

Idiopathic sclerosing encapsulating peritonitis (abdominal cocoon) presenting with abdominal pain and palpable mass: a case report

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Introduction: Sclerosing encapsulating peritonitis (SEP), also known as abdominal cocoon, is a rare condition characterized by a fibrocollagenous membrane encasing the bowel, leading to symptoms ranging from vague abdominal discomfort to intestinal obstruction. It may be primary (idiopathic) or secondary, most commonly associated with prior peritoneal dialysis.

Case presentation: A 45-year-old Ethiopian man presented with acute periumbilical abdominal pain, nausea, and a palpable abdominal mass, with a background of chronic intermittent abdominal symptoms for 5 years. Exploratory laparotomy revealed small bowel loops encased in a thick fibrocollagenous membrane consistent with abdominal cocoon. Complete membrane excision was performed, with preservation of bowel viability. Histopathology confirmed fibrocollagenous tissue with sclerosis and inflammatory infiltrates. The postoperative course was uneventful.

Discussion: The pathogenesis of primary SEP remains unclear but is thought to involve chronic peritoneal inflammation. Due to its nonspecific presentation, diagnosis is often made intraoperatively, particularly in resource-limited settings. Surgical excision of the encapsulating membrane remains the mainstay of treatment.

Conclusion: This case highlights the diagnostic challenges of abdominal cocoon and emphasizes the importance of maintaining a high index of suspicion, especially in patients from resource-limited settings presenting with unexplained intestinal obstruction.

Graphical abstract available at: <https://links.lww.com/IJSCR/A143>

Keywords: abdominal cocoon, Ethiopia, rare cause of intestinal obstruction, sclerosing encapsulating peritonitis, SEP

Introduction

Abdominal cocoon was first described in 1907 by Owtschinnikow and was termed *peritonitis chronica fibrosa*^[1,2]. It was later characterized in a series of 10 young women who developed intestinal obstruction due to a fibrocollagenous membrane encasing the bowel loops, resembling a cocoon, from which the term *abdominal cocoon* was derived^[3]. This condition represents one of the rarest causes of intestinal obstruction and therefore poses a significant diagnostic challenge, as a high index of suspicion is required to establish a preoperative diagnosis^[4–6]. The condition is also referred to as primary or idiopathic peritoneal encapsulating sclerosis (PES) in several studies and is distinguished from secondary PES

HIGHLIGHTS

- Abdominal cocoon is a rare cause of bowel obstruction that is often diagnosed intraoperatively.
- Idiopathic sclerosing encapsulating peritonitis can occur without identifiable risk factors.
- The presentation may vary from recurrent, nonspecific abdominal symptoms to acute intestinal obstruction.
- Surgical excision of the fibrocollagenous membrane is the definitive treatment.
- Awareness in low- and middle-income settings may reduce diagnostic delay.

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by the absence of identifiable predisposing factors in the former^[5]. The scarcity of reported cases from low- and middle-income countries settings, along with the unique diagnostic challenges encountered in these environments, prompted us to present this case with the aim of increasing clinical awareness and preventing misdiagnosis, delayed diagnosis, and related complications. This report is written according to the SCARE criteria^[7].

Case presentation

The patient is a 45-year-old male of Ethiopian descent who presented with a 10-hour history of crampy, non-radiating periumbilical abdominal pain associated with nausea and one episode of vomiting of recently ingested material. He also noticed a painless abdominal mass to the right of the umbilicus. The

symptoms had been recurring intermittently for the past 5 years, alternating with episodes of epigastric pain, for which he had been treated multiple times for dyspepsia. He denied any change in bowel habits, abdominal distension, or fever.

The patient had no history of abdominal surgery, peritonitis, tuberculosis, peritoneal dialysis, or abdominal trauma. On physical examination, he appeared acutely ill and in pain, but his vital signs were within normal limits. Abdominal examination revealed a firm, painless 5 × 4 cm swelling in the left lower quadrant, just lateral to the midline.

Laboratory investigations revealed leukocytosis of $13 \times 10^3/\mu\text{L}$ with neutrophil predominance (88.5%), suggesting an inflammatory process. Hemoglobin and platelet counts were within normal limits at 13.7 g/dL and $246 \times 10^3/\mu\text{L}$, respectively. Abdominal ultrasonography demonstrated dilated, fluid-filled bowel loops in the left lower quadrant, with a gaseous abdomen and a trace of reactive free intraperitoneal fluid, raising concern for bowel obstruction. A plain abdominal radiograph was equivocal (Fig. 1). A computed tomography (CT) scan was not available at our institution at the time of presentation.

