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Case report

Primary Sclerosing Encapsulating Peritonitis in an Adult Male: A Case Report

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Abstract

Introduction: Primary sclerosing encapsulating peritonitis (PSEP), also referred to as "cocoon abdomen", is a rare but potentially devastating disease. It is defined as the encapsulation of a segment of the small bowel by a fibro-collagenous membrane leading to acute or sub-acute small bowel obstruction. Though the exact etiology and pathophysiology of this debilitating medical condition are still controversial, PSEP has been reported to be associated with multiple congenital developmental abnormalities. Diagnosis of PSEP is usually delayed as the disease is relatively rare and commonly present with non-specific symptoms associated with non-specific laboratory findings and non-conclusive radiologic features. ***Patient Concerns:*** We present a 42-year-old male patient presenting for elective abdominal laparoscopy for his four-year recurrent abdominal pain. ***Diagnosis:*** Preoperative diagnosis of PSEP as the cause of his chronic abdominal pain was not made until at laparotomy where a thick fibrotic peritoneal sac encasing the entire small bowel was found. ***Interventions:*** Excision of the fibrotic sac with extensive adhesiolysis were done by the general surgeon. ***Outcomes:*** The patient experienced a ten-day period of post op ileus after which he had a complete recovery. ***Conclusion:*** With the increasing number of cases suffering from this condition being reported in literature, surgeons must be aware of this debilitating disease in the differential diagnosis of abdominal obstruction.

Keywords: ***Encapsulating peritoneal sclerosis, cocoon abdomen, small bowel obstruction, enterolysis.***

Introduction

Primary sclerosing encapsulating peritonitis (PSEP), or the so-called "Abdominal Cocoon" or idiopathic encapsulating peritonitis is a rare disease characterized by the encapsulation of either a segment or the entire small bowel by a thick grayish-white fibro-inflammatory collagenous sac leading to a clinical picture of acute or subacute intestinal obstruction [1-9]. Encapsulation may extend into other organs of the abdominal cavity including the large bowel, liver, and stomach [7,10]. SEP was first termed by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata" and later was divided by Foo in 1978 into primary (Idiopathic) and secondary, the former of which was described as "abdominal cocoon" [1-12]. In 1978, this medical condition was described as a disease closely linked to peritoneal dialysis and was later defined by the International Society for Peritoneal Dialysis as "a syndrome continuously, intermittently, or repeatedly presenting with symptoms of intestinal obstruction caused by adhesions of a diffusely thickened peritoneum" [2]. Though incidence is rare (0.9% to 7.3%), SEP is considered a debilitating syndrome given the associated high mortality rate that ranges between 43.5% and 78% [3]. Idiopathic SEP is mostly reported from tropical and subtropical areas with a mean age of 34.7 and a male: female ratio of 2:1 [3]. Congenital developmental abnormalities such as absence or pronounced shortening of greater omentum, uterine and adnexal dysplasia, cryptorchidism, congenital small bowel or colon malrotation and others have been reported to be associated with PSEP [13]. On the other hand, secondary SEP is linked to chronic peritoneal dialysis, intra-abdomen foreign body stimulation, recurrent peritonitis, autoimmune inflammatory diseases, administration of the β-blocker practolol, bacteria proof filter use, liver transplantation, tuberculous inflammation, ventriculoperitoneal and peritoneovenous shunts and carcinoid syndrome [10]. The majority of patients presenting with this condition present with a clinical picture of abdominal obstruction manifested mainly by long-standing abdominal pain and they are usually diagnosed intra-operatively [5]. Radiological studies, especially CT scan, is considered highly valuable as it can facilitate pre-operative diagnosis [10].

