Protein-losing Enteropathy Caused by Spontaneous Reduction of Intussusception with Meckel's Diverticulum

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Protein-losing enteropathy (PLE) is a relatively rare condition. In this article, we report the case of a 6-year-old boy diagnosed with PLE who developed intussusception, in whom at operation Meckel's diverticulum was identified in his intestine. Spontaneous reduction of intussusception is thought to relate to the mechanism of PLE.

Key words: Protein-losing enteropathy, Intussusception, Meckel's diverticulum, Spontaneous reduction of intussusception

INTRODUCTION

Intussusception is a frequent cause of bowel obstruction in children. The incidence of a definite anatomic lead point ranges from 2 to 12% in reported series [1]. These include Meckel's diverticulum, the appendix, polyps, carcinoid tumors, submucosal hemorrhage resulting from Henoch-Schdnlein purpura, non-Hodgkin's lymphoma, foreign bodies, ectopic pancreas or gastric mucosa, and intestinal duplications. The most common pathological lesion is a Meckel's diverticulum. The incidence of an anatomic lead points increases in proportion to age [2]. With the improvements in resolution and quality of ultrasound images, spontaneous reduction of intussusception was encountered more commonly than previously reported [3−6]. Those patients are often asymptomatic unless there is complete obstruction. On the other hand, Protein-losing enteropathy (PLE) is a relatively rare condition characterized by loss of protein in the intestines causing hypoalbuminemia, hypoproteinemia, edema and occasionally diarrhea [7]. However, it is not well known that PLE could be caused by intussusception. Here, we present a case report that has a rare combination of intussusception and PLE.

CASE REPORT

A 6-year-old boy in previously good health was admitted to the hospital with vomiting, abdominal pain. Physical examination revealed intermittent lower abdominal pain and slight peripheral edema. Laboratory tests showed increased white blood cell count (17500/μl) and hemoglobin (18.3 g/dl) with dehydration. Other laboratory tests showed low serum total protein level (4.1 g/dl) with hypoalbuminemia (albumin, 2.5 g/dl). There was no evidence of proteinuria on urinalysis. Laboratory findings are summarized in Table 1. The ultrasound revealed slight ascites and edema of the intestine. The scintigraphy using 99mTc-human serum albumin revealed protein loss around the ileocecal lesion and the fecal alpha-1 anti-trypsin clearance showed abnormal (66.5 ml/day). These findings indicated he

Table 1 Laboratory findings

【CBC】		Glucose	141 mg/dl
WBC	17500 /μl	Na	139 meq/l
Hb	18.3 g/dl	K	5.1 meq/l
Ht	52.9 %	Cl	105 meq/l
Plt	33.9×10^4/μl	CRP	0.126 mg/dl
【Blood Chemistry】		Serum protein electrophoresis	
TP	4.1 g/dl	Alb 61.7	α 1 8.2 α 2 15.1
Alb	2.5 g/dl	β 10.2	γ 4.8
AST	26 U/l	【Urinalysis】	
ALT	10 U/l	Protein	(−)
γ GTP	7 U/l	Glucose	(−)
BUN	25 mg/dl	SG	1.029
Cr	0.60 mg/dl	Ketones	1+

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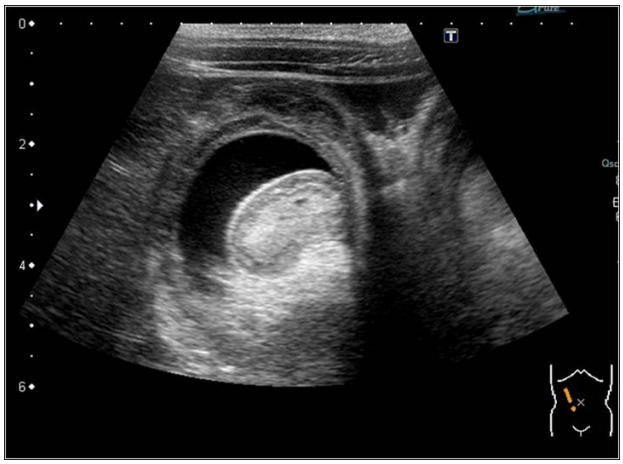

Fig. 1 Ultrasound revealing the typical "target sign" which is a characteristic image in the intussusception and fluid in the intestine.

