

IDIOPATHIC SCLEROSING ENCAPSULATING PERITONITIS CAUSING ACUTE INTESTINAL OBSTRUCTION AND GANGRENE: A CASE REPORT

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ABSTRACT

INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a relatively rare cause of intestinal obstruction resulting from encasement of variable lengths of bowel by dense fibro-collagenous membrane. It is more common in young females, and shows tropical and sub-tropical distribution. The idiopathic cases of SEP, which lack any identifiable cause from clinical, radiological and histopathological findings, are also reported under the descriptive term "abdominal cocoon syndrome". SEP presents with acute or sub-acute intestinal obstruction with or without a mass. In the era of laparoscopic surgery, inadvertent damage to the small bowel at insertion of the trocar and cannula can occur by being unaware of this condition resulting in unnecessary bowel resection. Persistent untreated SEP may advance to bowel gangrene or intestinal perforation, representing life threatening conditions.

We report the clinical presentation of a 75-year-old female presenting with signs of intestinal obstruction whose imaging findings revealed abdominal cocoon with bowel gangrene leading to perforation and the same confirmed at surgery. Surgical excision of the fibrotic sac encasing the bowel, resection of gangrenous bowel segment and end ileostomy was performed. Histopathology of the excised membrane confirmed sclerosing encapsulating peritonitis.

To our knowledge, only a few cases of abdominal cocoon with perforation have been reported in literature so far. Radiologists should be aware of this relatively rare cause of intestinal obstruction, its imaging findings and complications, as preoperative diagnosis will prevent delay and aid in treatment planning to the surgeon. Identification of soft tissue density membrane encasing congregated small bowel loops into a single area on computed-tomography gives diagnostic clue. Surgical excision of sac, release of bowel loops and adhesions with partial intestinal resection when necessary is the treatment.

KEYWORDS

Sclerosing Encapsulating Peritonitis, Intestinal Obstruction, Abdominal Cocoon, Peritonitis, End Ileostomy.

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INTRODUCTION: SEP is a rare disease entity characterised by total or partial encasement of small bowel by a thick fibrous sac, resulting in intestinal obstruction. This condition was first described in 1907 as "Peritonitis chronic fibrosa incapsulata" by Owtschinnikow.¹

SEP is classified as primary (idiopathic) or secondary based on etiological factors. Idiopathic cases which lack any identifiable aetiology were reported under the alternative descriptive name of 'abdominal cocoon syndrome', first described by Foo et al in 1978.² It has been speculated that infections of the fallopian tubes or retrograde menstruation with subclinical peritoneal infection may be related to the disease onset and progression. The idiopathic form primarily affects young women from tropical countries.³ Presentation in an elderly aged woman, makes our case unique. The secondary form has been associated with long term peritoneal dialysis, β blocker therapy, abdominal

tuberculosis, previous abdominal surgery, peritoneal shunts and gastrointestinal malignancies.⁴

CASE REPORT: A 75-year-old woman presented to casualty with colicky abdominal pain, non-bilious vomiting, abdominal distension and not passing stools or flatus for 3 days. She had no past history of abdominal surgery, tuberculosis, peritoneal dialysis or hepatic disease. Clinical examination revealed diffuse tenderness over abdomen, distension and decreased bowel sounds. Per rectal examination was unremarkable. Vital signs were consistent with hypovolemia and sepsis. (blood pressure 90/60, pulse 130 beats per minute, body temperate 101 degrees F) Laboratory investigations showed elevated total count of 28,800 cells/mm³ (neutrophils-92%). Plain abdominal x-ray showed few to multiple air-fluid levels centrally located, without free intraperitoneal gas. Contrast-enhanced abdomen computed tomography (CT) showed characteristic findings of small bowel loops congregated to the center of the abdomen encased by a soft-tissue density mantle. Erect radiograph of abdomen (fig. 1) revealed dilated small bowel loops and multiple air-fluid levels. Abdominal sonography showed clustering of dilated fluid-filled small bowel loops

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with intervening fluid between them in mid abdomen. Axial and coronal images of contrast enhanced computed tomography (CECT) (fig. 2) showed thin soft tissue density membrane encasing small bowel loops in midline giving the appearance of cocoon, few dilated small bowel loops within, loculated fluid surrounding the loops within the cocoon.

Emergency exploratory laparotomy was performed which showed a thick shiny whitish membrane encapsulating the small bowel loops (fig. 3A-D). On incising the membrane, gangrenous small bowel loops and 700 mL of serosanguinous free fluid were noted within the cocoon. Twenty cm of terminal ileum up to five cm proximal to ileocecal valve was found gangrenous. Perforation was also noted in the ileum proximal to the gangrenous segment. Resection of gangrenous bowel with end ileostomy was performed. Post-operative recovery was uneventful.

Histopathological examination of the excised peritoneal capsule showed proliferation of the fibroconnective tissue with signs of nonspecific inflammatory reaction and no signs of tuberculosis/malignancy (fig. 4), suggestive of sclerosing encapsulating peritonitis. Histopathology of excised gangrenous bowel suggested acute ischaemic pathology. The serosanguinous fluid sent for culture and sensitivity revealed E. coli and Klebsiella growth.



