E-ISSN: 2708-1508
P-ISSN: 2708-1494
IJCRS 2024; 6(2): 24-27
www.casereportsofsurgery.com
Received: 21-05-2024
Accepted: 27-06-2024

Dr. Jinal M Thakkar
Third Year Resident, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

Dr. Jay Pitroda
Assistant Professor, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

Dr. Gunvant H Rathod
Professor and HOD, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

Dr. Bhavin Kothari
Professor, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

Dr. Devarshi Bhagora
Senior Resident, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

Corresponding Author:
Dr. Jinal M Thakkar
Third Year Resident, Department of General Surgery, DR. M. K. Shah Medical College and Research Centre, SMS Multispeciality Hospital, Chandkheda, Ahmedabad, Gujrat, India

A case of concealed danger: Subacute intestinal obstruction due to idiopathic sclerosing encapsulating peritonitis

Dr. Jinal M Thakkar, Dr. Jay Pitroda, Dr. Gunvant H Rathod, Dr. Bhavin Kothari and Dr. Devarshi Bhagora

DOI: https://doi.org/10.22271/27081494.2024.v6.i2a.108

Abstract
Sclerosing encapsulating peritonitis (SEP) is characterized by partial or complete encasement of small intestine by a thick fibrocollagenous membrane. Depending on underlying causes, it is divided into primary and secondary forms. Primary or idiopathic also known as abdominal cocoon syndrome. Herein we presented a case of idiopathic sclerosing encapsulating peritonitis (abdominal cocoon syndrome). A 39-year-old male patient presented to our emergency department with signs and symptoms of subacute intestinal obstruction. Patient's history, physical examination findings, patient's age and abdominal radiography were consistent with subacute intestinal obstruction. Explorative laparotomy revealed a fibrous capsule encasing intestines as well as dense adhesions between intestinal loops. Adhesiolysis was done. Post-operative patient was discharged without complication. Despite advances in radiological techniques, the exact diagnosis in many cases is still made according to intraoperative findings and histopathological properties of the excised membrane. While some cases of ACS remain asymptomatic for years, most cases are characterized by recurrent bouts of acute, subacute or chronic intestinal obstruction.

Keywords: Sclerosing encapsulating peritonitis (SEP), subacute intestinal obstruction, abdominal cocoon syndrome (ACS), abdomen

Introduction
Idiopathic sclerosing encapsulating peritonitis (abdominal cocoon syndrome) is a rare disease that is defined as idiopathic partial or total encapsulation of the bowel within a fibrocollagenous membrane and is considered as a rare cause of small bowel obstruction (SBO) [1].

It was first termed as peritonitis chronic fibrosain capsulata by Owtschinnikow in 1907 and finally abdominal cocoon by Foo in 1978. It is most commonly seen in adolescent girls of tropical and subtropical region though few cases of male have also been reported in literature [2].

Case report
39 year male patient presented to emergency with complain of generalised sever abdominal pain and abdominal distension since 2 days. It was associated with nausea, vomiting and obstipation. Not associated with fever, diarrhea and weight loss. Patient has similar episode before 3 month for 1 day which was relived by conservative management. Patient had no other significant operative history or comorbidities. On examination there were generalized abdominal distension, generalized tenderness, gurding present in left iliac and hypogastric region with increased bowel sounds. After some conservative management patient had passed flatus and stool with some releved in symptoms.

In investigations X ray abdomen standing suggestive of intestinal obstruction. In USG abdomen visualized small bowel loop mildly dilated (maximum diameter 34 mm) and show sluggish peristalsis.

