CASE REPORT

Ileal atresia secondary to antenatal strangulation of Littre's hernia in an exomphalos minor

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ramnik@doctors.org.uk**SUMMARY**

We describe an unusual case of ileal atresia, resulting from antenatal strangulation of a Meckel's diverticulum in an exomphalos minor. This is a rare antenatal complication of Littre's hernia, which has not been previously described. A small exomphalos minor at the base of umbilical cord and late onset vascular accident at the narrow defect leading to ileal atresia may be missed in the prenatal diagnosis. The triad of exomphalos minor, Meckel's diverticulum and terminal ileal atresia secondary to antenatal strangulation of Littre's hernia is very rare to find in the same patient. A small exomphalos minor at the base of the umbilical cord can be missed even in the postnatal period. Our case lends direct evidence of vascular accident as a cause of ileal atresia and supports vascular theory. Whenever possible umbilicus should be preserved naturally and exomphalos sac should be opened higher up for excellent cosmetic results.

BACKGROUND

Although the association of exomphalos minor and Meckel's diverticulum is well known,¹ this particular triad of congenital anomalies (exomphalos minor, Meckel's diverticulum and ileal atresia) has rarely been reported.²⁻³ Meckel's diverticulum occurs in 1.2% of the population.⁴ Littre's hernia is the presence of a Meckel's diverticulum in a hernial sac, most usually an inguinal or femoral defect. Littre's hernia at an umbilical site is rare at any age.⁵⁻⁶ and prenatal occurrence is exceptional.

CASE PRESENTATION

A baby girl presented to our department on day 5 of life, with a 24 h history of bilious vomiting. She had been born at 38+4 weeks gestation by spontaneous vaginal delivery, following an uncomplicated pregnancy. A routine anomaly scan at 20 weeks had been unremarkable. She had been discharged home at 24 h of life. Except for a smear of meconium at 48 h, she did not pass anything per rectum and, on day 4 of life, developed abdominal distension, accompanied by bilious vomiting.

On admission, examination revealed a distended, non-tender abdomen, with visible and palpable bowel loops in the upper abdomen. There was a small 25 mm exomphalos minor at the base of umbilical cord, with a 15 mm defect and a normally sited patent anus.

INVESTIGATIONS

An abdominal radiograph demonstrated multiple dilated loops of bowel proximally, and a

Figure 1 Plain abdominal and control film showing low bowel obstruction, contrast enema showing microcolon and microileum.

gasless rectum (figure 1A). A lower gastrointestinal (GI) contrast study was performed, which revealed a narrow calibre microcolon, with the caecum displaced medially and contrast refluxing into the distal collapsed small bowel (figure 1B). Proximal to this point, there was no further bowel filling, despite several attempts and a change in catheter and position. The findings were consistent with distal small bowel obstruction; possibly an ileal atresia.

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of exomphalos minor with distal ileal atresia, meconium ileus or total colonic Hirschsprung's disease was considered.

TREATMENT

Exploratory laparotomy was undertaken. A grossly dilated proximal ileal loop was seen. This ended blindly, after giving rise to a short Meckel's diverticulum. The Meckel's diverticulum was sitting within the exomphalos sac, to which it was firmly adherent. Beyond the Meckel's diverticulum, there was type 3 ileal atresia, with a mesenteric gap. The Meckel's diverticulum was freed and resected, the exomphalos sac was excised and the abdominal wall defect was closed. Fifteen centimetres of dilated ileum was resected, and an ileoileal anastomosis formed to restore bowel continuity.

OUTCOME AND FOLLOW-UP

Histological examination confirmed a Meckel's diverticulum, with no evidence of gastric mucosa.

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The postoperative course was uneventful, and by postoperative day 6 she was on full feeds. Six years after surgery, she is thriving, with excellent cosmetic and functional results.

DISCUSSION

Late intrauterine vascular catastrophe is a well-documented cause of small bowel atresia^{7–8} and this case clearly demonstrates such a pathogenesis. The presence of Meckel's diverticulum alone has been implicated in causing such a vascular catastrophe, by acting as a lead point for antenatal intussusception in utero.^{9–10}

The narrow abdominal wall defect of exomphalos minor is known to be prone to both antenatal and postnatal incarceration.¹¹ and iatrogenic injury is another postnatal hazard.^{2–12} In our case the obstetric team had noted the thick and wide base of the umbilical cord and applied the umbilical clamp higher up, avoiding any postnatal iatrogenic injury leading to fistula at the base secondary to clip application.

