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Isolated subdiaphragmatic extralobar pulmonary sequestration: masquerading as suprarenal mass with spontaneous resolution

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Summary

Sub-diaphragmatic extralobar pulmonary sequestration (PS) is an extremely rare malformation defined as a portion of lung tissue that is totally discontinuous from the tracheobronchial tree and usually has its own pleural covering with an anomalous systemic blood supply. It is usually associated with other congenital malformations. The authors present an extremely rare case of isolated sub-diaphragmatic PS which was antenatally detected and followed up with postnatal ultrasound, where it masqueraded as a suprarenal mass; it was totally asymptomatic with spontaneous resolution at age of two years. This case emphasizes to add a differential diagnosis of malformation in congenital supra-renal masses, which remain stable in size and appearance, with possible spontaneous resolution and hence avoid immediate intervention and surgery.

Key words: Extralobar; Pulmonary sequestration; Sub-diaphragmatic; Suprarenal mass.

Introduction

Pulmonary sequestrations (PS) are rare thoracic malformations of unknown exact etiology. Sub-diaphragmatic PS which belong to extralobar type are extremely rare, and are most commonly associated with other congenital malformations [1-4]. The authors report a rare case of sub-diaphragmatic extralobar sequestration without other associated malformations, which was detected antenatally and masquerading as a suprarenal tumor and then resolved spontaneously at two years of age.

Case Report

A 43-year-old multigravida was referred to the present Maternal Fetal Medicine Center at 30 weeks of gestation because of gestational diabetes on insulin; her clinical history was otherwise unremarkable. The obstetric ultrasound examination was performed for the first time in the present center; it showed a well-defined echogenic rounded lesion measuring (1.92×1.94 cm) located superior to left kidney and below the dome of left diaphragm with the left adrenal gland being compressed (Figures 1, 2). Left kidney was normal. Small arteries were seen supplying the lesion likely arising from the upper abdominal aorta (Figure 3). There were no other fetal abnormalities. The fetal growth and welfare parameters were within normal ranges.

Follow-up ultrasound every two to three weeks until delivery showed no change in the lesion size, site or texture with normal fetal growth and welfare parameters. A spontaneous vaginal delivery of a male infant with weight of 3,550 grams was achieved at 38 + 4 weeks of gestation in good condition, without any respiratory problems, and did not require any resuscitation. After birth, vital signs were stable and oxygen saturation was maintained above 95% during feeding. Initial upper abdomen ultrasound followed by CT scan of chest and abdomen with IV contrast were performed for the infant at days 4 and 5 of life, respectively, and confirmed the authors' antenatal findings (Figures 3, 4). The baby remained well without any problems. Urinary catecholamines were normal. Follow-up ultrasound was performed every six months with no gross difference in the size of the lesion. At two years of age the abdomen ultrasound revealed no evidence of any suprarenal masses which was again confirmed after six months.

Discussion

The prenatal diagnosis of PS has been documented by many authors. It is defined as a mass of non-functioning lung tissue that has no connection to the normal tracheobronchial tree and receives its blood supply from the aorta. Because the systemic arterial supply is a pathognomonic feature of PS, identification of a feeding artery arising from the descending aorta is useful for diagnosing and differentiating this condition from congenital cystic adenomatoid malformation of the lung (CCAM), which lacks such a blood supply [1-4].

On the basis of the morphologic patterns, two types of PS are distinguished. Intralobar sequestration is more common (75%) and is localized within the normal lung parenchyma and shares visceral pleura of the involved lobe. The extralobar type is uncommon (25%), and has its own pleural investment and is separate from the normal lung [2, 4].

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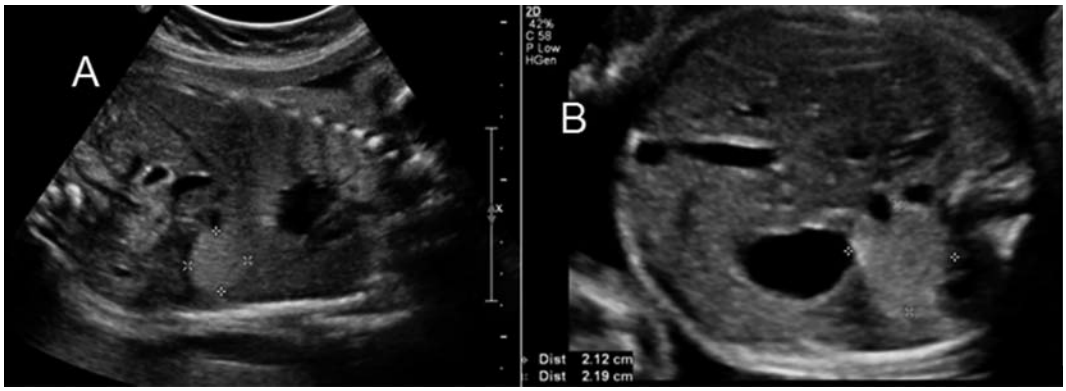

Figure 1. — Antenatal ultrasound image showed a left sub-diaphragmatic suprarenal lesion in a 30-week fetus (longitudinal and cross section view).

Extralobar PS are most commonly found intra-thoracically within the pleural space and usually on the left side in 95% of cases [1]. They may be rarely found in the mediastinum or pericardium. Extralobar sequestrations in intra-abdominal location are exceptionally rare and they are mainly sub-diaphragmatic in the suprarenal area [5]. They are usually associated with other congenital anomalies in more than 50% of cases, such as congenital diaphragmatic hernia, congenital cystic adenomatoid malformation, congenital heart disease, and rarely genitourinary and gastrointestinal anomalies. This is why more than half of patients present in the first six months of life with respiratory distress, congenital heart failure or feeding difficulties [2, 3].

