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Enteritis Cystica Profunda Presenting as Ileoileal Intussusception in a Child: Report of a Case

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Abstract
We report the case of a 3-month-old male infant with small bowel intussusception caused by enteritis cystica profunda (ECP). The baby was admitted because he was refusing to feed, and was passing "redcurrant jelly" -like stools. A palpable mass was identified, and abdominal ultrasonography showed a mass with a lumen and lumen appearance. We performed laparotomy and resected the segment of bowel containing the mass. The resected segment had enteritis cystica profunda, which was considered to have precipitated the intussusception. A review of the English medical literature revealed only three other cases of children with similar symptoms in the last 30 years.

Key words Enteritis cystica profunda · Intussusception

Introduction

Enteritis cystica profunda (ECP) is a rare disorder in which benign epithelia-lined cysts develop in the mucosa and submucosa of the small intestine. A mucous cyst of the colon was first described in 1766 by Stark,[1] then in 1863, Virchow reported a second case of submucosal cysts and proposed the term "colitis cystica polyposa."[2] This type of cyst usually develops in the colon and is called "colitis cystica profunda" (CCP).[3] Colitis cystica profunda of the small intestine (ECP) is very rare and to the best of our knowledge, only three other cases of children have been documented.[4–6] We describe a case of ECP causing ileoileal intussusception in a 3-month-old boy.

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Case Report

A 3-month-old infant, who had previously been healthy, was admitted to our hospital because he had passed a "redcurrant jelly"-like stool and was refusing to feed. On admission, he appeared well nourished and well developed. He had a fever of 37.3°C but there was no vomiting, constipation, or diarrhea. Physical examination revealed an elastic-soft, mobile, abdominal mass, about 5 × 4 cm in size, located to the right of the umbilicus. His abdomen was slightly distended and bowel sounds were exaggerated. Laboratory tests revealed a minimal elevation of the white blood cell levels to 12.1×10^9/l (normal range $<9.5 \times 10^9$/l), but all other laboratory findings were within normal limits. A plain abdominal X-ray showed a small amount of intestinal air without any intrapelvic colonic air. Abdominal ultrasonography (Fig. 1) and a computed tomographic scan (Fig. 2) showed an axial bowel-within-bowel appearance. An air enema X-ray study revealed gaseous distention of the ileum and a complete obstruction, based on which a diagnosis of intussusception was made. We performed a laparotomy through a transverse abdominal incision with the patient under general anesthesia. The intussuscepted segment was located 27 cm proximal from the ileocecal valve and it could not be reduced. This segment was almost completely necrosed, whereas all other areas of the intestine were normal. We resected this segment, which measured 24 cm, and performed an ileoileal anastomosis. We also found a submucosal nodule, 1.4 × 1.0 × 0.5 cm in size, considered to be the lead point of the intussusception. The cut surface of the nodule contained many cystic structures (Fig. 3). The intussuscepted segment of the ileum had intramural cystic ducts in the submucosal layer, and an intestinal type columnar epithelium lined the inside surface of these ducts with mucosal stroma and the muscular layer of the mucosa (Fig. 4). The lesion did not form a muscular layer similar to the intestinal muscle wall or follow

the continuity of the true intestine. These histopathological findings suggested the ECP may have caused the ileal intussusception.

Discussion

Enteritis cystica profunda is a rare lesion of the small intestine, which has the same pathological features as CCP of the colon. Macroscopically and histologically

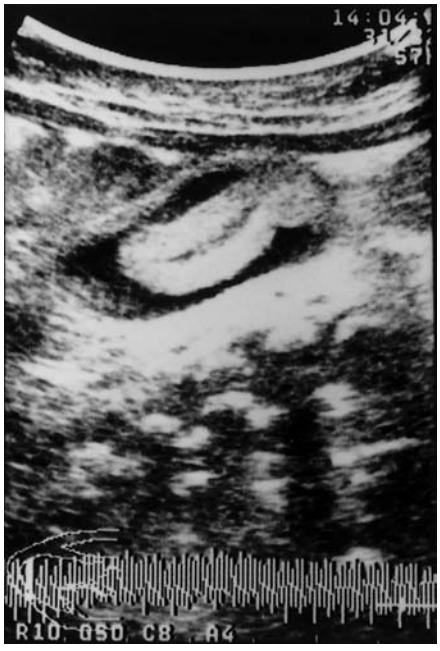


Fig. 1. Preoperative longitudinal ultrasound showed a bowel-within-bowel appearance

