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Segmental intestinal dilatation associated with omphalocele

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1. Introduction

Segmental intestinal dilatation (SID) is a rare congenital malformation, defined by the presence of a limited bowel dilatation with an abrupt transition between normal and dilated bowel, in absence of an extrinsic or intrinsic stop and with a normal pattern of neuronal plexus [1]. Concomitant SID and omphalocele is extremely rare, and sometimes associated with Meckel's diverticulum. We describe a patient with SID associated with perforated Mekel's diverticulum and omphalocele. After a literature review (PubMed database) we found 16 cases with the association between SID and omphalocele.

2. Case report

A 1-day-old boy (weight 3.1 kg), born through vaginal delivery in our hospital at 37 weeks of gestation, was evaluated for a leakage of meconium from the umbilical cord. An ultrasound performed at 33 weeks referred a cyst of the umbilical cord. An ultrasound revealed a minor omphalocele (2.5 cm of diameter). Based on clinical and radiological evaluation we put a suspicion on perforated omphalocele and we planned a surgery. At surgery a perforated Meckel diverticulum within the abdominal defect (Fig. 1) and a dilated bowel loop at 20 cm from the ileocecal valve (Fig. 2) were found. Our surgical procedure included: (i) resection of Meckel diverticulum and end-to-end anastomosis, (ii) resection of segmental bowel dilatation and end-to-end anastomosis, (iii) umbilicoplasty (Figs. Figs 3–5). Postoperative course was uneventful. Patient started orally on 8th postoperative day. Stool pass was on 6th post-operative day. At last follow up, after 9 months, patient is healthy.

3. Discussion

Segmental intestinal dilatation (SID) is a rare congenital malformation. It usually involves the distal ileum, and any cases of colon and jejunum are described [2,3]. It is usually located at 12–25 cm from the ileocecal valve [4]. Our child, the first in our experience, had a SID about 20 cm from the ileocecal valve as it is described. Resection and anastomosis is the surgical approach described in literature as we performed in our child.

Swenson and Rathauser described firstly the intestinal dilatation segmental (SID) and established the diagnostic criteria that are used to date: (i) limited bowel dilation with three or four times higher Al: dimensions, (ii) sharp transition between normal and dilated intestinal (iii) any extrinsic or intrinsic causes of obstruction (iv) normal neuronal plexus (v) clinical presentation intestinal obstruction [5].

It can be associated with other malformations such as intestinal malrotation, herniated Bochdaleck, intestinal atresia, spina bifida and bladder exstrophy. However, the omphalocele is the most frequent association (85–90%).

Based on literature review, we found 16 cases of SID-omphalocele association, as shown in Table 1 [6–13].

In most of cases, as in our patient, the SID was found during surgery at birth, but in a few cases it was undiscovered until a

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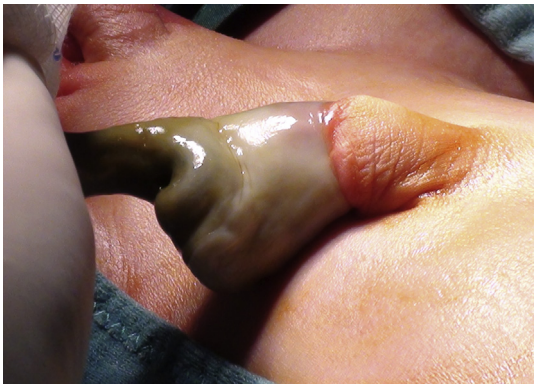

Fig. 1. Minor omphalocele.

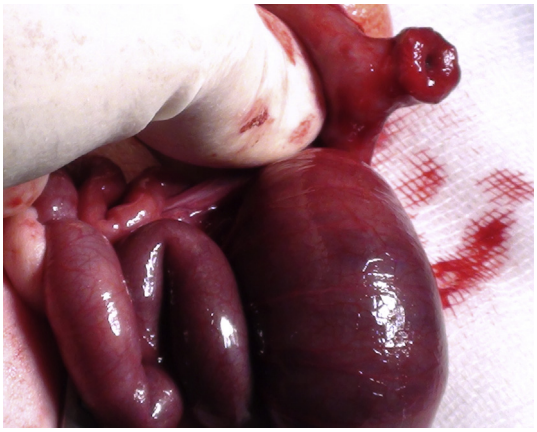

Fig. 2. Perforated Meckel's diverticulum within the minor omphalocele.

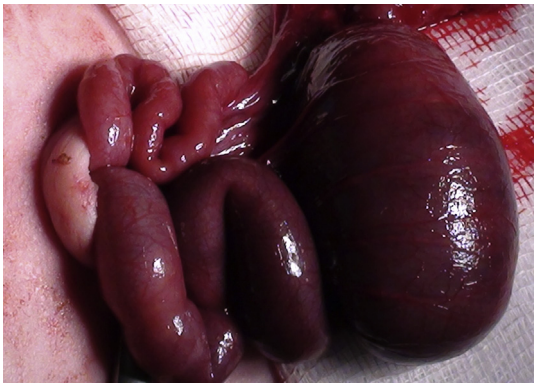

Fig. 3. Segmental intestinal dilatation (SID) at nearly 20 cm from the ileocecal valve.

symptomatic intestinal occlusion/sub-occlusion [14]. In two patients, SID was discovered due to the appearance of intestinal occlusion symptoms, after surgical closure of the omphalocele. In one patient, after the closing of the omphalocele and the execution of a biopsy, symptoms of intestinal occlusion appeared and so he was subjected to intestinal resection.

