Rare disease

A mesenteric hernia complicated with a triple necrotic volvulus

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Summary

A 6-year-old girl was admitted to the paediatric emergency department with colicky abdominal pain. She had a significant medical history, with four previous admissions due to recurrent abdominal pain in the past year. On examination the abdomen was soft, there was no rebound tenderness and Rovsing's sign was negative. Her blood tests revealed a raised white cells count, although her C reactive protein was within the normal range. Abdominal x-ray revealed small bowel obstruction. During her assessment the patient rapidly deteriorated and seemed to go into shock. Her clinical state in addition to the radiological findings meant that she was taken to theatre for surgical exploration. This showed a triple volvulus with necrotic bowel loops that had herniated through a mesenteric defect. The necrotic bowel was subsequently resected.

BACKGROUND

A transmesenteric hernia is a protrusion of bowel through a congenital defect in the mesentery. Although transmesenteric hernias are due to congenital defects, they usually present in adulthood;¹ only a small proportion of symptomatic cases have been reported in children and newborns.^{1–3} The rarity of this condition, in addition to the lack of specific radiological and laboratory findings means that diagnosis is often delayed in the paediatric population. Compared with other forms of hernia, transmesenteric hernias have a higher tendency to cause a volvulus or strangulation resulting in ischaemia. Incidences of these complications are reported to be between 30% and 40%, respectively; they are frequently misdiagnosed, which leads to a delay in surgery. Death rates for patients who are treated surgically range from 30% to 50%; this increases to 100% if patients do not receive appropriate treatment.^{1–6} We present the case of a girl admitted with acute abdominal obstruction; this was the result of a mesenteric hernia complicated by a triple necrotic volvulus.

CASE PRESENTATION

A 6-year-old girl, with severe abdominal pain and vomiting, was presented to our paediatric emergency department (ED). Previous to this she was admitted to the paediatric ED with recurrent abdominal pain on four separate occasions in the same year.

On examination, the girl was afebrile, pale and visibly upset. She was not dehydrated, her heart rate was 142 beats/min and her blood pressure was 105/75 mm Hg. The abdomen was soft with mild tenderness in the right upper quadrant. On palpation, no abdominal masses were present. The remaining physical examination was normal. Following admission the pain gradually diminished and there was complete resolution of symptoms. However, during radiological investigations, when she changed position from lying to standing, her clinical condition

suddenly deteriorated. She developed a syncopal episode with extremely slow recovery, despite prompt clinical assistance. At the clinical examination the girl appeared extremely pale, with increased body temperature (38.5°C), persistently high heart-rate (153 beats/min), but normal blood pressure. Shortly after, two episodes of haematemesis were reported without modification of her clinical abdominal evaluation. Radiological findings demonstrated intestinal obstruction; due to the patient's unstable clinical condition, abdominal exploration was indicated. A triple necrotic volvulus was revealed, the direct consequence of a mesenteric hernia.

INVESTIGATIONS

Initial laboratory investigations revealed an increased white blood cell (WBC) count ($15 \times 10^9/l$) with neutrophilia (80.3%); however C reactive protein (CRP) was normal (0.63 mg/dl). All other blood tests were normal. Sonography revealed thicker bowel loops with a hyperaemic wall in the right upper quadrant; there were reduced peristaltic movements and a moderate fluid film in the abdomen. Plain abdominal x-ray showed signs of acute small bowel obstruction: abnormal dilatation of the proximal small bowel loops with complete dislocation in the left hemiabdomen (figure 1). A lateral abdominal view revealed multiple air and fluid levels without extraluminal gas (figure 2).

Following clinical deterioration the patient had a raised WBC count ($24 \times 10^9/l$) although CRP remained normal.

DIFFERENTIAL DIAGNOSIS

Radiological imaging revealed that the patient's signs and symptoms were due to acute intestinal obstruction. In children intestinal obstruction is usually the result of intussusception, incarcerated hernia, midgut volvulus with or without bowel malrotation and postoperative adhesions.

Intussusception is the most frequent cause of intestinal obstruction in children between 3 months and 5 years of

Figure 1 Abnormal dilatation of proximal small bowel loops with completely dislocation in the left hemiabdomen.

age; it has a peak incidence in children aged between 5 and 7 months.⁷ All internal hernias, excluding those created following laparotomy, account for only 1.78%.⁴

Radiological findings excluded many of the above diagnoses. Sonography did not reveal target or whirlpool images, which may have been suggestive of intussusception or midgut volvulus, respectively.⁸⁻⁹ There was no history of abdominal surgery and therefore postoperative adhesions could be excluded.

Peritonitis was unlikely as there was no rebound tenderness on clinical examination and CRP was not raised. The high WBC count was considered to be a non-specific manifestation, possibly the result of the multiple vomiting episodes.

A CT scan was not performed as the dramatic decline in the patient's clinical condition required immediate laparoscopic exploration.

TREATMENT

Owing to the complexity of the surgical situation, a decision was made to convert laparoscopic exploration to an open procedure and an exploratory laparotomy was performed. This revealed a triple volvulus consisting of

Figure 2 Air-fluid levels without extraluminal gas.

Figure 3 Necrotic bowel herniated and twisted into the mesenteric defect, as it appeared to surgery.

