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# Idiopathic Sclerosing Encapsulating Peritonitis: A case report and review of literature

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***Abstract :*** *Idiopathic Sclerosing Encapsulating Peritonitis (Abdominal Cocoon) is a rare cause of small bowel obstruction. We present a case of acute small bowel obstruction in a young male preoperatively diagnosed as Idiopathic Sclerosing Encapsulating Peritonitis on computed tomography. A high index of clinical suspicion of such condition which can present with recurrent episodes of subacute intestinal obstruction supported by radiological investigations can help in preoperative diagnosis of such cases as most cases are incidentally diagnosed during laparotomy.*

***Keywords -*** *Idiopathic Sclerosing Encapsulating Peritonitis, Abdominal Cocoon, laparotomy, small bowel obstruction*

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## I. Introduction

Sclerosing encapsulating peritonitis is a rare cause of small bowel obstruction and can be idiopathic or secondary to chronic ambulatory peritoneal dialysis. The idiopathic type, also known as *abdominal cocoon* was first described by Foo et al in 1978. [1]It affects mainly young females from tropical and subtropical regions and is characterized by a thick, fibrotic, cocoon-like membrane, partially or totally encasing the small bowel. Clinically, it presents with recurrent episodes of acute or subacute small bowel obstruction, weight loss and sometimes with a palpable abdominal mass. [2] Most cases are diagnosed incidentally at laparotomy although a preoperative diagnosis is possible due to radiological investigations. We present a similar case of Idiopathic SEP in a 36 year old male who presented as acute small bowel obstruction.

## II. Case Report

A 36 year old male with no prior medical illness presented to the emergency department as acute colicky abdominal pain and history of multiple episodes of bilious vomiting. There was past history of similar episodes on multiple occasions which resolved on conservative management. On examination, patient was afebrile and hemodynamically stable. Per abdomen examination revealed generalized tenderness, abdominal distension and hyper peristaltic bowel sounds. A presumptive diagnosis of acute small bowel obstruction was made.

Laboratory investigations showedHb-11.8gm% TC-11800cell/mm3 with a neutrophilic predominance of 62%.Other laboratory parameters like renal and liver function tests were within normal limits. X-ray erect abdomen showed multiple air fluid levels and centrally located bowel loops without free intraperitoneal air. Ultrasound abdomen revealed dilated small bowel loops with maximum diameter of 3.1 cm. CT abdomen detected dilated clumped small bowel loops with maximum transverse diameter of 3.2 cm. Large bowel was collapsed but there was no transition zone noted.

Patient was taken for emergency laparotomy after adequate fluid resuscitation. Operative findings showed bowel loops encased in a fibrous capsule associated with the presence of interloop adhesions (Figure1). Incision of thick membrane and extensive adhesiolysis was performed without bowel resection. Histopathology revealed fibrocollagenous tissue without inflammation. Diagnosis of idiopathic sclerosing encapsulating peritonitis was made. Post operative period was uneventful and patient was discharged on 12$^{th}$ day of admission.