While the association SID/omphalocele is common and known, the co-presence of SID and Meckel's diverticulum is rare [15–17].

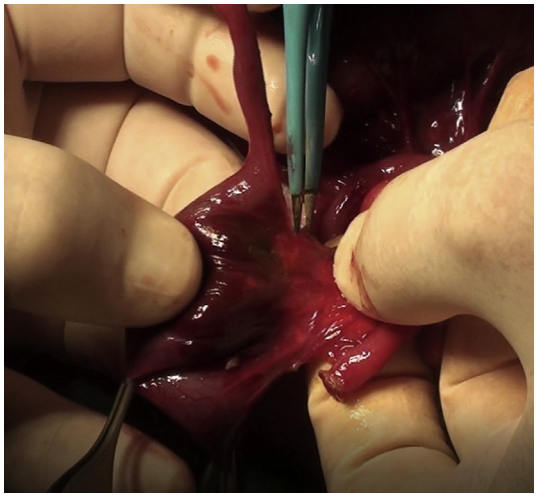

Fig. 4. Resection of segmental bowel dilatation.

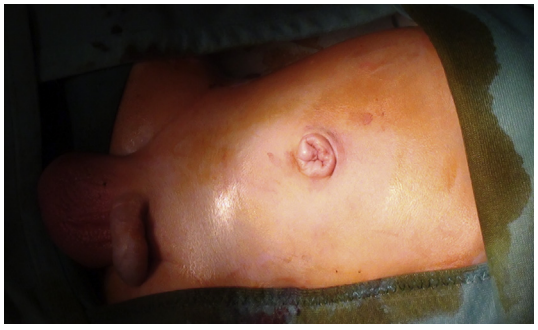

Fig. 5. Umbilicoplasty.

Table 1 shows 5 patients with Meckel's diverticulum, of which 2 had a perforation.

These data show the importance of an exploration of the abdomen and bowel in the case of omphalocele.

The treatment of congenital segmental dilatation of intestine is resection of the dilated segment and end-to-end anastomosis [18] Temporary stomas can be made in critically sick patients [19]. Our review shows only one patient treated with temporary ileostomy [14].

Histology of the resected segment is usually normal in most of the cases. We found 4 cases where histological examination showed mucous hypertrophy, 2 with mucous thinning, 1 with thickening, 2 with ectopic gastric mucosa (one in the context of the Meckel's diverticulum). In all cases, the ganglion cells were normally represented.

In conclusion, nevertheless SID is considered a less frequent malformations, it should be suspected in all patients with omphalocele.

The presence of a SID in association with omphalocele justifies the execution of a single surgical intervention with resection and intestinal anastomosis, in complete safety.

Table 1
This table summarizes the largest series of SID [11–18].

Series	N° of cases	Age of presentation	Symptoms	Site of dilatation	Associated malformations	Treatment	Histological findings
Molinaro et al.	1	Neonatal	Perforated Meckel's diverticulum in omphalocele (intestinal perforation)	Ileum	Meckel's diverticulum Omphalocele minor	Surgical resection and end-to-end anastomosis	Normal
Sam CJ, 2011	1	8 y.o.	Abdominal pain, bilious vomiting failure to pass stools	Ileum	Omphalocele, Meckel's diverticulum	Surgical resection and end-to-end anastomosis	Normal bowel musculature and normal ganglion cells with ectopic gastric mucosa in the region of Meckel's diverticulum
Wei CH et al., 2010	1	Neonatal	Omphalocele, vomiting	Ileum	Omphalocele	First operation, closure of omphalocele and intestinal biopsy, second operation surgical resection and end-to-end anastomosis	Normal
Thambidorai CR et al., 2009	1	Neonatal	Perforated Meckel's diverticulum in omphalocele (intestinal perforation)	Ileum	Meckel's diverticulum, Omphalocele minor	Surgical resection and end-to-end anastomosis	Normal
Brahim MB et al., 2006	8 (6 with omphalocele and SID)	7 Neonatal 1 y.o.	6 Omphalocele, 1 Neonatal obstruction, 1 Constipation	ileum	6 Omphalocele, 1 Esophageal Atresia, 1 Hirschsprung disease	Surgical resection and end-to-end anastomosis	3 hypertrophic muscolaris, 4 normal, 1 heterotopic gastric mucosa 1 Thin muscolaris
Basaran UN et al., 2005	1	Neonatal	Omphalocele	ileum	omphalocele	Ileostomy, Surgical resection and end-to-end anastomosis	Normal ganglion cells, Hypertrophic and thin muscolaris layer
Balik E et al., 1993	1	4 y.o.	Abdominal distension, bilious vomiting, failure to pass stools	ileum	Small omphalocele	Surgical resection and end-to-end anastomosis	Normal ganglion cells, thickening of muscular layer
Bell MJ et al., 1982	7 (2 with omphalocele and SID)	2 Neonatal 5 Children	2 Omphalocele, 5 pallor, fatigue, and anemia, intestinal obstruction and recurrent abdominal pain	Ileum	2 Omphalocele 2 Meckel's diverticulum	Surgical resection and end-to-end anastomosis	No lumenal stenosis
Aboulola M et al., 1979	2	2 Neonatal	2 Omphalocele	ileum	2 Omphalocele, 2 intestinal malrotation	Surgical resection and end-to-end anastomosis	–

Conflict of interest

None declared.

Source of support

Nil.

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