



Fetal Intra-abdominal Cyst: An Unusual Presentation

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Abstract A 26-year-old woman with spontaneous conception had “low risk” result in first-trimester screening. At the time of anomaly scan a large intra-abdominal cyst was seen in the fetus. However, the exact origin of the cyst could not be confirmed. Serial monitoring of the cyst was done by ultrasound scan and the size of the cyst remained stable throughout the pregnancy. The mother had a cesarean section at 36 weeks for obstetric reason. Investigations on the newborn suggested choledochal cyst but the laparotomy revealed dilated duodenum with multiple atretic bowel loops. Ladd’s procedure was performed but the baby died postoperatively.

Keywords Fetal abdominal cyst · Intestinal atresia · Prenatal diagnosis · Choledochal cyst · Fetal mesenteric cyst · Short-gut syndrome

Introduction

Fetal intra-abdominal cysts are not uncommon findings during routine antenatal scans. The cysts can arise from any organ but commonly they originate from gastrointestinal or urogenital systems. Cystic masses can be easily identified on ultrasound scan, owing to the hypoechogenic nature of the lesion. However, precise diagnosis can be quite challenging with respect to the origin of the mass [1]. We report a case of “intra-abdominal cyst” detected

antenatally during second trimester, which was later found to be due to multiple bowel atresias at laparotomy.

Report of Case

A 26-year-old woman (G3P0), from a nonconsanguineous marriage was sent to our unit at 12 weeks of gestational age for first trimester screening. Her combined first trimester risk was “low”. Her first pregnancy was a spontaneous miscarriage at 16 weeks and she had an ectopic pregnancy in the left fallopian tube during her second pregnancy for which she had a salpingectomy.

She returned for an anomaly scan at 19 weeks, during which the ultrasound scan revealed a cystic structure within the fetal abdomen. The cyst was sausage-shaped measuring 10 × 10 mm across the abdomen and was seen separately from the stomach and the bladder (Fig. 1). The left kidney was well visualized. However, the right kidney was not clearly seen, most likely due to the presence of the abdominal cyst. The exact origin of the cyst could not be ascertained. In addition, the umbilical cord had a single umbilical artery. Fetal growth and amniotic fluid were normal. There were no other obvious structural defects or significant markers of chromosomal abnormalities. Fetal cardiology had normal connections and flow patterns.

The mother was re-scanned in 3 weeks and the cyst persisted, crossing the midline as depicted in the image (Fig. 2).

We continued to monitor the cyst at regular intervals of 3–4 weeks throughout the pregnancy and the cyst remained stable in size extending across the abdomen. The cyst had clear contents and peristaltic movements were not seen within the cyst and hence a provisional diagnosis of a mesenteric or a pseudocyst was made (Fig. 3).

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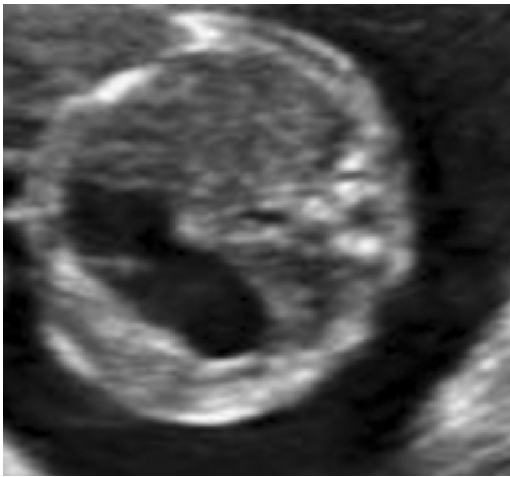


