An Uncommon Case Report of Abdominal Cocoon

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Abstract

The term “encapsulating peritoneal sclerosis” (EPS) refers to the uncommon and rare cause of intestinal blockage that is caused by abdominal cocoon. Retrograde menstruation-related viral peritonitis is the primary cause of EPS, and it is the reason for the history of peritoneal dialysis in young adolescent girls from tropical and subtropical countries. Most suffer from either stomach pain, which is most likely caused by a subacute intestinal blockage, or no symptoms at all. Results from imaging tests, such as computed tomography (CT), magnetic resonance imaging, and ultrasound, can be ambiguous. When intraabdominal fibroinflammatory processes are detected, the small intestine is partially or fully encased in a thick fibrocollagenous membrane that encircles it like a cocoon. The membrane impairs the movement of the gut, forming a fibrous tissue sheet that covers, fixes, and finally constricts the gut, resulting in symptoms that are commonly associated with an abdominal cocoon.

Keywords: Abdominal cocoon, Encapsulating peritoneal sclerosis, Fibrocolla-genous membrane

Introduction

Encapsulating bowel disease is a very rare but serious condition. It is known that EPS can occur in a wide range of unique ethnic and geographic contexts. EPS is caused by long-term intraabdominal fibroinflammatory processes, and it is typified by the bowel being completely or partially encased in a thick fibrocollagenous membrane that surrounds the small intestine like a cocoon [1]. Fibrous tissue sheets consequently form, covering, stabilizing, and eventually restricting the gut and reducing its motility. Most patients present with either no symptoms at all or with (recurrent) abdominal pain, most likely due to a partial or complete obstruction of the bowel. Most patients have had prior experiences with similar episodes that resolved on their own. The development of complete sclerosis caused by the cocoon formation is a clear sign of mechanical ileus.

The International Society for Peritoneal Dialysis recommends using ESP in cases where common inflammatory processes leading to cocoon formation are caused by peritoneal dialysis. These days, the majority of writers refer to both the secondary form of EPS and its primary, idiopathic form as abdominal cocoonos. Young adolescent girls from tropical and subtropical countries are typically described as having the primary type, which is predominantly idiopathic in nature. Several theories have been proposed for the primary form, including the etiopathogenesis of viral peritonitis, retrograde menstruation with viral infection superimposed, and cell-mediated immune tissue damage caused by gynecological infections. These theories may not completely explain the etiopathogenesis in all patients, despite the fact that this condition can also affect men, women who are not yet menstruating, and children. The secondary form of the condition can also be brought on by other predisposing factors, such as peritoneal dialysis, systemic lupus erythematosus, liver cirrhosis, endometriotic cyst, abdominal trauma/surgery, abdominal tuberculosis, or cancer [2, 3]. The term “peritoneal sclerosis, peritoneal fibrosis, sclerosing peritonitis, sclerotic thickening of the peritoneal membrane, sclerosing obstructive peritonitis, or encapsulating peritonitis” is the most commonly used term for this condition. The EPS and congenital peritoneal encapsulation (CPE) can be easily distinguished from one another. The first mention of CPE dates back to Cleland’s 1868 report [4]. It is caused by an intestinal deformity that happens during embryonic development when the yolk sac retracts into the abdominal cavity, resulting in the creation of an auxiliary membrane that covers the colon. This should be differentiated from EPS, which is much more prevalent than CPE and was initially referred to as “peritonitis chronica fibrosa incapsulata” by Owtschinnikow [5] in 1907. When an established cause could not be found for primary or idiopathic EPS, Foo et al. used the term “abdominal cocoon” [6].

The abdomen is often palpably soft in clinical settings. A soft, nontender mass that feels like clumped-up bowel loops may be palpable in the middle of the abdomen. Imaging results from CT, ultrasound, and magnetic resonance imaging are often nonspecific [7]. While they have been discussed, other imaging modalities such as barium studies or positron emission tomography have not demonstrated any appreciable advantage in the diagnosis of EPS. Consequently, it often takes surgery to diagnose this illness, which is subsequently verified by histological examination [8].

It is clear that EPS and CPE have different histologies. The thin membrane on the visceral peritoneum aids in the formation of the intestinal encapsulation of EPS. Histologically, the majority of the membrane is composed of organized fibrin, possibly derived from plasma exudation, within the peritoneal microvasculature. Peritoneal
fibroblasts not only have an enlarged and more cellular appearance, but they also express several markers linked to activation and proliferation. Persistent inflammatory changes also predict the onset of EPS [9].

**Case Report**

The patient came to op with chief complaints of vomiting and abdominal pain for 8 months.

