Case Report

Congenital Mesenteric Defect with Transmesenteric Hernia in Children: A Case Series

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Abstract

Transmesentric hernias are a type of internal hernia, in which there is herniation of bowel loops through a defect in the mesentery. They present with a wide variety of symptoms without any specific radiological features, leading to a delay in diagnosis and high mortality rate. Here, we present a case series of four patients with this rare but fatal cause of small bowel obstruction. Three children presented to the emergency department with small bowel obstruction. The other baby was a preterm neonate with an antenatal scan showing small bowel obstruction. All children underwent emergency laparotomy and were found to have a mesenteric defect with herniation and gangrene of the small bowel. Resection anastomosis of the gangrenous segment and closure of the mesenteric defect were done. The differential for small bowel obstruction in children should include transmesentric hernia. Laboratory or imaging investigations are often inconclusive. Timely exploration can save lives in this rare but life-threatening condition.

Keywords: Mesentric defect, small bowel gangrene, transmesentric hernia

Introduction

Transmesentric hernias are a type of internal hernia, in which there is herniation of bowel loops through a congenital defect in the mesentery.[1] The absence of a sac characterises these hernias. Although the defects are present since birth, the condition is rare in the paediatric age group, with most of the series reported in adults. In the limited paediatric case reports, the usual presentation is in the neonatal age group with gangrene. They can present with intestinal obstruction, strangulation and gangrene or chronic symptoms secondary to intermittent herniation. A varied presentation without a definitive radiological picture can lead to delayed diagnosis with associated morbidity and mortality. The mortality can be as high as 45%, as reported in one series.[2] We present our experience with this rare but fatal cause of small bowel obstruction.

Case Series

A retrospective descriptive study of four children treated for congenital mesenteric hernia in our department. The

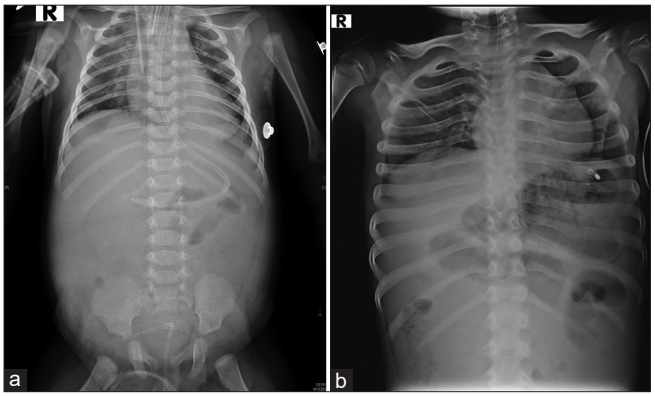


Figure 1: Plain X-ray abdomen - Erect. (a) 54 days old with distended abdomen and paucity of bowel gas, (b) 2 years old with distended abdomen showing dilated proximal loops and paucity of distal bowel gas

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Table 1: Clinical and operative details of children included in the study

Age	Sex	Presentation	Diagnosis	Intra operative finding	Outcome
54 days	Male	Septic shock	Clinical	Mid-ileal mesenteric defect with gangrene of 70 cm of ileum	Fully recovered
3 days	Female	Antenataly detected small bowel obstruction	Antenatal scan	Mid-ileal mesenteric defect with gangrene of 40 cm of ileum	Re-laparotomy for adhesiolysis Recovered
7 years	Male	Peritonitis, septic shock	Clinical	Traves field defect with gangrene of 70 cm of ileum	Fully recovered
2 years	Male	Peritonitis	Clinical	Traves field defect with gangrene of 30 cm of ileum	Fully recovered

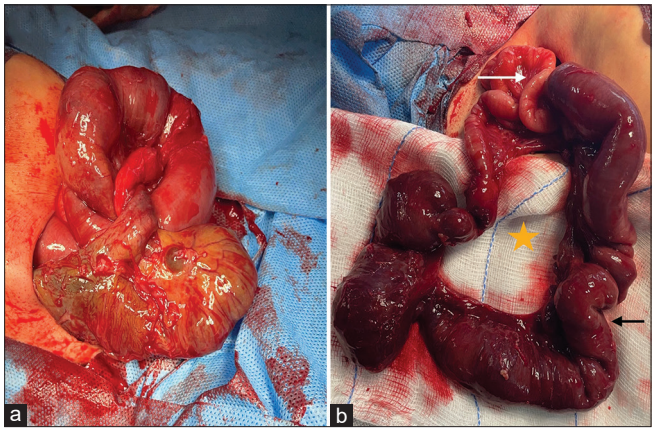


Figure 2: Intraoperative findings in a 3-day-old child: (a) Volvulus with gangrenous bowel loops seen on laparotomy (b) Mid ileal mesenteric defect with around 40 centimetres of ileal gangrene. Yellow star-mesenteric defect; White arrow-proximal healthy small bowel; Black arrow-gangrenous small bowel

