

Idiopathic Sclerosing Encapsulating Peritonitis: A rare abdomen cocoon presentation

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Abstract

Background: Abdominal cocoon is characterized by small bowel encapsulation and is a rare cause of intestinal obstruction.

Case report: We describe a young man who had recurrent subacute intestinal obstruction. On elective exploration of abdomen, the entire small bowel was found to be encapsulated inside a white dense fibrous sac. The peritoneal sac was excised, followed by lysis of the interloop adhesions. The outcome was satisfactory.

Conclusions: The clinical diagnosis of abdominal cocoon requires a high index of suspicion because of the rarity, nonspecific clinical picture and often noncontributory imaging findings. Surgical treatment consists of peritoneal sac excision and adhesiolysis.

Keywords: Abdominal cocoon, intestinal obstruction.

Introduction

Small-bowel obstruction (SBO) is a common surgical problem. The commonest cause is adhesion which is seen in 60% of patients with SBO. Intestinal stricture, hernia, and neoplasm are other common causes. Unusual causes are encountered in only 6% of patients. Abdominal cocoon (AC) or sclerosing encapsulating peritonitis is one such unusual cause of SBO.¹ Although a preoperative diagnosis requires a high index of clinical suspicion.

Case report

A 38-year-old white man presented with recurrent colicky abdominal pain, nausea, vomiting, and abdominal distension. He reported having had three such episodes over the previous 6-7 months which required hospitalization and resolved with conservative treatment. He had neither prior abdominal surgery nor had history of exposure to tuberculosis. There was central abdominal distension with a tender, lump palpable in the central abdomen intermittently with colicky pain. Plain upright abdominal X-ray showed presence of multiple air fluid levels with no free gas under the dome of the diaphragm. Ultra sonogram (USG) abdomen showed hypo-peristaltic, thickened small gut loops with intergut fluid. Barium meal follow through study showed spikling of small bowel loops along mesenteric border with delayed transit of contrast (Fig 1).



Fig 1. Barium contrast film showing small gut in centre

Contrast-enhanced computed tomogram (CECT) of the abdomen revealed multiple dilated gut loops with intergut fluid. Ascitic fluid examination revealed 95% lymphocytes with mesothelial cells and few red blood cells and neutrophils. Ascitic fluid proteins were 2.4 gms/dl and sugar was 47 mg/dl. Ascitic fluid deaminase was 20.70 U/L (normal < 30 U/L). Tubercular gold test was negative. Erythrocyte sedimentation rate (ESR) was 50 mm in one hour. Total leukocyte count was 6500/mm³ with 66% neutrophils and 30% lymphocytes. Decision for elective laparotomy was taken on the basis of clinical and radiological findings. On laparotomy, a thick white membrane was found encasing whole of the small gut with dense intergut adhesions (Fig 2). Approximately 150 ml of straw colour ascitic fluid was present. Excision of the cocoon membrane and lysis of intergut adhesion was undertaken with care to avoid injury to intestine. Histopathology of the cocoon wall revealed non granulomatous inflammation. Patient did well in postoperative period and discharged on eighth postoperative day. The patient was symptom-free on follow-up over a period of 3 months.

Discussion

Sclerosing encapsulating peritonitis is an unusual cause of SBO. It was first described by Owtschinnikow in 1907 as “peritonitis chronica fibrosa incapsulata”³. SEP can be classified as idiopathic or secondary. The idiopathic form is also known as abdominal cocoon, was first described by Foo et al in 1978.^{3,4} The primary or the idiopathic form is seen characteristically in young adolescent girls of 4–18 year age group from the tropical or subtropical countries.⁵ The more common secondary form is associated with prolonged beta blocker therapy, local irritation of the peritoneum by trauma, peritoneal dialysis, peritoneal–venous shunting, and ventriculoperitoneal shunt.⁶ The plausible hypothesis for pathogenesis of AC is recurrent low grade or subclinical peritonitis, during which the patients had no significant abdominal signs, leading to sclerosis and membrane formation with subsequent development of a cocoon.⁷

AC is characterized by partial or total encasement of the small gut by a fibrocollagenous sac that looks like a cocoon.⁵ Histologically, the peritoneum shows a proliferation of fibro-connective tissue, inflammatory infiltrates, and dilated lymphatics, with no evidence of foreign body granulomas, giant cells, or birefringent material.⁸⁻¹⁰ “Sclerosing” refers to the progressive formation of sheets of dense collagenous tissue; “encapsulating” describes the sheath of new fibrous tissue that covers and constricts the small bowel and restricts its motility; and “peritonitis” implies an ongoing inflammatory process and the presence of a mononuclear inflammatory infiltrate within the new fibrosing tissue.¹⁰

The clinical presentation of abdominal cocoon includes acute, subacute, or chronic intestinal obstruction, abdominal distension, nausea, and vomiting.³ A high index of suspicion is required to make preoperative diagnosis based on clinical presentation combined with relevant imaging findings and lack of other etiologies. Most cases are diagnosed incidentally at laparotomy. Imaging finding includes a combination of barium follow-through (concertina pattern or cauliflower sign and delayed transit of contrast medium), ultrasound (trilaminar appearance of bowel, tethering of bowel to posterior abdominal wall, ascites and dilatation and fixation of bowel loops), and computed tomography of the abdomen (small bowel loops congregated to the center of the abdomen encased by a soft-tissue density mantle).¹¹⁻¹⁴

Surgery (membrane dissection and extensive adhesiolysis) is the treatment of choice, and there is usually no need for bowel loop resection, especially when a preoperative diagnosis is feasible. Resection of the bowel is unnecessary and it increases morbidity and mortality. Resection is indicated only if the bowel is non-viable. An excellent long-term postoperative prognosis is most of the times guaranteed.¹⁵

Conclusion

Idiopathic sclerosing encapsulating peritonitis, although rare, may be the cause of a small bowel obstruction. A high index of clinical suspicion may be generated by the recurrent character of small bowel ileus combined with relevant imaging findings and lack of other plausible etiologies. Clinicians must rigorously pursue a preoperative diagnosis, as it may prevent a "surprise" upon laparotomy and result in proper management. CECT is mostly useful in clinching the diagnosis and planning elective surgery in experienced hands. Meticulous dissection of the cocoon membrane from the gut to release the entrapped intestine and separation of the inter loop adhesions is the treatment of choice. Iatrogenic injury to bowel and consequent bowel resections are associated with high morbidity and are to be avoided.

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