Fig. 1 Intra-abdominal cyst seen at 19 weeks

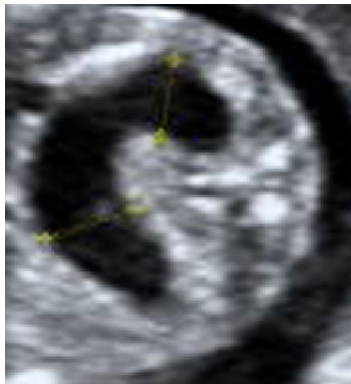


Fig. 2 The same cyst seen at 22 weeks

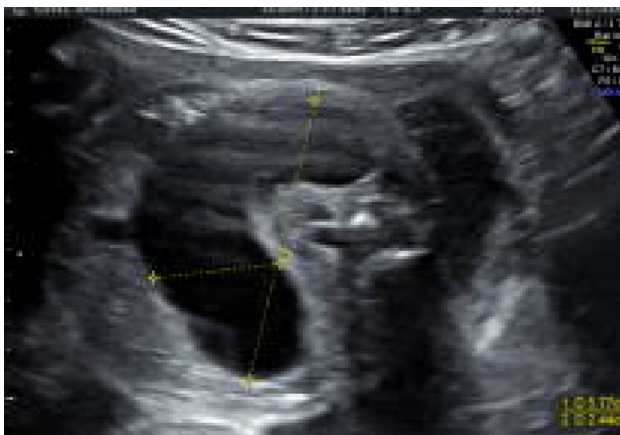


Fig. 3 Cyst seen at 35 weeks

Meanwhile, the mother developed gestational diabetes mellitus requiring insulin. At 36 weeks, a female baby weighing 2.16 kg was delivered by cesarean section for lack of progress in labor. The baby had normal APGAR score at delivery and remained stable. In view of the antenatal detection of the abdominal cyst, the baby

underwent radiograph, ultrasound, CT, and MRI scans. The following differential diagnoses were considered in the newborn baby:

- Gastric duplication cyst with compression of common bile duct (CBD)
- *Radiograph* suggested dilated stomach with gasless intestines
- *Ultrasound scan* suggested
 - intrahepatic biliary ductal dilatation with associated fusiform cystic dilatation in right hypochondrium extending to midabdomen
 - choledochal cyst (type 1 with involvement of proximal CBD)
- *CT scan* suggested a choledochal cyst (type 1 or 2) or an omental cyst. However, this was unlikely to be a gastric duplication cyst as it was not thick walled
- *MRI scan* suggested a choledochal cyst (type 1 or 2) or a bilioma secondary to ruptured CBD

The baby was stable in early postnatal period. However, in view of the discrepancies in the diagnosis, she was taken for an exploratory laparotomy on the third postnatal day. During laparotomy, it was observed that the duodenum was dilated with multiple intestinal atretic segments. So a Ladd's procedure was performed which involves untwisting the intestine, dividing the adhesive bands, widening the mesentery so that the bowel is replaced back in the safe nonrotated position. The poor prognosis of the baby was explained to the parents and they opted not to escalate the treatment and unfortunately the baby died on the third postoperative day.

Discussion

Fetal cystic abdominal lesions are not infrequent findings during antenatal ultrasound. They may originate from various organs of the fetal abdomen like gastrointestinal, ovarian, hepatobiliary, renal, adrenal, splenic, or pancreatic tissue. These may also represent meconium cysts, hydrometrocolpos or umbilical vein varices. This wide variability of origin of cysts often leads to difficulty in classifying them and predict the prognosis [2].

Most frequent intra-abdominal cysts in the female fetuses are ovarian in origin. Certain causes like immature hypothalamopituitary ovarian axis fetal gonadotropins and human chorionic gonadotropins are known to be involved in their pathogenesis [3]. Abdominal cysts often resolve or remain small and are usually associated with good outcome. However, rarely, there can be a more serious underlying gastrointestinal pathological condition and hence, close surveillance in the perinatal period is recommended [4].

Fetal intestinal atresia is a congenital defect in which part of the intestine fails to canalize properly resulting in obstruction or blockage. Nearly 15% of abdominal masses seen in neonatal age group originate from gastrointestinal tract [1].

Bowel atresias are caused by vascular insult to the developing gut leading to failure to canalize. The ischemic injury may be by primary vascular accident or secondary to mechanical obstruction due to in utero volvulus or inspissated tenacious meconium ileus. Nearly 30% of bowel obstructions are associated with cystic fibrosis [1]. Atresias of the large intestine can occur, albeit rarely.