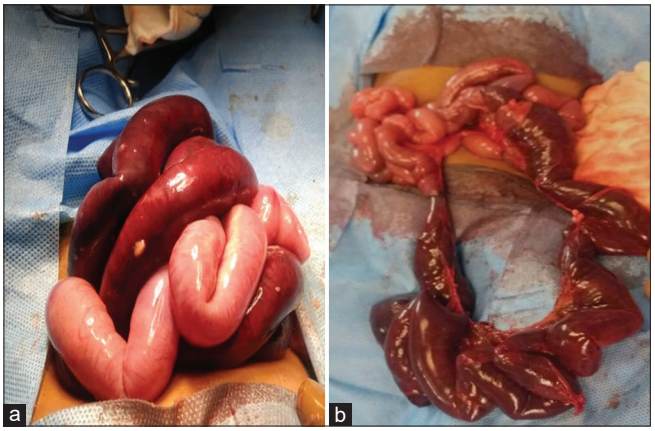


Figure 3: Intraoperative findings in a 54-day-old child: (a) Gangrenous bowel loops seen on laparotomy (b) Mid ileal mesenteric defect with around 70 cm of ileal gangrene

clinical details of the children are summarised in Table 1. Three children presented to the emergency department with acute-onset abdominal pain, distension and bilious vomiting. There were signs of peritonitis with shock in two children, while the other had severe dehydration. Blood investigations showed raised counts and lactate levels. X-ray was done on two children, showing a paucity of gas in the abdomen [Figure 1]. Initial resuscitation was done with inotropes required in children who presented with shock, followed by emergency laparotomy. The other child in the study had suggestions of small bowel obstruction in antenatal scans at 34 weeks of gestational age (GA). The child was delivered at 35 weeks of GA due to premature rupture of membranes. A nasogastric tube was inserted at birth with persistently high bilious aspirates. Contrast studies showed microcolon with no other diagnostic feature. The child was taken up for laparotomy on day 2 of life.

Laparotomy showed mesenteric defect with herniation of bowel loops in all cases. There was a volvulus of the herniated small bowel loops with gangrene of varying lengths ranging from 30 to 70 cm. The defect was in the terminal ileum in two patients and mid ileal in the remaining. The diameter was more than 10 cm in children with mid-ileal defects [Figures 2 and 3]. There were no other associated anomalies such as malrotation. Resection of the gangrenous bowel with primary end-to-end anastomosis was done in all cases. The immediate post-operative period was uneventful, with full feeds established within 5 days. One child developed adhesive intestinal obstruction 3 months following primary surgery requiring laparotomy and adhesiolysis. All children are doing well on long-term follow-up.

Discussion

Transmesentric hernia caused by a congenital defect in the mesentery was initially described by Rokitsansky in 1836, in which the caecum had herniated through a defect of the mesentery located near the ileocaecal valve.[3] The most common site for transmesentric herniation is near the ileocaecal region, as described by Treves in 1985.[4] He described an area in mesentery outlined by the ileocolic branch of the superior mesenteric artery and its anastomosis with the last ileal artery, which was called as Treves' field.[5] This area lacks fat, blood vessels or lymph nodes and is inherently weak.

Multiple etiological hypotheses were proposed. According to Federschmidt, there is a partial regression of the dorsal mesentery leading to the defect.[6] Menegaux postulated that the defect was due to inadequate vascularity.[6] This is thought to be similar to the pathogenesis of ileal atresia, the former representing a lesser severe vascular insult. Macklin postulated that coalescence inevitably takes place when two epithelial layers are opposed with a deficient intervening supporting stroma of connective tissue, with the development of a space or defect.[7]

The presentation can be acute or chronic. Acute presentations include bilious vomiting and abdominal distension, primarily

seen in neonates and infants. The necrosis may set in as early as 6 h, finally leading to shock and sudden death. The chronic presentation occurs when the bowel intermittently herniates, leading to abdominal pain, distension, nausea, vomiting and constipation. Tenderness on palpation may be present. Preoperative diagnosis is rare in chronic cases. Three cases presented to us acutely with rapid worsening of symptoms.

Blood investigations are typically normal; however, metabolic acidosis and leucocytosis may occur once necrosis sets. Therefore, in the emergency setting, it is imperative to correlate the clinical signs with the blood lactate levels and blood gases to suspect bowel gangrene.

No radiographic investigation has been proven to be valid. A plain X-ray may show air-fluid levels or signs of perforation. Signs to look for in abdominal contrast computed tomography (CT) include a constriction around the clustering of the small bowel with closely approximated afferent and efferent limbs of the herniated bowel and stretching or displacement of mesenteric vessels as they pass through the defect to supply the herniated segment.[8,9] These are not established criteria, and none of the reports diagnosed the condition pre-operatively. It leads to delays in exploration as well. In our experience, no time was wasted on radiological diagnosis, and it was decided to proceed with surgery based on clinical signs alone. In one case, intestinal obstruction was suspected on the antenatal ultrasound itself, but the diagnosis of mesenteric defect could be made at laparotomy only.

At laparotomy, the mesenteric defects are usually 2 to 3 cm in size.[9] In two of our cases, the defect was large (more than 10 cm) and located at the mid-ileum, while two others had a Treves field defect. Furthermore, herniation of the long length of the small bowel, between 30 and 70 cm, was noted. While minor defects can lead to vascular constriction and gangrene, large defects generally result in a volvulus, as seen in our series. The early and rapid onset of gangrene can also be attributed to the already precarious blood supply of the bowel at the level of the defect. Management consists of reduction of the herniated bowel loops, resection of the unhealthy bowel and restoration of bowel continuity. The mesenteric defect is closed. One has to look for associated rotational defects and atresia before closing. Despite normal-looking bowel, small bowel strictures have been noted on follow-up.[10]

Strangulated transmesenteric hernias are a rare cause of intestinal obstruction, especially in the paediatric population. To establish the diagnosis and provide immediate therapy, prompt open exploration based on clinical signs is mandatory. This can reduce morbidity and mortality in this rare life-threatening condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents/guardians have given their consent for images and other clinical information to be reported in the journal. The parents/guardians understand that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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