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An uncommon cause of well-known symptoms: acute abdomen in a 2-year-old boy with intestinal malrotation. Case report and literature review

Rzadka przyczyna dobrze znanych objawów: ostry brzuch u 2-letniego chłopca z nieprawidłowym zwrotem jelit. Opis przypadku i przegląd literatury

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Abstract

Midgut malrotation is the most frequent congenital defect of the small intestine. The clinical manifestation can vary from being asymptomatic to presenting acutely as volvulus with bilious vomiting.

Presented here is a case of a 32-months-old boy with abdominal pain and several emetic episodes before admission. The patient was diagnosed with ileus and needed emergency surgery which showed the presence of volvulus due to malrotation of the midgut. Extensive partial resections of the jejunum, ileum and colon beginning in the upper part of the rectum up to ascending colon were performed.

Intestinal malrotation is rarely a symptomatic abnormality, however, when it occurs severely it can result in life-threatening complications. Ultrasonography may be a helpful screening tool for early diagnosis, but it needs the experience of the doctor. Treating significant malrotation almost always requires surgery. The timing and urgency depend on the child's condition.

Key words: acute abdomen, child, congenital abnormality, intestinal malrotation, volvulus

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Introduction

Intestinal malrotation is the most frequent defect of the small intestine that develops during foetal life and is estimated to occur in 1 in 200–500 live births, with a male predominance. However, only 1 in 6000 live births have a symptomatic course of the disease [1–3]. Between the fourth and eighth week of foetal life, the midgut rotates in a counterclockwise direction around the superior mesenteric artery (SMA). It protrudes through the yolk stalk and makes a 90-degree rotation, intensively increasing its length. Eventually, the midgut retracts to the abdominal cavity to perform another 180-degree turn in the same direction [4]. This phenomenon ensures optimal digestive tract arrangement, though its complexity may lead to the occurrence of abnormalities in four different ways: non-rotation, incomplete rotation, reverse rotation and anomalous fixation of the mesentery. The most common symptoms are vomiting and abdominal pain [5]. Herein a case is presented of a 32-months-old boy with intestinal malrotation with volvulus, who needed a vast segmental resection of the bowel and developed short bowel syndrome.

Case report

A 32-months-old boy was presented with abdominal pain and several emetic episodes prior to admission. His

general condition was severe. The patient had a history of infancy *E.coli* sepsis. He had also been hospitalized because of an ileus suspicion. Recent recurrent abdominal pain was also reported.

Physical examination revealed a pulse rate of 130/min and crepitation at the bases of both lungs. The abdomen was dolorous, and excessively flatulent presenting no peristalsis on auscultation. Per rectum examination disclosed coprostasis. Laboratory investigations revealed: hyperglycaemia (656 mg/dl), hyponatraemia (122.7 mmol/l), hyperkalaemia (5.74 mmol/l), slightly increased C-reactive protein level (1.6 mg/dl), increased serum creatinine (1.14 mg/dl), low red blood cells count (3.8 $10^6/\mu l$), low haemoglobin (9.1g/dl), high total white cell count (22.3 $10^3/\mu l$), acidic pH (7.18), high pCO_2 (78.9 mm Hg).

After being treated in a paediatric diabetic department because of diabetes and metabolic acidosis, the patient was admitted to the Department of Paediatric Surgery, Traumatology, and Urology in Poznan with suspicion of ileus and dyspnoea due to increasing obstruction of the gastrointestinal tract and enlarged abdominal circumference. Abdominal ultrasonography was performed revealing a very large amount of intestinal gas. Intestinal loops were 4–5 cm wide, with walls up to 10 mm thick, with no evident peristalsis, and filled with liquid and faeces. Six spleens were localized. Fluid was detected in the left pleural cavity; however, an anteroposterior chest X-ray was unremarkable. Eventually, an abdominal X-ray demonstrated clear signs of ileus: air-fluid levels.