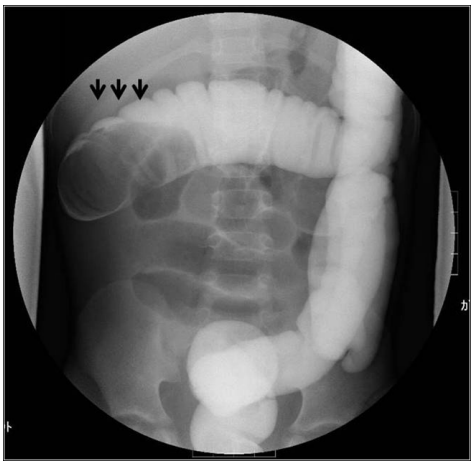

Fig. 2 Enema revealing "crab's claw sign" (arrows) in the transverse colon.

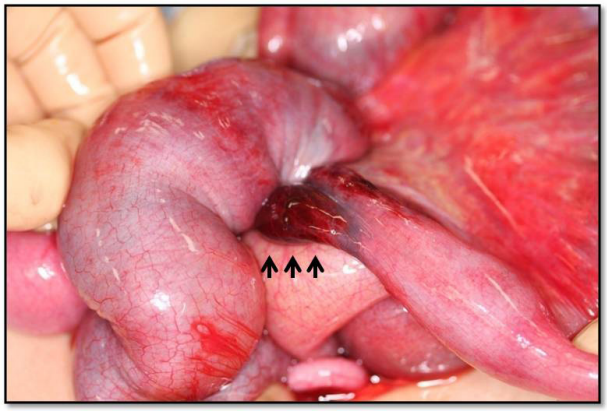

Fig. 3 Intraoperative photograph showing distal ileum invaginated into the ascending colon (arrows).

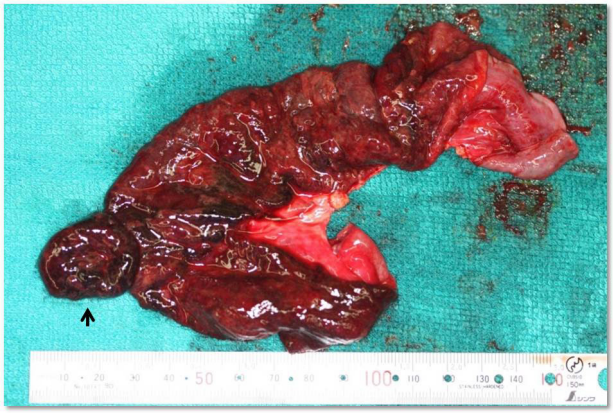

Fig. 4 Resected specimen revealing necrotic intestine with Meckel's diverticulum (arrow).

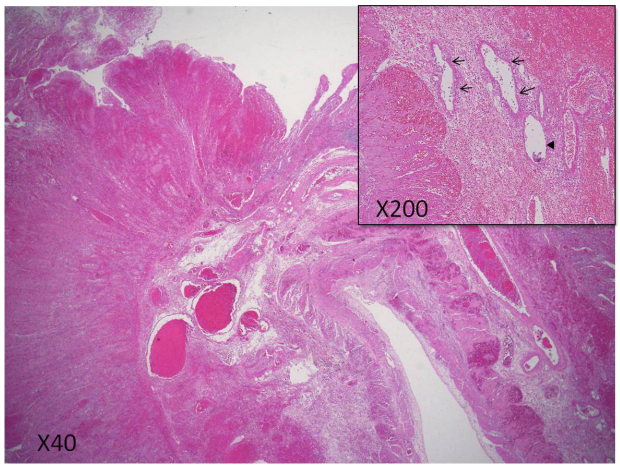

Fig. 5 Histopathological examination revealing the dilation of the lymphoducts (arrows) and vessels (triangle) in submucosa of the resected intestine. (H & E)

was in the condition with PLE. However, human albumin transfusion didn't make any improvement of his laboratory tests. A week after conservative treatment, he had an acute cramping abdominal pain accompanied with hematochezia and the intussusception was identified with ultrasound (Fig. 1). Although Hydrostatic reduction was attempted repeatedly (Fig. 2), successful enema reduction of the intussusception didn't completely exclude the lead point. Laparotomy revealed ischemic bowel loops caused by intussusception. Part of the distal ileum was observed to be invaginated into the ascending colon (Fig. 3). Since attempting resolve the intussusception using Hutchinson's maneuver was not successful, an ileocecal resection was performed. A gross examination showed an ileocecal intussusception with the lead point at Meckel's diverticulum (Fig. 4). Histopathological examination showed that submucosa of the resected intestine had dilation of the lymphoducts and vessels (Fig. 5). The necrotic tissue of the Meckel's diverticulum was observed. The postoperative course was uneventful and the hypoproteinemia was gradually recovered without albumin infusion.

