

# Idiopathic encapsulating sclerosing peritonitis

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## ABSTRACT

Sclerosing encapsulating peritonitis (SEP)/abdominal cocoon syndrome is a rare condition that is generally identified in young females. The exact cause is still unknown. Timely and accurate imaging and diagnosis play a critical role for morbidity and mortality. It is usually diagnosed during surgery. The initial treatment should be conservative, and aggressive surgical approach should be avoided as much as possible. Herein, we aimed to review the clinical features of SEP based on a patient who underwent surgery in our clinic.

**Keywords:** Idiopathic, encapsulating sclerosing peritonitis, abdominal cocoon, explorative laparotomy, corticosteroids

## INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a rare condition referring to complete or partial encapsulation of the small intestine by a fibro-collagen membrane, forming a cocoon appearance, which may lead to complications such as obstruction, bleeding and perforation (1). It is more common in young adult women living in tropical areas (2). Owtschinnikow first defined this entity as "peritonitis chronica fibrosa capsulata" in 1907. Its preoperative diagnosis is very difficult and is usually diagnosed during surgery (3). We aimed to review the clinical features of SEP based on a patient who underwent surgery in our clinic.

## CASE PRESENTATION

A 38-year-old woman presented to our general surgery department with complaints of episodic abdominal pain and weight loss for the last six months. The patient did not have any known chronic diseases, medication use, or previous surgery. Her physical examination was normal except a reduced umbilical hernia. Her bowel sounds were normo-active, and the digital rectal examination was normal. Her laboratory values were within normal range except blood urea (50 mg/dL) and creatine (3 mg) levels. The contrast-enhanced abdominal tomography (CT) revealed diffuse intraperitoneal fluid that was loculated in some areas, and widespread infiltration of the omentum and mesentery. There was thickening in the bowel wall. The signs were interpreted as peritonitis carcinomatosa or tuberculous peritonitis (Figure 1a, b).

The abdominal magnetic resonance imaging (MRI) findings were similar to that of CT. Additional imaging techniques such as gastroscopy and colonoscopy were not diagnostic either. Therefore, the patient was scheduled for explorative laparotomy with the presumptive diagnosis of tuberculous peritonitis-carcinomatosis peritonei. On exploration, there was diffuse ascites, a fibrous sac was encasing the entire small intestine, and there were dense fibrous adhesions within bowel loops as well as peritoneal implants. Biopsies were taken from the small intestine mesentery and peritoneum, and the operation was terminated. The histological diagnosis of tissue samples was reported as 'idiopathic sclerosing encapsulating peritonitis'. Postoperatively the patient was started on maintenance and corticosteroid therapy. She was discharged on the eighth postoperative day after improvement in her complaints and laboratory values. Steroid therapy was continued for three months. Seven months after surgery the patient's urea and creatine levels began to rise again. Upon deterioration of her renal functions despite medical treatment, she was planned for dialysis. She died on the eight postoperative month due to multiple organ failure. The patient had been informed in detail about the disease, the planned and executed procedures and a consent form had been obtained. Required consents to perform scientific studies were obtained from patients' relatives.

## DISCUSSION

Sclerosing encapsulating peritonitis, which is also called abdominal cocoon, is a rare condition. It usually occurs in the young and adolescent population. Intraperitoneal release of fibrin-like material by fibrogenic cytokines is involved in the pathogenesis (4). Primary and secondary types of SEP have been

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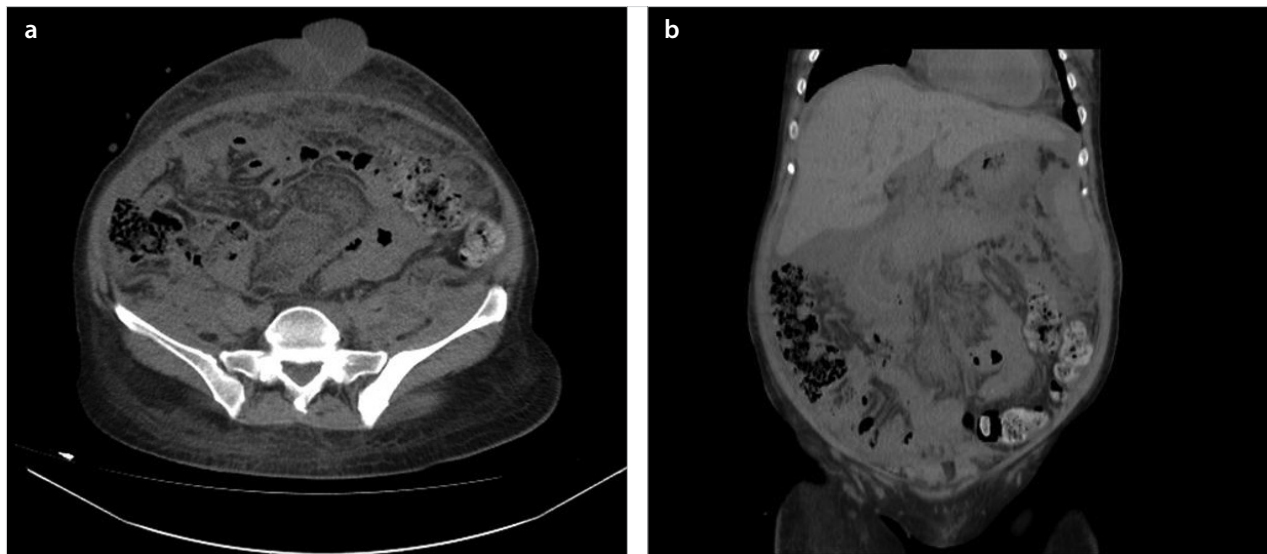