Exploratory laparotomy was performed on the day of admission. The abdominal cavity was filled with turbid, faecally-stinking fluid. The colon and the small intestine were dilated, swollen and dark in colour. Midgut malrotation with short mesenteric root, in which vascularization ran to the entire jejunum, ileum and colon, was diagnosed. After the detorsion of the intestines, the major length of the bowel was completely affected by necrosis with blood clots occluding big vessels (Fig. 1).

Extensive segmental resections of the jejunum, ileum and colon beginning in the upper part of the rectum up to ascending colon were performed. Then surgeons made:
— ileostomy 30 cm distal to duodenum;
— closure of ileum, 30 cm proximal to the ileocecal valve;
— colostomy of the ascending colon.

An appendectomy was not performed. Intestinal blood circulation was uncertain as the bowels were partially darkened. After surgery, the patient in severe condition was admitted to the paediatric intensive care unit. Next, he was transferred back to the surgical department on the 12th postoperative day after his biochemical, morphological and USG parameters stabilized. His general condition was good. He was equipped with two stoma bags on both sides of the abdomen. On the 16th postoperative day, the patient was in good general condition, physical examination was unremarkable. He was on parenteral nutrition with good results in introducing oral nutrition for two days (Fig. 2). When the patient's condition stabilized, he was discharged home. After 3 months the child was transferred to a hospital in Warsaw, to have the digestive tract continuity restored surgically. The patient presented short bowel syndrome (SBS) and is in a need to be fed mostly parenterally ever since.

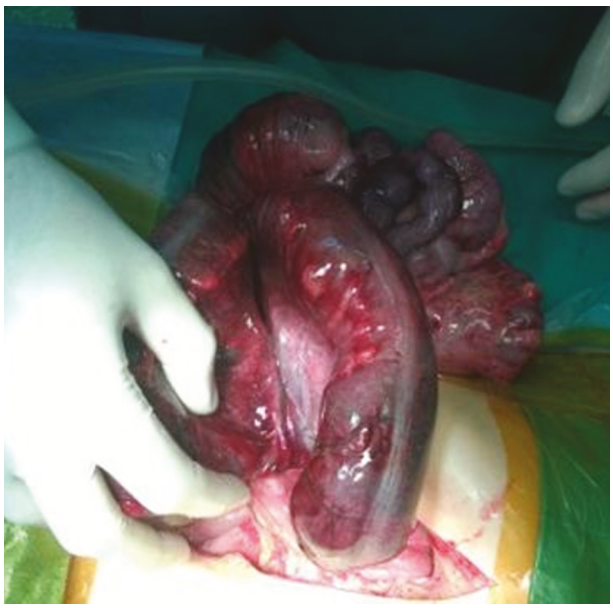

Figure 1. Necrotic bowel found during laparotomy

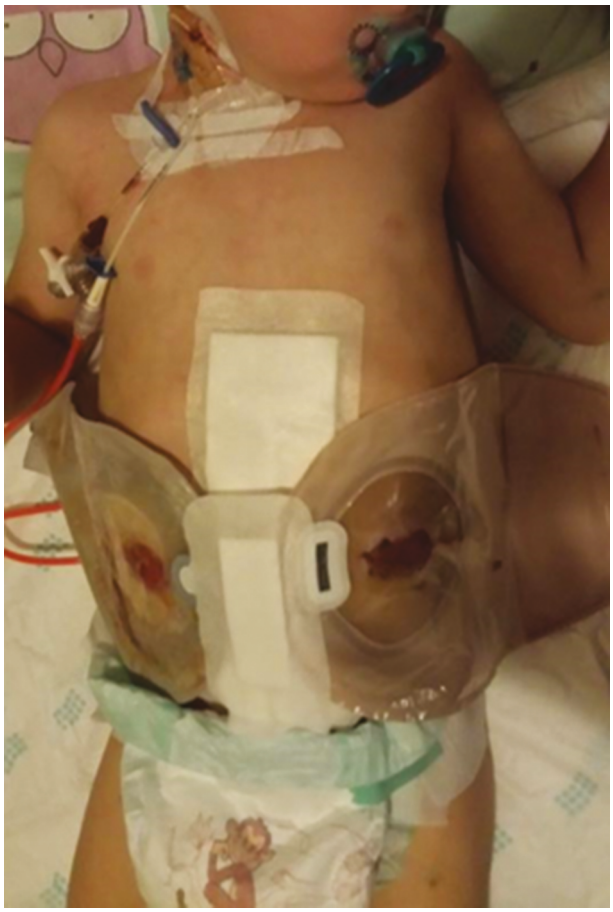

Figure 2. Patient 2 weeks after extensive intestinal resection, with intestines supplied with two stoma bags

