

Crohn's disease of the terminal ileum in a patient with congenital peritoneal encapsulation of the small intestine: case report

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Abstract. *The authors present a rare case of peritoneal encapsulation of the small bowel in a 24-year-old male found incidentally during laparotomy for the Crohn's disease. The clinical and diagnostic aspects of the case are discussed.*

Keywords: *peritoneal encapsulation, Crohn's disease*

Tachecí I, Rejchrt S, Jaroš E, Langr F, Bureš J. Crohnova choroba terminálního ilea u nemocného s vrozenou peritoneální enkapsulací tenkého střeva: kazuistika. Folia Gastroenterol Hepatol 2004; 2 (3): 139 - 142.

Souhrn. *Autoři popisují vzácný případ peritoneální enkapsulace tenkého střeva u 24-letého muže, která byla nalezena náhodně při laparotomii pro Crohnovu chorobu. Jsou diskutovány klinické a diagnostické aspekty případu.*

Klíčová slova: *peritoneální enkapsulace, Crohnova choroba*

Peritoneal encapsulation of the small bowel is a rare congenital developmental anomaly of the abdominal cavity. The case of small bowel encapsulation associated with Crohn's disease of the small bowel is described.

Case report

A 24-year-old male presented with repeated abdominal pain (namely in the right lower quadrant) and weight loss in the last one year. The ultrasound examination and enteroclysis raised suspicion of Crohn's disease affecting terminal ileum and leading to stricture and entero-colic fistula, see Fig. 1. The colonoscopy identified congested, ulcerated mucosa

on the ileocaecal valve and the orifice of the enterocolic fistula in the transverse colon. The laboratory inflammatory markers were high.

The patient was operated on (after preoperative medical preparation) and a peritoneal encapsulation involving the major part of the small bowel was found, see Fig. 2. After incision of the peritoneal sac (see Fig. 3), inflammatory involvement of the terminal ileum with the enterocolic fistula was confirmed. The major part of the peritoneal sac, terminal ileum and right hemicolon were resected, with subsequent ileo-transversoanastomosis. Histological examination confirmed the diagnosis of Crohn's disease. On follow-up, the patient is well after 7 months since the



Figure 1
Enteroclysis. Irregular inflammatory stenosis in distal 15 cm of the terminal ileum (arrows). There are no radiograph signs of small intestinal encapsulation.
Enteroklýza. Nepravidelná zánětlivá stenóza distálních 15 cm terminálního ilea (šipky). Nejsou žádné radiologické známky nasvědčující peritoneální enkapsulaci tenkého střeva.

operation, without any symptoms of recurrence of the disease.

Discussion

There are three medical terms for small bowel encapsulation in the abdominal cavity used promiscuously. Each of them is a distinct pathological entity (19).

The first term is abdominal cocoon (7, 11, 17, 22, 24, 30). This is a total or partial encapsulation of the small bowel in a thick, opaque, dense fibrous membrane presents in adolescent girls from tropical or subtropical countries as acute or chronic bowel obstruction. The aetiology is unknown, an infective aetiology (tuberculosis) (17) or immunopathology (i.e. amyloidosis) (12) are suggested. Histopathological examination of the encapsulating membrane shows thickened vascular fibrocollagenous tissue, with or without areas of lymphocyte and plasma cell infiltrates. The earliest description of this disease was made by Owtschinnikow in 1907, entitled "peritonitis chronica fibrosa incapsulata" (ref. from 17). The term "abdominal cocoon" was first applied by Foo et al. in 1978 (7).

The second entity is sclerosing encapsulating peritonitis. It is a rarely observed complication of chronic peritoneal dialysis (8,9,13,14,21,26-28), the prolonged use of the β -adrenergic blocker practolol (4,5,27) or is idiopathic (15,31,32). Patients develop general

