Fetal Ovarian Cyst

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This case report demonstrates a complex fetal ovarian cyst. A fetal ovarian cyst is a fluid-filled ovarian tumor. Ovarian cysts are rarely detected in infants but are slightly more common on fetal sonography. The majority of cysts are benign corpus luteal cysts. The size and appearance of the cysts may vary. The largest cyst could possibly occupy the entire abdomen. The cyst will most likely be anechoic, but it can be complicated due to hemorrhage or torsion. Difficulties may arise when trying to differentiate a large ovarian cyst from other cystic lesions, such as enteric or mesenteric cysts. Both placental and maternal hormones can cause excess stimulation of the fetal ovary. A decrease in hormone stimulation after birth causes most neonatal ovarian cysts to spontaneously regress.

Key words: fetal ovarian cyst, complex cyst, 2D sonography, 3D sonography

Case Presentation

A patient in her late 20s, gravida 1, para 0, first visited our office for a fetal survey and amniocentesis due to an abnormal AFP that indicated an increased risk for Down syndrome. She was 17 weeks and 1 day, and sonography revealed no abnormalities. The patient also elected to have an amniocentesis that produced a normal 46, XX karyotype. She was referred back to our diagnostic clinic 16 weeks later because a questionable fetal ovarian cyst was seen on a sonogram in her doctor's office. The 2D sonographic examination was performed using a GE Voluson 730 Expert with a curved-array 2- to 7-MHz transducer. The sonogram revealed a 2.1 × 1.9-cm cyst of mixed echogenicity located on the left side of the fetal pelvis. There appeared to be layering within the cyst (Fig. 1). The cyst was identified separate from the left kidney and bladder (Fig. 2). Three-dimensional imaging of the cyst was also performed, which re-

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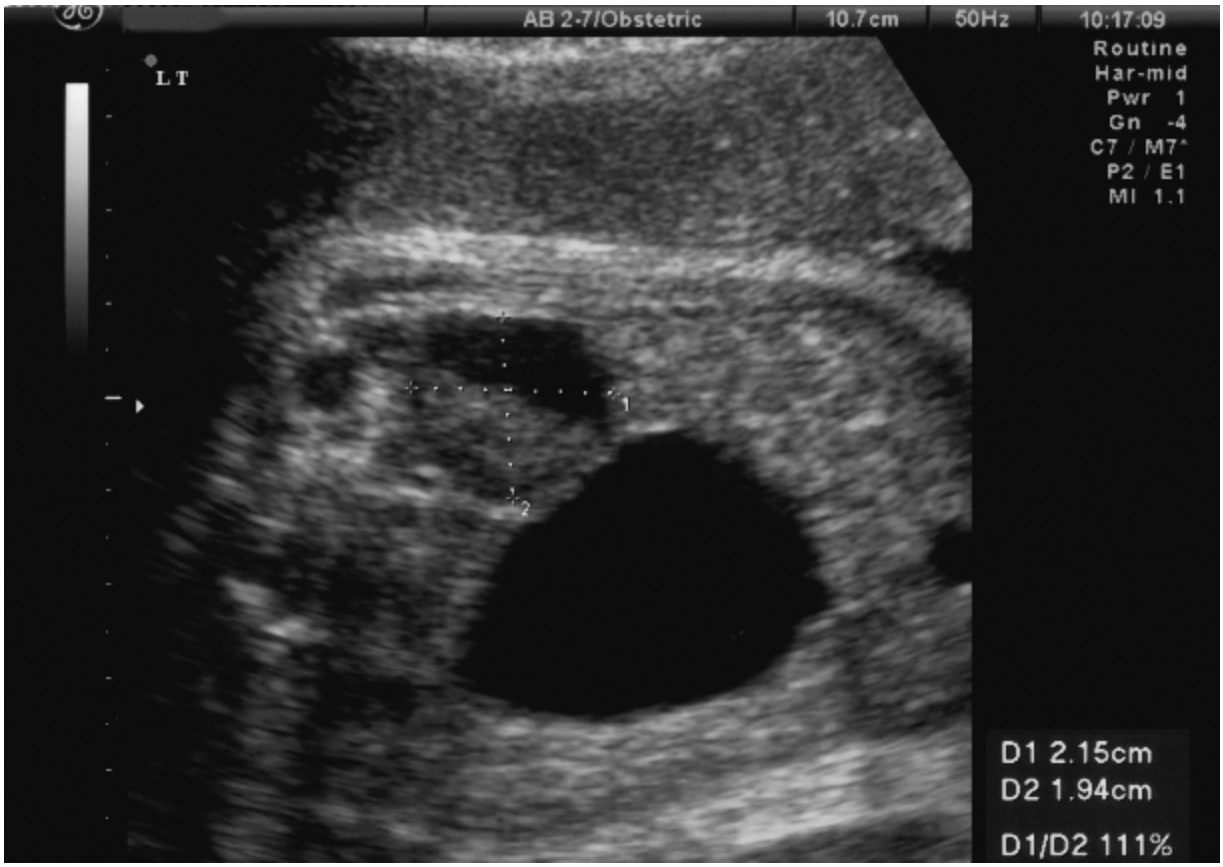

