trimester (5). In our case, postnatal CT scan with intravenous contrast was helpful in further defining the mass as an ELS by visualizing the aberrant feeding arteries of the sequestration arising from the left gastric artery and celiac trunk with venous drainage into the portal confluence. An ipsilateral normal adrenal gland identified on postnatal MRI effectively excluded an adrenal hemorrhage or an adrenal tumor.

A period of simple observation appears warranted for these lesions, particularly for subdiaphragmatic ELS.7 In cases of subdiaphragmatic ELS or when a systemic arterial blood supply cannot be demonstrated, operation may be advocated because of the lesion may represent a neuroblastoma. Some groups have used percutaneous needle biopsy to differentiate ELS from neuroblastoma (8).

Treatment remains operative resection according to Danielson et al. (9) Surgical removal gives the opportunity to confirm the diagnosis, eliminate the risk of malignancy and ensure that a neuroblastoma is not being left untreated. The associated anomalies such as congenital diaphragmatic hernias are also searched by surgical exploration. Danielson et al advocate a laparoscopic approach to these lesions because it pro-
vides definitive treatment with the benefits associated minimally invasive surgery (9).

Joyeux et al. (10) also stated that minimally invasive surgery for ectopic ELS in small children is a feasible and safe technique. In our patient, an operative resection was recommended. The parents opted for clinical monitoring with imaging methods. Despite the persistence of the lesion, the parents have not yet consented for surgery.

Intra-abdominal ELSs usually remain asymptomatic throughout life; they may be safely observed without surgery unless they become symptomatic or show any change in the characteristics of the radiologic appearance. Clinical observation with imaging methods may be recommended in infants with ELSs. The debate regarding the indications for surgery and its timing compared to simple observation will continue until long-term outcome studies encompassing large numbers of patients can be finished.

References