Multiple air fluid level suggestive of intestinal obstruction

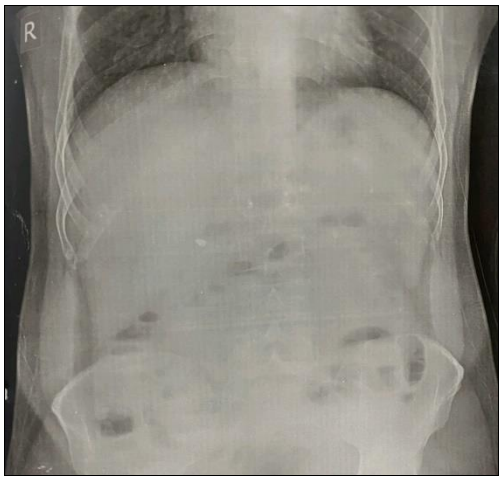


Fig 1: X-ray abdomen standing

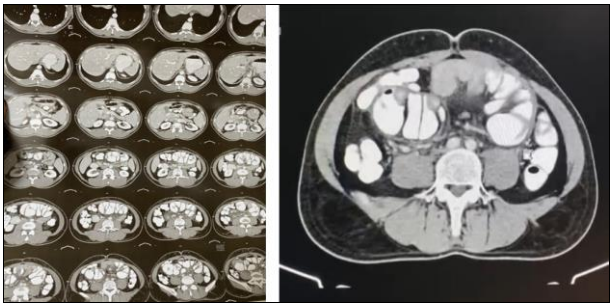


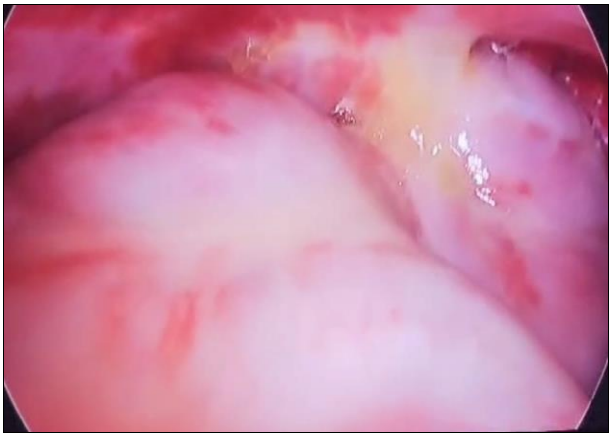
Fig 2: CECT abdomen and pelvis

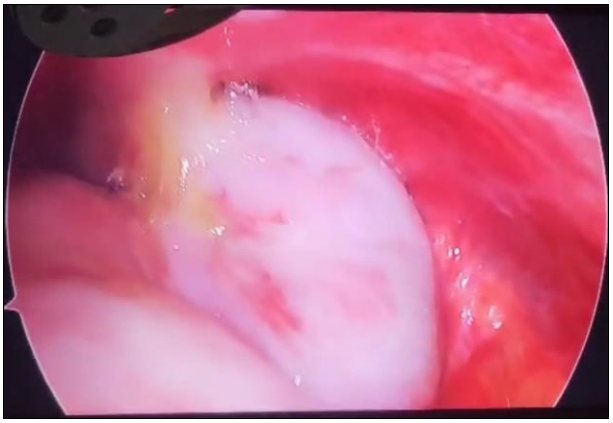
In CECT abdomen pelvis there was few of small bowel loops in lower abdomen and pelvis region appear dilated (maximum diameter of 34 mm) and appear crowded with adjacent area of peritoneal thickening, mild mesenteric haziness and inter-bowel fluid with bowel loops opacified and contrast reaching up to rectum and anal canal suggestive of subacute bowel obstruction possibility due to peritonitis.

On basis of investigation our patient diagnosed with subacute intestinal obstruction and scheduled for diagnostic laparoscopy and sos procedure. In diagnostic laparoscopy there were two encapsulated small bowel mass with a thickened white, cocoon-like fibrous membrane, including the distal whole small bowel from duodeno-jejunal flexure to ileo-cecal junction. So laparotomy done for further surgical management. In laparotomy there were two small bowel cocoon present big one in left flank and left iliac region and small one in right iliac region which was attached with anterior abdominal wall. Appendix was separate. A careful sharp dissection and excision of the dense membrane with lysis of the severe adhesions between the bowel loops were performed without any bowel injury during dissection. Fibrous membrane sent for histopathological examination, culture and CBNATT examination. ADK no. 28 placed in pelvic cavity.

In postoperative period patient was vitally stable. Liquied were started from POD 2 and soft diet started from POD 3. Patient tolerating food and drain removal done on POD 5 without any complications. Postoperative period was uneventful and patient was discharged on post-operative day 8.