The most common site of intestinal atresia is the small intestine (jejunum and ileum). The incidence of jejunal and ileal atresia ranges from one in 1500 to 12,000 births [5].

Duodenal atresia occurs in one in 10,000 to 40,000 births. Approximately 30% of infants with duodenal atresia have a chromosomal anomaly, primarily Down syndrome (DS) [5]. Fifty to seventy-five percent of newborns with DS will have other birth defects, like congenital heart disease, renal, and gastrointestinal abnormalities.

Cystic fibrosis and meconium plugs in the fetal bowel can mimic intestinal atresia. Dilated fetal bowel is a sonographic finding that is associated with meconium ileus, a feature of cystic fibrosis (CF). Prenatal diagnosis of CF is possible through analysis of the cystic fibrosis transmembrane regulator gene mutations [6].

Technical refinements and increasingly sophisticated equipment have led to higher sensitivity in prenatal diagnosis of congenital malformations; however, such progress may be accompanied by decreased specificity because of the high false positive rate regarding diagnoses of certain anomalies, unnecessary psychological burden to prospective parents may ensue. This issue should be dealt with sensitively [7].

Fetal intestinal atresia can be diagnosed by antenatal ultrasound scan and the evaluation of the bowel should be a part of routine second trimester anomaly scan. The evaluation of the fetal bowel is usually subjective. In a prospective study of 130 fetuses, the normal ultrasonographic appearance of fetal colon and small bowel were studied [8]. The colon appeared as a continuous tubular structure in the periphery of the abdominal cavity. The diameter of the colon gradually increased with the gestational age reaching maximum of 18 mm at term. Whereas the small bowel was located centrally and never exceeded 7 mm in diameter or 15 mm in length. In addition, peristalsis was routinely seen in small bowel but was not seen in the colon [8].

Fetal MRI has been used as a complimentary imaging modality to ultrasound scan in prenatal evaluation of gastrointestinal anomalies. It can be considered a valuable tool

not only for confirming or excluding but also for providing additional information to fetal ultrasonographic findings [9].

The causes of bowel obstruction and level of atresia cannot be accurately determined by ultrasound scan. They are usually associated with polyhydramnios, particularly when the level of obstruction/atresia is high. However, lower intestinal obstruction/atresia may not have associated polyhydramnios, like in our case. Other associated abnormalities and aneuploidies are rare. When abdominal cysts are diagnosed antenatally, particularly in early gestational ages, then possibility of bowel atresia must be considered. If there is a strong suspicion, the parents need to be counseled by pediatric surgeon so that they know what to expect when the baby is born. The baby should be delivered in a tertiary care center where facilities are available to handle such babies.

In most of the cases, the atretic segments are removed and the healthy segments are reconnected. Long-term effects depend on the amount of the intestine that is removed and the associated defects. These children need to be diligently followed up postnatally for development of symptoms of “short gut syndrome”, which occurs as a result of removal of parts of the small intestine by surgery. The nutrients are not adequately absorbed and hence, these children may present with diarrhoea, fatigue, swelling of the legs, and weight loss. These children may need symptomatic treatment in the form of high calorie diet, vitamin, and iron supplementations on regular basis. Rarely these children may even require small bowel transplantation [10].

Conclusion

Fetal abdominal cysts usually present in the late second or third trimesters. Bowel atresia is relatively rare in fetuses. Presentation of bowel atresia as a “fetal abdominal cyst”, particularly in the early second trimester, is an unusual occurrence. We emphasize the importance of considering bowel atresia as a differential diagnosis, especially when the abdominal cyst extends into both sides of the fetal abdomen and presents in the midtrimester. Bowel atresias, particularly when multiple levels are involved, have much more serious consequences to the baby postnatally. Additional modalities of imaging antenatally, may be of limited value. Hence, index of suspicion should be high, considering the high mortality and morbidity associated with the condition.

Compliance with Ethical Standards

Conflict of interest None.

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