

Idiopathic Peritoneal Sclerosis: Case Presentation, And Literature Review

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1. Abstract

1.1. Background: Most of the literature regarding peritoneal sclerosis is derived from nephrology literature surrounding peritoneal dialysis as the main and primary cause of this very rare and devastating disorder. The primary aim of this abstract is to encounter a case presentation of idiopathic peritoneal sclerosis and elaborate further on this rare condition.

1.2. Case Presentation: 54 years old male patient presented with 9 months' history of intermittent progressive vomiting and significant weight loss in 2014. Clinically his abdomen was soft not tender but distended. His blood tests showed elevated inflammatory markers. CT abdomen and pelvis showed sub-acute distal small bowel obstruction. He didn't respond to conservative management. He underwent a laparotomy that revealed a frozen, matted small bowel loops which were thickened and leathery feeling. All structures at the root of the mesentery were covered by a thick sheet of fibrous tissue including the mesentery.

Peritoneal biopsies were taken for histology and microbiology. The report of which was consistent with encapsulating peritoneal sclerosis. Notably, it was negative for TB.

Patient was referred to tertiary center for further management and was discussed internationally with peritoneal sclerosis expertise. He recovered on conservative measure and tapering doses of steroids.

He is still experiencing recurrent small bowel obstruction which usually resolve using steroids and conservative measures.

1.3. Conclusion: Idiopathic Peritoneal Sclerosis is a rare condi-

tion with high mortality and morbidity. It poses a very challenging management dilemma.

2. Introduction

Most of the literature regarding Encapsulated Peritoneal Sclerosis (EPS) is derived from nephrology literature surrounding peritoneal dialysis as the main and primary cause of this very rare and devastating disorder [1]. The primary aim of this abstract is to present a case of idiopathic peritoneal sclerosis and elaborate further on this rare condition with exceptionally low incidence.

3. Case Presentation

54 years old male patient with background medical history of traumatic brain injury and alcoholic liver disease, presented with 9months history of intermittent progressive vomiting, constipation and weight loss in 2014.

Clinically, his abdominal examination was unremarkable.

Blood investigations showed elevated inflammatory markers.

CT abdomen and pelvis was reported as subacute distal small bowel obstruction (Figure 2).

He didn't respond to conservative management.

Subsequently, He underwent an exploratory laparotomy that revealed a cocooned matted small bowel loops resulting in a frozen abdomen. The small bowel was thickened and had a leathery feel. All structures at the root of the mesentery were covered by a thick sheet of fibrous tissue including the mesentery (Figure 1).

Peritoneal specimens were taken for microbiology and histology, report of the latter was consistent with encapsulating peritoneal sclerosis. Notably, it was negative for TB (Figure 3).



Figure 1: During laparotomy of a patient with EPS [2]. Notice the cocoon appearance of the small bowel with leathery looking.

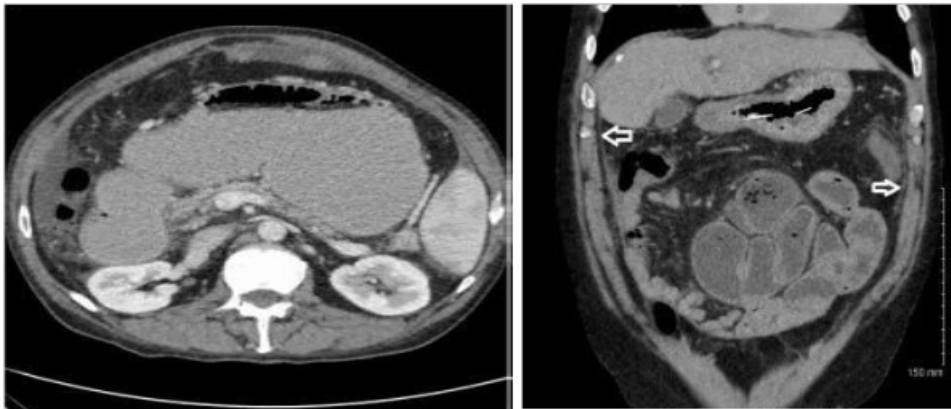


Figure 2: The CT showing the thickened peritoneum (arrows) in both coronal and axial cuts and the cocooned small bowel in the coronal cut)

