**Case Report**

# Idiopathic abdominal cocoon: An enigma

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## Abstract

Abdominal cocoon is often described by various terminologies like encapsulating peritoneal sclerosis (EPS) or sclerosing encapsulating peritonitis or peritonitis chronica fibrosa incapsulata is defined as syndromes associated with symptoms due to formation of a fibro-collagenous peritoneal membrane involving commonly the small intestinal loop. Clinical presentation ranges from abdominal pain to features of intestinal obstruction which may be acute or sub-acute. It is believed to be mesenchymal transition of mesothelial cells. This condition is commonly associated with tuberculosis, peritoneal dialysis and previous abdominal surgeries but may also be idiopathic. Though a wide range of medical management has been tried for conservative management of the patient, surgery is the preferred choice of treatment to alleviate the persisting symptoms. This is one condition where on table intra op diagnosis supersedes the imaging and histological diagnosis. Here, we discuss the case report of 32 year old male, known diabetic for 4 years, who presented with complaints of abdominal pain, intermittent for over 5 years, with no evidence of intestinal obstruction. Imaging showed abdominal cocoon of small bowel loops and mid gut rotation anomaly with internal hernia. Laparoscopically the cocoon sac was removed and adhesiolysis was done. This case report is to add richness to limited amount literary resources available about abdominal cocoon syndrome.

## Key words

Abdominal cocoon syndrome, Encapsulating peritoneal sclerosis, Sclerosing encapsulating peritonitis, Peritonitis chronica fibrosa incapsulata, Mesenchymal transition, Tuberculosis, Peritoneal dialysis.

## Introduction

Abdominal cocoon is often described by various terminologies like encapsulating peritoneal sclerosis (EPS) or sclerosing encapsulating peritonitis or peritonitis chronica fibrosa incapsulata [1] is defined as syndromes associated with symptoms due to formation of a fibro-collagenous peritoneal membrane involving commonly the small intestinal loop. Clinical

presentation ranges from abdominal pain to features of intestinal obstruction which may be acute or sub-acute. It is believed to be mesenchymal transition of mesothelial cells [2]. This condition is commonly associated with tuberculosis, peritoneal dialysis and previous abdominal surgeries but may also be idiopathic [3]. Though a wide range of medical management has been tried for conservative management of the patient, surgery is the preferred choice of treatment to alleviate the persisting symptoms. This is one condition where on table intra op diagnosis supersedes the imaging and histological diagnosis [4].

Abdominal cocoon refers to complete or partial encapsulation of small bowel loops by fibro collagenous membrane. It may be primary or idiopathic and secondary with a definitive cause. It was first described by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata" and termed "abdominal cocoon" by Foo in 1978 [5]. Various pathogenesis have been proposed for the primary Sclerosing Encapsulating Peritonitis, though concrete answers could not be obtained, yet. Secondary causes could tuberculosis, drugs (practolol, propranolol), chronic ambulatory peritoneal dialysis, ventriculoperitoneal shunts, and diseases such as sarcoidosis, SLE, liver cirrhosis, uterine leiomyomas, endometriotic cysts or tumours of the ovary [6].

We reported here a rare of case abdominal cocoon seen in 32 year old male, presenting with recurrent abdominal pain with no evidence of obstruction.

## Case report

A 32 year old male presented to our department with non-radiating colicky type of abdominal pain for 1 week duration. Patient gave history of non-bilious vomiting containing food particles since 3 days. There was no history of altered bowel or bladder habits. Patient gave past history of similar intermittent complaints for 5 years. The symptoms were relieved on medications. There was also previous admission in a private hospital, where he was treated conservatively 2 years back. There was no history of significant loss of weight or loss of appetite. Patient is known case of type 2 diabetes mellitus on OHA and no history of contact with tuberculosis or no previous surgeries.

On presentation, patient was afebrile, adequately hydrated, moderately built and well nourished. His vitals were normal and all basic investigations were within normal limits. Abdomen was soft, with no palpable swelling and diffuse minimal tenderness present over all quadrants. Ultrasound abdomen and pelvis showed superior mesenteric vein above and left of superior mesenteric artery- consistent with mal rotation and mild splenomegaly (13.1 x 5.5cm). Contrast Enhanced Computed Tomography (CECT) showed sac like clumped and moderately dilated small bowel loops noted in mid abdomen, showing stretched and engorged mesenteric vessels within it, 3$^{rd}$ and 4$^{th}$ part of duodenum not crossing the midline and mid gut rotation anomaly with internal hernia. Colonoscopy showed diminutive polyp within transverse colon.