Case Report

A 42-year-old male patient with no known comorbidities and no significant surgical and family history was admitted to the hospital for an elective abdominal laparoscopic exploration to define the reason for his on and off symptoms of more than four years duration presenting as abdominal pain and intermittent bloating. Gastroscopy and colonoscopy done before admission were normal. On admission, the patient was afebrile and hemodynamically stable. The physical exam was unremarkable with no signs of peritonitis. Bowel sounds were normal. Initial laboratory investigations were unremarkable. Four years prior to presentation, Magnetic Resonance (MR) Enterography revealed a 150mm dilated small bowel loop in the mid pelvic cavity showing mild and diffuse mural thickening associated with fat accumulation (creeping) and fibro-fatty proliferation of the adjacent mesenteric fat. Three consecutive MR Enterographies performed after several months (one after three months and two after seven months) revealed a regression of the previously described mural thickening with persistence of a mild dilation of the jejunal loops and fibro-fatty proliferation of the adjacent mesenteric fat with minimal stranding and evidence of collapsed loops, hence concerning for either an inflammatory bowel process (Crohn's Disease was considered first) or an internal supra-vesical hernia. No imaging evidence of bowel ischemia was noted. With the persistence of symptoms despite administering medical treatment for Crohn's disease, the patient was finally referred for elective laparoscopy. Intra-operatively, an encapsulation of the entire small bowel in a thick layer of peritoneum was observed along with thickening of parietal peritoneum, and since excision of the capsule with extensive adhesiolysis was not possible by laparoscopy, the latter was converted into midline laparotomy. Full running of the small bowel showed no Crohn's disease or segmental stenosis. Ablation of the visceral peritoneum and adhesiolysis were subsequently done along with decompression of the dilated bowel loops. The excised sac was sent for pathology. The patient experienced a delayed post-operative ileus for ten days during which he was conservatively managed until full recovery. Follow-up after 5 years shows good general status and quality of life.

Discussion

Primary sclerosing encapsulating peritonitis (PSEP) is a rare clinical condition of unknown etiology and is characterized by a partial or total encasement of the small bowel by a thick grayish-white fibro-collagenous membrane [7,13]. The etiology of PSEP is still controversial with multiple hypothesis being formulated including retrograde menstruation with a superimposed viral infection, retrograde peritonitis, and cell mediated immunological tissue damage triggered by gynecological infection. However, since this condition has also been reported in males, premenopausal women and children, there seems to be little support for these theories [6,7,9]. This entity must be distinguished from peritoneal encapsulation, an incidental finding probably embryogenic in origin and is characterized by the encapsulation of the bowel by a secondary normal peritoneal layer derived from the yolk sac and attached to the ascending, transverse, and descending colon [5]. Patients diagnosed with this entity may have other associated mesenteric abnormalities and usually present with no or mild abdominal pain unlike the abdominal cocoon which often presents as acute or subacute small bowel obstruction manifested clinically as acute abdominal pain, nausea, vomiting, abdominal distention, weight loss, palpable abdominal or pelvic mass, and disappearance of bowel tone [5,10]. A study conducted by Yu et al on nine patients with abdominal cocoon in the Second Affiliated Hospital of Soochow University between January 1991 and January 2018 showed that 100% of patients presented with acute abdominal pain and that approximately two-third of them had a partial or complete abdominal obstruction that required surgical intervention [10]. Despite its chronic nature, it is not uncommon for patients with SEP to present with acute obstruction, perforation, or ischemia [1].

In relation with the extent of encapsulation, SEP is divided into three types: Type 1- partial encapsulation of the small bowel, Type 2- complete encapsulation of the small bowel, and Type 3- encapsulation of the small bowel with other organs such as the colon, appendix, stomach, etc [7].