In this case, incarceration occurred antenatally, disrupting the blood supply to the immediately distal ileal segment and resulting in atresia. Owing to the collateral ileal blood supply from the ileal branch of the ileocaecal artery, a short segment of terminal ileum survived beyond the atretic segment.

In this case, the exomphalos minor and Littre's hernia escaped attention at the antenatal ultrasound scan, although antenatal detection is possible.¹³ Indeed, although the admitting hospital did note a 'large umbilical cord', exomphalos minor was not recognised prior to the patient's surgical transfer. The presence of a small exomphalos minor, in the presence of clinical and radiographical evidence of distal ileal atresia raised the suspicion of a unifying underlying pathology.

This triad of anomalies should therefore always be considered in case of exomphalos minor with neonatal distal bowel obstruction.

In a fresh uncomplicated exomphalos minor, we prefer to open exomphalos minor through its sac about 5 cm away from the umbilical rim. We then inspect the contents and reduce them in the abdomen followed by repair of the hernial defect. The umbilical cord is then clamped over the site of the opening in the amniotic membrane covering of the exomphalos sac. This is in contrast to traditional approach of circular incision 2 mm away from the rim of the exomphalos on the normal skin surrounding the exomphalos minor sac which requires umbilicoplasty and the cosmesis is inferior to our new approach. However, in this case of complicated exomphalos minor with distal small bowel obstruction more exposure was needed to ascertain the anatomy. In addition, the exomphalos sac was completely dried up due to delayed diagnosis.

Learning points

- ▶ A small exomphalos minor at the base of umbilical cord and late onset vascular accident at the narrow defect leading to ileal atresia may be missed in the prenatal diagnosis.
- ▶ The triad of exomphalos minor, Meckel's diverticulum and terminal ileal atresia secondary to antenatal strangulation of Littre's hernia is very rare to find in the same patient.
- ▶ A small exomphalos minor at the base of the umbilical cord can be missed even in the postnatal period.
- ▶ Our case lends direct evidence of vascular accident as a cause of ileal atresia and supports vascular theory.
- ▶ Whenever possible umbilicus should be preserved naturally and exomphalos sac should be opened higher up for excellent cosmetic results.

Contributors All authors have contributed in the clinical management of the patient, researching the literature and preparation, editing and appraisal of the final manuscript.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Nicol JW, MacKinlay GA. Meckel's diverticulum in exomphalos minor. *J R Coll Surg Edinb* 1994;39:6–7.
- 2 Vassy LE, Boles ET. Iatrogenic ileal atresia secondary to clamping of an occult omphalocele. *J Pediatr Surg* 1975;10:797–800.
- 3 Salomon LJ, Benachi A, Auber F, et al. Omphalocele: beyond the size issue. *J Pediatr Surg* 2002;37:1504–5.
- 4 Zani A, Eaton S, Rees CM, et al. Incidentally detected Meckel diverticulum: to resect or not to resect? *Ann Surg* 2008;247:276–81.
- 5 Kremer RM, Morton JH. Strangulated Littre's umbilical hernia. *Am Surg* 1968;34:432–5.
- 6 Messina M, Ferrucci E, Meucci D, et al. Littre's hernia in newborn infants: report of two cases. *Pediatr Surg Int* 2005;21:485–7.
- 7 el-Shafie M, Waag KL, Spitz L. Ileal atresia secondary to antenatal strangulation by a ruptured omphalocele. *J Pediatr Surg* 1972;7:64–5.
- 8 Shigemoto H, Horiya Y, Isomoto T, et al. Duodenal atresia secondary to intrauterine midgut strangulation by an omphalocele. *J Pediatr Surg* 1982;17:420–1.
- 9 Lin CH, Wu SF, Lin WC, et al. Meckel's diverticulum induced intrauterine intussusception associated with ileal atresia complicated by meconium peritonitis. *J Formos Med Assoc* 2007;106:495–8.
- 10 Gündoğdu HZ, Senocak ME. Intrauterine intussusception due to Meckel's diverticulum as a cause of ileal atresia: analysis of 2 cases. *Eur J Pediatr Surg* 1996;6: 52–4.
- 11 Groves R, Sunderajan L, Khan AR, et al. Congenital anomalies are commonly associated with exomphalos minor. *J Pediatr Surg* 2006;41:358–61.
- 12 Chapman-Sheath P, Wilcox D, Mok Q, et al. Iatrogenic ileal obstruction: a complication of umbilical cord clamping. *BMJ* 1996;313:613–14.
- 13 Tourne G, Chauleur C, Varlet MN, et al. Prenatal discovery of an omphalocele associated with an inner umbilical cord Meckel's diverticulum. *J Matern Fetal Neonatal Med* 2007;20:427–30.

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