Patient underwent laparoscopy and diagnosed as a case of abdominal cocoon with internal herniation of distal ileum. A thin whitish membrane was encapsulating small bowel loops till terminal part of ileum. Around 15 cm from ileo colic junction, terminal ileum was found herniating within the cocoon and Duodenal jejunal flexure was shifted to the right. The cocoon sac was then removed and adhesiolysis of flimsy strands was done along the terminal ileum. The excised sac was sent for histopathological examination, which revealed proliferation of the fibroconnective tissue with signs of nonspecific inflammatory reaction and no signs of tuberculosis or malignancy. Post-operative period was uneventful and patient recovered well. Regular follow up was advised for the patient (**Figure – 1, 2, 3**).

**Figure – 1:** Ultra sound abdomen showing clumped large echogenic mass- small bowel loops.

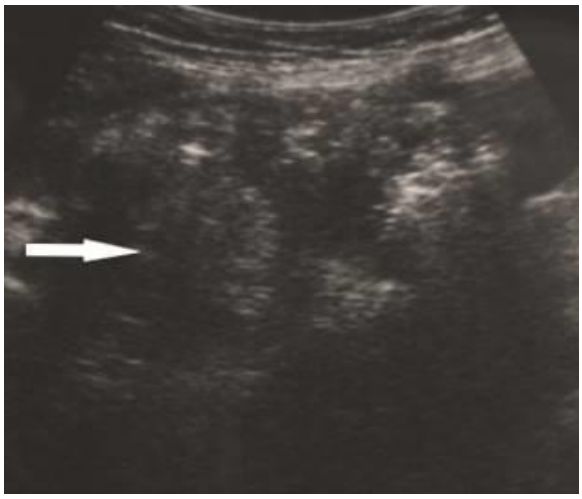


**Figure – 2:** Contrast-enhanced abdomen computed tomography (CT) demonstrating small bowel loops congregated to the centre of the abdomen encased by a soft-tissue density mantle with minimal ascites.

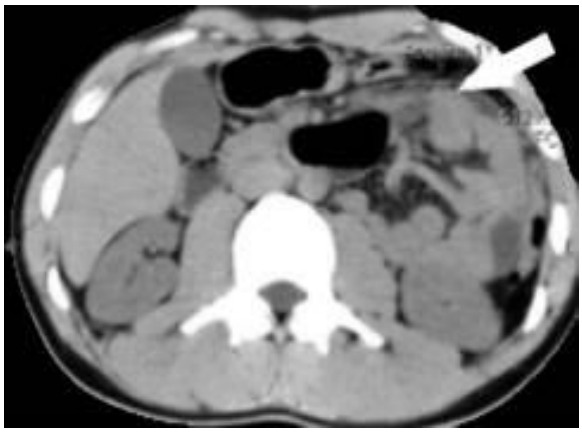
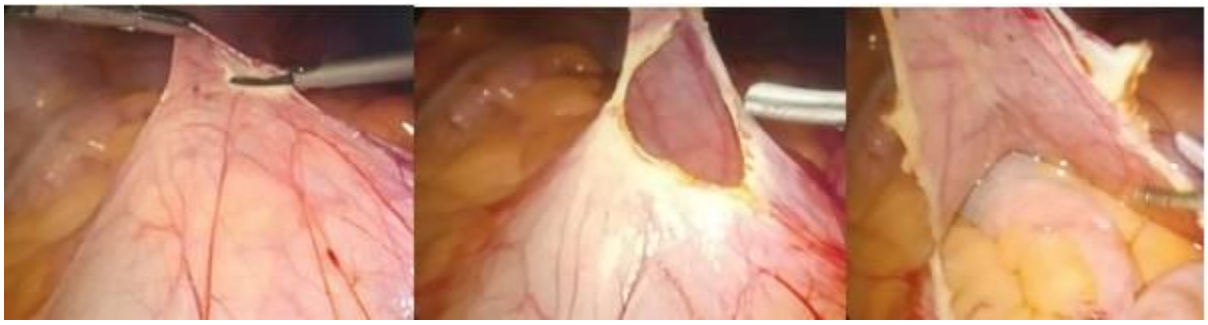


**Figure – 3:** Intra operative images showing abdominal cocoon- thin membrane extending till terminal part of ileum. The cocoon sac was opened and excised.