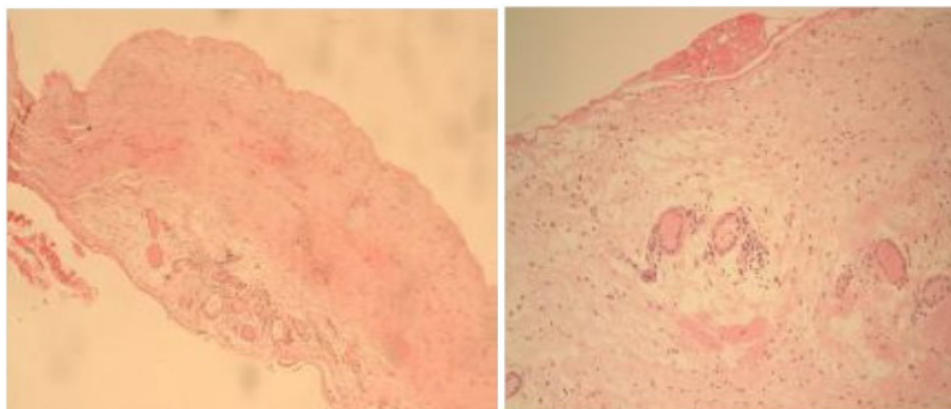


Figure 3: The histopathology showed thickened peritoneum with dense hyalinisation in a laminated pattern associated with mild vascular congestion and patch mononuclear inflammation consistent with EPS

He had a prolonged post-operative course. He showed a good response to reducing doses of steroids. He was discharged home with dietary advice and symptomatic treatment.

He had a stationary course of the disease for 5 successive years but recently, he presented with recurrent attacks of subacute small bowel obstruction which resolved with conservative management measures and reducing doses of steroids. He is being followed up in our outpatient clinic.

4. Discussion

EPS is a rare syndrome, first defined by Foo et al. in 1978 [3]. It mostly affects the small bowel. In this syndrome, there is intra-abdominal fibro-sclerosis and peritoneal adhesions that surround the bowel, creating as a cocoon that causes acute or chronic intestinal obstruction [3].

EPS can be divided into primary and secondary. Primary EPS is also defined as idiopathic (also called abdominal cocoon), and the cause is still unknown. Secondary EPS is related to many conditions; the most common is continuous ambulatory peritoneal dialysis (CAPD) (4). About 1% of CAPD patients may be inflicted with SEP, with the risk increasing with years of peritoneal dialysis [5]. Failure of ultrafiltration, loss of weight, and symptoms of recurrent intestinal obstruction should prompt the exclusion of SEP [6].

The pre-eminent clinical signs of this disease are abdominal pain, nausea, vomiting, occasional constipation. Malnutrition and anemia are evident in longstanding cases [4].

The radiological diagnosis preoperatively remains challenging as this is a rare condition and radiologists may not have this condition in their differential diagnosis. In a high percentage of cases, the diagnosis is reached at the time of procedure [4].

The radiological diagnosis workup comprises abdominal Xray, showing bowel distention with fluid level. The sonographic features may show the small bowel loops encased in a thick membrane, made visible by minimal ascites. The small bowel loops are shown within the sac, arranged in a concertina shape with a narrow posterior base [7].

CT scan will facilitate the diagnosis upon visualization of the characteristic encasement of variable length of the small bowel loops in the central part of the abdomen with a membrane like sac, causing small bowel obstruction [8, 9]. Other findings are nonspecific and include peritoneal thickening, peritoneal calcification, fixation of intestinal loops, free fluid, and reactive lymphadenopathy [8].

Differential diagnosis includes the causes of small bowel obstruction, particularly adhesions and internal hernias, especially trans-mesenteric and Para duodenal, when differentiation may be challenging [10]. Extrinsic masses such as carcinoids, lymphoma, peritoneal carcinomatosis, appendicitis, and diverticulitis should also be considered. Intrinsic causes such as adenocarcinomas,

Crohn's disease, tuberculosis, intramural intussusceptions or hemorrhage, radiation enteropathy, intraluminal bezoars, and intestinal malrotation are also considered in the differential diagnosis [11].

The Diagnosis is definite during laparotomy or laparoscopy [12, 13]. Surgical release of the entrapped bowel via removal of the fibrotic membrane with adhesiolysis is the treatment of choice for this condition [14].

Histopathology of this disease is not pathognomonic, and a diagnosis after the resected specimen is still achieved in concomitance to the clinical and radiological findings [4]. It shows a characteristic fibrin deposits, associated inflammatory cell components are often seen [15].

Current Medical Treatment modalities focuses on the inflammatory and fibrotic responses of the disease. Some clinical experience in patients with EPS is only available with Tamoxifen and prednisone [16]. However, partly due to the low number of patients with EPS, there is a lack of large randomized-controlled trials. However, Surgical treatment is difficult and should be delayed to the last resort [17].

5. Conclusions

- EPS is a rare condition. It poses a particularly challenging diagnosis and management dilemma with high incidence of morbidity and mortality.
- Recognition of this entity and awareness of the typical radiological findings may aid in proper management.
- A gap in knowledge still exists in our understanding of the underlying pathogenesis of EPS and fibrosis and will need to be bridged in order to develop effective therapies.

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