

A Rare Cause of Recurrent Intestinal Obstruction: Abdominal Cocoon

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ABSTRACT

Abdominal cocoon (AC) is a rare disease in which the small intestine is completely or partially surrounded by a thick fibrous membrane. A 56-year-old male patient was admitted to the hospital with complaints of abdominal pain, nausea and vomiting. It was observed that the small intestine segments from the Treitz ligament to the cecum were surrounded by a thick, fibrous peritoneum in the patient who was operated due to intestinal obstruction. The peritoneal sac was excised. Adhesions between the small intestine loops were separated and the small intestines were completely freed. The patient, whose complaints regressed in the postoperative period, was discharged on the seventh day. AC is a rare disease that causes intestinal obstruction. Preoperative diagnosis is based on a high index of suspicion along with imaging modalities because of clinically nonspecific findings.

Keywords: Abdominal cocoon, intestinal obstruction, diagnosis, treatment.

Introduction

Encapsulating peritoneal sclerosis (EPS) is a chronic inflammatory disease in which the small intestine is surrounded by a dense, fibro-collagenous membrane. This disease is divided into primary (idiopathic) and secondary EPS, caused by an underlying etiological factor.¹ Abdominal cocoon (AC), also known as idiopathic EPS and first described by Foo et al. in 1978, in which the small intestine is surrounded by a fibrous membrane. The disease has also been called idiopathic sclerosing peritonitis, primary sclerosing peritonitis, and sclerosing encapsulating peritonitis.² Clinical findings vary according to the severity of the disease, its duration, and the immunological status of the person.¹ There are usually signs and symptoms of acute or subacute intestinal obstruction. The etiology and pathogenesis are not exactly known. Preoperative diagnosis is difficult due to the lack of specific findings and is usually made by laparotomy and biopsy.^{2,3}

In this article, the preoperative findings of a case with AC operated for intestinal obstruction, are presented and discussed in the light of literature.

Case Report

A 56-year-old male patient was admitted to the hospital with complaints of abdominal pain, nausea and vomiting. On physical examination, bowel sounds were normoactive, there was some abdominal tenderness but no defense or rebound was detected. He had no history of previous abdominal surgery or trauma. The patient did not have any comorbidity and did not use any medication. Standing, direct abdominal X-ray was evaluated as normal. On laboratory tests, leukocyte values were within normal limits. Abdominal ultrasonography showed free fluid between the intestinal loops in the lower abdominal quadrant. The patient's oral intake was discontinued. Intravenous (IV) fluid therapy was started and a nasogastric tube was inserted. After the partial regression of the patient's complaints, it was decided to continue medical treatment. In contrast enhanced (IV and oral) abdominal computed tomography (CT), the stomach was distended with the contrast agent, and the third and fourth parts of the duodenum were dilated. Minimal prominence in the jejunal structures and also fluid densities adjacent to the jejunal loops were observed. After the patient's complaints



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regressed, oral intake was started and the patient, who tolerated resumed oral intake, was discharged.

The patient presented with complaints of abdominal pain, nausea and vomiting one month later and was re-evaluated. Oral intake of the patient, whose leukocyte, urea and creatinine levels were found to be high, was discontinued. A nasogastric tube was inserted, IV fluid therapy and antibiotic therapy were started. Multiple air-fluid levels were observed at the small intestine level on the standing direct abdominal X-ray. Contrast-enhanced abdominal CT showed increased wall thickness at the stomach pyloric level and lesser curvature, and dilatation in the first, second and third parts of the duodenum. In the fourth part of the duodenum the aorto-mesenteric calibration was decreased, jejunal loops showed a tortuous course in the left upper quadrant of the abdomen, there was increased wall thickness in the jejunum and encapsulated fluid accumulation were observed (Figure 1). The patient underwent endoscopy under sedation anesthesia, which revealed that the gastric mucosa was edematous and the second and third segments of the duodenum were dilated. The jejunum was reached, but the lumen could not be opened in the proximal jejunum (Figure 2). Since the findings of intestinal obstruction did not regress, the decision was taken to proceed to surgery. The abdomen was entered with a midline incision. In the exploration, it was observed that a dense membrane surrounded the small intestines from the Treitz ligament to the cecum and the small intestines were intertwined, resembling an accordion (Figure 3, 4). Membranes were separated by blunt and sharp dissection and all small intestines were freed. The histopathology result reported