FIG. 1. A 2D image of the complex fetal ovarian cyst located on the left side.

vealed a complex mass with smooth borders (Fig. 3). The 3D image helped to better differentiate the ovarian cyst from the bladder and kidney. It also demonstrated the mixed echogenicity of the cyst. We recommended a follow-up sonogram on the baby after birth and suggested that the parents meet with a pediatric surgeon before the baby was born in case surgery was necessary. Sonography was performed on the baby one month after birth, and the cyst was still present. The surgeon recommended that the mother stop breastfeeding the baby so that there would be no effect of maternal hormones. He asked to see the baby again about 2 months later to see if the cyst had resolved. If it had grown to greater than 5 cm in size, then he would consider intervention, including a needle aspiration of the cystic fluid. If the follow-up sonogram demonstrated that the cyst was complicated, then laparoscopic or surgical management might be needed to exclude a tumor and intestinal duplication or a torsed ovary. We are awaiting the follow-up assessment.

Discussion

A fetal ovarian cyst is a fluid-filled ovarian tumor. The epidemiology is unknown. Most of the cysts are follicular in origin. Fetal pituitary gonadotropins such as the follicle-stimulating hormone (FSH) and luteinizing hormone (LH), maternal and fetal estrogens, and placental human chorionic gonadotropin (hCG) all influence fetal follicle and therefore cyst development. Several theories have described the hormonal control of fetal cyst formation. The theories link cyst formation to (1) fetal gonadotropin, (2) placental hCG, and (3) a preterm or immature hypothalamic-pituitary-ovarian axis (gonadostat). Most support has come from the first theory. There is proof that gonadotropins (LH and FSH) are needed for follicle and