Fig. 1: X-ray Abdomen (erect)-Shows Dilatation of Small Bowel Loops with Multiple Air-Fluid Levels



Fig. 2A

Fig. 2B

Fig. 2A & B: Axial CECT images of abdomen shows clustered and dilated small bowel loops with diffuse wall thickening and centralisation surrounded by fibrous capsule. Marked ascites is also noted.

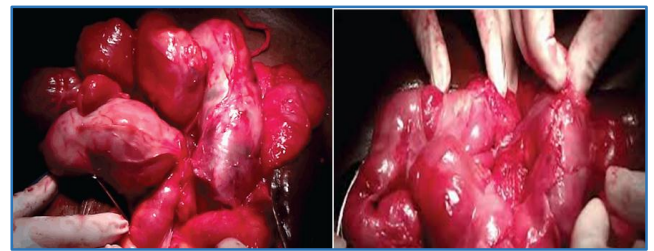


Fig. 3A

Fig. 3B

Fig. 3A & B: Encapsulated part of the small intestine. Loosening of encapsulated segments (3A, 3B)



Fig. 3C: On Excising the Fibrotic Membrane, Gangrenous Bowel is Seen (3C)



Fig. 3D: On Total Excision of the Membrane and after Freeing the Bowels, 100% Oxygen was Given. Perforation was Noted in Ileum Proximal to the Gangrenous Segment (3D)

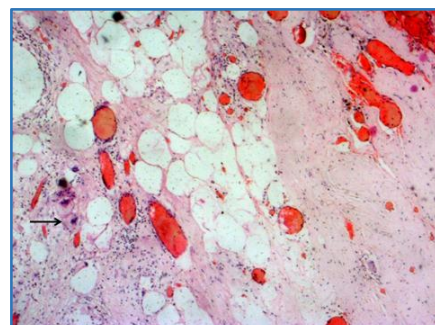


Fig. 4: Histopathology Shows Areas of Sclerosis with Foreign Body Giant Cells (Haematoxylin and Eosin x4)

DISCUSSION: Abdominal cocoon syndrome, also known as idiopathic sclerosing encapsulating peritonitis, is a rare cause of acute intestinal obstruction in which the small bowel becomes encased (or cocooned) by a dense fibrocollagenous membrane.

While some patients with SEP remain asymptomatic, majority present with recurrent attacks of acute, subacute or chronic intestinal obstruction, weight loss, loss of appetite, palpable abdominal mass or ascites. Gastrointestinal perforation is a relatively rare complication of SEP.⁵ The current case had perforation at proximal ileum with gangrene of small bowel, in a case of primary SEP, making this case unique in the literature. To our knowledge, only two cases of SEP related perforation has been reported. One of them had high jejunal perforation in a case of SEP secondary to tuberculosis⁶ and the other had ileal perforation in a case of primary SEP with coexisting incarcerated Meckel's diverticulum in inguinal hernia.⁵

Imaging plays an important role in preoperative diagnosis and further disease management. Erect abdominal radiographs may show evidence of small bowel obstruction, with dilated bowel loops and air-fluid levels. The classic finding on a barium meal follow through study as described by Sieck et al,⁷ is a serpentine or concertina-like configuration of dilated small bowel loops in a fixed U shaped cluster giving a cauliflower appearance. Maguire et al,⁸ suggested that delayed transit time is more diagnostic. Characteristic sonographic findings include altered peristalsis, adherence of the bowel to the anterior abdominal wall, intraperitoneal echogenic strands and membrane formation during the late stages of the disease. Classic computed tomography (CT) findings include small bowel loops congregated in a single area or in the midline encased by a soft tissue density envelope. Ancillary CT findings include ascites, loculated fluid collection thickening, enhancement or calcifications of peritoneum, peritoneal deposits, thickening of omentum bowel wall, tethering or fixation of bowel loops and abdominal lymphadenopathy. Presence of the additional CT findings may suggest secondary form of SEP due to causes like tuberculosis, malignancy. Prior to the era of CT imaging, definitive diagnosis was usually made during surgery. Currently CECT is the investigation of choice as it gives more accurate information on the degree of obstruction, types of bowel loops involved and associated complications.⁹

Surgical intervention is the treatment of modality. It includes freeing of adhesions, total excision of the membrane, or partial intestinal resection when necessary¹⁰ as in our case.

In our case, clinical, laboratory and radiological findings were concordant with abdominal cocoon. Because the additional clinical and radiological findings of tuberculosis peritonitis, peritoneal carcinomatosis, previous abdominal surgery, dialysis, long term use of praxolol, chronic disease history were not present, and she had normal test results from biochemical (erythrocyte sedimentation rate) and microbiological (blood and peritoneal fluid culture) assays, idiopathic abdominal cocoon was presumed. Preoperative

diagnosis was made on CECT, as it showed classic findings and complications of gangrene and perforation were suspected resulting in early emergency surgical intervention of the patient.

Acute intestinal obstruction secondary to abdominal cocoon with bowel gangrene and perforation presenting in an elderly aged woman makes our case unique and rare in literature. Radiologists should be aware of this relatively rare cause of intestinal obstruction, its imaging findings and complications, as preoperative diagnosis will prevent delay and aid in treatment planning to the surgeon. Identification of soft tissue density membrane encasing congregated small bowel loops into a single area on CECT gives diagnostic clue. Surgical excision of sac, release of bowel loops and adhesions with partial intestinal resection when necessary is the treatment.

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