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Case Report

An omphalocele containing a surprised congenital segmental intestinal dilatation: A case report and literature review

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Abstract

This report describes the characteristics and management of segmental dilated small intestine, a rare condition that resembles the liver in an omphalocele. After the dilated intestine was resected and end-to-end anastomosis was performed, the abdomen could be closed simultaneously without immediate complications. Our review of the literature on this condition shows that although its nature is still unknown, it is associated with excellent operative outcomes. Finally, awareness of congenital segmental intestinal dilatation in omphalocele leads to proper management.

Keywords: Congenital segmental intestinal dilatation, omphalocele, segmental small bowel dilatation

Segmental intestinal dilatation (SID) is a rare congenital anomaly characterized by a localized and abnormal dilation of the intestine, often resulting in functional obstruction without a known intrinsic or extrinsic cause. [1 - 9] The dilated segment shows an abrupt transition between normal and affected bowel, which can complicate diagnosis and management, especially when it occurs together with other congenital anomalies. Omphalocele, a condition in which abdominal contents protrude through a defect in the abdominal wall, can further complicate cases of SID, as the management of both conditions may require simultaneous surgical intervention. Whereas omphalocele repair often involves staged closure to prevent abdominal compartment syndrome, the presence of SID typically necessitates immediate resection of the dilated bowel segment to prevent obstruction and associated complications. [10] This report highlights the rare occurrence of SID within a giant omphalocele and discusses how its presence can alter the standard surgical approach. [11]

Case report

An eighteen-year-old mother gave birth to a 3020-g male at 39 weeks of gestational age via vaginal delivery. He was diagnosed with omphalocele, which had a 6.5-cm pink and liver-like content. The abdominal wall defect was 1.5 cm in diameter at the neck of the omphalocele **(Figure 1);** it had no abnormal features or other anomalies. Pre-operatively, the content of the omphalocele appeared pink, resembling the liver. The abdominal x-ray showed an abnormal bowel dilatation and a hypodense lesion within the omphalocele retrospectively compatible with a dilated bowel **(Figure 2).** The primary closure operation seemed possible given the large proportion of his body relative to the omphalocele. After adequate resuscitation, he underwent surgery on the first day of his life.

After dissecting the amniotic membrane from the abdominal wall, the entire omphalocele content measured 6.5 × 6.0 × 6.0 cm segmentally dilated small bowel. The remaining small bowel length was 100 cm. The segmental dilated bowel was located at the terminal ileum, 10 cm distal to the Meckel diverticulum

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and 20 cm proximally to the cecum. The intestine was incompletely rotated, the cecum and the appendix were in the right upper quadrant, and the DJ junction was in the right upper quadrant. A thin band adhesive between the cecum and the dilated bowel caused a narrow mesentery base **(Figure 3).**

The dilated segment was resected, before end-to-end anastomosis was performed. The segmental dilated bowel contained meconium. The Meckel diverticulum was resected. An appendectomy was performed. The band between the cecum and cyst was divided to widen the mesenteric base. The small bowel was placed in the right abdomen, and the large bowel was placed in the left abdomen. Primary fascial abdominal closure was performed along with umbilicoplasty **(Figure 4A).** The baby started feeding on the fifth postoperative day and reached full feed a week later. As there was no sign of intestinal dysfunction, and he was discharged two weeks later. His well-being was proper and unevenly well at eight months **(Figure 4B)** and two years of age, which was last follow-up.

The pathologic result showed a segmental cystically dilatated small bowel filled with meconium content, measuring 6.0 × 5.5 × 5.0 cm, attached to the middle part of the herniated umbilical cord **(Figure 5).** The histologic revealed unremarkable small bowel mucosa and muscularis propria, containing ganglion cells in the myenteric and submucosal plexuses. The appendix also contained ganglion cells in the myenteric and submucosal plexuses. Heterotopic tissue was not observed in the Meckel diverticulum or the dilatated small bowel.

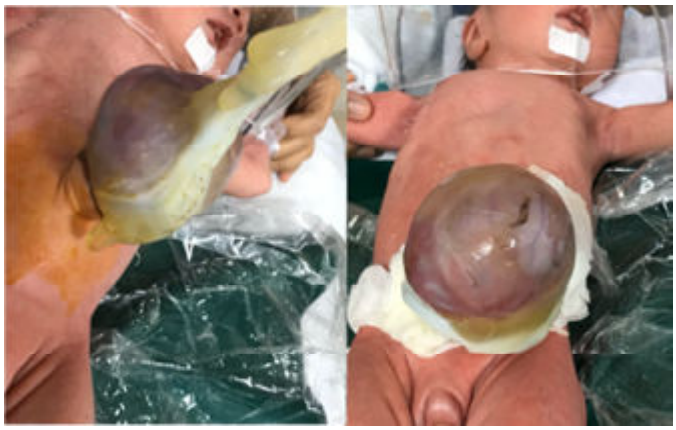

Figure 1. Omphalocele with a 6.5 cm pink liver-like content. The abdominal wall defect was 1.5 centimeters in diameter at the neck of the omphalocele.

