CASE REPORT

Isolated ascites in a newborn with 'apple peel' jejunal atresia

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SUMMARY

Isolated fetal ascites was diagnosed at 20 weeks in a primiparous woman with no significant medical history. Progressive fetal ascites worsened after 28 weeks and resulted in fetal hydroceles. Delivery was by caesarian section at 33 weeks, preceded by reduction of fetal ascites under ultrasound guidance. Following delivery, the baby required further reduction of abdominal fluid and endotracheal intubation to provide respiratory support. An extensive set of investigations, including metabolic and genetic screening, was performed; all results were negative. On day two of life, the baby developed bilious aspirates and an abdominal radiograph suggested intestinal obstruction. At laparotomy, an 'apple peel' jejunal atresia, abnormal mesentery with precarious blood supply and a proximal perforation were identified and the perforation 'sewn over'. The postoperative course was unremarkable, with Monogen feeds tolerated three weeks later. The baby continued to thrive at one year, tolerating increasing amount of long-chain fatty acids in diet.

BACKGROUND

'Apple peel' jejunal atresia is a rare condition and may associate multiple anomalies, being characterised by atresia and wrapping of the distal small intestine in a spiral pattern around a thin vascular pedicle. These babies present with neonatal intestinal obstruction, requiring surgical intervention, usually resection and primary anastomosis. The prognosis is guarded,¹ and depends on the existence and extent of bowel necrosis, usually due to the precarious blood supply. In our case, the presentation was unusual, with the intestinal obstruction causing fetal ascites. The mechanism was likely vascular, although possible inflammation due to perforation followed by meconium peritonitis may be considered. The management required a joint team of obstetricians, paediatricians and surgeons.

CASE PRESENTATION

Isolated fetal ascites was diagnosed in a primiparous woman (blood group O negative, no antibodies), with no significant medical history, at 20 weeks gestation. Progressive fetal ascites and polyhydramnios of uncertain aetiology gradually worsened after 28 weeks and resulted

in large fetal hydroceles. Infection screening was negative. The fetal ultrasound did not inform with regards to aetiology and no peritoneal calcifications were visible. In view of massive polyhydramnios, worsening ascites and concerns about fetal well-being, delivery was by caesarian section at 33 weeks gestation. To facilitate delivery, this was preceded by reduction of fetal ascites under ultrasound guidance during which 440 mL of serous fluid was aspirated from the fetal abdominal cavity. After delivery, the baby required further aspiration of abdominal fluid (<100 mL) along with endotracheal intubation for respiratory support. Due to drainage of ascitic fluid, the baby developed low serum albumin and generalised oedema, which subsequently resolved. An echocardiography was normal, as was the postnatal abdominal ultrasound which only demonstrated significant ascites and did not offer information regarding the possible aetiology (figure 1). An extensive set of investigations was performed; however, all results remained negative. Screening for congenital infections, metabolic screening, genetic testing and cranial ultrasound were normal. The ascitic fluid did not show an increase in the level of triglycerides and lactate dehydrogenase value and liver function tests were normal. Increasing abdominal distension and bilious aspirates on day two of life were investigated with abdominal x-ray which showed a paucity of bowel gas, raising the possibility of bowel obstruction (figure 2). In view of the clinical picture suggesting abdominal obstruction correlated with x-ray findings, the baby underwent exploratory laparotomy.

At laparotomy, an 'apple peel' jejunal atresia, abnormal mesentery with precarious blood supply and a proximal perforation were identified. The distal bowel appeared volvulated and no calciform ligament was observed. The perforation was closed by primary anastomosis and mesentery was straightened. No bowel was resected. The postoperative course was unremarkable, with Monogen (medium chain triglycerides feeds) being tolerated three weeks later prior to discharge home. At one year of age, the baby continues to thrive and long-chain fatty acids are progressively introduced in diet.

