

## A Case of Intestinal Obstruction Caused by Abdominal Cocoon in an Adult Male: A Case Report

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### Abstract

Abdominal cocoon also known as “Encapsulating peritoneal sclerosis” (EPS) is a rare cause of intestinal obstruction presenting as a surgical emergency. It is described as thick, white, fibrous “cocoon-like” membrane, formed by chronic intra-abdominal fibro-inflammatory process that results in formation of fibrous tissue sheets, which formed a “second abdominal cavity” containing variable amounts of bowel segments that cover, fix and ultimately constrict the gut compromising its motility. We present a case of acute intestinal obstruction due to primary idiopathic Sclerosing Encapsulating Peritonitis (SEP). The diagnosis was established and early laparotomy was done with surgical release of the entrapped bowel and complete removal of the fibrotic membrane.

**Keywords:** Abdominal cocoon; Encapsulating peritoneal sclerosis; Sclerosing encapsulating peritonitis; Intestinal obstruction.

### Background

Abdominal cocoon also known as “Encapsulating Peritoneal Sclerosis” (EPS) is a rare cause of intestinal obstruction presenting as a surgical emergency. This entity was first described in 1907 by Owtschinnikow under the term “peritonitis

chronica fibrosa incapsulata”. A detailed description and first use of term “abdominal cocoon” was made in 1978 by Foo et al.<sup>1</sup>

It is a total or partial encapsulation of the small bowel by a fibro-collagenous membrane with local inflammatory infiltrate leading to acute or chronic bowel obstruction.

Here we report a case of 25 year young male who presented in our surgical emergency with intestinal obstruction caused by abdominal cocoon.

### Case Report

A 25 year young male presented with a chief complains of pain abdomen for last 3 days and not passing stool or flatus for last 2 days. The pain was acute in onset, colicky in nature, present diffusely in whole abdomen, associated with progressively increasing abdominal distention and multiple episodes of bilious projectile vomiting. The patient had not been through any previous abdominal surgery.

On clinical examination his temperature was normal, pulse rate 110/min, BP = 120/80 mm Hg, Respiratory rate = 25/min. On systemic examination he had abdominal distension with central abdominal tenderness with guarding and hyperperistaltic bowel sounds on auscultation and without any palpable lump. On DRE, the rectum was empty and collapsed, no ballooning, mucus on examining finger.

The patient was initially managed conservatively with IV fluids to correct dehydration and antibiotics. A nasogastric tube was inserted which had bilious

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output of 600 ml. Blood parameters were normal. The straight X-ray abdomen revealed multiple air fluid levels with dilated small bowel loops suggestive of small bowel obstruction (Fig. 1).



Fig. 1: Straight X-ray abdomen - AP view

A CECT of abdomen was obtained which showed “dilated small bowel loops with air fluid levels and collapsed terminal ileal loops. Ill defined kinking is seen in distal ileal loops. A well-defined suspected abdominal cocoon formation is seen with minimal ascites” (Fig. 2).

A provisional diagnosis of acute intestinal obstruction was made and an emergency exploratory laparotomy was conducted. Intra-operatively it was seen that the entire small bowel loops including IC junction and part of cecum and appendix was encapsulated in a transparent fibrous membrane forming “a cocoon” like structure having a sac with a constricting ring at its mouth. Adhesiolysis was done and the constriction was released along with excision of the membranous sac (Fig. 3).

His postoperative recovery was unremarkable and the patient was discharged on postoperative day 7 with return of normal bowel habits and in stable condition.

## Discussion

Abdominal cocoon or “Encapsulating Peritoneal Sclerosis” (EPS) was previously termed as Sclerosing Encapsulating Peritonitis (SEP) and is an



Fig. 2: CECT abdomen showing a well-defined collection of bowel loops “abdominal cocoon”.

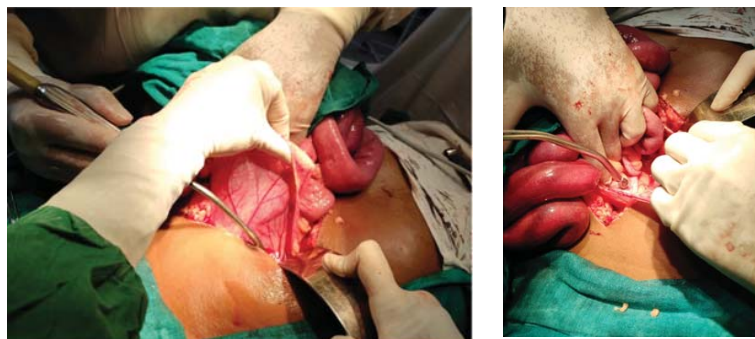


Fig. 3: The membranous sac like covering containing bowel loops (left). The mouth of the sac forming constricting band causing intestinal obstruction (right).