As we mentioned above, SEP is divided into primary (Idiopathic) or secondary forms. Literature reports an association between the primary (Idiopathic) form and congenital dysplasia [10] whereas the secondary form has been linked to infection, medications, organ transplantation, endometriosis, luteinizing gynecologic neoplasms, dermoid cyst rupture, chemical or mechanical peritoneal irritants, and systemic inflammatory or rheumatologic conditions [2]. All these factors can lead to peritonitis and trigger the release of pro-inflammatory and pro-angiogenic cytokines, the most important of which is transforming growth factor β1 (TGFβ1) responsible for the trans differentiation of peritoneal mesothelial cells to mesenchymal cells which in turn activates profibrotic genes expression mainly COL1A1 leading to fibrogenesis and extracellular matrix deposition [2]. Microscopically, peritoneal mesothelial cell layer in SEP patients is denuded by proliferating fibroblasts residing in a fibro-collagenous background rich in fibrin and harboring a mononuclear inflammatory infiltrate [11]. These features overlap between simple peritoneal sclerosis and SEP, the latter being distinguished by the presence of podoplanin (in a diffuse pattern) and smooth muscle actin double positive cells having a fibroblastic appearance located in the prominent fibrous tissue [14].

The diagnosis of SEP is mainly clinical and is confirmed radiographically or by surgery, with the majority of patients presenting with non-specific symptoms of abdominal obstruction [2]. The largest case series of idiopathic SEP states that the average duration of symptoms was 3.9 years prior to presentation and states that 52.3% to 100.0% of admitted patients were diagnosed intra-operatively and only 16.7% to 48.7% were diagnosed during their preoperative workup [1]. Comparably, our patient presented with a four-year duration of non-specific abdominal symptoms and was also diagnosed intra-operatively.

Laboratory speaking, literature confirms that no biomarker can predict SEP development [15]. Laboratory findings such as leukocytosis (i.e., elevated WBC count), elevated C-reactive protein level, hypoalbuminemia and anemia may be detected in patients with PSEP [16].

As we mentioned above, imaging helps confirm the diagnosis of SEP. Erect abdominal X-rays can display features of abdominal obstruction such as largely dilated small bowel loops with multiple air-fluid levels. However, these features may sometimes be absent and are non-specific for PSEP. Transabdominal ultrasound can show dilated intestinal loops enclosed within a membrane, peritoneal thickening and ascites. Barium meal films can feature the characteristic serpentine or cauliflower appearance of the distal dilated bowel loops [12, 13]. So far, CT scan has been considered the imaging modality of choice as it can give the whole image of the entity while excluding all other conditions that can mimic it clinically. The definitive radiologic feature of SEP is the centrally congregated small bowel loops encased by a soft tissue density mantle that cannot contrast enhance. Other features that can aid in diagnosis include localized or diffuse peritoneal calcifications, peritoneal thickening, peritoneal enhancement, loculated ascites, reactive lymphadenopathy, serosal intestinal wall calcification, thickening of the bowel wall, tethering of intestinal loops, and proximally dilated bowel loops [5,6,7,12,17]. If the latter finding is associated with transmural enhancement or thickening, it is indicative of active inflammation or transition to transmural fibrosis [18].

The management of SEP is controversial and differs somehow between the primary and the secondary types. For the idiopathic (primary) type, surgical treatment is warranted as by most

authors [7]. In fact, surgery carries both a diagnostic and a therapeutic approach for such cases as diagnosis is often delayed until laparotomy in many of the patients [7]. During surgery, a grossly white fibrotic sac or membrane is found to encapsulate the bowel loops which, upon sectioning, present many loculated abscesses due to local perforations [7]. Surgical modalities vary from those aimed to cure, such as enterolysis (ablation of fibrotic tissue and lysis of adhesions), to those intended at targeting a specific complication such as limited adhesiolysis to free the bowel or resection of perforated or ischemic bowel. Though less time consuming, the latter techniques are less preferred due to their associated higher frequency of symptom recurrence. Enterolysis is generally preferred but still remains technically difficult and carries a risk of bowel injury [19-21]. In asymptomatic SEP, surgical treatment is not required. Some of the complications reported after surgical intervention include intra-abdominal infections, perforation, and entero-cutaneous fistula [22].