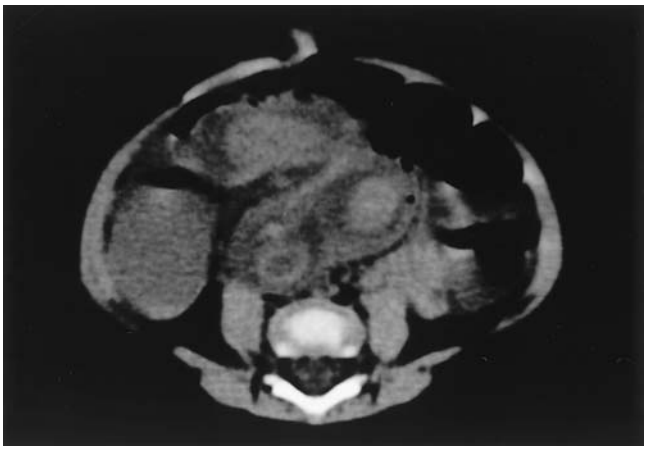


Fig. 2. Preoperative computed tomography scan showed a manifest bowel-within-bowel appearance

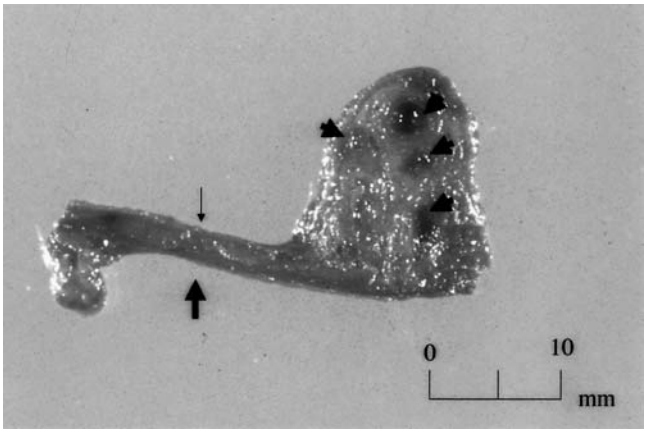
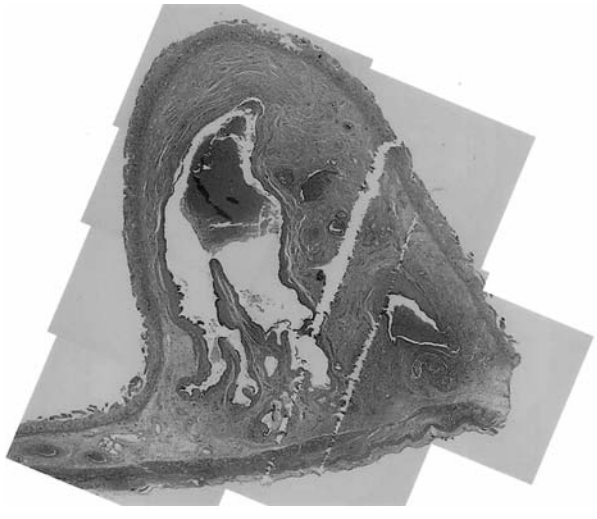


Fig. 3. Macroscopic view of the intramural nodule. *Fine arrow*, mucosa; *bold arrow*, adventitia; *arrowhead*, intramural cystic lesion



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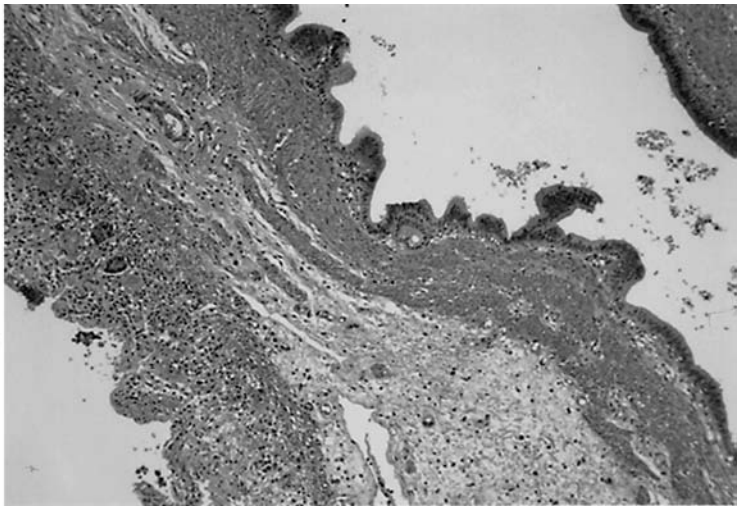