uncommon cause of intestinal obstruction.<sup>1</sup> It occurs due to chronic intra-abdominal fibro-inflammatory process that results in formation of fibrous tissue sheets that cover, fix and ultimately constrict the gut compromising its motility.<sup>2</sup> The entity was first described by Foo et al. in 1978 in adolescent girls from tropical and sub-tropical areas it was called primary or idiopathic EPS as the etiology was obscure. This was proposed to be retrograde peritonitis from spread of infection through fallopian tubes and retrograde menstruation with subsequent immunologic damage.<sup>3</sup> 75% of patients with primary SEP are men, premenstrual women or children and idiopathic SEP is twice as common in men than in women.<sup>4</sup> It is described as thick, white, fibrous "cocoon-like" membrane, which formed a "second abdominal cavity". The entity was categorized into 3 Types according to the extent of the encasing membrane: Type I-(1) The membrane encapsulated partial intestine; (2) Type II-The entire intestine was encapsulated by the membrane; and (3) Type III-The entire intestine and other organs (e.g. appendix, cecum, ascending colon, ovary, etc.) were encapsulated by the membrane.<sup>5</sup> There have been many hypothesis for etiology. The causes have been divided into primary and secondary. Most cases have reported cause to be idiopathic.<sup>6</sup> The most common secondary cause is continuous ambulatory chronic peritoneal dialysis.<sup>4,7-9</sup>

In underdeveloped countries abdominal tuberculosis is an important etiological agent of secondary SEP. In a case series of 18 cases reported by Brijendra Singh et al. encapsulating membrane sent for histopathological examination, revealed tubercular aetiology in 9 cases.<sup>10</sup>

The other less frequent causes of secondary SEP are Systemically induced by Beta adrenergic blocking agents (Practolol, Timolol, Propanolol), other drugs such as Methotrexate, Protein S deficiency, Exposure to asbestosis, Abdominal trauma, Abdominal surgery, Liver transplantation, Peritoneo-venous shunt, Ventriculoperitoneal shunt, Peritoneal sarcoidosis, Liver cirrhosis, Familial mediterranean fever, Systemic lupus erythematosus, Gastrointestinal malignancy, Intraperitoneal chemotherapy, Fibrogenic foreign body, Endometriosis, Dermoid cyst rupture, Luteinized ovarian thecomas, Cytomegalovirus peritonitis, Recurrent peritonitis, Granulomatous peritonitis related with parasitic infestation.<sup>8,11,12</sup>

The most common clinical presentation is with signs and symptoms of intestinal obstruction in emergency departments. Although rare, a painless, soft abdominal mass can be palpated in some

patients.<sup>13-15</sup> However, palpable abdominal lump was absent in our case. Ultrasound of abdomen may also show "sandwich appearance" due to the presence of echogenic membrane around the bowel loops.<sup>3,16</sup>

Contrast-enhanced CT is the most helpful imaging modality for the diagnosis of abdominal cocoon. CECT typically shows small bowel segments that are conglomerated at the midline and encased by a dense capsule with a contrast-free periphery.<sup>3,12,17</sup>

Fibrosis results in retraction of the root of the mesentery causing the bowel to clump together leading to obstruction and dysfunction. Retraction of the mesentery can lead to a characteristic appearance of the tethered small bowel loops that we have dubbed the "gingerbread man" sign.<sup>3,4,8</sup>

As most often the diagnosis is made intraoperatively, the most widely used surgical procedure is complete resection of membrane and adhesiolysis, wherever possible, cautiously avoiding inadvertent bowel injury. A simple surgical release of the entrapped bowel and removal of the fibrotic membrane is all that is required to free the bowel if no other cause of obstruction, such as a stricture, is found. Intestinal resection is only performed in non-viable gut but there are very high chances of postoperative fistula formation.<sup>8</sup>

Role of laparoscopy in management of SEP is yet to be established. Laparoscopy can be used for both diagnostic and therapeutic purposes in patients with a doubtful diagnosis but also carries risk of iatrogenic bowel injury during trocar entry and hence, insertion of the trocar into the abdomen via the open technique is recommended.<sup>18-20</sup>

In secondary SEP in dialysis patients, the management consists of cessation of peritoneal dialysis, nutritional support, and surgery if intestinal obstruction develops. Recently, it has been seen that use of steroids and tamoxifen and Angiotensin II inhibitors are beneficial.<sup>21-23</sup> If the cause of SEP is established to be of tubercular origin then postoperatively the patients were given with Anti tubercular treatment.<sup>10,24</sup> A recent report from India suggests that the use of antitubercular therapy in patients with tubercular abdominal cocoon may help avoid surgery in a subset of patients.<sup>25</sup>

## Conclusion

Abdominal cocoon is a rare cause of small bowel obstruction. Ours was a case of primary idiopathic SEP. The preoperative diagnosis requires a high

index of suspicion, supported by clinical data and imaging findings indicative of the condition. However, most cases are diagnosed at exploratory laparotomy due to lack of awareness about this entity. In our case, the preoperative imaging not only helped taking decision of early surgical intervention but also plan the steps to be taken intraoperatively. Early surgical intervention gives an excellent outcome with an excellent long-term postoperative prognosis. There is little to no risk of recurrence in primary SEP treated surgically. In our country, TB is an important etiology hence, must be ruled out in all cases of SEP and anti-tubercular treatment to be given in cases established to be of tuberculosis. The TB workup in our patient was negative, hence it was ruled out as an etiology. As India is one of country with highest reports of this condition, Abdominal cocoon should be included in differential diagnosis where other causes of intestinal obstruction cannot be established.

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