A ventral hernia, most likely a Spigelian hernia—was considered the initial diagnosis based on the patient's history of intermittent, crampy abdominal pain and the presence of a left lower-quadrant paraumbilical mass measuring approximately 5 × 4 cm on physical examination. Although ultrasonography is generally considered adequate for detecting Spigelian hernias, the examination did not demonstrate a fascial defect or other findings suggestive of a hernia. However, given the operator-dependent nature of ultrasonography, the absence of CT imaging to establish a definitive diagnosis, and the strong clinical suspicion based on physical examination findings, a Spigelian hernia remained an important differential diagnosis. The patient was counseled regarding management options. He consented to surgical exploration.

The patient was then taken to the main operating room and placed in the supine position under general anesthesia, and the abdomen was initially accessed through a lower midline incision. Because of inadequate exposure, the incision was extended superiorly to a full midline laparotomy.



Figure 1. Plain abdominal radiograph showing a few dilated bowel loops without significant air–fluid levels.

Intraoperatively, after fascial dissection, multiple abdominal wall swellings were noted, measuring approximately 3 × 3 cm and 4 × 4 cm in the right lower quadrant, and 5 × 4 cm in the left lower quadrant, just lateral to the midline. Exploration revealed multiple small bowel loops clumped together and encased within a dense, whitish, fibrinous membrane, approximately 4 mm thick (Fig. 2A,B). An intraoperative diagnosis of abdominal cocoon was made.

Careful excision of the encapsulating membrane was performed, revealing viable small bowel loops. During dissection of the encapsulating membrane, a dense interloop adhesive band was identified approximately 100 cm distal to the ligament of Treitz. The band tightly compressed a segment of small bowel, resulting in dusky discoloration suggestive of compromised perfusion. Adhesiolysis was performed, and division of the band resulted in restoration of normal bowel color and viability. Small residual portions that were densely adherent to the bowel serosa were intentionally left in situ to avoid iatrogenic bowel injury. The operative time was approximately 2 hours, and the estimated blood loss was less than 200 mL.

The excised membrane was sent for histopathological examination, which revealed fibrocollagenous membranous tissue consistent with SEP, showing sclerosis, thickening, and inflammatory cell infiltration predominantly composed of lymphocytes. No granulomas or malignant cells were identified, providing no histopathological evidence of granulomatous inflammation (including abdominal tuberculosis) or malignancy as an underlying secondary cause.

Postoperatively, the patient received adequate analgesia and was started on oral sips 12 hours after surgery. He showed marked clinical improvement and was discharged on postoperative day 5. Follow-up visits at 1 week, 2 weeks, and at 1 month and 6 months confirmed an uneventful postoperative recovery. The timeline for major clinical events is summarized in the graphical abstract.

Discussion

Sclerosing encapsulating peritonitis (SEP) can be classified as idiopathic (primary) or secondary. Secondary SEP occurs in patients with identifiable predisposing factors, the most common being peritoneal dialysis^[1,5]. Other, less frequent associations include viral peritonitis^[8], use of beta-blockers, cirrhosis, familial Mediterranean fever^[9], abdominal tuberculosis^[10], prior abdominal surgery, ventriculoperitoneal shunt, sarcoidosis, intraperitoneal chemotherapy, liver transplantation, dermoid cyst rupture, gastrointestinal malignancy, luteinized ovarian thecomas, endometriosis, and protein S deficiency^[1].

The pathophysiology of SEP, particularly the idiopathic type, remains poorly understood. Earlier hypotheses suggested a role for retrograde menstruation^[3], but recent studies indicate a male predominance^[1,5,6,11]. Genetic factors have also been proposed, as evidenced by the coexistent absence of the greater omentum in a two-patient case series^[12]. Idiopathic SEP is thought to result from chronic, low-grade inflammation of uncertain etiology, potentially due to subclinical, episodic peritonitis, which eventually leads to peritoneal sclerosis and fibro-collagenous membrane formation. Cytokines and fibroblasts are also believed to influence peritoneal fibrosis and angiogenesis^[5,13].