Finally, regarding secondary SEP, treatment relies on targeting the underlying condition: For example, in Peritoneal dialysis associated SEP, the treatment involves substituting peritoneal dialysis by hemodialysis [23]. If SEP is drug-induced, withdrawal of the medication is recommended. The same applies to SEP induced by infectious or inflammatory etiologies. However, in most of the preceding conditions, SEP is unlikely to resolve given its chronic and fibrotic nature hence necessitating treatment for the ongoing inflammation and fibrosis [2]. In general, treatment modalities include nutritional support, immunosuppression, anti-fibrotics, and surgery [2]. Among immunosuppressants, corticosteroids are the most studied, but the evidence of their use is limited to observational studies with different formulations, duration, dosing, and results. Surely, the use of steroids cannot be initiated unless infection is ruled out. The same applies to the use of anti-fibrotics such as Tamoxifen, a selective estrogen receptor modulator (SERM) which anti-fibrotic activity lies in its ability of inhibiting TGF-β, an important regulator in the mechanism of fibrosis [24]. Its use is confined to observational studies in patients on peritoneal dialysis.

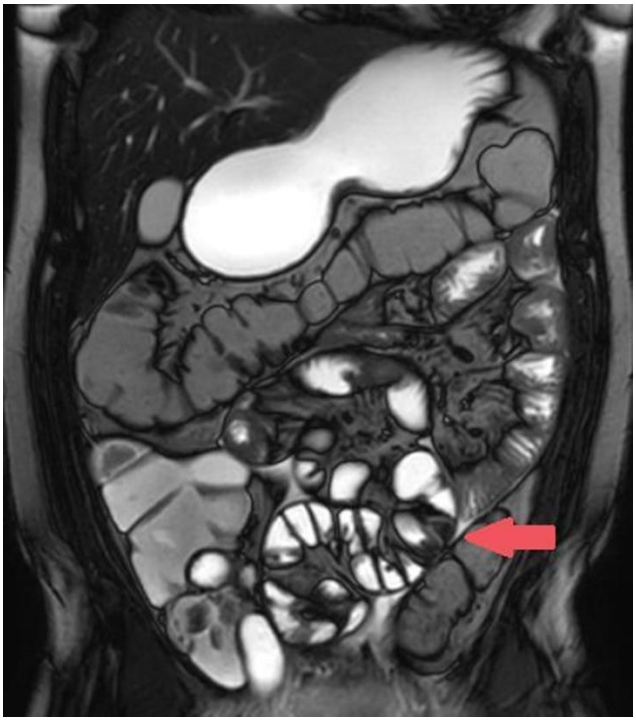


Figure 1: The red arrow indicates a thin layer of peritoneum surrounding the small bowel and possibly causing partial obstruction and adhesions.