In investigations tissue culture were negative for gram's stain and ZN stain. In CBNATT *M. tuberculosis* not detected. In histopathological examination there were fibrocollagenous tissue with inflammatory infiltrates, consisting of lymphocytes and plasma cells along with mild degree of vascularity; no malignancy suggestive of chronic non-specific inflammatory changes. Depanding on all investigations and intraoperative finding diagnosis of idiopathic sclerosing encapsulating peritonitis (abdominal cocoon syndrome) were made.

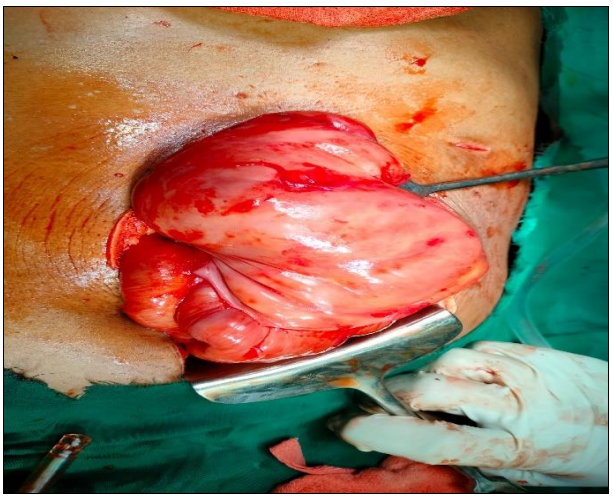




Laparoscopic view of small bowel cocoon present in left flank and left iliac region.

Laparoscopic view of small bowel cocoon in right iliac region which was attached with anterior abdominal wall

Fig 3: Laparoscopic view of abdominal cocoon syndrome



Small bowel mass with a thickened white, cocoon-like fibrous membrane, including the distal whole small bowel from duodeno-jejunal flexure to ileo-cecal junction

Fig 4: Laparotomy view of abdominal cocoon syndrome

Discussion

Intra-abdominal inflammation, recognized as sclerosing encapsulating peritonitis (SEP), stands as an uncommon yet consequential condition precipitating intestinal obstruction and the peril of gangrene. This affliction delineates a scenario where abdominal organs become enveloped or encased within a dense fibrous tissue, evoking the imagery of a protective
'Cocoon' [6]. Depending on the underlying causes, SEP is divided into primary (idiopathic) and secondary forms. The idiopathic form of the disease was named as 'abdominal cocoon syndrome' in 1978 [5]. In its first description 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. Apart from the primary variant, EPS is classified as secondary when there are identifiable underlying triggering factors that may include continuous ambulatory peritoneal dialysis (CAPD) with recurrent peritonitis which can be due to bacterial infection or sterile chemical peritonitis, peritoneal tuberculosis, use of certain drugs like beta blockers, methotrexate, asbestos, use of intraperitoneal chemotherapy, LeVeen shunt, ventriculoperitoneal shunts, systemic lupus erythematosus, sarcoidosis, luetinized thecoma of ovary, ruptured dermoid cyst among others [3].

Abdominal cocoon syndrome is believed to be a result of a 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. This eventually leads to a marbled, thickened, leathery fibro-connective tissue sheath like structure that envelopes the small intestine in the form of a cocoon. A possibility of developmental abnormality cannot be ruled out, as abdominal cocoon has been associated with omental hypoplasia and mesenteric vascular malformation [3].

It is encountered equally in both genders and presents over a wide age range of ages from young as well as old age. The hallmark of EPS is the intermittent nature of its presentation. The initial symptoms are usually related to the altered gut motility and transit and altered peritoneal permeability. With the development of complete sclerosis due to formation of cocoon there are overt signs of intestinal obstruction (partial/ complete).

small intestinal obstruction presents with abdominal pain, nausea, vomiting, constipation, and ascites with or without abdominal mass with signs of severe malnutrition [3]. In present case report middle age male patient presented with subacute intestinal obstruction.