Figure 1a, b. (a) (Axial view), (b) (Coronal view): Diffuse infiltration in the omentum and mesentery, bowel loops with partial wall thickening

described. The etiology of primary disease is unknown, thus it is called idiopathic. Generally, it is more common in young women living in tropical or subtropical regions. Several authors suggested that meconium peritonitis, sarcoidosis, cadaveric liver transplantation, long-term abdominal catheter, and tuberculous peritonitis play a role in the etiology of secondary SEP (5).

Abdominal pain and non-specific clinical signs such as bloating and nausea occur in both types (6). Consequently, the diagnosis and treatment of these patients is delayed. In addition to these findings, patients may also present with complications such as mechanical intestinal obstruction, bleeding or perforation (7). Our patient did not have additional risk factors that may have played a role in the etiology, and she experienced non-specific symptoms such as abdominal pain episodes and weight loss. The reason for admission to the hospital was umbilical hernia, which is thought to be due to diffuse intra-abdominal ascites.

The preoperative diagnosis of SEP is difficult. There are no specific physical examination and laboratory findings suggestive of SEP (8). Dilated bowel loops, thick fibrous membranes and loculated ascites that surround the small intestine are detected on ultrasonography (US). In addition, cauliflower-like intestinal loop, or sacs formed by accordion-like fibrous membranes may be viewed. Computed tomography plays an important role in ruling out other causes that lead to intestinal obstruction, in addition to viewing the band of collagen that may affect other abdominal organs. Additional signs such as obstruction, bowel wall thickening, ascites, localized fluid collections, peritoneal thickening, peritoneal or wall calcifications, and reactive lymphadenopathy may occur (9). Like CT, MRI can also be used for diagnosis. Magnetic resonance imaging may be more sensitive in showing the sclerotic capsule. However, these findings only guide us; they are not specific for a diagnosis. In general, a definitive diagnosis is made during the operation. In our case, CT just guided us and MRI did not

offer any additional information as compared to CT. The definitive diagnosis could only be established with the intraoperative findings and histological diagnosis of peritoneal biopsies taken.

The differential diagnosis includes peritonitis carcinomatosa, pseudomyxoma peritonei, peritoneal mesothelioma, and tuberculous peritonitis according to findings on physical examination, clinical and imaging methods. The presumptive diagnoses were peritonitis carcinomatosa or tuberculous peritonitis in our patient based on CT findings, and screening was performed accordingly.

Medical or surgical treatment can be preferred depending on the patient's clinical condition (10). Bowel rest and nutritional support should be started initially (10). Recently corticosteroids, tamoxifen and immunosuppressive agents are also used in treatment. It is reported that these drugs reduce inflammation and peritoneal fibrosis (10). Surgical intervention should be reserved for a potential complication. If SEP is considered as part of differential diagnosis during surgery, dissecting the membranes and separation of adhesions is recommended with as minimal damage as possible, if it is possible to do so. However, if there are no complications requiring urgent intervention or if there is diffuse involvement then only biopsies should be taken and the operation should be completed. Bowel resection is unnecessary unless there is an ischemic segment. Our patient underwent diagnostic exploratory laparotomy since we were unable to conclude on a definitive diagnosis in the preoperative period. Diffuse fibrotic adhesions and conglomeration of small bowel loops were observed on laparotomy. There were no perforated, ischemic or dilated bowel loops. Therefore, the operation was completed after obtaining samples for diagnostic biopsy. The patient was started on intensive enteral nutritional support and corticosteroids. However, the supportive therapy and corticosteroids did not prevent the patient from progressing to multiple organ failure since the patient was admitted at advanced disease stage.

## CONCLUSION

Sclerosing encapsulating peritonitis is a rare disease with dismal prognosis. It can lead to serious complications such as severe malnutrition, sepsis and death if not promptly diagnosed and treated, as in our patient. The review of the literature suggests that surgery should be avoided if the disease is diagnosed preoperatively, and that aggressive surgery should be avoided if diagnosed peroperatively.

**Informed Consent:** Written informed consent was obtained from patient's parents who participated in this case.

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