

Ileal perforation in segmental intestinal dilatation associated with omphalocele

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ABSTRACT

Localised dilatation of a segment of the intestine without any macroscopically-identifiable cause is rare, and has been reported in association with omphalocele in only 14 children up to 2006. In most of these cases, the segmental intestinal dilatation (SID) was either diagnosed incidentally, or due to presentation with partial or complete intestinal obstruction. We report, for the first time, a 37-week-old neonate with bowel perforation in SID associated with omphalocele. In our case, a long thin vessel that resembled the mesodiverticular vessel of a Meckel's diverticulum was present in the dilated segment, supporting the view that SID and Meckel's diverticulum may be embryologically related.

Keywords: bowel perforation, Meckel's diverticulum, omphalocele, segmental intestinal dilatation

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INTRODUCTION

Segmental intestinal dilatation (SID) is characterised by localised dilatation of the intestine to about 3–4 times the width of the adjacent normal bowel, and by an abrupt transition between the normal and dilated segments. SID usually presents with features of complete or partial bowel obstruction in the absence of any obvious intrinsic or extrinsic cause for the dilatation. The dilated segment has a normal distribution of intestinal neuronal plexus, and a complete recovery occurs after resection of the dilated segment.^(1,2) SID with omphalocele minor has been recorded in 14 children up till 2006, but perforation of the bowel in association with SID and omphalocele has not been reported to date.⁽²⁻⁹⁾

CASE REPORT

A 37-week-old female Chinese baby, in whom omphalocele was detected antenatally at 33 weeks of gestation, was born by normal vaginal delivery. The amniotic fluid was stained with meconium, and at delivery, a small defect was present in the omphalocele

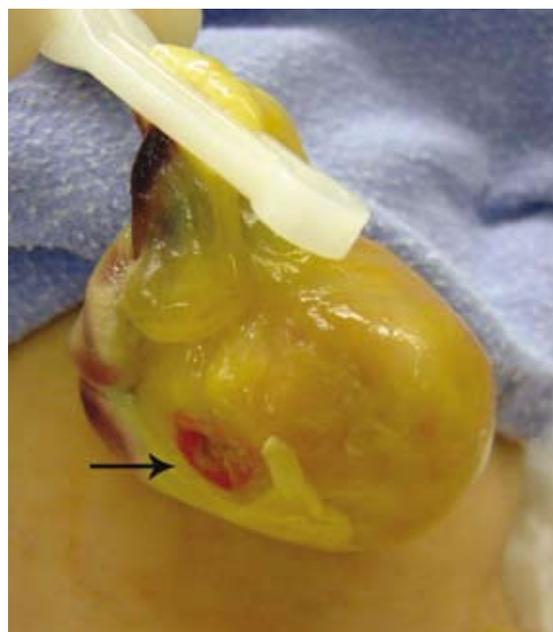


Fig.1 Photograph shows a disruption in the omphalocele sac (arrow), with bowel perforation underneath.

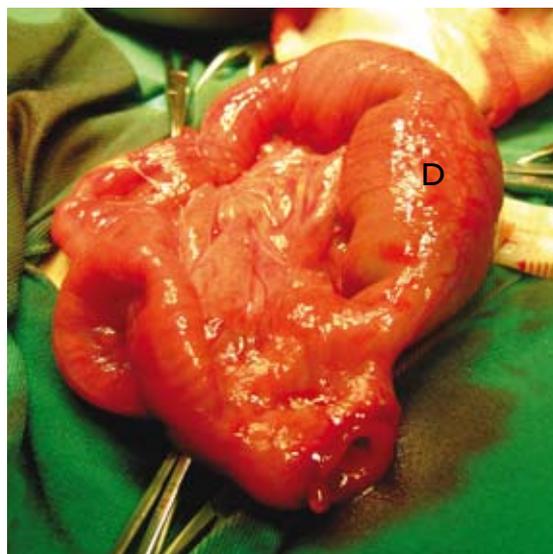


Fig.2 Photograph shows segmental dilatation of the ileum (D), with perforation near its distal end. Normal calibre bowel is seen proximal and distal to the dilated segment.

sac, with perforation of the bowel underneath it (Fig. 1). At laparotomy, a length of 10 cm of the distal ileum, located about 10 cm from the ileocaecal junction, was noted to be dilated and flabby (Fig. 2). The ileum proximal