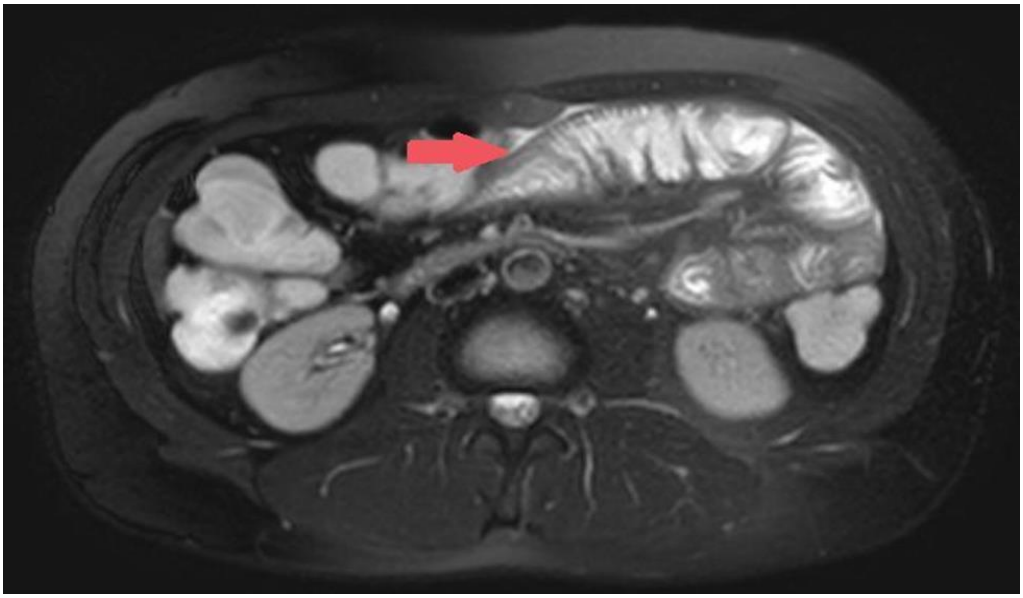


Figure 2: The red arrow indicates a narrowed small bowel loop with edema and surrounding free fluid.

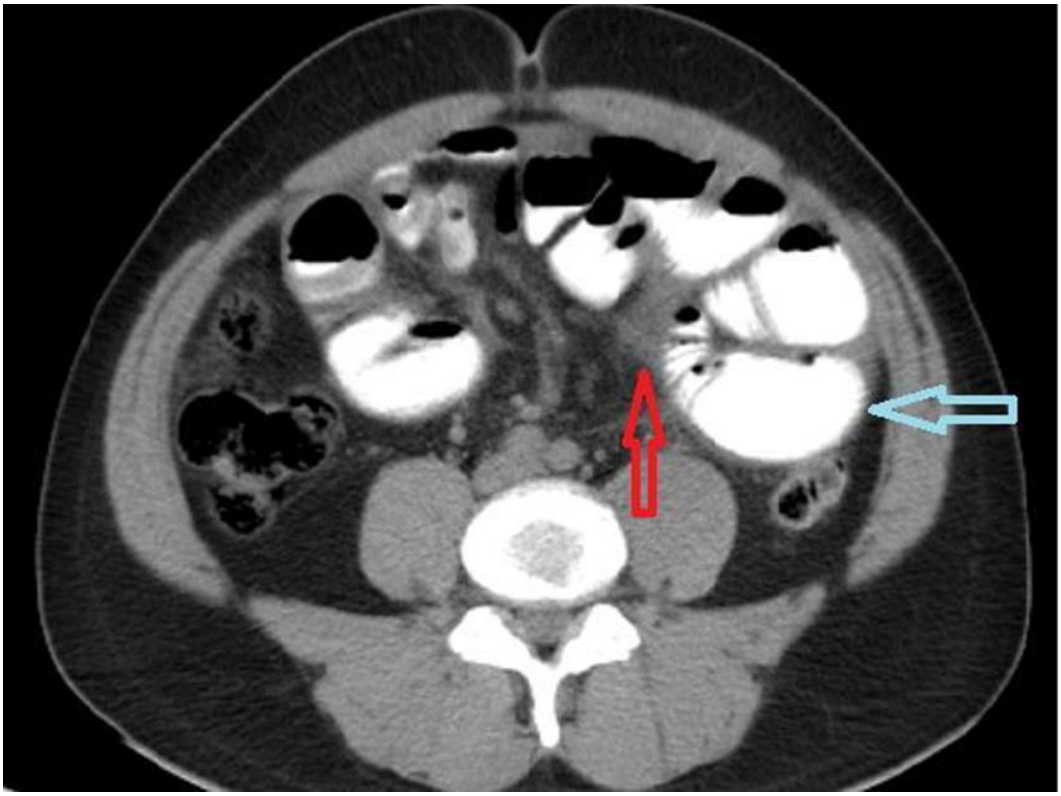
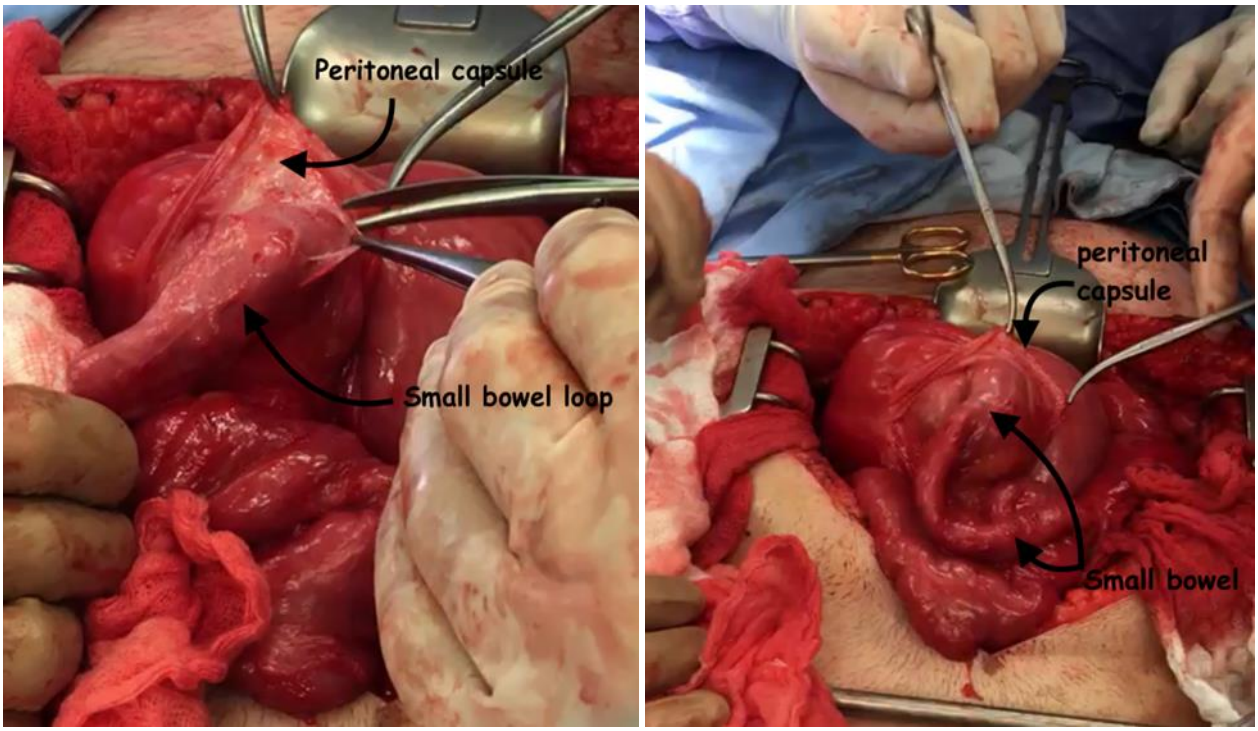


