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Intraabdominal extrapulmonary sequestration in an infant diagnosed by color Doppler ultrasound

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ABSTRACT

Intra-abdominal extralobar pulmonary sequestration is seen very rarely. When an echogenic mass with cystic spaces below the diaphragm in an infant is seen with ultrasound, color Doppler technique should be used to demonstrate the feeding artery. We present a case of intra-abdominal extrapulmonary sequestration in an infant that was diagnosed by color Doppler ultrasound.

Intra-abdominal extralobar pulmonary sequestration (PS) is a rare, benign congenital malformation in the differential diagnosis of suprarenal lesions [1]. We report a case of infant presenting with an infradiaphragmatic retroperitoneal mass diagnosed as extrapulmonary sequestration by color Doppler ultrasound.

1. Case report

A 28-year-old pregnant woman having uneventful prenatal course underwent a routine ultrasound examination at 28 weeks gestation, which showed a 34 × 24 mm hyperechoic lesion located superior to the left kidney and extralobar sequestration or CCAM were considered in the differential diagnosis. A fetal magnetic resonance imaging study was performed at 30 weeks gestation. Congenital cystic adenoid malformation (CCAM) was considered as a prenatal diagnosis (Fig. 1).

After normal delivery at term gestation, the patient remained asymptomatic and was discharged in satisfactory condition after 24 h of close observation. The plain chest radiography was normal. At the age of one month, abdominal ultrasound revealed an echogenic mass with cystic spaces located below the diaphragm, above the left kidney and medial to the spleen (Fig. 2). A thin feeding artery originating from the aorta and passing to the lesion was

identified by Doppler ultrasound simultaneously (Fig. 3). These findings suggested the presence of extralobar infradiaphragmatic PS and the patient underwent laparotomy surgery at the age of 2 months. The patient had an uneventful recovery and was discharged from 3 days later.

2. Discussion

Pulmonary sequestrations [PS] are rare lesions constituting 1.1%–1.8% of all pulmonary resections, firstly defined by Pryce in 1949 [1,2]. PS are areas of non-functioning lung which have no connection to the bronchial tree. These anomalies can be classified as intra-lobar or extra-lobar. An intralobar sequestration is located within the lung parenchyma, while the extralobar sequestration is separate from the lung, including a separate pleural covering. These lesions possess a systemic arterial blood supply which often arises from the aorta, and the venous drainage may drain into the pulmonary veins, the azygous system or the inferior vena cava [3].

Embryologically, an accessory lung bud develops from anterior part of primitive foregut which is supplied by primitive splanchnic vessels. The initial connection with the foregut usually regresses but may persist as a fibrous pedicle that accompanies the feeding and draining vessels [4]. In some cases, connection to the foregut persists and allows communication with the gastrointestinal tract [2].

Intralobar sequestrations are more common, forming 75–85% of all PS, while only 25% are extralobar form [5]. Intralobar sequestration is usually considered an acquired abnormality of the lung,

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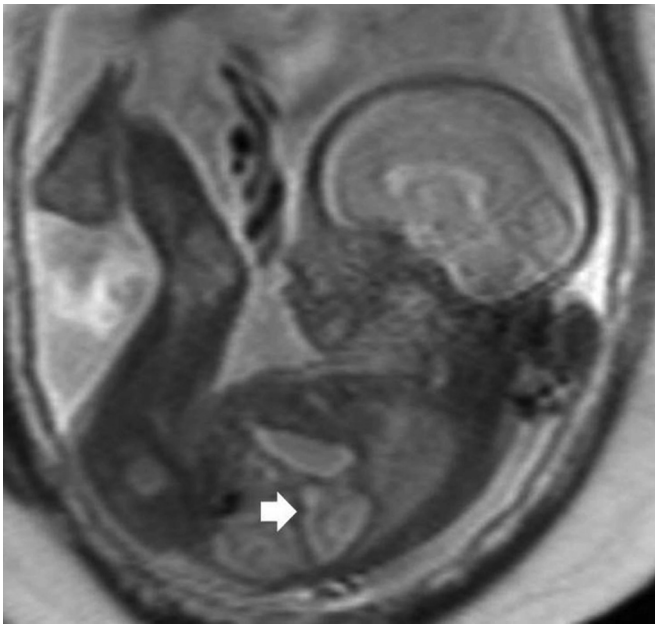*E-mail address:* gokhan.duygulu@gmail.com (G. Duygulu).