**Present history**

A 30-year-old gentleman was apparently alright 8 months back. Then he started complaints of vomiting after eating food with food as content. Associated with colicky type of pain abdomen after eating food. H/O constipation and straining while passing stools. No h/o cough, fever. No h/o diarrhea. No H/o Pulm Kochs in the past

**Past history**

No comorbidities

**On examination**

PR: 84/min; BP: 110/80 mmHg; Temp: 98.20 °F; RR: 18/min SpO2 : 98%.

**Diagnosis**

Sub-acute intestinal obstructions and abdominal cocoon.

**Surgery**

Diagnostic laparoscopy. Converted to exploratory laparotomy in view of abdominal cocoon with adhesiolysis.

**Intraoperative findings**

Abdominal cocoon - entire small bowel and large bowel within abdominal cocoon. Small bowel adhesions - multiple noted, small bowel within abdominal cocoon (Figure 1). Appendix normal, appendectomy done. Small + large bowel appears normal, minimal free fluid in peritoneal cavity. No peritoneal and omental nodule seen.

Post operatively managed with IV fluids, analgesics, antibiotics, and other conservative management methods. On POD 3, the patient started on liquid diet trial feed. The patient developed distension with vomiting on POD 4. Patient was kept on NBM...Started IV fluids. On POD 11, the patient was on liquid diet there were no complaints of vomiting or distension. Started on a soft diet. Patient improved symptomatically tolerating orally and is being discharged under stable conditions. Biopsy - negative for granuloma. Features suggestive of chronic inflammation and fibrosis.

**Condition at discharge**

Hemodynamically stable. Wound healthy.

**Discharge advice**

Tab. Acef 500 mg twice daily for 1 week. Tab. Esotrend-d daily twice daily before food for 1 week.Tab. Dolo 650 mg SOS if pain. Tab. Zofer 4 mg SOS if vomiting's. Tab Lesuride 25 ma twice daily for 2 days. Sup Gaviscon 15 ml thrice daily before food for 1 week. Syp. Cremaffin 20 ml per oral at night 1 h before sleep for 1 week. Abdominal binder daily application. Steam inhalations daily. Tri- ball spirometery daily.

**Discussion**

A thickened, grey-white membrane made of collagen and fibrous tissue resembles a cocoon and envelops the viscera of abdomen in Primary Sclerosing Encapsulating Peritonitis (SEP), a rare disease phenomenon [10]. The external sign of a mildly inflammatory illness called idiopathic or secondary SEP, also referred to as abdominal cocoon, is intestinal obstruction [11]. Without a known cause, SEP is believed to be a chronic inflammatory illness. Without any noticeable abdominal symptoms, it begins as recurrent subclinical or low-grade peritonitis, develops into sclerosis and membrane formation, and then forms cocoons. In [12] What sets this organ apart is the thick fibrocollagenous membrane that covers the small intestine and, occasionally, other intra-abdominal organs like the stomach, colon, and liver. It is possible to categorize the organs into three types based on the extent of encapsulation: type I, in which the membrane only covers a part of the intestine; type II, in which the membrane covers the entire intestine; and type III, in which the membrane covers the entire intestine along with extra organs (like the appendix, cecum, ascending colon, ovaries, etc.) [13].

Depending on the underlying cause, SEP is classified as primary (idiopathic) or secondary (acquired). Although fibroblasts and cytokines are important in the development of peritoneal fibrosis and neoangiogenesis, there is no evidence of an underlying cause for primary SEP. Within tropical and subtropical regions, where there is a 2:1 male-to-female population predisposition, the prevalence of idiopathic SEP is higher. The pathophysiology of Secondary SEP is now more fully understood [14]. Idiopathic SEP has been linked to a wide range of possible causes, including retrograde menstruation, retrograde peritonitis, omentum hypoplasia, and mesenteric vascular defect. Danford et al. claim that due to SEP’s rarity and the unpredictability of its etiologies, its overall incidence and prevalence are unknown [15]. Idiopathic SEP is less common than secondary SEP. The peritoneum becomes inflamed in secondary SEP as a result of a systemic or localized cause. Many factors can lead to peritoneal inflammation, and the most common cause in peritoneal dialysis patients are b-blockers, sarcoidosis, tuberculosis, ovarian tecomas, organ transplantation, and cirrhosis [14].