Diagnosis of intra-abdominal PS is difficult and is usually diagnosed histopathologically of surgically resected

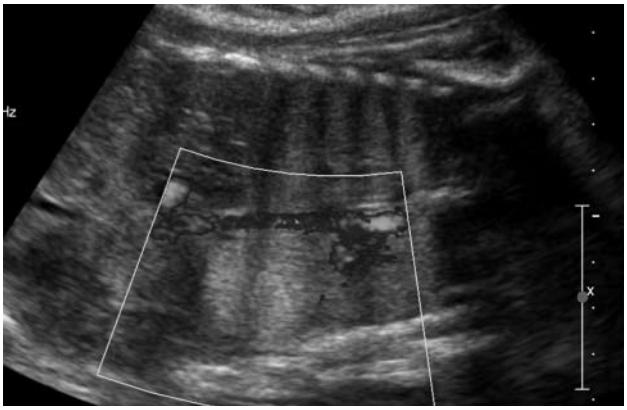

Figure 2. — Antenatal ultrasound image showing the origination of feeding vessel of the lesion from the upper abdominal aorta.

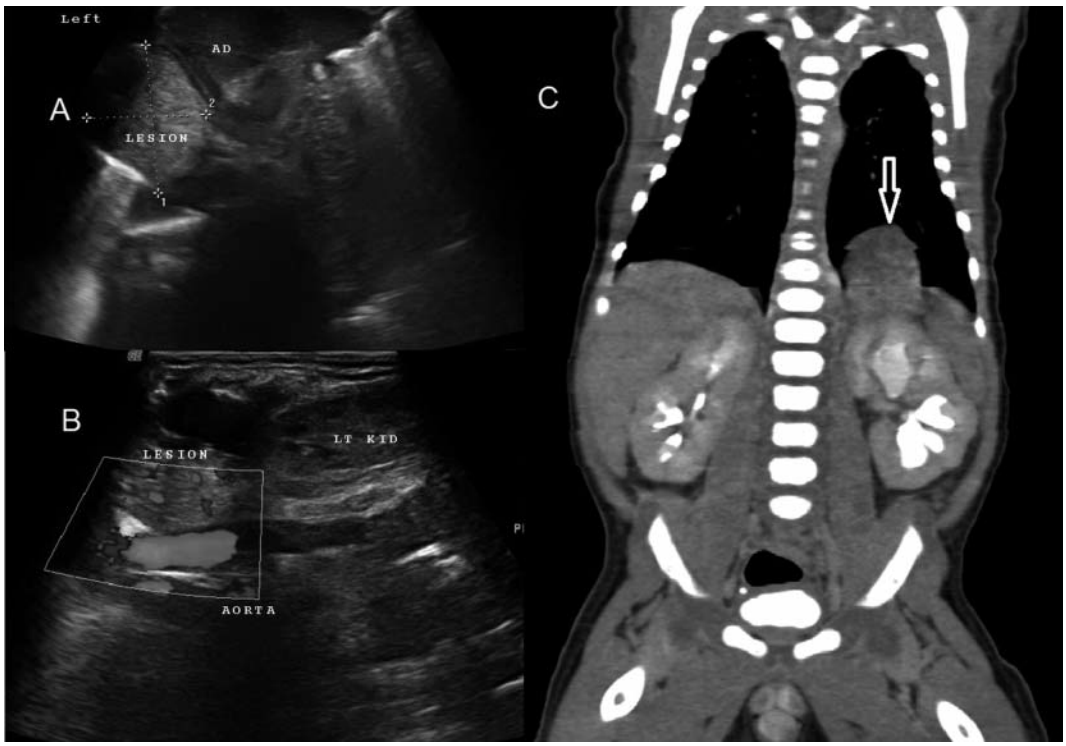

Figure 3. — A, B. post-natal ultrsound of infant's abdomen at day 4 of life showing a left supra-renal mass with the feeding vessels from the upper abdominal aorta. C. post-natal CT-scan chest and abdomen of infant at day 5 of life showing a left supra-renal mass.

lesions. While the radiological differential diagnosis of a suprarenal mass includes variable anomalies and tumors, including neuroblastoma, the presence of systemic feeding vessels on color Doppler in the absence of raised urinary catecholamines narrows the differential [3, 6]. CT or MRI facilitate identification of the anatomical relationships of the lesion, including the origin of the systemic feeding vessels, allowing surgeons to plan for an optimal surgical approach [3].

Spontaneous improvement and resolution of PS has been reported [1, 2]. Although many researchers have described a significant decrease in the size of PS with spontaneous resolution [6-8]. Few studies have concentrated on the natural changes of PS antenatally. Some researchers reported high incidence of significant growth of PS during pregnancy. However, their cases included both PS and microcystic CCAM, which are different entities from both pathogenetic and therapeutic points of view [9]. The present case showed that PS mass volumes remained constant in size antenatally and postnatally with no clinical implications until two years of age when it completely disappeared.

Conclusion

Sub-diaphragmatic extralobar PS as an isolated anomaly is extremely rare and should be a differential diagnosis in a fetus presenting with an unusual abdominal mass. Utilization of color Doppler will assist in making the diagnosis. An expectant management should be attempted in stable and asymptomatic cases as spontaneous resolution could occur.

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