DISCUSSION

PLE is a relatively rare condition in children. Most frequent causes of this condition are cardiac and gastrointestinal disorders. Gastrointestinal causes are divided into erosive gastrointestinal disorders, nonerosive gastrointestinal disorders, and disorders involving increased central venous pressure or mesenteric lymphatic obstruction [8]. PLE is mainly a diagnosis by exclusion. The diagnosis of PLE should be considered in patients with hypoproteinemia after other causes, such as malnutrition, proteinuria, and impaired protein synthesis due to cirrhosis, have been excluded. The most efficient examination is a fecal alpha-1 anti-trypsin clearance and a scintigraphy using 99mTc-human serum albumin. Primary intestinal lymphangiectasia which is nonerosive gastric disorder causing PLE is a rare disease usually diagnosed in children [9]. Initially this might be the case for our patient, but later we found out intussusception had occurred.

Intussuscepition in older children is more likely to be associated with a lead point such as a Meckel's diverticulum. Symptoms of obstruction, vomiting, abdominal distention, and cramp-like pain are usually present. The Meckel's diverticulum may be recognized at operation, or may be found unexpectedly in the resected specimen [10, 11]. The most common types of heterotopic mucosa are gastric and pancreatic [12, 13]. In our specimen the Meckel's diverticulum was necrotic due to the obstruction. Even though it had a heterotopic mucosa, it was not reasonable to hypothesize that PLE was caused by Meckel's diverticulum since heterotopic mucosa is not known as losing protein from intestine. Although it is not well known that intussusception is related to PLE, in our case the scintigraphy using 99mTc-human serum albumin revealed protein loss around the ileocecal lesion. That indicates protein was losing from ileocecal lesion where we found out intussusception had occurred. In addition, histologically, submucosa of the resected intestine had dilation of the lymphoducts and vessels which suggested the evidence of existing lymphatic obstruction at the lesion of intussuception. Even though the ultrasound didn't show the findings of intussusception when he initially presented to the hospital, we assume intussusception was occurred by the time since he had abdominal pain and vomiting. The mechanism of protein loss in this condition is thought to relate to spontaneous reduction of intussusception which could have been the unusual cause of lymphatic obstruction.

Spontaneous reduction of intussusception was reported in 1940, Goldman *et al.* [14]. These authors suggested that this finding represented spontaneous reduction of an intussusception and postulated that this occurrence was probably more frequent than commonly believed at that time. Recently, Kornecki A *et al.* retrospectively reviewed 50 children series of spontaneous reduction of intussusceptions [3]. These authors reported 42% of children who presented with symptoms or signs suggestive of an intussusception (abdominal pain, vomiting or rectal bleeding). In the other 58% of children, the intussusception was incidentally found by imaging examination. The causes of spontaneous reduction of intussusceptions such as Henoch-Schonlein purpura, previous ileocolic intussusception that had been successfully reduced, and post laparotomy status were identified. Obstruction from Meckel's diverticulum is usually sudden onset, not well known as developing chronic progress. In our case, since he had been asymptomactic with conservative treatment for a week, we assume his intussusception was reduced spontaneously during this period.

In our case there was no evidence of having any other cause for PLE. The patient was asymptomatic when intussusception was reduced spontaneously. Moreover, after the patient underwent surgery resection, protein loss from the intestine disappeared without albumin infusion. We suppose a large amount of mucoid material excreted secondary to intussusception was lost without reabsorption in the intestine. Although the combination of intussusception and PLE are rare, spontaneous reduction of intussusception could have been an unusual cause of the PLE. No such case was encountered in a thorough research of literature using the keyword of PLE and intussusception. To the best of our knowledge, this is the first report of a patient with the combination of intussusception and PLE in children with Meckel's diverticulum. Although uncommon, it is important to consider that spontaneous reduction of intussusception could cause PLE.

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