Intestinal obstruction is the presenting symptom of ACS due to the intestine’s kinking and compression within the enclosing membrane. Its symptoms, which include nausea, anorexia, vomiting, constipation, acute, subacute, or chronic abdominal pain, are therefore typical of ileus. An additional piece of evidence supporting the diagnosis is a history of similar symptoms that have previously gone away on their own. In the largest case series of idiopathic SEP, the majority of patients (75%; mean body mass index, 17.5 kg/m2) were malnourished, and the average duration of symptoms before presenting was 3.9 years. 29% of patients required “emergency surgery” at the time of initial presentation, despite the disease being subtle and chronic. This suggests that a significant number of patients have more severe obstruction, ischemia, or even perforation. In some circumstances, ascites and a palpable mass may be felt. This modality typically has no side effects. Nearly half of the patients received their diagnosis while undergoing surgical procedure, according to the data from a large case series which was conducted in China [15] and India [16], with 16.7 - 48.7% of patients receiving their diagnosis earlier. Nakamoto’s proposal divides the development of EPS into four phases, taking pathological and clinical findings into account.
The pre-EPS stage is characterized by ultrafiltration failure and/or changed solute transport status. There are subsequent phases that are obstructive, encapsulating, and inflammatory; the last phase is marked by intestinal blockage and the formation of an "abdominal cocoon." The literature pertaining to EPS associated with peritoneal catheterization has suggested a staging system that integrates radiographic, laboratory, and clinical findings. Nakamoto [17] classified patients with EPS into four stages: Stage 1 (pre-EPS), Stage 2 (inflammatory), Stage 3 (encapsulating), and Stage 4 (chronic), based on intestinal findings, inflammation, encapsulation, and abdominal symptoms. Depending on the disease's stage, a variety of conservative treatment strategies have been suggested, including nutritional support, immunosuppression—corticosteroids being the most well-researched—colchicine, azathioprine, mycophenolate mofetil, cyclosporine, and mammalian target of rapamycin. Moreover, antibiotics like tamofoxen can boost the effectiveness of medications that suppress the immune system. The CECT scan appears to be the investigation of choice for SEP, even if there isn't complete consensus on the optimal diagnostic and treatment strategy because the majority of papers reported are some case reports, and the available systematic review is from the year 2015. The CT scan's principal findings include ascitic fluid between bowel loops, adhesions, which provide the surgical team with information regarding the length and complexity of the procedure, and matted, dilated intestine segments with a thick peritoneal membrane (>2 mm). The smudged appearance of the large omentum, focal (or/and) diffuse calcifications on the membrane (or/and) lymph nodes, and Bottle Gourd sign [18-20], which is the dilated part of second and third section of the duodenum with encasing the distal duodenum and proximal part of the jejunum, are among the other findings observed. Reports of lymphadenopathy have been made, mainly as a secondary cause in cases of tuberculosis in the abdomen. When identifying complications like intestinal ischemia, or strangling, which is characterized by a lack of gut enhancement, a contrast-enhanced CT scan can be very helpful. Other grading schemes that are dependent on the CT results exist, but they are more appropriate for academic applications as they have no relationship to the outcome [8].

Additionally, a cauliflower sign on small intestine barium exams may hint at the diagnosis [21]. An abdominal erect X-ray may reveal a dilated bowel loop with multiple air-fluid levels; these findings are not specific to SEP but are consistent with bowel obstruction. Having said that, considering that SEP is a rare disease, the majority of the cases, which present in young population, are diagnosed during surgeries which are performed after failing to improve symptoms of the bowel obstruction by conservative management. A combination of all three evaluating modalities of clinical accumen, radiological evaluation, surgical exploration, and histo-pathological findings is the best way to have acertain diagnosis. The standard course of the treatment involves laparoscopic or open approach excision of the membrane and removal of adhesions. The age range of the patients was 10 to 18 years, and only one male instance was reported. Adolescent females made up the majority of the patients (80%). This may provide credence to a thin additional peritoneal sac enveloping the small bowel. It is usually asymptomatic, whereas abdominal cocoon is characterized by a thick fibrous membrane that causes intestinal obstruction [23]. Intestinal malrotation, voluminous invagination, and other causes of peritoneal adhesion are less common conditions to consider as differential diagnoses [24]. It is necessary to fully rule out tuberculous peritonitis in areas where tuberculosis is prevalent [25]. Since idiopathic SEP is an uncommon condition, most doctors either never see patients with it or fail to diagnose it when they do. It is very difficult to make an accurate preoperative diagnosis in affected patients and calls for a high index of clinical suspicion. Because the early clinical manifestations of SEP lack specificity, preoperative diagnosis is difficult. A high index of clinical suspicion is required [26]. Imaging techniques are essential for the diagnosis of SEP, in addition to the clinical presentation and patient history. Dilated small intestine loops and diffuse air-fluid levels can be seen on abdominal X-rays. The gathered and conglomerated bowel loops observed in the center of the abdomen during a small intestine barium transit may be indicative of the so-called “cauliflower sign” [27]. Extended travel time confirms the diagnosis as well. Abdominal ultrasonography can show free abdominal fluid, a thicker peritoneal layer, and dilated bowel segments surrounded by a thick fibrous membrane [28]. On CT, the most helpful modality for verifying the diagnosis, the appearance of small intestine segments conglomerated at the midline and encircled by a thick capsule with a contrast-free periphery is a characteristic sign [29]. Evaluations of peritoneal thickening are usually subjective, but a thickness larger than 2 mm seems to be a reasonable place to start, even though there is no perfect cutoff point. Other radiographic features include diffuse or localized peritoneal calcification, elevated mesenteric fat density, and located ascites [8]. Even though complex loculations may be a sign of intra-abdominal bleeding, they should raise suspicions of sepsis or perforation, particularly if they contain gas [30].