DISCUSSION

Isolated ascites represents abnormal fluid collection in the abdominal cavity without evidence

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Figure 1 Abdominal ultrasound.

of other fluid accumulation. It is a separate entity from hydrops fetalis, with a different pathophysiology and therefore different prognosis. The prevalence of isolated fetal ascites is not clear and an aetiology for fetal ascites is only identified in 70%–90% of cases.² Fetal ascites may be the result of various mechanisms, and intestinal obstruction with meconium peritonitis is one of the most common gastrointestinal disorders associated with isolated ascites.^{3,4} Ascites may consist of transudates (thin, low protein count and low specific gravity) or exudates (high protein count and high specific gravity). The quality of the ascitic fluid may suggest aetiology and inform management.⁵ For example, the existence of a meconium ascitic fluid may suggest perforation and prompt early laparotomy in the context of suggestive clinical picture.

The overall prognosis for babies with isolated ascites is guarded; many of the causes cannot be treated antenatally and there is little data on long-term outcomes. Nose *et al* demonstrated a strong correlation between prognosis and gestation at

Figure 2 Abdominal x-ray.

presentation, negative prognosis being associated with lower gestational age at presentation.⁶

Postnatal management is condition specific, and it may include nutritional interventions, surgical or medical treatment.

There are different types of intestinal atresia, such as duodenal atresia, jejunal atresia, ileal atresia or colon atresia. Duodenal atresia is the most common type, followed by ileal atresia. The most common cause of non-duodenal intestinal atresia is a vascular accident in utero, leading to decreased intestinal perfusion and ischaemia of the bowel, followed by narrowing, or complete obliteration of the intestinal lumen. The affected area of bowel takes a spiral configuration, 'apple peel' like. The disorder is uncommon and may associate proximal jejunal or distal duodenal atresia, agenesis of the dorsal mesentery and variable parts of the distal small bowel, agenesis of the distal superior mesenteric artery and possibly a malrotated microcolon. The 'apple peel' appearance can be sometimes seen on the preoperative contrast enema. The 'apple peel' atresia is a rare disease, constituting approximately 10% of all small bowel atresia,⁷ equivalent to one in 100 000 live births; it has occurred in families¹ and may be associated with other malformations. Ileal atresia can also result as a complication of meconium ileus. Most newborns with intestinal obstruction present with abdominal distension and bilious vomiting.

In a case series by Bishry *et al*, of 12 patients with isolated fetal ascites, 10 babies survived, two babies required bowel surgery and ascites resolved spontaneously in five cases.⁸

Zelop *et al* described a series of 18 cases, in which 14 out of 18 cases of isolated fetal ascites resolved in the antenatal period spontaneously. However, in this case series, anomalies were identified in 14 cases, including chromosomal abnormalities and infections; four cases were deemed as idiopathic.⁹ Seven cases presented with gastrointestinal concerns (one case of cystic fibrosis, four cases of meconium peritonitis, one case of jejunal atresia, one case of chylous ascites). In those cases, fetal ultrasound showed dilated loops of bowel and/or intra-abdominal calcifications.

Favre *et al* reported that fetal ascites was associated with an overall mortality of 57% in a series of 79 cases, with good survival for urinary ascites (14 out of 15 cases).¹⁰ The mortality rate ranged between 100% in metabolic storage disease (seven cases) and none for chylous ascites (three cases). In this case series, ascites before 24 weeks of gestation or in the presence of fetal hydrops indicates poor prognosis (respectively 78.6% vs 45% mortality rate after 24 weeks; $p < 0.01$; and 77% vs 50.8%

Learning points

- Isolated fetal ascites is a different entity from hydrops fetalis. When associated with late gestational onset, it may resolve spontaneously. An extensive evaluation to determine the aetiology is important, both for the patient and for future pregnancies. In our case, the suspected pathophysiology was likely vascular, with venous/lymphatic compromise.
- One should consider intestinal obstruction as possible cause of isolated fetal ascites. If the cause can be resolved by surgical intervention, the prognosis depends on the presence of other abnormalities and the postoperative course.

without hydrops; $p < 0.02$). The cause for ascites was defined by fetal ultrasound in 36 cases.

In our case, isolated ascites was the result of a vascular ischaemic event leading to 'apple peel' jejunal atresia. The mechanism of developing ascites was likely vascular, due to venous/lymphatic compromise.

The prognosis of 'apple peel' atresia depends on the initial vascular compromise and the occurrence of postoperative complications.

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