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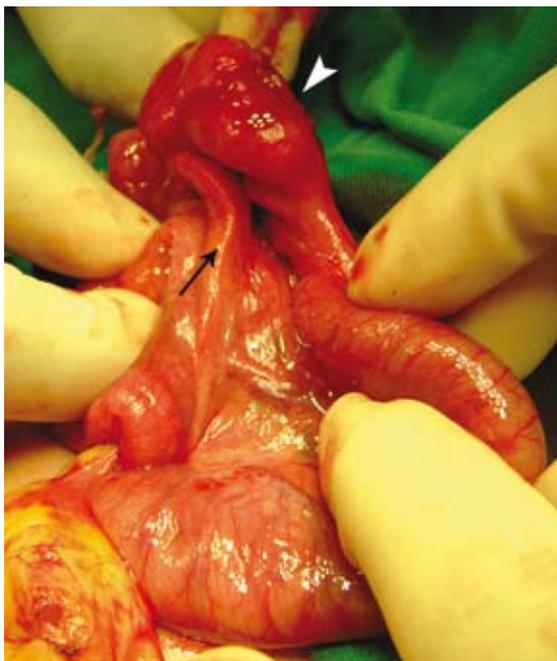


Fig. 3 Photograph shows segmental dilatation of the ileum with a prominent vessel (arrow) running from the mesentery and the site of the perforation (arrowhead).

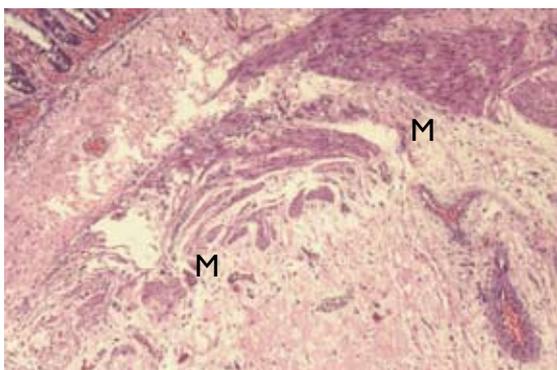


Fig. 4 Photomicrograph shows thinning of the circular and longitudinal muscle layers (M), with architectural distortion in the dilated segment (Haematoxylin & eosin, $\times 100$).

and distal to this segment was normal, with an abrupt transition between the normal and dilated segments. A long thin vessel, resembling the mesodiverticular vessel of a Meckel's diverticulum, passed from the ileal mesentery to the dilated bowel segment (Fig. 3). There was a perforation of about 1.5 cm in diameter near the distal end of the dilated bowel at the antemesenteric border. There was no macroscopically-identifiable cause for the localised ileal dilatation and for the perforation in that segment. The dilated ileum with a 2-cm margin on each side was resected, and the ileal continuity was restored. The patient was discharged well a week later. Microscopical examination of the longitudinal sections from the whole length of the resected small bowel showed areas of thinning of the muscularis propria, more towards

the distal end (Fig. 4). Both the inner circular and outer longitudinal muscle layers became thinner and markedly distorted architecturally near the perforated area. There was no feature of acute or chronic inflammation near the area of perforation. Ganglion cells were seen within the submucosa and intermyenteric plexus throughout the length of the bowel.

DISCUSSION

SID was first described in the colon and later in the duodenum, jejunum and ileum.^(1,2) The pathogenesis of SID is unclear. Entrapment of the bowel, with incomplete intestinal obstruction within the omphalocoele during gestation, has been postulated as the cause.⁽²⁻⁷⁾ Alternately, the development of SID may be the primary event, with the dilated bowel preventing closure of the umbilical ring. It has also been suggested that SID may be embryologically related to Meckel's diverticulum.^(2,7) The following findings, seen in some cases of SID, support the latter view: an abnormal vascular pedicle running from the ileal mesentery to the dilated segment, resembling the mesodiverticular vessel of a Meckel's diverticulum (as seen in our case); the eccentric dilatation of the SID, more conspicuous on the antimesenteric side of the bowel; and the frequent presence of vitellointestinal remnants in association with omphalocoele.^(2,7,10)

Histological examination is the most reliable criterion for the diagnosis of SID. The absence of features of acute or chronic inflammation, the presence of normal ganglion cells in the intermyenteric and submucosal plexus, and the altered architecture of the muscularis propria, are important features. The muscular layer in SID is usually thinned out or absent, but in some cases, may be hypertrophic.^(7,10) In our case, the bowel perforation possibly resulted either from ischaemia due to distention of the dilated segment within the omphalocoele, or from birth trauma. Of the 14 patients of SID with omphalocoele minor reported in the literature, nine were males. 13 of them were diagnosed as neonates, and one was diagnosed four years after omphalocoele repair.^(2,3,6-9) In all the patients, the SID was in the distal ileum, and its distal extent was about 12–25 cm from the ileocaecal valve. The associated anomalies reported were the presence of ectopic gastric or pancreatic tissue, enteric duplication, trisomy 13 and hemivertebrae. Resection of the SID and primary anastomosis with closure of the omphalocoele is the treatment of choice, as was done in all the patients reported.^(2,3,6-9) Inspection of the intestine to exclude SID is recommended at the time of omphalocoele minor repair.⁽⁷⁾

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