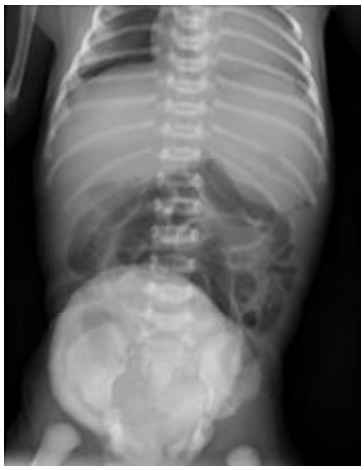

Figure 2. The x-ray showed an abnormal bowel gas pattern.

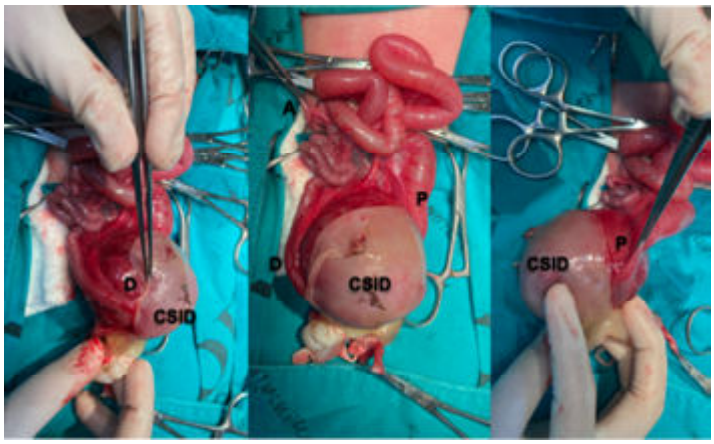

Figure 3. Intraoperative finding: congenital segmental intestinal dilatation (CSID) with continuously patent lumen (A, Appendix); (P, Proximal bowel); (D, Distal bowel).

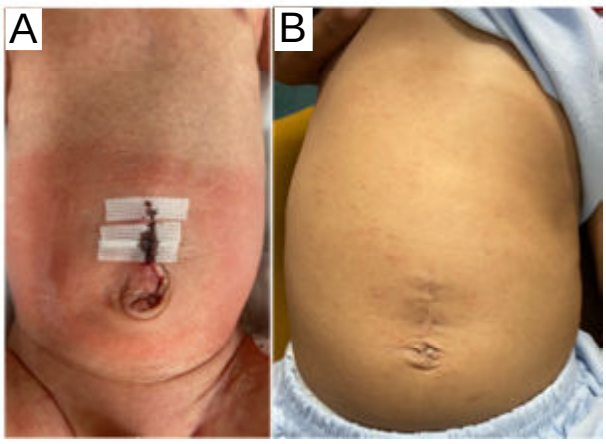

Figure 4. (A) Postoperative day 1; **(B)** 8 months old.

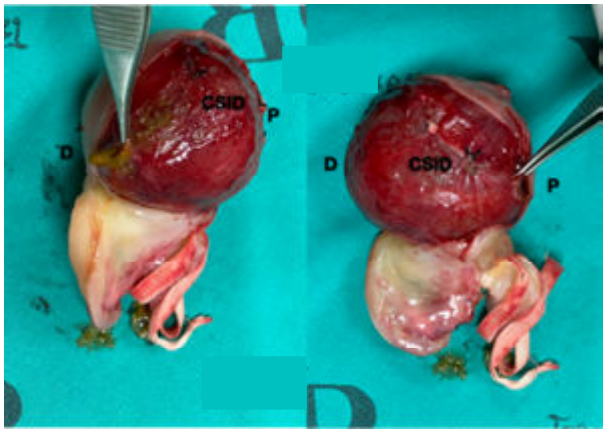

Figure 5. Gross specimen of congenital segmental intestinal dilatation (CSID); (P; Proximal bowel); (D; Distal bowel).

Discussion

In this report, we present a rare coincidental incidence of congenital SID and an omphalocele. The omphalocele content appeared similar to the liver. Additionally, the size of the omphalocele was controversial in some points of operative management. After scarification, the delayed abdominal closure may be suitable for a liver containing a large omphalocele to avoid the risk of anesthesia in the neonatal period and operative complications, especially postoperative intra-abdominal hypertension due to excessive pressure from the significant liver. On the contrary, a segmental dilated small bowel must be operatively resected.