Discussion

Intestinal malrotation is the most frequent defect of the small intestine that develops during foetal life. Most of the anomalies remain asymptomatic, though the presented patient had been hospitalized in the past, before admission to the hospital with a suspicion of ileus. A major part of patients present within the first year of life, however, this case refers to an over 2,5-year-old boy. Midgut malrotation coexists with other congenital defects such as congenital heart disease, heterotaxy (including polysplenia or asplenia) and omphalocele [6, 7]. Polysplenia was the only abnormality detected in the examination.

The deformation can be divided depending on its course: acutely, intermittently, or asymptomatically [6]. The present patient had an intermittent history with an acute ending. Nutrition disorders, growth disturbance and enteritis with protein loss may be indicative of a mild midgut malrotation course [8]. This was not observed in this patient. Typical acute presentation in children involves abdominal pain and bilious vomiting, secondary to bowel obstruction same as in the present patient. In infants and children, the presentation involves chronic or intermittent abdominal pain and constipation, reduced food intake, distention, recurrent vomiting, growth disturbance, chronic diarrhoea and malabsorption. It is essential to remember that most of the abnormalities remain asymptomatic [2, 9].

Diagnostic imaging is based on a contrast RTG study of the abdomen which defines duodenojejunal flexure in relation to the pylorus and vertebral column. Other findings can show the disposition of caecum or double bubble sign (dilated stomach and duodenum) and air-fluid levels in patients who develop severe midgut volvulus same as in this patient. Ultrasonography of the abdomen has almost 100% sensitivity in detecting neonatal malrotation and was suggested to be a good screening tool for effectively ruling out malrotation at risk of volvulus [10]. It shows the inversion of the SMA and superior mesenteric vein (SMV), which can be also revealed in the computed tomography study [11–13]. Additionally, USG may show dilated bowel loops which was the only ultrasound finding present in the present case. The present patient, however, was hard to diagnose until exploratory laparotomy was performed as the literature suggests [14–16].

Open Ladd's procedure is a gold standard for the management of midgut malrotation, though recent retrospective studies suggest benefits of the laparoscopic approach that need to be further investigated [17–19]. The procedure consists of detorsion of the volvulus, division of abnormal peritoneal Ladd's bands, widening of the mesenteric root, positioning of the bowel in a stable position and prophylactic appendectomy (optional) [20]. In the present patient, the Ladd's bands were not observed. A laparotomy and extensive intestinal resection were performed. Woo S. states that patients with white blood cells over 15,000/μl are at higher risk of needing an intestinal resection [21]. This theory confirms in the present patient with leucocytosis, who had a major length of bowel completely affected by necrosis. He presented with signs and symptoms of malabsorption such as weight loss, diarrhoea, dehydration, malnutrition, and electrolyte imbalance. SBS was caused by extensive small intestine resection due to volvulus. This complex condition can be life-threatening [22]. After surgery, the remaining intestine slowly adapts. Some children with severe short bowel syndrome will need long-term parenteral nutrition. This can sometimes cause problems. If this happens, a patient may need intestine transplantation [23]. The patient is under constant outpatient care, currently without any decision on transplantation.

Conclusions

Intestinal malrotation should be considered as a potential cause of bowel obstruction in the paediatric population. Abdominal ultrasound as a screening tool may be a good, non-invasive way to prevent severe presentations such as volvulus. In severe cases of volvulus with necrotic, ischemic intestine segmental resection of the bowel might be needed. Patients who develop malrotation with volvulus should be treated by a qualified multidisciplinary team. Management of SBS requires an interprofessional team led by a physician with gastrointestinal expertise.

Conflict of interest

None declared

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