Rare Clinical Presentation of Intestinal Malrotation After Neonatal Period: Protein-losing Enteropathy Symptoms Due to Chronic Midgut Malrotation

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Abstract

Protein-losing enteropathy is caused by a variety of diseases. However, it is rarely caused by chronic intestinal malrotation. A 1-year-11-month old male baby and a 12-year-old female presented with chronic diarrhoea and hypoproteinaemia. Both of the patients underwent an exploratory laparotomy, in which a midgut malrotation was discovered. Intestinal malrotation should be considered as one of the differentials when diagnosing protein-losing enteropathy.

Key words

Diarrhoea; Hypoproteinaemia; Intestinal malrotation; Laparotomy

Introduction

Intestinal malrotation is a congenital anomaly, which is mainly diagnosed during infancy. The presenting malrotation symptoms are mainly bilious vomiting, abdominal distention, and incomplete or complete intestinal obstruction. Protein-losing enteropathy (PLE) is rarely caused by chronic intestinal malrotation. Here, we present two cases, a 1-year-11-month old male baby and a 12-year-old female, who presented with chronic diarrhoea and hypoproteinaemia. Both of the patients underwent an exploratory laparotomy, which confirmed midgut malrotation.

Case Report

Case one was a 1-year-11-month old male baby with a chief complaint of recurrent diarrhoea for six months and a decreased level of serum albumin for five months. The patient had watery diarrhoea for six months, with 5-6 bowel movements per day, which was accompanied by a mild fever. After being admitted into the ward, the laboratory examinations revealed a decreased level of serum albumin (17.6 g/L), while the complete blood count was normal. The faecal tests for pathogens were all negative, and the liver and renal functions were normal. There was no significant finding during gastroendoscopy or colonoscopy. A gastrointestinal contrast indicated intestinal malrotation. An abdominal computed tomography (CT) scan confirmed that the mesenteric vessels presenting as a "whirl sign" (Figure 1). During the physical examination, his vital signs were stable, the weight of the patient was 12.4 kg, and the height was 89 cm. Lung and cardiac auscultation were normal. No abdominal tenderness was present during abdominal palpation, nor was there evidence of hepatosplenomegaly or oedema of the lower limbs. And then he underwent an exploratory laparotomy, in which a midgut malrotation with mesenteric swelling and narrowing was discovered. He underwent the Ladd procedure and recovered.

Case two was a 12-year-old female who presented with a primary complaint of an intermittent fever and diarrhoea occurring for six months and lower limbs oedema over a
period of 12 days. The patient had intermittent fever accompanied with diarrhoea for six months, 2-3 mushy stools per day, and mild abdominal pain. The laboratory test showed decreased serum albumin level 20 days ago, and she presented with lower limbs oedema 12 days ago. During the physical examination, her vital signs were stable at the time of admission, the weight of the patient was 33 kg. The lung and heart sounds were normal. The patient had abdominal distension with a positive shifting dullness. Her face, abdominal skin, and lower limbs had pitting oedema. The albumin level was 8.5 g/L, while the complete blood count was normal. The electrolyte analysis revealed mild hypokalaemia and hyponatraemia. There were no significant findings during the gastroendoscopy and the colonoscopy. She underwent surgery, which confirmed a midgut rotation with volvulus; the duodenum and ascending colon were compressed by Ladd's bands.

Discussion

PLE is caused by a variety of diseases, such as primary and secondary lymphangiectasia, intestinal inflammation, vasculitic disorders, and tumour. Intestinal malrotation is a congenital anomaly that results from an abnormal or incomplete rotation and fixation of the midgut during embryonic development. Approximately 75% to 85% of these patients are diagnosed during infancy. The symptoms of malrotation include bilious vomiting, abdominal distention, and incomplete or complete intestinal obstruction. About one-third of intestinal malrotation cases are diagnosed beyond the period of infancy. The chronic presentation is a diagnostic challenge. The chronic intestinal malrotation symptoms in older patients usually include either atypical symptoms, such as abdominal pain, nonbilious vomiting, failure to thrive, malabsorption, anaemia, chylous diarrhoea or a lack of any clinical symptoms, and is only discovered during surgery for other diseases. The pathophysiology of these chronic symptoms may relate to the compressive effects of the peritoneal bands running from cecum and ascending colon to the right lateral wall. In the study of Nilesh G, recurrent colicky abdominal pain (61.9%), nonbilious vomiting (38.1%), and failure to thrive/weight loss (33.3%) were the most common presentations of the older patients. Other older patients in this study presented with early satiety, abdominal bloating, acute pancreatitis or acute small intestinal obstruction, and some were diagnosed with malrotation intraoperatively.

The diagnosis of intestinal malrotation is mainly based on the typical clinical manifestations. An upper gastrointestinal contrast study is diagnostic for malrotation in most patients. Ultrasonography can reveal either an abnormal relationship of the superior mesenteric artery (SMA) and vein or a classic whirlpool sign of the midgut volvulus for some patients. Contrast CT scan is needed to confirm or differentiate a diagnosis, and sometimes is helpful in assessing mesenteric ischaemia. The characteristic appearance of a twisted mesentery, collapsed small bowel loops, and mesenteric fat wrapped around the SMA is pathognomonic for malrotation and is commonly referred to as the "whirl sign" or "clockwise whirlpool sign". Intestinal malrotation with chronic symptoms usually need surgery as soon as possible in case of intestinal volvulus. Emergence surgery would be more complicate than elective operation, including life-threatening bowel necrosis requiring an extensive small intestinal resection.

It is rare that intestinal malrotation presents with protein losing enteropathy symptoms. Protein losing enteropathy can be caused secondary to either lymphatic obstruction or intestinal lymphangiectasia as a result of malrotation. To our knowledge, there are only two case reports documenting a malrotation-induced protein-losing enteropathy. One case was a 17-month-old boy presenting with hypalbuninaemia, peripheral oedema, diarrhoea, and failure to thrive since 9 months of age. His laboratory analyses revealed a low serum level of albumin and a lymphopenia without sings of lymphangiectasia. A CT of the abdomen revealed a whirlpool sign and suggested an incomplete vascular volvulus. During the laparotomy, chronic midgut volvulus was discovered.

Figure 1  Abdominal CT scan: Part of the vessels branches are dilated obviously, mesenteric vessels presenting typical "whirl sign".
with a 180° twisting of the jejunum and the superior mesenteric vessels causing a lymphatic obstruction and leakage of milky white lymph into the cut surfaces of the mesentery. Morozov et al reported a young Russian male patient with protein-losing enteropathy, who was diagnosed a malrotation of the duodenum with recurrent midgut volvulus causing secondary intestinal lymphangiectasia. The two cases reported identified either lymphatic obstruction or lymphangiectasia. However, in our cases, we did not find any evidence of lymphangiectasia or lymphatic obstruction. Although intestinal malrotation is rare beyond the age of infancy, it should be considered as an atypical manifestation. For protein losing enteropathy, intestinal malrotation should be added as one of the causes of the disease.

**Conclusion**

To conclude, an intestinal malrotation should be suspected in all patients presenting with varied acute or chronic abdominal symptoms. Intestinal malrotation should be considered as one of the differentials when diagnosing protein losing enteropathy.

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**Conflicts of Interest**

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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