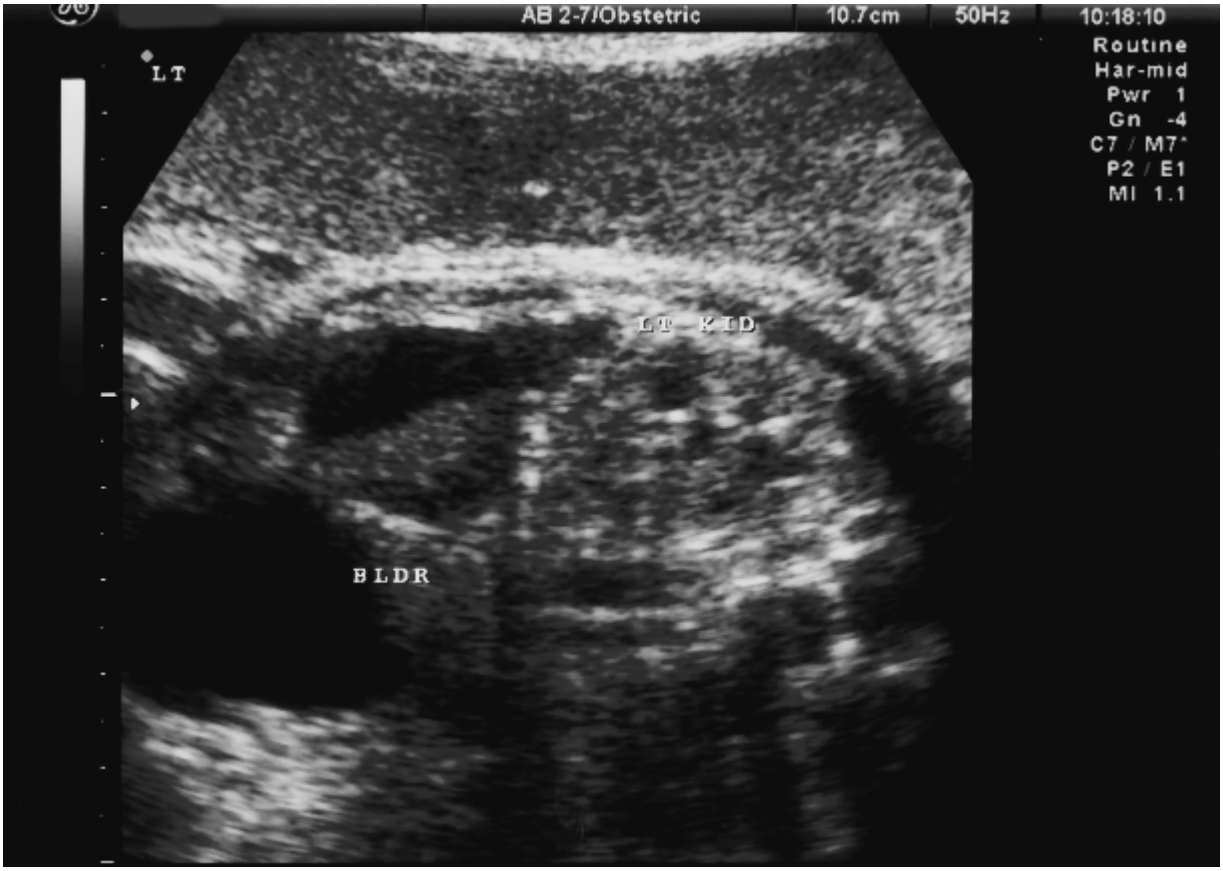

FIG. 2. A 2D image demonstrating the complex cyst separate from the left kidney (LT KID) and bladder (BLDR).

therefore cyst development. There may be an early FSH peak between the 20th and 30th weeks of gestation, which correlates with the timing of fetal cyst development. The second theory shows a relationship between placental hCG and fetal cyst formation. Support for this theory comes from evidence that there is an increased occurrence of fetal ovarian cysts in pregnancies complicated by Rh isoimmunization, gestational diabetes, and preeclampsia. These conditions are all linked with excessive levels of serum chorionic gonadotropins. Finally, it has been recommended that immature hypothalamic-pituitary-ovarian feedback may be associated with the formation of fetal cysts. The hypothalamic-pituitary-ovarian axis matures after 29 weeks' gestation in the presence of increased levels of fetoplacental estrogens. In very premature infants and term infants with partial maturation of the "gonadostat," Bryant and Laufer[1] postulate that ovarian cysts may occur as insufficient negative feedback to the pituitary causes a form of ovarian hyperstimulation and therefore increased cyst and follicle production.

Fetal ovarian cysts are rarely linked to other chromosomal or structural anomalies. Some cases may be associated with very rare genetic syndromes such as the McKusick-Kaufman syndrome. Associated findings with this syndrome are hydrometrocolpos, polydactyly, and congenital heart disease.[2] Congenital hypothyroidism can be an anomaly linked to fetal ovarian cyst. Evaluation of thyroid function in any neonate with an ovarian cyst has been recommended. Nevertheless, most cases of fetal ovarian cysts are sporadic.[1]

The prognosis is generally excellent. Most will suddenly resolve without any postnatal treatment. However, lethal complications can occur with very large cysts due to ovarian torsion and hemorrhage. In some studies, the incidence of torsion has been reported in as high as 50% to 78% of fetal/neonatal