Fig. 1. Fetal magnetic resonance imaging reveals a mass with cystic spaces (arrow) located cranial to the left kidney.

develops secondary to recurrent infections and bronchial obstruction [6]. Extralobar PS is often diagnosed in antenatal period or first 6 month of life, however intralobar PS is usually diagnosed in childhood period [7].

Although most of extralobar sequestrations are thorax located, 10% are intra-abdominal or infradiaphragmatic [5]. 90% of intra-abdominal PS are left sided. Intra-abdominal extralobar PS, is usually presented as retroperitoneal mass adjacent to stomach or associated with it. Arterial supply is almost always from aorta and venous drainage is via azygous system, inferior vena cava, or pulmonary veins (25%). 60% of fetuses with extralobar PS have also associated cardiac, diaphragm and other lung anomalies. Because of associated anomalies, extralobar PS is often diagnosed in the first 6
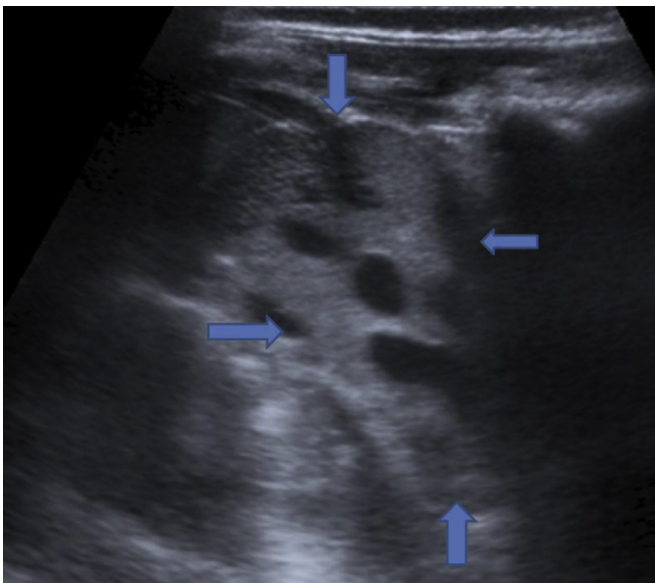

Fig. 2. Longitudinal real-time ultrasound scan of the left side of the abdomen reveals solid echogenic mass with cystic areas (blue arrows).

months of life. However only 14% of intralobar PS have other associated anomalies [8].

Extralobar PS affects males approximately 4 times more often than females. It occurs on the left in 95% of cases and 75% of these are found in the costophrenic sulcus. Mediastinum, pericardium, and within or below the diaphragm are other localizations of extralobar PS [9].

The diagnosis and characterization of congenital lung lesion (CLL) are made primarily by the use of ultrasound. The initial categorization of CLLs is as a solid or cystic mass. Cystic lesions are further classified as macrocystic, microcystic or hybrid lesions. Sequestrations appear as a solid, echogenic lesion. Using Doppler, the systemic feeding vessel can often be identified in these lesions. All centers recommend that fetal patients with CLL receive an initial ultrasound. Decisions regarding the utility of fetal MRI and fetal echocardiogram are based on the interpretation of the ultrasound. The CAM volume ratio (CVR), presence of hydrops, mediastinal shift, reversal of flow within the umbilical vein and abnormal cardiac echo are all useful parameters to aid in the determination of whether fetal therapy may be beneficial. Finally, the presence of placentomegaly (>5 cm in thickness) can be a harbinger of imminent in utero demise [3].