190 cm of necrotic and gangrenous small bowel loops that were subsequently resected (figures 3 and 4). The volvulus occurred as a consequence of bowel loops that had herniated through a mesenteric defect. The defect had a diameter of about 20 cm and was located in Treves' area (figure 5). The surgical procedure was completed with an end-to-end anastomosis and closure of the mesenteric defect. Incidentally, a Meckel's diverticulum was found in the necrotic bowel.

OUTCOME AND FOLLOW-UP

The patient recovered rapidly; normal bowel function was obtained within 4 days and she was discharged with no serious sequelae.

DISCUSSION

Internal hernias have an autopsy incidence of 0.2–0.9% and transmesenteric hernias account for 5–10% of these. In total, 35% of transmesenteric hernias result in clinical problems in the paediatric age group, ranging from recurrent

Figure 4 The resected tract of necrotic small bowel loops (190 cm).

Figure 5 Evidence of the huge mesenteric defect once reduced the internal hernia.

abdominal pain to small bowel obstruction. They are also the leading cause of internal hernias in children.¹⁰

The pathogenesis of the mesenteric defect can be related to three main hypotheses: a partial regression of the dorsal mesentery,¹¹ the coalescence of two opposed epithelial layers with a deficient intervening of connective tissue¹² or the consequence of inadequate vascularisation.¹³ The last hypothesis is also supported by two recent case reports.^{14 15} These authors noted specific vascular anomalies in patients with transmesenteric hernias: in one case there was a unique ileocolic branch surrounding the mesenteric defect. The other case demonstrated collateral circulation arranged in an 'apple-peel'-like configuration similar to that seen in ileal atresia. Transmesenteric hernias and ileal atresia are pathologies that seem to be strongly linked. Thus if ileal atresia is the result of an intrauterine vascular insult, transmesenteric hernias may be the manifestation of a less severe vascular accident. Treves' field is the area that is most commonly implicated with herniation. It consists of a portion of mesentery near the terminal ileum devoid of fat, visible blood vessels and lymph nodes. For this reason, the area is highly susceptible to injury during embryonic development. In our case, although the location of the defect was in Treves' area, vascular anomalies or other congenital defects were not detected.

In total, 30% of all internal hernias are clinically silent with no symptomatic manifestations of the disease.¹⁶ However, the majority of cases are characterised by recurrent abdominal pain caused by intermittent herniation and spontaneous reduction of the bowel loops.^{2 4} In addition, review of the literature suggested that 30–40% of paediatric patients with mesenteric hernias present with acute bowel obstruction.⁶

Over the past year, our patient was discharged on four separate occasions from the ED and was diagnosed with recurrent abdominal pain. During her last admission she developed acute abdominal obstruction; she appeared to be in a toxic shock-like state and this required immediate abdominal exploration.

The preoperative diagnosis of a transmesenteric hernia is challenging, as it is a rare condition with varied modes of presentation and non-specific laboratory or radiological findings.

Sonography is a valuable tool in the diagnosis of acute abdominal obstruction in paediatric patients. It can demonstrate specific features that are diagnostic of intussusception or volvulus.^{17 18} In our patient, the 'whirl-pool' sign—indicative of a volvulus—was not detected on abdominal ultrasound. However, there were thickened bowel loops and fluid in the abdomen, which are suggestive of ischaemia.

CT is the gold-standard imaging technique for the diagnosis of all internal hernias.⁶ The most important diagnostic indications for CT scans include: evidence of small bowel obstruction, clustering of the small bowel, displacement of other bowel segments and stretched displaced crowded or engorged mesenteric vessels.^{8 19 20} Accuracy of the CT image is dependent on the presence of a confining sac. A transmesenteric hernia that does not contain a sac has a variable appearance on CT, resulting in diagnostic uncertainties. CT scans are inconclusive in about 40–50% of cases, even when reevaluated following confirmatory surgery.^{6 8 20} Nakazawa *et al*'s² study showed that only one patient of a total of four, was diagnosed with CT prior to surgery. Delayed exploration in patients presenting with acute abdominal obstruction, may increase subsequent mortality from gangrenous bowel necrosis (27% of cases).⁴ For these reasons, CT scans should not be used to confirm transmesenteric hernias in critically ill children with acute bowel obstruction.

Surgical exploration is currently the gold standard for the definitive diagnosis of a transmesenteric hernia; it can also be a life-saving procedure if it is associated with any complications.^{1 2 9} In conclusion, repetitive ED admissions for acute abdominal pain should alert physicians to focus on rarer abdominal pathologies including, mesenteric hernias. Immediate surgical evaluation of critically ill patients with complicated transmesenteric hernias, allows an early diagnosis, which could be life saving and minimise the need for small bowel resection.

Learning points

- ▶ In paediatric patients, admitted to the emergency department for abdominal obstruction, commoner diagnosis has to be ruled out. After that, rarer pathologies, as our case of complicated mesenteric hernia, should be kept in mind.
- ▶ Preoperative diagnosis is very difficult because of common non-specific clinical and radiological findings.
- ▶ Prompt surgical exploration is the best way to reduce the risk of gangrenous necrosis and mortality in critically ill patients with acute abdominal obstruction with an unknown aetiology.

Competing interests None.

Patient consent Obtained.

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