CASE REPORT

Emergency abdominal wall defects in neonates: saved by distress

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SUMMARY

This report presents two cases of neonatal patients with abdominal wall defects requiring emergency intervention, a closing gastroschisis and a pedunculated exomphalos with eviscerated liver. Both presented as pre-partum fetal distress and were delivered in a tertiary centre, where they received antenatal care. Coordination in the multidisciplinary team and prompt surgical intervention prevented loss of the eviscerated abdominal contents and prevented mortality in both cases.

BACKGROUND

Gastroschisis and exomphalos are both well described congenital abdominal wall defects. The incidence of these conditions has been estimated at 3.7/10 000 and 2.1/10 000 live-births, respectively, and is thought to be increasing.¹ These two conditions are normally considered as separate clinical entities although both do share the features of externalised abdominal viscera. This presents to the right of the umbilicus without a covering sac in gastroschisis and at the umbilicus with an encasing sac in the case of exomphalos. Gastroschisis presenting with a closing abdominal wall defect is a rare occurrence with only 44 cases reported in the literature to date, and is associated with high rates of complications and mortality.^{2–10} Those cases of closing gastroschisis that do go on to survive are at high risk of severe complications including short bowel syndrome, long-term parenteral nutrition and potential for bowel transplantation. To date, there have been no reported cases of a narrow based exomphalos major.

This case report highlights two unusual cases of abdominal wall defects that required emergency surgical treatment soon after birth. The mothers of both cases had been reviewed in the monthly fetal surgical clinic as part of their antenatal care. In this clinic, patients are scanned and seen by the hospitals multidisciplinary team, which in our institution includes a paediatric surgeon, fetal medicine consultant, geneticist, neonatologist and fetal specialist nurse. Although separate clinical entities, in both cases, the prompt response of the multidisciplinary team, reacting to perinatal fetal distress, led to surgical intervention without delay and aided a positive outcome without severe complications.

CASE PRESENTATION**Case 1**

An expectant mother presented with spontaneous commencement of labour at 36 weeks gestation. She was admitted to hospital and actively

monitored, which revealed ongoing abnormalities in keeping with fetal distress. Previous antenatal imaging at 20 weeks had demonstrated exomphalos major involving liver and bowel. No other anomalies had been detected at this time and the cardiac structure had been noted as being normal. Delivery had been aimed to be performed electively at 38 weeks, however, due to fetal deterioration on monitoring, this was expedited.

A male infant, weighing 3.8 kg, was born by emergency C-section and required resuscitation. Examination revealed an exomphalos with the entire liver within the sac and a narrow pedicle in the cord that contained the liver vasculature and biliary system (figure 1). Kinking of the cord resulted in haemodynamic instability of the patient, thus the cord needed securing to the incubator to ensure continual vertical suspension during transfer. The defect measured less than 1 cm and the protrusion from the umbilicus had a surface that was not epithelialised. The liver within the defect was in keeping with the diagnosis of exomphalos major. The narrow lumen of the cord with entire biliary tree within, and clinical instability of the patient, led to urgent planning for surgery. There was evidence of neither associated abnormality nor a syndromic association. Following initial resuscitation, the infant was urgently transferred to theatre. Due to the hospital setting in a tertiary centre, this was performed almost immediately after initial stabilisation (within the hour). Although theatre was not delayed to assess whether the defect was reducing in size, the acute presentation of the infant with such a small pedunculated exomphalos led to concerns that this likened to a closing gastroschisis.

At surgery, a laparotomy was performed to increase the defect, and the liver was gently reduced to avoid kinking and placed into the abdominal cavity as far as anatomically possible; no fixation was performed on the liver. Due to respiratory instability intraoperatively following reduction and concerns over increased pressure compromising hepatic blood flow, primary closure could not be obtained. The decision to undergo a staged repair was made. A silo was utilised—a traditional (hand-sewn) silo in the initial phase—and the infant improved clinically when the abdominal contents were replaced. Over the following fortnight, gentle reduction did not achieve abdominal closure and the patient returned to theatre at day 14 of life, when a biological patch was used (biodesign—biological graft, Cook).

Postoperatively, the baby progressed well and was discharged home. At 10-year follow-up, he was

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Figure 1 A pedunculated exomphalos with narrow stalk, requiring constant elevation in order to prevent haemodynamic instability.

well with no other significant medical problems and had no long lasting liver abnormalities.

Case 2

The second patient had a gastroschisis noted on a 12-week antenatal scan and had been followed up regularly by the obstetric team for intrauterine growth restriction (5th centile on imaging). At 37 weeks of gestation, the mother suffered premature rupture of membranes and was induced. However, due to failure to progress and persistent fetal tachycardia, delivery was expedited by means of an emergency caesarean section.