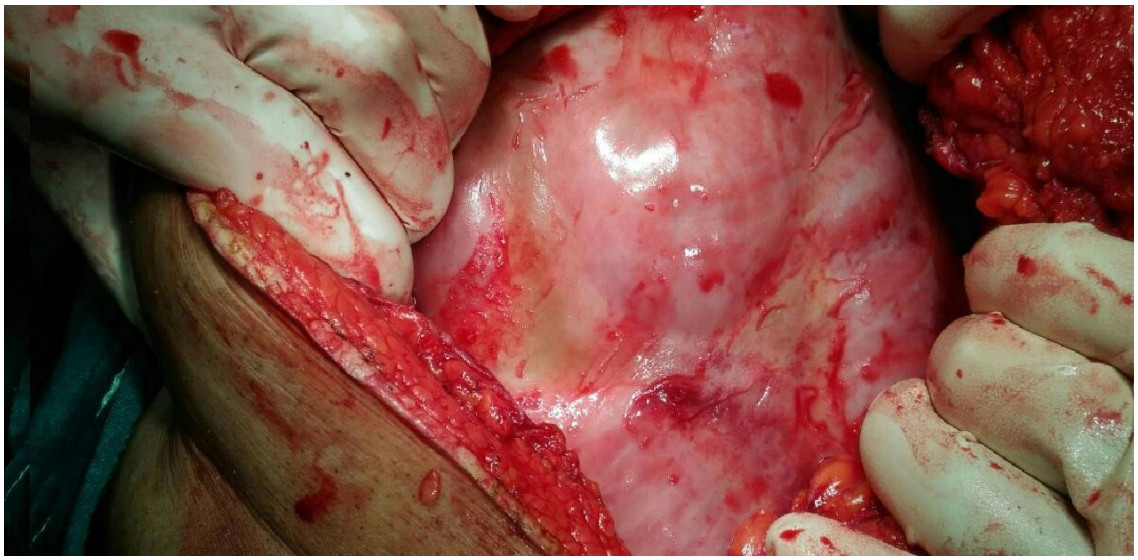

**Figure 1 Laparotomy showing an encapsulating thick adherent membrane encasing the small bowel loops**

## III. Discussion

SEP has been described in young adolescent females from the tropical and subtropical regions. [3] The proposed mechanisms include retrograde menstruation with a superimposed viral infection, retrograde peritonitis and cell-mediated immunological tissue damage triggered by gynecological infection. This is one of the rare cases presenting in a young male with no prior etiology contrary to the female predominance as in previously reported cases of SEP. [4] Developmental abnormality may also be a probable etiology as it is often accompanied by other embryologic abnormalities like greater omentum hypoplasia. [2] The secondary form of SEP has been frequently reported in association with continuous ambulatory chronic peritoneal dialysis. [5] Other rare causes of secondary form of SEP reported are prior abdominal surgery, tuberculosis, subclinical primary viral peritonitis, recurrent peritonitis, beta-blocker treatment (practolol), peritoneovenous shunting and pertioneoventricular shunting. [6]

Preoperative diagnosis requires a high index of clinical suspicion as earliest clinical features of SEP are nonspecific and can be easily missed. It presents with recurrent abdominal pain, nausea, vomiting, weight loss and recurrent episodes of acute, subacute or chronic small bowel incomplete or complete obstruction. Sometimes it can present with a palpable soft non tender abdominal mass. Although it is difficult to make a definite preoperative diagnosis, most cases are diagnosed incidentally at laparotomy.

Conventional radiographs may show dilated bowel loops and air fluid level. Contrast study of the small intestine shows varying lengths of small bowel tightly enclosed in a "cocoon'' of thickened peritoneum, proximal small bowel dilatation, and increased transit time. It may show a fixed cluster of dilated small bowel loops lying in a concertina like fashion, giving a cauliflower-like appearance("cauliflower sign"). [7]Ultrasound findings include a trilaminar appearance of the bowel wall, tethering of the bowel to the posterior abdominal wall, dilatation and fixation of small bowel loops, ascites, and membrane formation. [7]

The exact diagnosis is made by computed tomography of the abdomen demonstrating centrally located small bowel loops encased by a thick membrane. [8] This was further classified into 3 types according to the extent of the encasing membrane: (1) Type I - 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. Histologically, the peritoneum shows a proliferation of fibro-connective tissue, inflammatory infiltrates, and dilated lymphatics, with no evidence of foreign body granulomas, giant cells, or birefringent material. [9]

Management of SEP is debated. In most cases, the diagnosis is established at later stages of the disease at laparotomy when the patient develops partial or complete small bowel obstruction. Laparotomy reveals characteristic gross thickening of the peritoneum, which encloses some or all of the small intestine in a cocoon of opaque tissue. Fibrous bands form between the bowel loops, and when the mass of bowel is sectioned, many small loculated abscesses due to local perforations are found.

Various treatment options have been used such as subtotal excision of the membrane, enterolysis, small bowel intubation, bowel resection, and exploratory laparotomy in patients with high perforative risk. Stripping of the membrane with intestinal releasing without intestinal resection is the treatment of choice. [10] Bowel resection is indicated only if it is nonviable as it unnecessarily increases the complications. Surgical complications reported include intra-abdominal infections, enterocutaneous fistula and perforated bowel. [10] In

some patients, repeated adhesiolysis may be required. Laparoscopic approach has also been tried. [11] This clinical entity carries an excellent long-term postoperative prognosis with minimal risk of recurrence. No surgical treatment is required in asymptomatic SEP. Treatment for secondary SEP in dialysis patients is cessation of PD, nutritional support, and surgery for intestinal obstruction, if required. Studies have shown that steroids and tamoxifen or Angiotensin II inhibitors could be of benefit in treatment of abdominal cocoon. [12]

## IV. Conclusion

Idiopathic Sclerosing Encapsulated Peritonitis although rare should be kept as one of the differential diagnosis of acute to subacute intestinal obstruction especially cases with prior recurrent attacks. Although etiology is still unknown, a better knowledge of this rare entity supported by the radiological investigations can help in preoperative diagnosis of the condition and better management.

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