**Title Page**

**Title: Abdominal Cocoon Syndrome, A Rare and Interesting Cause of Intestinal Obstruction, Case Report.**

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**Abstract**

Abdominal cocoon syndrome, also known as encapsulating peritoneal sclerosis, is a rare cause of intestinal obstruction in which the small intestine may be entirely or partially wrapped in a thick sac of fibrous tissue that resembles a cocoon. We present a male Ethiopian patient, 60 years of age, who had a six-day history of symptoms of intermittent intestinal obstruction. Before his current presentation, he had a six-month history of sporadic vomiting and periodic abdominal pain. These symptoms would go away on their own. An exploratory laparotomy was performed for the preoperative diagnosis of small intestine obstruction secondary to primary small bowel volvulus after a plain abdomen x-ray confirmed the small bowel obstruction diagnosis. But during surgery, we discovered something unexpected: a mass formed by the encasing membrane over the small bowel. En bloc resection of the mass and distal ileum with ileo-transverse anastomosis was performed. The patient was discharged after 5 days of an uneventful post-operative stay. The morbidity and mortality of this rare instance can be decreased by awareness, prompt diagnosis, and appropriate intervention. We discuss diagnostic and therapeutic challenges encountered during the management of this patient.

**Keywords**:

abdominal cocoon syndrome, intestinal obstruction, intestinal perforation, encapsulating peritoneal sclerosis, case report

**Key Clinical Message**

Abdominal cocoon syndrome is a rare cause of bowel obstruction, with variable presentation. It needs a high index of suspicion for diagnosis. Surgical management with the release of adhesions is the preferred option for a healthy bowel. Iatrogenic bowel perforation is a possibility during bowel manipulation and the release of thick fibrous adhesions resulting in bowel resection.

**INTRODUCTION**

An abdominal cocoon is a very rare and life-threatening condition in which the small bowel is encased in a fibro-collagenous membrane that has a cocoon-like appearance. [1] It was first described by Owtschinnikow in 1907 as peritonitis chronic fibrosa encapsulata and as sclerosing encapsulating peritonitis by Deeb et al. in 1998. [1] It is considered a chronic condition in which the small bowel is partially or totally encased in a thick fibrous collagenous membrane. [2] It is more common in tropical and subtropical areas. Young adolescent females are the most commonly affected age group. [3]

The presentation of ACS is variable, and patients can present with acute, sub-acute, or chronic intestinal obstruction. [4] The clinical manifestations are not specific, preoperative diagnosis is difficult, and the majority of patients are diagnosed intraoperatively. In a systematic review of 118 patients reported by Machado et al., the most common clinical manifestations of ACS are abdominal pain, abdominal distension, abdominal mass, and nausea or vomiting. [5]

Although this condition is more common in tropical and sub-tropical areas, only a few cases have been reported from Africa, and to the best of our knowledge, this is the first case to be reported from Ethiopia. Here, we present a 60-year-old Ethiopian male patient who presented with intestinal obstruction features and whose intraoperative findings showed this unusual disease entity.

**CASE PRESENTATION**

**History**

A 60-year-old male patient presented to the emergency OPD with worsening abdominal pain of six days’ duration. The pain is periumbilical and colicky in nature. Associated with this, he had vomiting of ingested matter for several episodes, failure to pass feces, and flatus and abdominal distension of two days’ duration. The patient has been suffering from similar abdominal pain intermittently over the past 6 months, for which he was taking over-the-counter pain drugs. But this time it didn’t subside and forced him to seek medical attention. He has been hypertensive for the past 10 years on amlodipine 10 mg to be taken daily. Otherwise, the patient had no history of abdominal trauma, rectal bleeding, weight loss, contact with a known tuberculosis patient, or abdominal surgery.

**Examination**

On physical examination, he was acutely sick-looking; his vital signs were: Pulse rate: 112 beats per minute Respiratory rate: 20 per minute; blood pressure: 150/80 mmHg. He had dry lips and buccal mucosa. Abdominal examination revealed mild distention of the abdomen, which moves with respiration, and visible peristalsis. There was a hyperactive bowel sound, and the abdomen was hyper tympanic to percussion. There was a 20-cm by 10-cm soft, non-tender mass over the periumbilical area. On the digital rectal examination, there was a stool on the examining finger, but there was no blood or mass. The rest of the examination was unremarkable.

**Investigation**

Laboratory investigations, including a complete blood count and blood chemistry tests, were normal. An erect plain abdominal x-ray showed a centrally located distended bowel loop with multiple air-fluid levels (Figure 1).

**Differential diagnosis**

Preoperatively top on the list of our differential diagnoses was small bowel obstruction secondary to small bowel volvulus which is supported by clinical exam and abdominal x-ray findings. The patient has no history of abdominal surgery to consider post-operative adhesions, he has no history suggestive of underlying malignancy such as weight loss or rectal bleeding and there was no abdominal defect to suggest incarcerated hernia.

**Treatment**

With a preoperative diagnosis of complete small bowel obstruction, the patient underwent exploratory laparotomy. The intraoperative findings were 200 ml of reactive fluid in the general peritoneum; the distal half of the ileum was adherent, together with a whitish, thick fibrous band forming a mass (Figure 2). Reactive fluid was sucked out, and release of the thick adhesion was tried, but there was perforation of the distal ileum during manipulation. The mass (encapsulated small bowel) was resected, and the remaining distal ileum, which was only 4 cm, was closed in two layers, end to side, single layer. Ileotransverse anastomosis was done with 2.0 Vicryl and the abdomen closed in layers.

