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Sonographic Detection of Fetal Extrathoracic Pulmonary Sequestration

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The use of diagnostic ultrasonography during pregnancy has gained widespread acceptance. Unquestionably this diagnostic modality facilitates discovery of fetal anomalies that might otherwise remain unsuspected. Such discoveries can produce therapeutic dilemmas, but may permit the obstetrician to tailor antepartum and intrapartum care in accordance with potential deleterious effects of the anomaly on the fetus or newborn, and the emotional disturbance that such discovery provokes in patients and family.

We report a case of retroperitoneal pulmonary sequestration that was diagnosed during the antenatal period.

REPORT OF A CASE

A 23-year-old black woman, gravida 4, para 2, abortus 1, had two previous uneventful pregnancies with normal full-term spontaneous vaginal deliveries. At 37 weeks' gestation by dates she was referred for ultrasound examination because of questionable inappropriate size for dates. The examination showed a single, living, intrauterine fetus of 34 weeks' size. Fetal head to body ratio was normal, and femur length concurred with fetal age. A large retroperitoneal mass measuring approximately 3.5 cm in diameter was noted in the right upper quadrant. It was uniformly echogenic with smooth margins (fig. 1). Fetal kidneys, stomach, liver, and diaphragm were visualized as normal. The fetus was in vertex presentation with a grade II posterior placenta. Amniotic fluid volume was normal, and no ascites was noted. Follow-up examination three days later showed no change in the size and appearance of the mass. Because of the location of the tumor and the date in pregnancy at which the discovery was made, the diagnosis of a neuroblastoma was entertained. An at-term vaginal delivery in a tertiary care level institution was planned, and a pediatric surgeon was consulted. The parents agreed to a conservative treatment approach since no obvious deterioration of fetal health was evident. The patient's membranes ruptured prematurely one week after the ultrasound examination, and she spontaneously delivered an infant weighing 2466 grams, appropriate for gestational age, with Apgar scores of 8 and 9 at one and five minutes, respectively. The Ballard score corresponded to 34 weeks.

Physical examination of the newborn revealed no palpable abdominal mass. Chest, abdominal, and long-bone radiographs were normal. Intravenous pyelography showed flattening and slight inferior displacement of the right kidney consistent with a suprarenal mass. Ultrasound examination revealed a well-circumscribed, rather homogenously echogenic mass in the right suprarenal position. The mass appeared separate from the kidney, liver, and inferior vena cava with compression of the latter. These findings were confirmed by computed tomography (Figs. 2*A* and *B*).

The infant underwent exploratory surgery through a right subcostal incision. The mass was inferior and adherent to the liver, arising from the superior surface of an otherwise normal appearing adrenal gland. It was soft, pinkish in color, and solid. The arterial supply was systemic mainly from the right renal artery through the adrenal gland, and one large and several smaller branches from the aorta to the mass. Additional appreciable blood supply was derived from the inferior phrenic artery. Venous return was to the inferior vena cava through "normal" adrenal veins. No communication to the gastrointestinal tract was present. Because of the adherence of the mass to the adrenal gland and the common blood supply, an en bloc resection including the right adrenal was performed.

Pathologic examination revealed an ovoid, spongy, pink-gray mass, measuring 4 cm in diameter, weighing 12 g, and consisting of lung parenchyma at varying stages of maturity. Bronchioles lined with ciliated respiratory epithelium were seen. These findings were consistent with extralobar sequestered lung in the periadrenal location.

DISCUSSION

Pulmonary sequestration is a rare congenital anomaly of the developing lung which was first described over 100 years ago. The term was introduced by Pryce who described an ectopic, nonfunctioning pulmonary mass which lacked communication with the tracheobronchial tree and received blood supply from an anomalous systemic vessel.[1] The two classical forms of sequestration are: 1) intralobar, denoting a lesion that lies within the pulmonary visceral pleura, and 2) extralobar, denoting a lesion which lies outside the pulmonary visceral pleura. The arterial supply to an intralobar sequestration is systemic with pulmonary venous drainage, whereas extralobar lesions usually have