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Fig. 4. a The intussuscepted segment of ileum contained intramural cystic ducts in the submucosal layer (H&E, ×40). **b** The intestinal type of columnar epithelium lined the inside of those ducts with a mucosal stroma and a muscular layer of the mucosa (H&E, ×100)

Table 1. Reported cases of enteritis cystica profunda in children

First author (year)[Ref.]	Age	Sex	Clinical features	Location	Other disorders
Baillie (1970)[6]	20 m	F	Diarrhea	Jejunum	Polyposis
Karnak (1997)[4]	5 y	M	Intussusception	Ileum	No
Salman (1998)[5]	12 y	M	Abdominal pain	Jejunum	No
Present case (2004)	3 m	M	Intussusception	Jejunum	No

m, months; y, years

similar disorders have also been found in structures such as the esophagus, gallbladder, and stomach.[3] Although cystica profunda may arise in all parts of the gastrointestinal tract, it mainly develops in the large bowel. In 1766, Stark described benign mucosa-filled cysts of the colon, but it is only in the last 20 years that CCP has become readily recognized by pathologists and clinicians. Cystica profunda is a non-neoplastic condition associated with intramural mucus containing cysts. Sometimes mucus is observed to come out of the cyst, similar to the findings of our patient.[7] In adults, cystica profunda is often mistaken for mucinous carcinoma and invasive carcinoma. Sakurai et al. reported that CCP may also represent premalignant change or the initial pathological features of malignancy.[8]

The pathogenesis of CCP and ECP is not yet fully understood. Many investigators suggest that this disease has an acquired pathogenesis, but others believe it is cogenital. Cystica profunda occurs in various locations in the gastrointestinal tract; however, this cyst has not been noted in a large pediatric autopsy series. Cystica profunda has a high incidence of concurrence with other diseases that tend to irritate the bowel, such as ulcerative colitis, Crohn's disease, adenomatous polyps, Peutz-Jeghers syndrome, and spastic colitis.[7,9,10] Enteritis cystica profunda has been simulated in rats by suturing the small intestine to the abdominal wall and then exposing the mucosa externally.[11] Other hypothetical explanations for ECP are epithelial downgrowth through ulcerated mucosa, herniation through a weakened muscularis mucosa, re-epithelization of a submucosal abscess, and hemorrhage.[12]

Our research of the English-language medical literature revealed only three other children aged between 20 months and 12 years old with ECP (Table 1). Our patient and one other had cystica profunda in the ileum, which led to ileoileal intussusception.[4] Both these patients underwent resection because the intussuscepted segment was almost completely necrotic. One 12-year-old boy presented with acute abdominal pain and an ECP, 1.5 cm in diameter, was found in the terminal ileum.[5] Although the cause of his abdominal pain was not clearly defined, ECP was suspected to have caused the small bowel intussusception. A 20-month-old girl had multiple congenital cysts in other organs[6] and underwent a laparotomy for manual reduction of intussusception. Enteritis cystica profunda was found at the autopsy of this patient, although it does not appear to have caused her intussusception because the intussuscepted segment was located at a much lower position than the outlined ECP. It is notable that three of the four patients in Table 1 had no associated diseases. There have been many cases of ECP causing small bowel intussusception, similar to that observed in our patient; however, small bowel intussusception in pediatric patients is uncommon. In a reported series of 1126 pediatric intussusceptions, small bowel intussusceptions accounted for only 1.68%. Among these, clear lead points of the intussusception were found in only 44.4%. Complications secondary to small bowel intussusception, such as ischemic or necrotic bowel change, were also frequently encountered (36.8%).[13]

Enteritis cystica profunda is usually associated with other diseases, but our patient's ECP arose without any background factors. Thus we considered it to be of congenital origin, which would account for the intussusception. In conclusion, because ECP tends to cause small bowel intussusception in infants and young children, children operated on for small bowel intussusception should be checked for ECP.

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