**Conclusion**

Abdominal cocoon syndrome is the encapsulation of the intestines with a thick fibrous membrane and is a rare cause of bowel obstruction. Pre-operative diagnosis is often challenging, especially in resource-limited settings where patients come late and important radiological investigative modalities may not be available. A detailed history, thorough physical examination, laboratory tests, and a high index of suspicion are useful for the diagnosis of this condition. Surgical resection of the membrane and relieving obstruction is the usual treatment option.

The aim of this case report is to increase awareness of this rare cause of intestinal obstruction and to emphasize that surgical release of the fibrous membrane can rarely be complicated by intestinal perforation, as seen in our patient

**Outcome and follow-up**

The patient passed flatus, and normal bowel movement was restored on the third postoperative day. His nasogastric tube was successfully removed, and antibiotics were discontinued. He was discharged on the fifth postoperative day after tolerating oral diets without any nausea or vomiting. On 2 weeks and 1 month of follow-up, he reported no episodes of pain, vomiting, or abdominal distention, and he was in good health.

**Discussion**

Sclerosing Encapsulating Peritonitis (SEP), also known as abdominal cocoon syndrome (ACS), is an extremely rare cause of small bowel obstruction [6]. It is characterized by the formation of a dense, thick, grayish-white membrane made of fibrous tissues and collagen, which encloses abdominal structures partially or totally in a cocoon-like sac [3]. The pathogenesis is still unclear, and it is essential to distinguish it from congenital peritoneal encapsulation of the small bowel with a thin transparent membrane, which is often found incidentally at laparotomy and does not usually result in intestinal obstruction [7]. Abdominal cocoon can be caused by etiologies that can be classified as primary (idiopathic) or secondary [8]. The idiopathic SEP, which is predominantly seen in male populations with a male-to-female ratio of 2:1, is common in tropical and sub-tropical regions of the world [3]. In secondary SEP, the inflammatory process in the peritoneum is initiated by several local or systemic factors, including abdominal surgery, peritoneal dialysis, sarcoidosis, beta-blockers, tuberculosis, organ transplantation, ovarian tumors, and cirrhosis [9]. Our patient had no identifiable risk factor for the development of abdominal cancer, so we consider him to have idiopathic abdominal cancer.

Three types of abdominal cocoon have been described: type 1 is when a portion of the intestine is encapsulated; type 2 is when the entire intestine is encapsulated; and type 3 is when the encapsulation also involves other intra-abdominal organs like the appendix, caecum, ascending colon, and ovaries in addition to the small intestine [10]. The intraoperative finding in our patient indicated that only a portion of the small intestine is encapsulated, which is consistent with type 1 abdominal cocoon.

The clinical manifestation of ACS could be variable. In a systematic review of 97 articles on ACS, about 2% of patients with the condition were asymptomatic. They can also present with acute or chronic abdominal pain and signs of intestinal obstruction, and most of these patients had chronic symptoms for several months before they presented with acute intestinal obstruction [11]. However, in some cases, patients may present more acutely with symptoms of intestinal obstruction, bowel ischemia, or even perforation [12]. In a single-center review of 65 patients, the average duration of symptoms before presentation was 3.9 years in idiopathic ACS [12]. The median age of presentation is 39 years [11, 12]. Our patient had a chronic history of intermittent abdominal pain before his current acute presentation, with clinical features of small bowel obstruction.

The diagnosis of ACS requires a combination of a carefully taken medical history, a complete physical examination, and evidence from a variety of laboratory and radiological investigations. It also needs a high index of suspicion from the physician. [13] Although radiological investigations are crucial in making the diagnosis, they are not usually helpful in distinguishing abdominal cocoon from other causes of small intestinal obstruction. [14]

Radiological modalities like plain abdominal X-ray, abdominal ultrasound, and abdominal CT, with the abdominal CT scan being the most useful modality, can help in the diagnosis of this rare entity. [17]. Considering the acute onset and severe condition of our patient, we proceeded with the surgical approach without an abdominal CT scan and reached a favorable outcome, including the resolution of all symptoms on follow-up.

The management of abdominal cocoons is complex. For patients who presented with less severe symptoms, nasogastric tube decompression, keeping NPO, and fluid administration could be enough. But for patients who presented with a severe sign of bowel obstruction, adhesiolysis and excision of the covering membrane to free the trapped intestinal loop in the operating room are the best management options. [18]

Intraoperatively, the main principle of the operation is to remove the capsule, release adhesion, and relieve obstruction [14]. In cases where the bowel is gangrenous or perforated, resection of the bowel with primary anastomosis or stoma creation is indicated. Our patient had a thick and tightly adherent membrane encasing the distal part of the ileum. We tried to lyse the adherent membrane from the bowel wall, but it was very difficult, and there was ileum perforation during manipulation, so en bloc resection of the membrane along with the encased bowel, which was the distal part of the ileum, was done, and then we performed ileo-transverse anastomosis.

Patients have a very good prognosis after surgical treatment [18]. Our patient had a smooth post-operative course and was discharged on the 5th day. He was also seen one month later for a follow-up, and he reported no abdominal pain or symptoms of intestinal obstruction.

**AUTHORS CONTRIBUTION**

Endeshaw Menberu: conceptualization, original draft writing, validation Solomon Guteta: Original draft writing, Simeon Mulugeta: Writing, review, editing, and analysis, Tesfaye Bekele: writing review, Yonathan Aliye: validation and editing, MergaDaba: original draft writing, investigation, and analysis, and Abdulhamid Mustefa: Investigation and Editing

**Data Availability Statement**

All data regarding the case have been reported in the manuscript. Kindly contact the corresponding author if you require any further information.

**Ethical Approval**

The author’s institution does not require ethical approval for the publication of a single case report.

**Consent for Publication**

The patient provided written informed consent for the publication of details, including history, physical findings, laboratory reports, and images.

**Disclosure**

We authors have no conflict of interest.

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