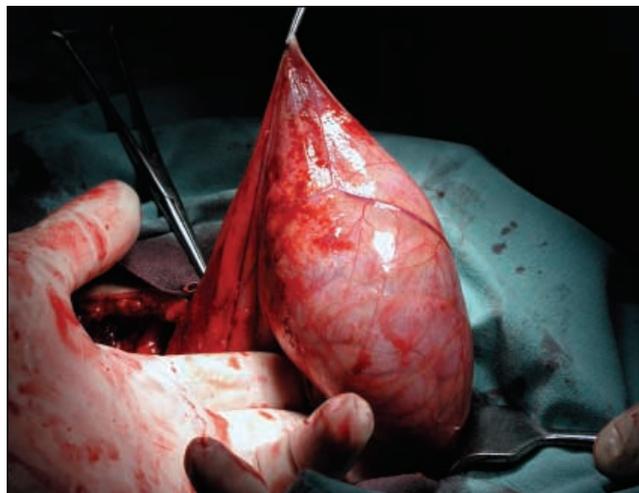


Figure 2
Intraoperative photograph shows encapsulation of the small bowel. The peritoneal membrane is otherwise normal. The sac is still to be opened.
Intraoperační fotografie zobrazuje enkapsulaci tenkého střeva. Peritoneum je normálního vzhledu. Vak dosud nebyl otevřen.

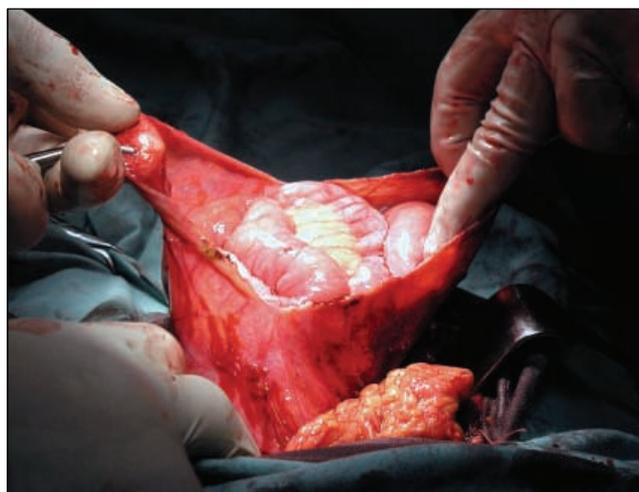


Figure 3
Intraoperative photograph after incision of the peritoneal sac. Normal intestinal loops are seen.
Intraoperační fotografie zachycuje situaci po incizi peritoneálního vaku. Jsou patrné kličky tenkého střeva normálního vzhledu.

peritoneal fibrosis and a part of (less frequently the entire) small bowel is encased in a fibrous membrane. This disease is presented with ascites or intestinal obstruction.

Last is the congenital peritoneal encapsulation of the small bowel (2,20,24,28,29), an exceedingly rare developmental abnormality. The entire small bowel (seldom only part of it) is lying behind an accessory but otherwise normal transparent peritoneal membrane between the omentum and mesocolon (19). It is probably developed from the yolk sac in the 12th embryological week. In this time the peritoneum of

the physiological umbilical hernia is drawn into the abdominal cavity along with the midgut (23). Histological examination reveals normal peritoneum without signs of inflammation. It could be associated with midgut malrotation and a vascular anomaly. The peritoneal encapsulation was first documented by Cleland in 1868 (ref. from 17).

As illustrated by this case report, small bowel encapsulation can be completely asymptomatic, found during laparotomy or autopsy (11). In our patient, this rare condition was diagnosed incidentally during laparotomy performed for Crohn's disease of the terminal ileum. However, intermittent abdominal pain, acute intestinal obstruction and, rarely, aortic occlusion have been reported (1,3,10,18,25).

The preoperative diagnosis is difficult when the patient is asymptomatic. In patients with non-strangulating small intestinal obstruction, two clinical signs are described (11,19). The first sign is asymmetrical, fixed distension of the abdomen, which does not change with peristaltic activity due to the unvarying position of the fibrous capsule (19). The second is the

difference in the consistency of the abdominal wall to palpation. The flat area is rigid, due to the dense fibrous capsule and the distended area soft, due to the thin walled distended small bowel (11,19). These signs are certainly non-specific and could only lead to the suspicion of this rare condition. In our case the clinical signs were modified by Crohn's disease with small bowel stenosis.

Radiographic studies are usually normal or non-specific. The correct diagnosis could sometimes be made by means of the CT (6,16).

It should be treated surgically by excision of the peritoneal sac. The resection of a necrotic ileal loop and intestinal reconstruction by an anastomosis in case of intestinal obstruction is necessary.

In summary, peritoneal encapsulation of the small bowel is an extremely rare intraabdominal developmental anomaly. It is associated with no typical clinical features, which may cause diagnostic difficulties. In our case the clinical picture was modified by Crohn's disease and it was recognised incidentally during laparotomy.

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