It is not currently known of any biomarker that can be used to anticipate the occurrence of SEP. Infection, starvation, and inflammation are linked to the nonspecific laboratory results from SEP [31]. It was found that the dialysate of SEP patients had higher levels of inflammatory cytokines than that of peritoneal dialysis controls, even years before the clinical development of SEP [32]. The vague histologic features of SEP can be confused with those of simple peritoneal sclerosis or infectious peritonitis. By binding inflammatory cytokines, a transmembrane glycoprotein called podoplanin, which is present in peritoneal mesothelial cells, aids in the differentiation of EPS from peritoneal sclerosis and peritonitis [33]. The histopathological examination, which makes the final diagnosis, shows that the peritoneum histologically shows fibroconnective tissue growth, inflammatory infiltrates, and dilated lymphatics without any evidence of giant cells, birefringent material, or foreign body granulomas [34].

Conservative care is the best course of action for those with mild symptoms. This includes enteral or parenteral nutritional support, bowel rest, and nasogastric decompression. Treating nutritional disorders is an important part of treatment for many patients with recurrent abdominal complaints. Improving the nutritional status of these patients is critical as it may prevent surgical complications like fistulae and infection or improve their response to conservative care. If conservative treatment is not successful, patients may be treated
with drugs such as tamoxifen, steroids, colchicine, azathioprine, and mycophenolic acid.

Tamoxifen plus steroids may be helpful in the prevention and/or treatment of SEP according to human studies. The exact pharmacological mechanism by which corticosteroids act on SEP is unknown. However, the immunosuppressive and anti-inflammatory effects of the drug may be to blame. The concept of using tamoxifen to treat SEP in its early stages is particularly intriguing. Patients with long-term Parkinson’s disease who have ultrafiltration failure with a high transport status but do not have the characteristic SEP imaging symptoms may find this useful. Moreover, these medications may be beneficial for individuals whose postoperative symptoms are still bothersome.

Surgery is only recommended for patients who have failed conservative, medical therapy and, if possible, in centers with expertise in such operations due to the time-consuming, technically complex, and dangerous nature of surgical techniques for SEP. Surgery can be done to cure a specific complication, like enterolysis (ablation of fibrous tissue and lysis of adhesions), or it can be done to address a specific complication, like limited lysis of adhesions or ablation of perforated or ischemic bowel. A laparoscopy or laparotomy should be performed on patients who demonstrate severe intestinal obstruction or who are diagnosed with SEP during surgery.

In cases of gastrointestinal injury, membrane excision plus anastomosis—either with or without a protective enterostomy—is the standard procedure. Peritoneal deterioration [12], which has been demonstrated to get worse with the course of Parkinson’s disease, particularly in cases that have lasted longer than ten years, is one of the most significant predictors of postsurgical outcomes. Because of the poorly defined intestinal wall and capsules in these patients, improper enterolysis can easily lead to intestinal perforation. If there is peritoneal calcification around capillaries, there may be a higher chance of perforation during excision. If the intestinal surface membrane can be entirely removed, the risk of recurrence is extremely low [35].

Antiadhesive compounds placed between the bowel loops before closing the abdomen after total excision of the membrane may lower the risk of postoperative adhesions; however, the practicality of these materials for patients with partially excised membranes remains questionable. Improving secondary SEP cases requires treating the underlying cause. Therefore, the surgical procedure and histopathological examination of the peritoneum serve as the foundation for both diagnosis and treatment in cases of idiopathic SEP.

In cases of SEP, intestinal transplantation and intestinal rehabilitation programs have spared patients from the complications associated with total parenteral nutrition.

Conclusion

The thick fibrocollagenous membrane that surrounds the small bowel is a sign of the inflammatory illness ACS. It is a rare cause of small bowel obstruction and is categorized as an idiopathic or secondary syndrome. When making a differential diagnosis for small bowel obstruction, it is important to consider this, especially if the obstruction has repeatedly occurred. Laparoscopy or laparotomy is still the gold standard for diagnosing SEP, despite the growing popularity of CT in preoperative SEP diagnosis. Surgery seems to be the most beneficial therapeutic option in cases of severe SEP or when conservative treatment is unsuccessful.

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Conflict of Interest

None.

References


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