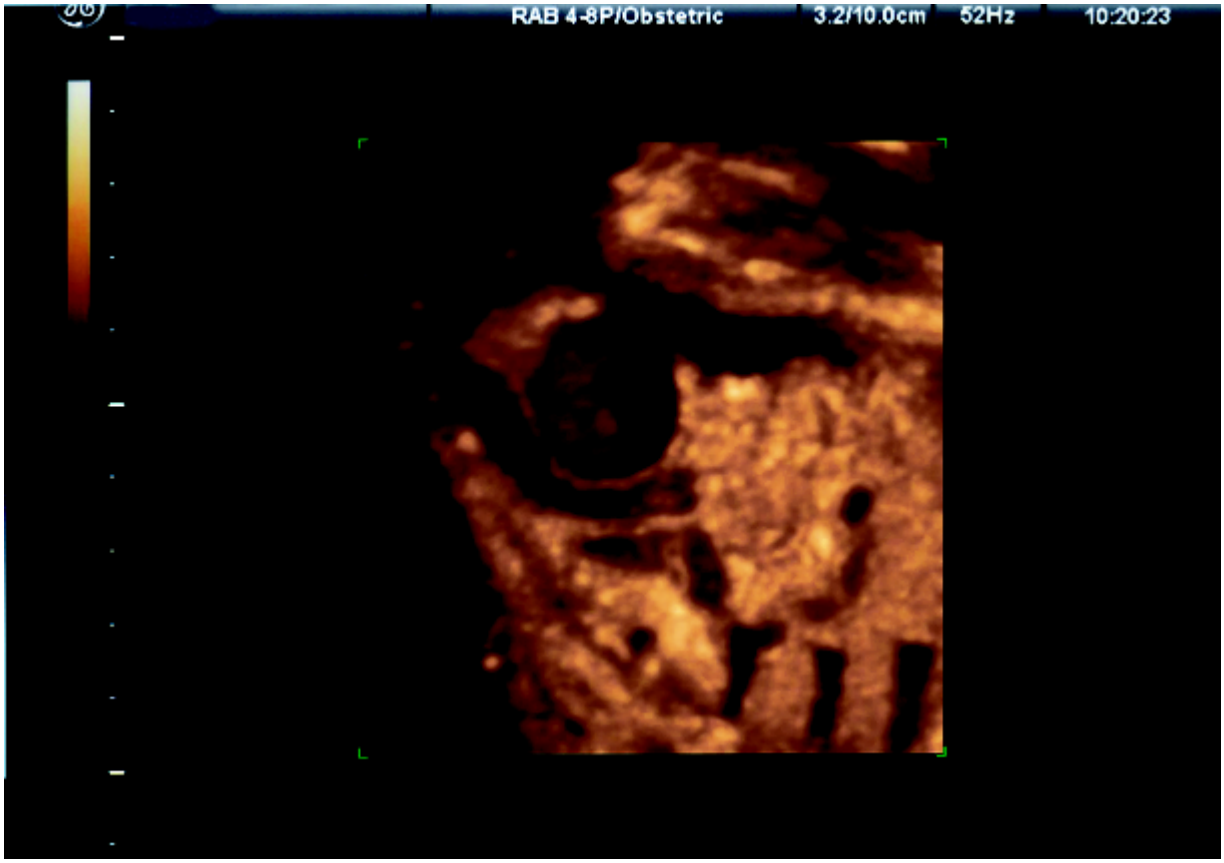

FIG. 3. A 3D image revealing a complex mass in the left fetal pelvis.

cysts. This report has led to a proposal for early surgical intervention. It is thought that more torsion takes place prenatally than postnatally; therefore, many authors think that it is reasonable to intervene with in utero cyst aspiration and decompression in simple cysts >2 cm. Torsion can cause loss of the ovary, affect future fertility, and cause secondary complications that can be even more serious. It is important for the physician to understand all the risk factors when trying to determine whether to intervene. Controversy does exist over intervention. Some groups are conservative in regard to all simple, asymptomatic incidental fetal ovarian cysts, regardless of size, and some are more aggressive. The goal of intervention is to protect ovarian tissue, prevent complication, address acute issues, but limit invasive surgery.[1]

The sonographic appearance is anechoic in an uncomplicated cyst. Sonographically complicated cysts appear complex, containing echogenic debris, septa, or retracted clot. Echogenic walls may be produced by dystrophic calcification. Difficulties may arise when trying to differentiate a large ovarian cyst from other cystic lesions.[3] Diagnosis should be based on four criteria. One is to confirm female sex. The next is to identify the presence of a cystic structure regular in shape and located off the midline. The third criterion is to identify normal urinary tract anatomy, and the last is to identify normal gastrointestinal tract structures.[1] Intestinal obstruction can occur when there is a very large cyst. Because the broad ligament in utero is stretchable, it can be found at any location in the abdomen. The amniotic fluid should be normal unless there is gut obstruction. In this case, polyhydramnios may develop. Development of ovarian cysts should generally not occur before 23 weeks.[2]

There are several differential diagnoses to consider: hydronephrosis, distended urinary bladder,

renal cyst, cystic renal dysplasia, urinoma, choledochal cyst, and liver cyst.[4]

Conclusion

The prognosis of fetal ovarian cysts is excellent. Most will spontaneously regress. Serial sonograms may be performed to evaluate cysts less than 4 cm. They usually resolve within 3 to 4 months. If a cyst is greater than 4 cm, there is a higher risk of torsion.[3] If the cyst is still present after serial postnatal sonograms, surgery may be necessary. Operative indications include cysts greater then 6 cm in diameter, evidence of torsion or hemorrhage, or failure of regression.[2] New treatment options do exist and include in utero aspiration, laparoscopic intervention, and neonatal aspiration. The risk of cyst complications may be decreased by these procedures. Considering the controversy that continues over how and when to intervene, more trials comparing treatment options and intervention criteria are needed.[1]

Several issues should be considered in the immediate newborn period. Resuscitation may be necessary if the cyst is so large that severe abdominal distention prevents efficient breathing. In that case, the infant should be transported to a tertiary facility with a pediatric surgeon. An abdominal sonogram should be performed to confirm the prenatal diagnosis. If a very large mass is present, additional contrast studies of the urinary and gastrointestinal tracts may be necessary.[2]

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