SEP is classified into three types based on the extent of intestinal involvement: **type I**, affecting only a portion of the small

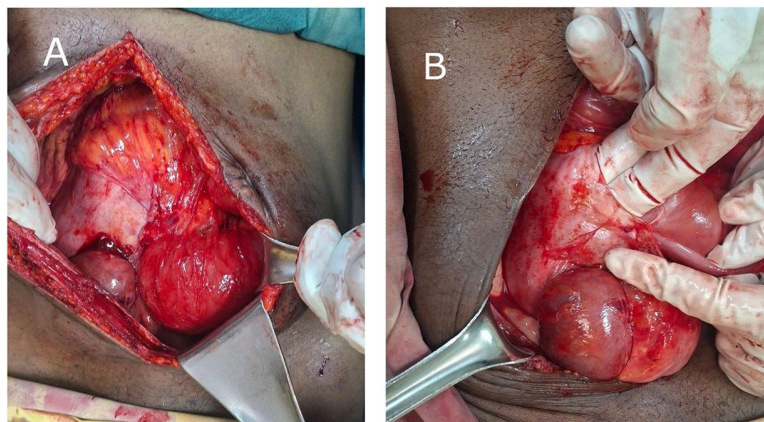


Figure 2. Intraoperative findings of sclerosing encapsulating peritonitis. (A and B) Multiple small-bowel loops clumped together and encased within a dense, whitish, fibrinous membrane approximately 4 mm thick, forming a characteristic encapsulating sac consistent with an abdominal cocoon.

intestine; **type II**, involving the entire small intestine; and **type III**, in which the entire small intestine and additional organs such as the stomach, liver, appendix, cecum, ascending colon, and others are encapsulated^[4,14]. The exact incidence of idiopathic SEP is unknown, but it predominantly affects populations in tropical and subtropical regions, with lower prevalence in temperate areas^[5].

Clinically, SEP may present with acute, subacute, or chronic symptoms of intestinal obstruction, including abdominal pain, nausea, vomiting, obstipation, and occasionally a painless abdominal mass^[1,14,15]. Imaging modalities for SEP may include plain abdominal radiographs, barium studies, and abdominal CT scans. Plain radiographs may demonstrate nonspecific signs of bowel obstruction, whereas barium studies may show a conglomeration of multiple cauliflower-like bowel loops adherent to one another and surrounded by a sac-like structure. Abdominal CT is a more useful diagnostic tool and may demonstrate conglomerated small bowel loops located in the mid-abdomen, surrounded by a thick capsule with a contrast-free peripheral rim^[11,14].

Most cases of idiopathic SEP, however, are definitively diagnosed intraoperatively upon identification of the characteristic encapsulating membrane^[4,6,11]. Nonoperative management has been described for patients without obstruction^[6], but because the abdominal cocoon frequently causes an acute abdomen, surgical intervention with careful excision of the encapsulating membrane is usually required to relieve obstruction^[16]. Bowel resection may be necessary in cases with gangrenous segments^[17].

In our case, the patient was a middle-aged male, consistent with the reported male predominance in idiopathic SEP^[1,5,6]. He had no secondary predisposing factors, and his symptoms had recurred over 5 years, close to the mean duration of 3.9 years reported in one study on idiopathic SEP^[4]. CT imaging was not available at our institution. Referral to another facility for CT scanning was considered; however, this was deferred because the patient was unable to afford the out-of-pocket cost. SEP was not suspected preoperatively, and the working diagnosis remained a Spigelian hernia based on the clinical presentation. Given the concern for an underlying mechanical obstruction, surgical exploration was undertaken, during which the unexpected

diagnosis of SEP was established by identifying the typical fibro-collagenous membrane encasing the bowel. The patient had **type III** SEP, with the entire small intestine, descending colon, and sigmoid colon encased.

Conclusion

Abdominal cocoon is more frequently reported in tropical and subtropical regions; however, cases from Africa remain scarce, likely due to underreporting. Our patient presented with subtle, nonspecific symptoms but benefited from timely surgical management. There is a need for further studies to evaluate long-term outcomes in patients undergoing surgical excision, particularly regarding the incidence of adhesion-related complications following extensive membrane removal.

Ethical approval

Institutional Review Board approval was not necessary, as this study describes a single case report.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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None.

Author contributions

M.C.K.: Writing – original draft; Writing – review & editing; Conceptualization. R.T.K.: Writing – original draft; Writing – review & editing; Supervision; Data curation. N.A.B.: Validation; Writing – review & editing. K.A.D.: Data curation; Validation; Writing – review & editing. M.A.G.: Resources; Writing – review & editing. P.E.I.: Writing – original draft; Writing – review & editing.

Conflicts of interest disclosure

The authors declare that they have no known competing financial interests or personal relationships that could have influenced this work.

Research registration unique identification number (UIN)

This study did not require registration in a publicly accessible registry, as it is a single-patient case report and does not involve interventional research.

Guarantor

Dr. Robel Tibebe Kasaye.

Provenance and peer review

Not applicable.

Data availability statement

All data generated or analyzed during this study are included in this published article.

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