## Discussion

Abdominal cocoon syndrome, also known as idiopathic Sclerosing Encapsulating Peritonitis (SEP), is a rare cause of intestinal obstruction when bowel loops get entangled by fibro collagenous membrane. Though most of the patients are asymptomatic it might present with features of intestinal obstructions, pain, loss of appetite, palpable mass or ascites [7]. Abdominal cocoon may be classified into primary or idiopathic and secondary. Primary abdominal cocoon occurs mainly in young women from tropical and subtropical zones. Although retrograde menstruation with or without viral infection of the fallopian tubes has been suggested as a possible cause [8], it may not account for the occasional occurrence of abdominal cocoon in males [9]. Secondary abdominal cocoon is associated with factors like recurrent peritonitis, intake of intra peritoneal irritants, including antibiotics and beta blockers, chronic ambulatory peritoneal dialysis (CAPD), sarcoidosis, familial Mediterranean fever, carcinoid syndrome, exposure to asbestos, and autoimmune disease [10]. In sub-tropical countries like India cocoon syndrome is usually associated with extra pulmonary tuberculosis. In spite of advancement in imaging and bio chemical analysis the pre-operative diagnosis of abdominal cocoon syndrome is difficult. It's usually intra operative findings which provide the final diagnosis of the disease followed by histopathological confirmation.

The ultra sound imaging of abdomen in the patients usually shows clumped bowel loops (sandwich appearance) [11] and varied degree of

dilation and vascularity based on the level obstruction with or without ascites. The Contrast imaging shows small bowel loops congregated to the centre of the abdomen encased by a soft-tissue density mantle. This is often referred to as 'cauli flower sign' [12] on sequential films.

The management of abdominal cocoon is still under debate. Though conservative management is preferred to alleviate the symptoms, most authors accept that surgery is the choice of treatment [13]. Depending on the adhesions, vascularity of bowel loops either open or laparoscopic surgery is preferred by the surgeon. During surgery, in addition to careful dissection and excision of the covering membrane, dense inter bowel adhesions also need to be freed for complete recovery [14]. The surgical complications expected include intra-abdominal infections, enterocutaneous fistula and perforation.

## Conclusion

To conclude, the abdominal cocoon syndrome continues to be an enigma to diagnose and treat. In spite of recent advancements in bio chemical analysis and imaging technique pre-operative diagnosis of the disease remains difficult. Early identification may possibly delay the progression of disease. However, in patients with recurring symptoms, surgery remains as the cornerstone of management.

## References


1. Foo KT, Ng KC, Rauff A, et al. Unusual small intestinal obstruction in adolescent girls: the abdominal cocoon. Br J Surg., 1978; 65: 427-30.
2. Lewin K, McCarthy LJ. Peritoneal encapsulation of the small intestine. Gastroenterology, 1970; 59: 270-2.
3. Xu P, Chen LH, Li YM. Idiopathic sclerosing encapsulating peritonitis (or abdominal cocoon): a report of 5 cases. World J Gastroenterology, 2007; 13: 3649-51.
4. Akbulut S. Accurate definition and management of idiopathic sclerosing encapsulating peritonitis. World J Gastroenterology, 2015; 21: 675-87.
5. Machado NO. Sclerosing Encapsulating Peritonitis: Review. Sultan Qaboos Univ Med J., 2016; 16: e142-51.
6. Singh B, Gupta S. Abdominal cocoon: a case series. Int J Surg., 2013; 11: 325-8.
7. Li N, Zhu W, Li Y, et al. Surgical treatment and perioperative management of idiopathic abdominal cocoon: single-center review of 65 cases. World J Surg., 2014; 38: 1860-7.
8. Santos VM, Barbosa ER, Jr., Lima SH, et al. Abdominal cocoon associated with endometriosis. Singapore Med J., 2007; 48: e240-2.
9. Awe JA. Abdominal cocoon syndrome (idiopathic sclerosing encapsulating peritonitis): how easy is its diagnosis preoperatively? A case report. Case Rep Surg., 2013; 2013: 604061.
10. Tannoury JN, Abboud BN. Idiopathic sclerosing encapsulating peritonitis: abdominal cocoon. World J Gastroenterol., 2012; 18: 1999-2004.
11. Ti JP, Al-Aradi A, Conlon PJ, et al. Imaging features of encapsulating peritoneal sclerosis in continuous ambulatory peritoneal dialysis patients. AJR Am J Roentgenol., 2010; 195: W50-4.
12. Wang Q, Wang D. Abdominal cocoon: multi-detector row CT with multiplanar reformation and review of literatures. Abdom Imaging., 2010; 35: 92-4.
13. Habib SM, Betjes MG, Fieren MW, et al. Management of encapsulating peritoneal sclerosis: a guideline on optimal and uniform treatment. Neth J Med., 2011; 69: 500-7.
14. Lewin K, McCarthy LJ. Peritoneal encapsulation of the small intestine. Gastroenterology, 1970; 59: 270-2.