

## Case Report

### Abdominal Cocoon: Case Report and Literature Review

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**Abstract:** Abdominal cocoon is a rare condition characterized by total or partial encapsulation of the small bowel by a fibro collagenous membrane (cocoon) leading to acute or chronic bowel obstruction. Diagnosis is usually incidental at laparotomy. Imaging plays an important role in disease management. There are well-defined radiologic clues in the diagnosis of abdominal cocoon. 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, as it may prevent a “surprise” upon laparotomy and result in proper management.

**Keywords:** Abdominal cocoon, Idiopathic sclerosing encapsulating peritonitis, small bowel ileus, imaging.

#### INTRODUCTION

Abdominal cocoon is a rare condition characterized by total or partial encapsulation of the small bowel by a fibro-collagenous membrane (cocoon) leading to acute or chronic bowel obstruction [1, 2]. The term abdominal cocoon was coined by Foot *et al.* in 1978 [3], however condition was first described as ‘peritonitis chronic fibrosaincapsulata’ by Owtshinnikow way back in 1907 [4]. Deeb *et al.* in 1998 [2] named condition as ‘sclerosing encapsulating peritonitis’.

Diagnosis is usually incidental at laparotomy. 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, as it may prevent a “surprise” upon laparotomy and result in proper management [5].

#### CASE REPORT

A 40yr old male patient presented with history of episodic abdominal pain and vomiting and was referred to our department with pre-diagnosis of intestinal obstruction. He did not have history of fever or altered bowel habits. He had no previous history of surgery, chronic illness or long term use of any medication. Physical examination revealed non-tender abdomen with slight distension in epigastric and right hypochondrium with slightly increased bowel sounds. Blood tests showed elevated levels of C-reactive protein and WBC count.

Plain-X-ray-Abdomen-Erect revealed few air-fluid levels, centrally located, without free intra-peritoneal gas (Fig. 1). Sonography was done on a Philips iU22 scanner with broad band convex probes of 2 to 5 and alinear probe of 3 to 9MHz. On sonography there was localized collection of small bowel loops with mild dilatation and increased peristalsis in umbilical region. Because a localized collection of small-bowel loops with symptoms of obstruction can be seen in an abdominal cocoon or an internal hernia, a diagnosis of an abdominal cocoon with a differential diagnosis of an internal hernia was made. Contrast-enhanced MDCT of the abdomen showed clustered small-bowel loops extending from the umbilical region to right lumbar region and encased within a thin membrane (Fig. 2). There was no omental caking, ascites or abdominal adenopathy. A provisional diagnosis of abdominal cocoon was made on the basis of the clinical and radiologic findings. On surgery, a fibrous capsule encasing whole of small bowel loops was revealed (Fig. 3), with the presence of inter-loop adhesions. The greater omentum looked hypoplastic and encased in fibrous tissue (Fig. 4). Incision of the thick membrane and extensive adhesiolysis of small bowel loops were performed without loop resection. Histology of the membrane showed thickened fibro collagenous tissue without inflammation. A diagnosis of idiopathic sclerosing encapsulating peritonitis (*abdominal cocoon*) was established. Postoperative recovery was uneventful and patient was discharged on 10<sup>th</sup> post-operative day.



**Fig. 1:** X-ray-Abdomen-A.P.-Standing showing air-fluid levels in umbilical region. No free gas under diaphragm

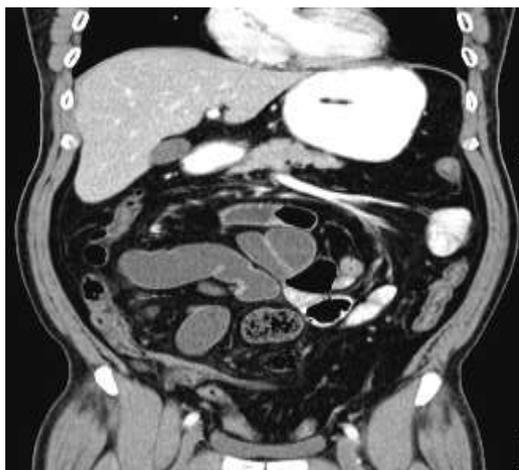


Fig. 2(a)



Fig. 2(b)

**Fig. 2(a ,b):** CECT Abdomen. Axial Images showing cluster of dilated bowel loops encased in a membrane arranged in a concertina shape.



3(a)



3(b)

**Fig. 3:** CECT Abdomen.(a)Coronal image showing cluster of dilated bowel loops in mid abdomen displacing SMA superiorly and to the left.(b) Sagittal image clustered, dilated, small bowel loops encased in a membrane.