The female infant was born at a weight of 2.2 kg and required two boluses of intravenous fluids at delivery, for persistent tachycardia. She was otherwise in good condition and did not require ventilatory support. Examination revealed a small abdominal defect (0.5–1 cm) with the surrounding skin adherent to the external bowel. A large amount of small and large bowel was visible, including the appendix. At initial assessment, the bowel was still viable but was collapsed and dusky. The infant was diagnosed as having a closing gastroschisis and was immediately transferred to theatre. At operation, a closed defect was found with compressed, collapsed bowel at the skin level. On full assessment, it was estimated that a large amount of the small and large bowel was external to the abdomen except 15 cm of proximal jejunum, which was intra-abdominal. The bowel was assessed as viable after warm packs and replaced into the abdomen with a comfortable primary closure.

Postoperatively, the infant progressed slowly with generalised improvement in the first 2 weeks after operation. At this time, the patient suddenly deteriorated and presented with significant abdominal distension and high-volume aspirates in keeping with bowel obstruction, while still nil-by-mouth and on parenteral nutrition. Abdominal radiography demonstrated proximal

dilation and she proceeded to a second laparotomy. An obstruction at the point of an adhesional band was identified and an adhesionolysis without resection was performed; there was evidence of neither atresia nor bowel necrosis at this time. The patient recovered well, building up to full feeds, and she was discharged at 1 month of age. The infant returned 2 days later with fevers, loose stools and vomiting. Pneumatosis was present on abdominal radiography with no evidence of free air, and so she was treated conservatively for necrotising enterocolitis with antibiotics and a period of bowel rest. Following this, she was slowly built up on feeds and, after 1 month of admission, discharged home. On the last outpatient review, at 6 months of age, she was thriving on full feeds, with bowels opening regularly, and her weight had improved to above the 9th centile.

DISCUSSION

The presentation of these two cases of acute abdominal wall defects had positive outcomes following prompt intervention in response to fetal distress. Although the diagnoses of these two cases differ, they both represent acute neonatal emergencies in anterior abdominal wall defects and highlight similar important learning points. In both of these cases, the access to multidisciplinary care in the perinatal setting and prompt management of pathology that appeared time critical were of paramount importance in preserving precarious abdominal viscera and preventing severe complications.

In the case of the pedunculated exomphalos, although a defect was still visible at the level of the umbilicus, the content of the entire liver with its vascular and biliary system in the cord highlighted the potential for severe complications in this patient. There are no reports in the literature of a closing exomphalos, and the acute deterioration of the infant did not allow time to measure the umbilical defect over time to ascertain if the defect was closing, but our unit's experience of closing gastroschisis was likened to a similar presentation. The infant's emergency delivery and prompt transfer to theatre meant the blood supply was not compromised, suspension of the sac aided the prevention of compression at the small umbilical ring. Following prompt management, the infant progressed well and was without serious consequence at 10 years of age. This presentation of exomphalos is rare and this report aims to highlight the importance of prompt surgical management to preserve the abdominal viscera for a clinician faced with a similar case in the future.

Closing gastroschisis is a rare presentation of this condition and is estimated to affect 6% of infants born with gastroschisis.⁴ Early reports showed that this condition was associated with a high mortality rate. A review by Tawil *et al*⁶ in 2001 reported a case of a stillborn infant and reviewed seven further case reports of closing gastroschisis—all of which were fatal. This is in contrast to a more recent case series by Houben *et al*,⁴ where survival was achieved in eight of the nine cases reported, and only one patient required long-term parenteral nutrition. It should be noted that this success was attributed to the close monitoring by regular antenatal ultrasound to identify bowel dilation early, and urgent delivery if cause for concern was found.⁴ There is still a great deal of debate about whether early delivery is beneficial in the antenatal diagnosis of gastroschisis. It is of interest that in the case of gastroschisis the delivery was at 37 weeks of gestation, the same timing recommended in a recent retrospective study by Carnaghan *et al*¹¹ that did not find benefit in early delivery. Our experience in both of these cases was that expedited delivery and prompt access to definitive care was of vital

importance, rather than gestational age, in these time-critical conditions.

Overall, these cases highlight two unusual cases of anterior abdominal wall defects. They demonstrate the importance of close monitoring in patients with an antenatal diagnosis of congenital conditions, and swift handover between clinical teams. In both of these cases, the patients required input from radiological, obstetric, neonatal and paediatric surgical teams in quick succession, and, through a coordinated multidisciplinary approach, a positive outcome was achieved.

Learning points

- This report highlights two cases of rare emergency abdominal wall defects in the neonatal period.
- We describe a rare presentation of exomphalos with a pedunculated defect containing the liver vasculature and biliary tree. The suspension of the sac to the incubator was a suitable method to protect the precarious perfusion of the organ.
- Our experience in these cases is that the gestational age did not seem to affect outcome but, rather, the close antenatal monitoring and prompt delivery in the tertiary setting in response to distress aided a positive outcome in both cases.

Competing interests None declared.

Patient consent Obtained.

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