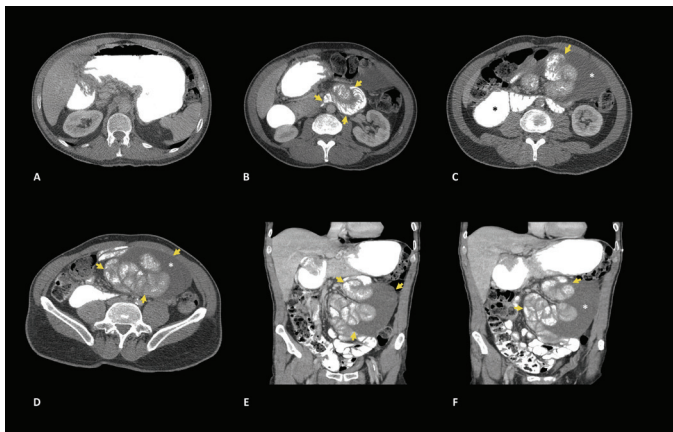


Figure 1. Axial (A-D) and coronal plane (E, F) contrast-enhanced abdominal computed tomography examination. Dilatation in the second part of the duodenum (black asterisk, A-C), decrease in the calibration of the fourth part of the duodenum at the aortomesenteric level (C), starting from the duodenojejunal junction, encapsulated with the jejunal loops (yellow arrows) in accordion form and surrounded by the fluid density (white asterisk). Slight thickening of the jejunal loops and wall (B-F)

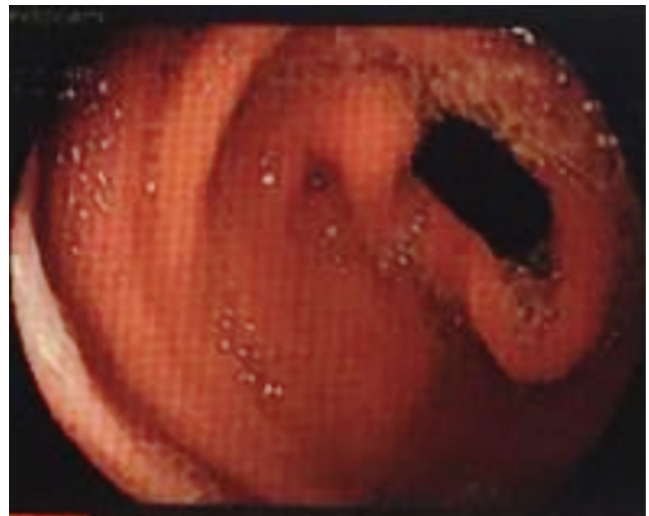


Figure 2. Jejunal segment stenosis was seen on endoscopy



Figure 3. From the ligament of Treitz, there are thick dense membranes that completely surround the jejunal loops (arrow) and calcifications on the serosal surfaces (duodeno-jejunal junction: asterisk)

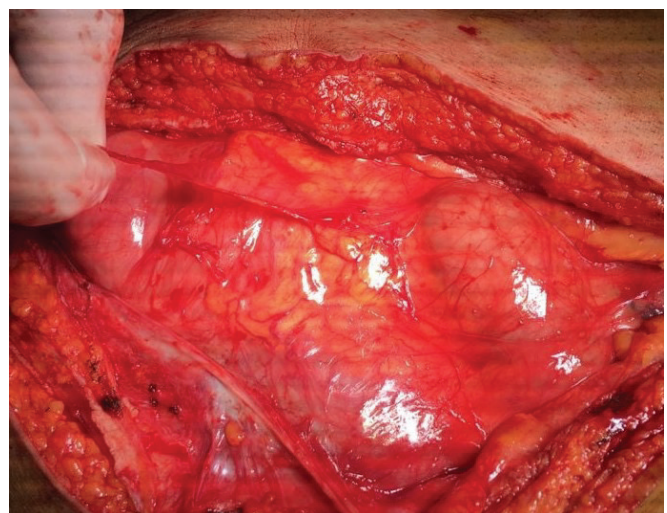


Figure 4. The dense membrane surrounding the small intestine is seen

tissue compatible with calcified, fibro-hyalinized cyst wall and capsule wall surrounding the small intestine.

The patient's complaints regressed in the postoperative follow-up, oral intake was started on the fifth postoperative day and the patient was discharged without any problem. At the time of writing the patient is in the sixth postoperative month and has no complaint. The patient was informed and informed consent was obtained.