4 (a)



4(b)

**Fig. 4(a, b): Intraoperative photographs: a fibrotic, cocoon-like membrane covering small bowel loops.**

## DISCUSSION

Sclerosing encapsulated peritonitis (abdominal cocoon) is a rare cause of small bowel obstruction. It is characterized by total or partial encasement of the small bowel by a thick fibrous sac, resulting in intestinal obstruction. The term “sclerosing” refers to the formation of sheets of dense collagenous tissue, whereas “encapsulating” describes the new fibrous tissue that covers the small bowel and restricts its motility and “peritonitis” refers to the inflammatory changes characterized by a mononuclear inflammatory infiltrate [6].

Sclerosing encapsulated peritonitis (abdominal cocoon) has been classified as primary and secondary based on whether it is idiopathic or has a definite cause [2]. The etiology of the primary form is still uncertain with various hypotheses proposed, although it is probably caused by a subclinical peritonitis leading to the formation of a cocoon [1, 3, 7]. Foo *et al.* detected the condition in 10 young girls with symptoms of bowel obstruction 2 years after menarche and postulated that a chemical peritonitis was caused by retrograde menstruation, leading to the formation of a cocoon [3]. Secondary causes include the placement of Le Veen shunt for refractory ascites [1, 7], continuous ambulatory peritoneal dialysis (CAPD) [8, 9], ventriculoperitoneal shunts, peritoneovenous shunts [4, 10], systemic lupus erythematosus [11], use of povidone iodine for abdominal wash-out [12], praxolol therapy [13], sarcoidosis [14] and peritoneal tuberculosis [15, 16]. Retrograde peritonitis and retrograde menstruation [3] have also been suggested as the cause. The prevalence of abdominal cocoon in patients undergoing CAPD ranges from 0.5% to 2.8%. In the only large prospective study conducted to date, abdominal cocoon was diagnosed in 2.5% of 1958 Japanese patients treated with CAPD over a 4-year period. Surprisingly, one-half to two-thirds of the patients acquired the disorder within an average of 4 months after termination of CAPD therapy; in some cases, the delay was as long as 4 years [17, 18].

Clinical features of abdominal cocoon include episodes of bowel obstruction, abdominal pain, nausea, vomiting, weight loss, and the presence of a non-tender soft abdominal mass detected by palpation [19]. Laboratory findings are nonspecific; slightly elevated WBC count and C-reactive protein level, hypoalbuminemia, and anemia may be detected [20].

Imaging plays an important role in disease management. There are well-defined radiologic clues in the diagnosis of abdominal cocoon [21]. Plain abdominal X-ray film may show dilated bowel loops with air-fluid levels [22]. Barium studies show a fixed U-shaped cluster of dilated small-bowel loops lying in a serpentine or concertina-like fashion, with delayed small-bowel transit time [6, 16, 19]. Some authors have described a ‘cauliflower sign’ on barium small bowel series [23]. On ultrasonography characteristic appearance of abdominal cocoon has been described by S. Boopathy *et al.* [24] as encasement of most of the small-bowel loops in a thick fibrous membrane and arrangement of the loops in a concertina shape with a narrow posterior base, having the overall appearance of a cauliflower. The CT findings described are peritoneal thickening, signs of obstruction, tethering/angulation, fixation of loops, mural thickening, ascites and loculated fluid collections [25]. Containment of loops within a sac, the walls of which may show enhancement is also reported [2, 15, 16, 19]. The imaging features are, however, not pathognomonic and preoperative diagnosis requires a high index of clinical suspicion.

A localized collection of small bowel loops with symptoms of obstruction can also be seen in internal hernia on sonography and it should be considered in *differentials*. The computed tomographic features of an internal hernia are evidence of small-bowel obstruction: clustering of the small bowel and stretched, displaced, crowded, and engorged mesenteric vessels [26]. It has been observed that CT findings of a membrane enveloping loops of small bowel were also

seen in some paraduodenal hernias and peritoneal encapsulation. However, clinical and pathological features of these entities are different [27]. Peritoneal encapsulation is described as a developmental anomaly where the whole of the small bowel is encased in a thin accessory membrane. The clinical symptoms of this condition differ from those of the abdominal cocoon syndrome, in that the patients are mostly asymptomatic and the findings are incidental and late in life [7, 27].

Differential diagnosis are internal hernia and peritoneal encapsulation.

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 [5, 28].

In conclusion, Idiopathic sclerosing encapsulating peritonitis or *abdominal cocoon*, although a rare cause of a common surgical emergency such as small bowel ileus, may be responsible, especially in cases with recurrent attacks of non-strangulating obstruction in the same individual. A high index of clinical suspicion may be generated by the recurrent presentation of small bowel ileus combined with relevant imaging findings and lack of other etiologies. Clinicians must rigorously pursue a preoperative diagnosis, as it may prevent a "surprise" upon laparotomy and unnecessary procedures for the patient, such as bowel resection.

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