Figure 3: The red arrow indicates fat stranding, and the blue arrow indicates distended small bowel loop.



Pictures 1,2: Encapsulating Peritonitis.

Conclusion

Primary sclerosing encapsulating peritonitis (PSEP) is a rare but serious medical condition that necessitates a high index of suspicion using a constellation of clinical and radiologic findings. Though our understanding of this entity has increased in the recent decades and efforts have been made to increase the physicians' awareness of its early diagnosis, the latter is often delayed till the intra-operative stage upon visualization of the encapsulating fibrotic sac which is adherent to underlying bowel loops. Improving the resolution of CT imaging in the future will help clinch the diagnosis preoperatively.

Ethical statement

Ethics approval was not required for this case report.

Abbreviations

PSEP: Primary Sclerosing Encapsulating Peritonitis
SEP: Sclerosing Encapsulating Peritonitis
CT: Computed Tomography
MR: Magnetic Resonance
TGFβ: Transforming Growth Factor
COL1A1: Collagen, type I, alpha 1 gene

WBC: White Blood Cells
SERM: Selective Estrogen Receptor Modulator

Author contributions

Alaa Haydar contributed in data collection and analysis. Hussein Kaouk, Alaa Haydar, Marita Saliba and Reina Ibrahim contributed in performing the literature search, review, and drafting of the manuscript. Zeina Sawaya and Claude Tayar supervised the article's redaction.

Conflict-of-interest statement:

There are no financial disclosures or conflicts of interest in the production of this manuscript.

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