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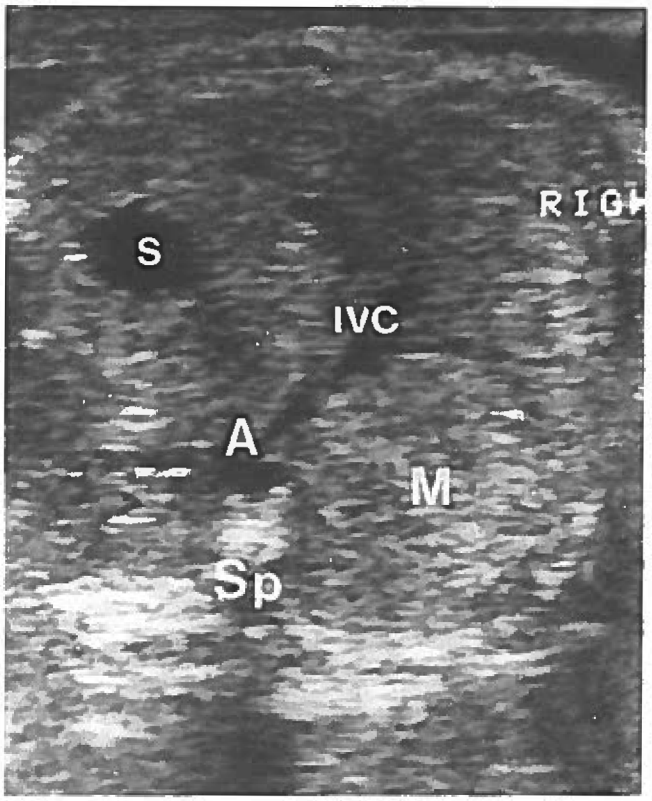
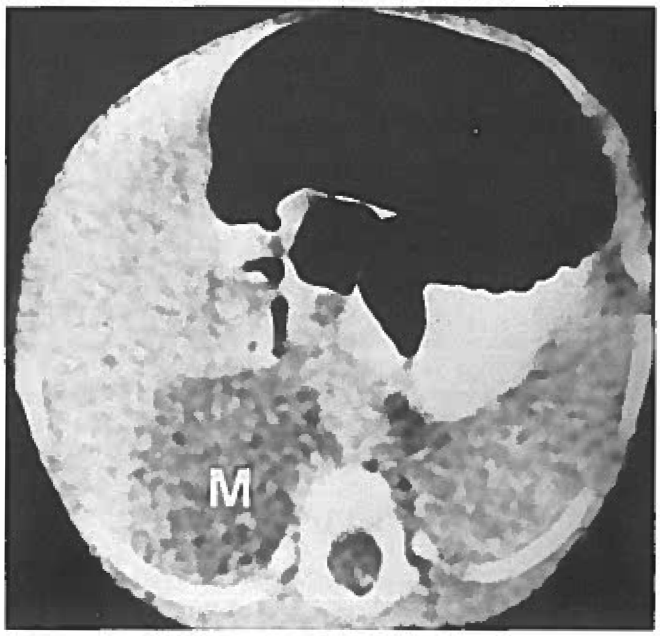


Figure 1 (*left*). Transverse section through fetal abdomen at the level of the fetal stomach showing the retroperitoneal mass. M, mass; SP, spine; A, aorta; IVC, inferior vena cava; s, stomach.



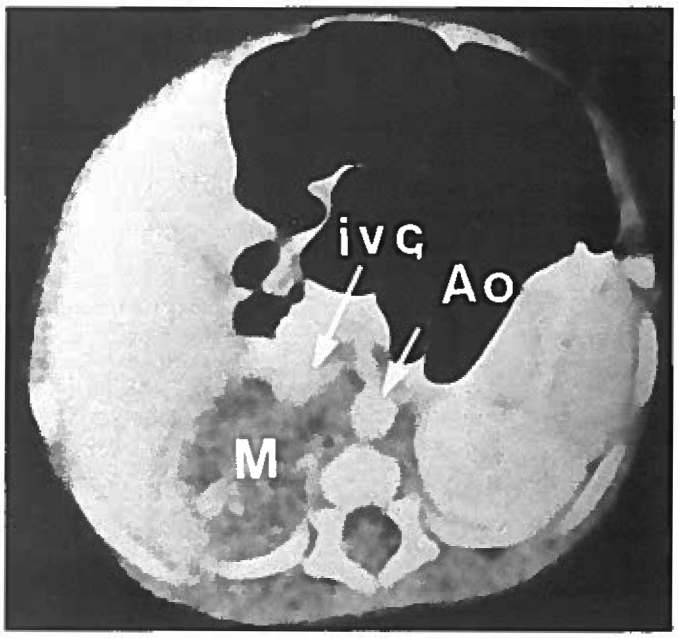


Figure 2 *A* (*lower left*). Unenhanced computed tomographic (CT) scan of the neonatal abdomen demonstrates a low attenuating mass in the right upper quadrant. No calcifications are seen within the mass. M, mass. *B* (*lower right*), enhanced CT scan of the neonatal abdomen demonstrates a mass with peripheral and focal enhancement displacing the inferior vena cava anteriorly. M, mass; Ao, aorta; IVC, inferior vena cava.

both systemic arterial and venous supply. Although the etiology of pulmonary sequestration remains controversial, it is accepted that extralobar and intralobar sequestrations are bronchopulmonary foregut malformations.[2] Twenty-five per cent of pulmonary sequestrations are extralobar;[3,4] the majority of these are left-sided with male predominance. Approximately 5 per cent are located below the diaphragm.[5,6]

The present case depicts an extralobar pulmonary sequestration located in the right suprarenal position. It was first identified prenatally by ultrasonography, thus enabling serial examinations of the lesion and the taking of adequate precautions at delivery. It appeared to be a solid, dense mass with a few hypoechogenic areas. It displaced the right kidney inferiorly and inferior vena cava anteriorly and towards the midline (figs. 1 and 2*B*). The incorrect initial working diagnosis was a result, in part, of the location of the mass. The most common le-

sion in the differential diagnosis given this presentation would be neuroblastoma. Adrenal hemorrhage has been thought to be a postnatal event and to our knowledge not previously recognized in utero. Retroperitoneal teratomas could occur in this area but would be anticipated to be more cystic in appearance.

Pulmonary sequestration and diaphragmatic hernia must be considered with the need for aggressive neonatal support for the latter. Knowledge of the sonographic appearance for postnatal identification of a pulmonary sequestration by ultrasonography is essential since the clinical and radiologic appearance may mimic other lesions such as cystic adenomatoid malformation or bronchogenic cyst. In conclusion, accurate antenatal diagnosis of pulmonary sequestration is feasible. This permits adoption of precautionary measures to deal with any postnatal complications including congestive heart failure, or, rarely, respiratory distress when shunting is present.[7,8] Conservative therapy in the event of pulmonary sequestration can be considered if there is no evidence of fetal deterioration.

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