The etiology of this abnormal bowel is unclear. This lesion is characterized by an abrupt change in diameter, approximately 3–5 times larger, before returning to a standard size, and the internal lumen remaining patent, but often causing functional obstruction. [1-3] The diagnosis criteria for congenital SID are[4-10]: A) limited bowel dilatation with a three- to four-fold increase in size; B) a sharp transition between the dilated and normal bowel; C) no intrinsic or extrinsic barrier distal to the dilatation; D) a clinical picture of intestinal occlusion or sub-occlusion; E) normality of the neuronal plexus, and F) complete recovery after resection of the affected segment. Our patient met all the diagnosis criteria.

Congenital SID can be an isolated lesion with a challenging diagnosis and mandatory differentiation from other common causes of intestinal obstruction, such as intestinal atresia. [5] Hirschsprung's disease [5] or gut duplication [4] segmental intestinal dilation with omphalocele has been mentioned, mostly with the dilated bowel tapped in the omphalocele sac. [1,5,6] Sometimes, the dilated bowel occurs in the abdomen. [4,7] Irving and Lister [1] opine that the involved dilated segment in the omphalocele sac could result from entrapment by the umbilical ring or this lesion causing omphalocele. This possibly explains the etiology in other isolated lesions caused by temporary intestinal obstruction at an early stage of intestinal development (before intestinal involution). As the segmental bowel dilatation with omphalocele is primarily located in the terminal ileum, the congenital band adheres to the dilated bowel. [4,6] Some authors suggest that congenital SID is a variation of Meckel's diverticulum. [4,6,8] Our findings did not support this theory, although the band adhered between the cecum and the dilated bowel. We also found Meckel's diverticulum 10 cm proximal to the segmental dilatation.

Apart from omphalocele, other associated anomalies have been reported, such as esophageal atresia [5], neural tube defect [1], congenital heart disease [3], duplication cyst [2,10], Hirschsprung disease [5], intestinal atresia, [3,9] and malrotation. [8,9] The histopathological findings of the SID also show a normal mucosal and muscular layer. [5,10] Heterotopic tissues such as cartilage [1], pancreatic [8], or gastric tissue [5] rarely occur in the dilated bowel. [1] There is no evidence of aganglionosis and neuronal intestinal dysplasia in all reported cases. [1-8,10]

Congenital SID occurs in many locations of the alimentary tract, occasionally in the ileum [1,5] and colon [3], but rarely in the duodenum and other parts of the intestine. [3] The clinical presentation depends on the intestinal location. Additionally, the onset of the clinical relies on the length of the involved segment. Dysmotility of the bowel mainly causes clinical intestinal obstruction during the neonatal period. [1-3] Retro-sigmoid lesions must be differentiated from Hirschsprung's disease. Terminal ileal lesions may be misdiagnosed as total colonic aganglionosis or intestinal atresia. [5] Some are eventually incidental findings in laparotomy. [5] One patient presented with clinical midgut volvulus. [8]

The recommended operative management for congenital SID is complete resection of the involved bowel and restorative intestinal continuity. [1-10] For congenital segmental intestinal obstruction with omphalocele, simultaneous complete resection with end-to-end anastomosis and the closure of the abdomen are acceptable with minor complications. [4,6] There is a report of a patient with a small omphalocele suffering from a partial intestinal obstruction recurrence due to a remaining SID. [7] If complete resection is impossible, the tapering procedure resulting in proper bowel function is normally used. [3] A patient's abdominal distention can be temporarily improved after irrigation with gastrografin enema. However, surgical intervention is ultimately required. [1]

Besides congenital SID and omphalocele, malrotation in omphalocele is a challenge. Intestinal malrotation and non-rotation are associated with omphalocele and gastroschisis. Recommendations of its management vary. In this case, the content in omphalocele had only congenital SID. The rest of the intestine was grossly normal. After resection of the dilatation bowel, the cecum and DJ were finally located in the same position, causing a narrow mesenteric base compatible with intestinal malrotation. Therefore, we decided to perform the modified Ladd's procedure and appendectomy.

Conclusion

Congenital SID associated with the omphalocele is rare. A rising awareness of this coincidental lesion has led to its proper management. Although the etiology of the congenital SID is still unclear, definite resection is a standard treatment and is usually safely done simultaneously with the abdominal closure operation.

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Conflict of interest statement

Each of the authors has completed an ICMJE disclosure form. None of the authors declare any potential or actual relationship, activity, or interest related to the content of this article.

Data sharing statement

Data generated or analyzed for the present report are included in this published article. Further details are available from the corresponding author on reasonable request after deidentification of the patient whose data are included in the report.

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