There are no specific physical examination and laboratory findings suggestive of SEP [7]. The examination of barium-contrast X-ray, USG, CT, and occasionally contrast-enhanced magnetic resonance imaging (MRI) might be helpful for the definitive preoperative diagnosis of idiopathic sclerosing encapsulating peritonitis to some extent. Plain X-ray findings are not specific to the diagnosis of idiopathic SEP, which shows diffuse air-fluid levels and dilated small intestinal loops in the abdomen. Barium-contrast X-ray studies are better, manifesting the accumulated, and conglomerated intestinal loops at the center of the abdomen. Barium studies may not be possible in patients with prominent signs of intestinal obstruction. Dilated bowel loops, thick fibrous membranes
and loculated ascites that surround the small intestine are detected on ultrasonography (US) [7]. Multidetector computer tomography technology has greater accuracy because it allows for multiplanar (axial, sagittal, and coronal) reconstruction. This characteristic appearance was that the intestine was centralized in a capsule, termed as the "cauliflower sign" or "accordion pattern." A prolonged transit time may also aid in the diagnosis. There was almost similar outcome between using contrast enhanced MRI and CT imaging, but the encasing membrane was more obvious in the MRI examination. Abdominal CT scans tell us the degree of obstruction with AC. Histologic examination of the membrane tissues showed proliferation of fibrocytes and enrichment of collagen fiber, with nonspecific inflammatory reaction and vascular proliferation in some cases [4]. Same histological finding were present in our case report.

Surgery is considered to be the first choice for patients with recurrent acute or chronic intestinal obstruction. During surgery, we can find entire or partial intestine encapsulated in a thick, white, fibrous "cocoon-like" membrane. Membrane excision and adhesiolysis should be applied to all encased intestinal segments.[4] In present report on basis of symptoms of subacute intestinal obstruction laparoscopy followed by laparotomy done and same intraoperative finding were present as described erlier and ahesiolysis done.

Postoperative complications including intra abdominal infection, intestinal fistula, short bowel syndrome, and bowel perforation are quite rare in patients with ACS. Early postsurgical activity was encouraged. Acupuncture, physical therapy can be used to promote enterokinesia [4].

Conclusion

Idiopathic sclerosing encapsulating peritonitis forms a minority of unusual conditions that lead to acute or subacute intestinal obstruction. Preoperative diagnosis is a true challenge and most reported cases have been incidentally diagnosed during laparotomy**.** This study is to raise awareness of this rare type of intestinal obstruction. The combination of a careful history, physical examination, and radiological signs specially on X-ray abdomen standing, ultrasonography abdomen, CT abdomen; along with high suspicious index by clinicians, may be helpful in achieving a definitive diagnosis. The management of this disease is currently considered to be surgery. Adhesiolysis with excision of the membrane is needed.

Conflict of Interest

Not available.

Financial Support

Not available.

References

1. Schauki M, Hashi Salad A, Abdullahi A. Abdominal cocoon syndrome, a diagnostic challenge affecting the treatment: A rare case report from Somalia. J Surg. Case Rep. 2023;3:1-4.
2. Jagdale S, *et al*. Abdominal cocoon - A rare etiology of intestinal obstruction. J Fam Med Prim Care. 2017;6(3):357-359.
3. Mandavdhare HS, *et al*. Abdominal cocoon: An enigmatic entity. Trop Gastroenterol. 2016;37(3):156-167.
4. Xia J, *et al*. Abdominal cocoon with early postoperative small bowel obstruction. Medicine, 2018, 97(25).

5. Yavuz R, Akbulut S, Babur M, Demircan F. Intestinal obstruction due to idiopathic sclerosing encapsulating peritonitis: A case report. Iran Red Crescent Med. J, 2015 May, 17(5).
6. Vipudhamorn W, Juthasilaparut T, Sutharat P, Sanmee S, Supatrakul E. Abdominal cocoon syndrome - A rare culprit behind small bowel ischemia and obstruction: three case reports. World J Gastrointest Surg. 2024 Mar 27;16(3):955-965.
7. Acar T, Kokulu İ, Acar N, Tavusbay C, Hacıyanlı M. Idiopathic encapsulating sclerosing peritonitis. Ulus Cerrahi Derg. 2015;31:241-243.

How to Cite This Article
Thakkar JM, Pitroda J, Rathod GH, Kothari B, Bhagora D. A case of concealed danger: Subacute intestinal obstruction due to idiopathic sclerosing encapsulating peritonitis. International Journal of Case Reports in Surgery. 2024;6(2):24-27.