Discussion

The etiology of AC is not known exactly. Although AC has classically been described in young adolescent women in tropical regions, it has also been reported in adult cases in temperate regions.⁴ EPS is divided into primary and secondary according to its etiology and pathological features of the fibrocollagen membrane. Primary EPS, the idiopathic form, is known as AC, and although there is no underlying cause in this form, cytokines and fibroblasts have an effect on the formation of peritoneal fibrosis and neoangiogenesis. In secondary EPS, local or systemic factors initiate inflammatory processes in the peritoneum. Secondary EPS usually develops into chronic, asymptomatic peritonitis, such as endometriosis, retrograde menstruation, peritoneal dialysis, abdominal tuberculosis, abdominal trauma, and/or liver cirrhosis.^{1,3,5}

Although there are clinical findings, such as recurrent abdominal pain, nausea, vomiting, loss of appetite, weight loss, malnutrition, acute or chronic intestinal obstruction attacks, and painless abdominal mass, some cases may be asymptomatic.⁴ In the presented case, there were complaints of recurrent abdominal pain, nausea and vomiting for about a month.

On diagnostic imaging, air-fluid levels and enlarged bowel loops are seen on direct abdominal X-ray. Ultrasonography may show a three-layered bowel wall, dilated and adherent bowel loops to the posterior abdominal wall, loculated stomach acid and fibrous adhesions, and intertwined accordion-shaped bowel segments. However, CT may be more useful in early diagnosis and planning of optimal treatment. On CT, small bowel loops surrounded by thick membrane can be seen in the midline of the abdomen, helping to confirm a more precise diagnosis.⁴ Findings such as peritoneal calcifications, peritoneal thickening, peritoneal clarification, loculated fluid collection, conglomerated bowel loops with fluid in the peritoneal cavity, small bowel loops surrounded by a thin membrane, thickening of the intestinal wall, and calcifications on the serosal surface of the intestine may be seen. It also provides information about complications and other causes of intestinal obstruction. In our case, diffuse air-fluid levels at the level of the small

intestine were observed in the abdomen on direct X-ray. On contrast-enhanced CT, conglomerated small intestine segments in the left upper quadrant of the abdomen were detected.^{4,6}

Preoperative diagnosis is challenging because the clinical findings are non-specific for diagnosis and thus a high index of clinical suspicion is required. Exclusion of other etiological factors that may cause obstruction in non-strangulated intestinal obstruction cases combined with clinical suspicion are important in preoperative diagnosis. Preoperative diagnosis helps for planning optimal treatment.⁴ However, although all imaging methods, such as ultrasound or CT are used, the diagnosis is made incidentally during laparotomy in most cases.

Although it is often argued that surgical treatment is necessary, the treatment management of EPS is still a matter of debate. According to the literature, in cases with minimal symptoms, oral intake, resting the bowels, nasogastric decompression and parenteral nutrition can be done.⁷ Many treatment options are recommended, such as surgical subtotal excision of the membrane, enterolysis, intubation of the small intestine, and medical treatment after exploratory laparotomy in cases with a high risk of bowel resection and perforation.⁴ In order to avoid mortality and morbidity, such as postoperative anastomotic leakage and short bowel syndrome, it has been advised to free the intestines by separating the membranes with as little resection as possible in cases with normal intestinal viability.^{4,8-10} In the current case, air-fluid levels were found in the small intestines on direct abdominal X-ray. No obvious pathology was detected in the abdominal ultrasound. In the CT of the abdomen with IV-oral contrast, dilated segments of the duodenum were detected and a tortuous appearance was evident in jejunal segments. Loculated fluid density was observed adjacent to the jejunal segment. The patient was taken to operation because his complaints did not regress despite medical treatment. During exploration, AC was considered and the small intestines were freed. No resection procedure was applied to the small intestines.

Conclusion

EPS is one of the rare causes of intestinal obstruction. Preoperative diagnosis is challenging due to the nonspecific clinical findings. Preoperative diagnosis is possible with the combination of high clinical suspicion and imaging findings. Based on the patient's history, peritoneal dialysis and peritoneal tuberculosis should be considered among the secondary causes. Cases without findings suggesting strangulation can be followed up with medical treatment. However, in cases where no diagnosis can be made or surgery is chosen based on clinical findings, we think

that enterolysis and membrane excision is sufficient while avoiding resection as far as is possible and unless absolutely necessary.

Ethics

Informed Consent: The patient was informed and informed consent was obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: T.S., K.Ç., N.T.K., Concept: K.Ç., T.S., S.Ö.G., Design: K.Ç., T.S., H.S., S.Ö.G., Data Collection or Processing: K.Ç., N.T.K., H.S., T.S., Analysis or Interpretation: K.Ç., H.S., N.T.K., Literature Search: K.Ç., N.T.K., S.Ö.G., Writing: K.Ç., T.S., N.T.K.

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