PS are seen as well circumscribed, echogenic, solid masses in antenatal ultrasound examination. It is difficult to differentiate from CCAM. CCAM is supplied by pulmonary arteries, while systemic arteries supplies PS. Thus, demonstration of feeding artery from aorta narrows differential diagnosis [8]. In our case, lesion evaluated with antenatal MR examination was considered as left basal thorax located and CCAM was the first choice of differential diagnosis. After delivery, post-partum color Doppler ultrasound examination (as ultrasound has higher spatial resolution than MR examination) detected infradiaphragmatic lesion localization and feeding artery arising from abdominal aorta. Prior diagnosis was PS according to US findings and this diagnosis was approved histopathologically.

The CVR or CAM volume ratio has emerged as one of the most useful tools in predicting outcomes in prenatally diagnosed pulmonary lesions. The CVR is a measurement of tumor volume normalized by gestational age and is calculated by using ultrasound to measure the pulmonary lesion in 3 dimensions (length, width, height). This volume is multiplied by a constant (0.52) and divided by head circumference (which normalizes for gestational age). The CVR equation is $(L \times H \times W) \times 0.52$/head circumference. The initial report documented that 80% of fetuses with a CVR >1.6 went on to develop hydrops (Crombleholme). A more recent study described a similar predictive value of CVR, but with a cutoff of 2.0 (Cass). In this series, 56% of fetuses with CVR >2.0 required prenatal intervention compared to 3% of the fetuses with CVR<2.0. Clearly, the CVR is a useful prognostic tool for CLLs, but one which is continuing to be studied and refined [3].

The initial intervention that should be considered in patients with large, symptomatic solid or microcystic congenital lung lesions (CLL) is the administration of prenatal steroids. This therapy has been described in several case series, yielding a variable but definite response. Although there are currently no definitive recommendations and no conclusive data to support the use of prenatal steroids for symptomatic CLLs, several centers have independently noted lesion regression and hydrops reversal after administration. The administration of steroids is a two-dose maternal regimen of betamethasone given 24 h apart. The exact mechanism as to how betamethasone induces lesion regression is not clearly defined [3].

In treatment of PS, most authors suggest surgical resection due to concern for infection, malignant degeneration and difficult differention from other tumors. However, some authors recommend management without surgical resection or embolization of feeding artery [4,10].
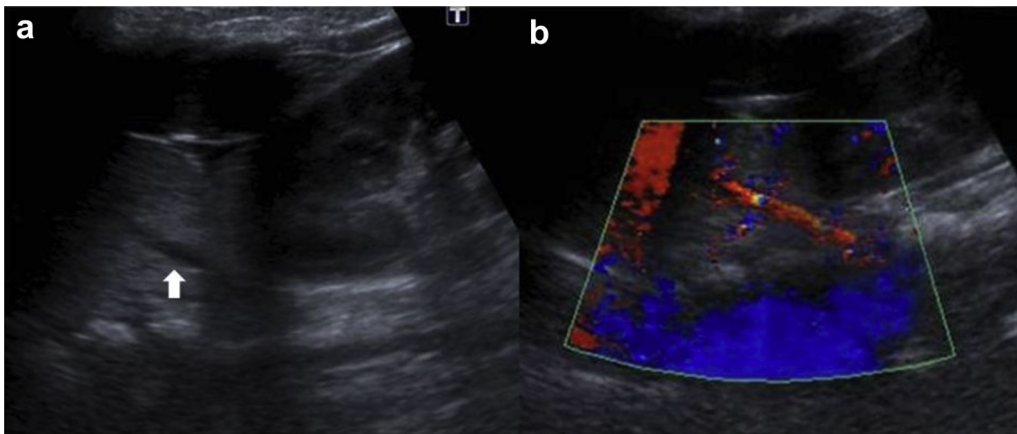
Fig. 3. B-mode (a) and color Doppler sonography (b) demonstrates the mass is located cranial to the left kidney and has a feeding artery (arrow).

3. Conclusion

Color Doppler US is an extremely useful imaging method in differential diagnosis of intraabdominal PS. It should always be applied to patients with suspect of PS in antenatal or neonatal period. When an echogenic mass with cystic spaces below the diaphragm in an infant is seen with ultrasound, color Doppler technique should be used to demonstrate the feeding artery.

Conflicts of interest

The authors declare that there are no conflicts of interest.

Financial disclosure

The authors declare no competing financial interests.

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