Prenatal diagnosis of a bronchogenic cyst in an unusual site

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ABSTRACT

We report a rare case of a subdiaphragmatic cyst, diagnosed prenatally during routine ultrasound screening at 25 weeks' gestation. Serial ultrasonographic evaluation demonstrated a slight increase in the size of the cyst during pregnancy. On the basis of the anatomic site and the sonographic features of the lesion, four diagnostic hypotheses were postulated: cystic neuroblastoma, adrenal hemorrhage, duplication of the inferior third of the esophagus and retroperitoneal cystic lymphangioma. A healthy baby, weighing 3300 g and with Apgar scores of 8 and 9, was delivered vaginally at term. He underwent successful surgery at the age of 3 months. The postoperative course was uneventful and the baby is currently doing well. Histology revealed a bronchogenic cyst.

INTRODUCTION

The sonographic finding of an intra-abdominal cystic lesion renders problematic the formulation of the differential diagnosis, with regard to both the nature of the cyst and the organ of origin. In fact, abdominal cystic malformations of mesenteric, omental, pancreatic, cholecdochal, hepatic and ovarian origin, and secondary to intestinal pathology, have been reported in the literature. In the present article, the prenatal ultrasound appearance of an ectopic, intra-abdominal bronchogenic cyst is described. Problems and issues with regard to pathogenesis in addition to differential diagnosis are discussed.

CASE REPORT

A 23-year-old woman, gravida 1, para 0, presented with a normally developing pregnancy, in the course of which she was given vitamin and iron therapy. An ultrasound scan carried out at 25 weeks had demonstrated the presence of a male fetus, whose growth parameters were within the normal range expected for gestational age. An anomaly scan had revealed no malformations, apart from evidence within the abdomen of an anechoic cyst, measuring approximately 16 mm in diameter. The patient was seen for the first time at our tertiary referral unit at 28 weeks' gestation. An ultrasound scan at this time confirmed normal fetal growth (biparietal diameter 71 mm; transabdominal diameter 72 mm; femur length 52 mm; humerus length 48 mm) and the presence of a cystic lesion in the right hypochondrium, in the paracavovascular area. This lesion measured 16 mm in diameter, was entirely anechoic, and had a regular outline and clearly defined margins (Figure 1). The ipsilateral kidney was of normal dimensions and sonographic structure. The other abdominal organs (stomach, liver, gallbladder, bowel loops) also appeared normal with regard to both size and structure. No diaphragmatic defect was detected.

A further ultrasound scan at 38 weeks' gestation showed continuing normal fetal growth (biparietal diameter 90 mm; transabdominal diameter 100 mm; femur length 70 mm; humerus length 61 mm). The cyst now appeared to be clearly subdiaphragmatic in position. It showed a mild increase in size (diameter 27 mm), was anechoic and negative to color Doppler. Its dimensions and structural characteristics did not change with variation in the scanning plane. No peristaltic-type movements were observed internally. The ultrasound features of the lesion suggested a differential diagnosis of cystic neuroblastoma, adrenal hemorrhage, duplication of the inferior third of the esophagus and retroperitoneal cystic lymphangioma. The baby was delivered vaginally at term, weighing 3300 g and with Apgar scores of 8 and 9 at 1 and 5 min, respectively. His clinical condition at birth was good, without signs of respiratory distress. Three months postnatally, follow-up of the baby at our Department of Neonatal Surgery confirmed a good clinical condition, with absence of symptoms. A chest X-ray revealed an isolated opacity measuring 27 mm in diameter, located under the right lung. A computed tomography (CT) scan of the thorax and abdomen was therefore carried out. This showed a low-density (fluid-filled) lesion, with dimensions of 35 × 25 × 43 mm. It was localized above, and medial to, the right kidney, between the vena cava and the aorta, displacing and indenting the vena cava (Figure 2). Adrenal
mucus-secreting, ciliated respiratory-type epithelium, resting on a thin layer of connective tissue. Carriage may also be found within the lesion. The arterial supply to the cyst is independent from the pulmonary arteries. Pulmonary bronchogenic cysts are typically centrally positioned, although they may be located in any area of the parenchyma. These cysts appear well-circumscribed and hypoechogenic on ultrasound. If located in the lung, they can secondarily cause pulmonary hyperechogenicity. Although rare, ectopic bronchogenic cysts have been described in various anatomical regions in the literature. Detection of cervical cysts in the fetus as well as in children, and detection of retroperitoneal and abdominal cysts in the adult, have been reported. As regards the abdominal site, the pinching off of irregular lung budding from the ventral foregut, with abnormal migration into the abdomen before fusion of the diaphragm, has been hypothesized. However, the pathogenesis of abdominal cysts is unknown.

We report for the first time the prenatal ultrasound observation of an intra-abdominal bronchogenic cyst. The site and sonographic characteristics of the lesion made us consider the differential diagnosis of cystic renal hamartoma, adrenal hemorrhage, duplication of the inferior third of the esophagus and retroperitoneal cystic lymphangioma. The same ultrasound features are common to all these diagnostic options. The diagnostic hypothesis of an intra-abdominal bronchogenic cyst was not considered prenatally or postnataally. A definitive preoperative diagnosis in either a neonate or an adult appears to be difficult to make. The diagnosis can result only from the histological examination. It is clear, therefore, that definitive diagnosis is not possible prenatally. A prenatal diagnosis does not indicate any variation in the management of either the pregnancy or its delivery. Furthermore, with the exception of adrenal hemorrhage that does not require surgery, prenatal diagnosis does not change the indication for neonatal surgery. In the case of any of the other hypothesized lesions, surgery is indicated to enable a precise diagnosis to be made, and to prevent or eliminate any possible symptoms caused by compression. The risk of a malignant transformation is also excluded in some cases.

Our clinical case, whilst raising once again the problems in defining the etiology and pathogenesis of these lesions, confirms the diagnostic difficulty presented by sonographic findings. However, the prenatal diagnosis of a possible bronchogenic cyst enables postnatal diagnostic assessment and surgical treatment to be initiated. Thus, in spite of the rarity of our observations, this report suggests that an ectopic bronchogenic cyst needs to be considered in the differential diagnosis of intra-abdominal cystic lesions, especially when located in the upper abdominal cavity. In addition to the previously stated risks, early elective surgery of an intra-abdominal bronchogenic cyst prevents other potentially life-threatening complications, such as infection and perioperative hemorrhage.

REFERENCES

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