Sclerosing Encapsulating Peritonitis: A mysterious cause of small bowel obstruction

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We do not have any conflict of interest

Introduction:

SEP incidence and prevalence has not been studied. However, older reports indicate a prevalence between 0.54%-7.3%. Additionally, 10%-20% of patients receiving peritoneal dialysis have been seen to develop this rare condition [6, 7]. Although not increasingly prevalent, literature suggests a similar pattern of modalities towards diagnosis of SEP. In fact, diagnosis is usually confirmed by direct visualization of abdominal contents since imaging findings have a low specificity [8]. Diagnosis should be considered in patients with unexplained recurrent episodes of small bowel obstruction and sac-like encasing of intestines with interloop adhesions found on imaging. The purpose of this report is to create awareness of this condition by contributing to the proper elaboration of diagnostic and treatment guidelines. In response to the increasing mortality caused by recurrent inflammation and fibrotic nature of this disease, this report can assist in development of appropriate conservative and/or surgical intervention.

Case presentation:

Case of a 45-year-old male with previous medical history of multiple small bowel obstruction comes to the ER biliary colic symptoms. Patient describes postprandial abdominal pain lasting for several hours before resolving on its own accompanied by 6 episodes of vomiting. Upon further questioning, he referred previous admissions due to nonspecific abdominal pain and diagnosis of small bowel obstruction. Bowel movements were unaffected and he denied malignancy, peritoneal dialysis, and tuberculosis. Upon physical examination, abdomen was soft and depressible, nondistended but moderately tender to palpation especially in the right upper quadrant. Bowel sounds were positive. There was no rebound tenderness, guarding, or additional peritoneal signs present. Abdominal CT scan with oral and IV contrast findings showed evidence of cholelithiasis and bowel wall thickening (Figures A & B). Our patient was then scheduled for laparoscopic cholecystectomy to ameliorate symptoms. Upon gas insufflation of the abdominal cavity poor visualization was achieved due to extensive adhesions and tethering of intestinal loops to adjacent structures. After arduously identifying the relevant anatomy, the procedure was converted to an open approach which was when the abnormal anatomy was directly visualized. Bowel loops were organized in a clumped fashion surrounded by extensive adhesions and fibro collagenous membranes.

Patient was evaluated 2 weeks post-operatively at clinic. He endorsed improvement of symptoms. Had been tolerating diet, passing flatus and bowel movements consistently. He was lost to follow-up since then.

This case was studied retrospectively while analysing the abdominal CT scans from previous hospital admissions, the first one occurring nine years ago. During this stay, an abdominopelvic CT scan was performed which showed findings consistent longstanding small bowel obstruction evidenced by small bowel dilation, bowel wall thickening, and calcifications (Figure C & D). Conservative management with intestinal rest and nasogastric tube decompression was initiated and resulted in clinical improvement. Patient then visited different hospitals for similar presentations.

Treatment guidelines include the removal of the inciting factor, for which was not clear in our case, bowel rest, and TPN. The latter two however are not sufficient to combat the chronic nature of this condition. Many trials have been reported indicating use of immunosuppressive therapy including corticosteroids, colchicine, azathioprine and cyclosporine. These interventions however, are indicated for patients who have a clear and active focus of inflammation. One medical therapy that has proved to be effective in reducing the mortality rate of SEP is Tamoxifen. The latter contains anti-fibrotic properties that contributed to a significant decrease in mortality rate from 74.4% to 45.8% [9]. The exact dosage indicated for this case is not clearly defined. When exploratory laparotomy is indicated in a deteriorating patient, the most common procedures performed is the ablation of fibrotic tissue, adhesiolysis, and resection of perforated or necrotic bowel. To reduce risk of recurrence, it is advised to continue steroid or tamoxifen therapy.

Discussion:

Sclerosing Encapsulating Peritonitis has previously been described in literature as a rare condition causing the small intestine to be wrapped in a fibrocollagenous membrane [2].  Several risk factors have been identified including prolonged intraperitoneal dialysis, abdominal surgery, use of beta blockers, TB, systemic lupus erythematosus, and retrograde menstruation but can be idiopathic. These inciting events cause the release of pro-inflammatory (TGF-β, IL-6), angiogenic (VEGF) cytokines and the subsequent expression of profibrotic genes (COL1A1, fibronectin 1, sulfatase 1) which contribute to peritoneal fibrosis, thickening and adhesion formation [1]. This patient’s case seems to be primarily idiopathic as history taking did not reveal exposure to any risk factors associated with development of SEP. Although, our patient’s recurrent obstructive symptoms might have exposed the abdominal contents to prolonged inflammation and start of fibrinous cascade towards development of this disease.

The diagnostic strategy can be tailored to each particular case. SEP can be part of the differential diagnosis when evaluating the history and physical findings which present recurrent intestinal obstruction, anorexia, abdominal pain, nausea and vomiting, and the presence of an inciting factor such as previous surgeries or peritoneal dialysis. Imaging can be helpful when evaluating SEP. Small bowel dilation, tethering, bowel wall thickening, calcifications, and a thickened peritoneum can be indicative of this condition. One case reported a patient with a CT-scan demonstrating stomach and jejunal dilation, and bowel wall thickening encased in a peritoneal sac indicating abdominal cocoon syndrome [2]. Many reports indicate that SEP may show the “cauliflower sign” on CT-scan which is highly suggestive of the anomaly [1,10, 11]. Because of this, imaging must be carefully studied especially in patients suffering from recurrent SBO. When surgical intervention is indicated, the diagnosis is confirmed during direct visualization of the  bowel loops encased in a cocoon-like cyst with extensive adhesions.

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| ***A picture containing porcelain  Description automatically generated***  a  b  ***A close-up of a fossil  Description automatically generated with low confidence A close-up of a human brain  Description automatically generated with low confidence***  d  c  Coronal IV and oral contrast CT scans (a,b) of patient’s most recent visit showing cholelithiasis and bowel wall thickening (white arrows). In comparison, CT scans from previous small bowel obstruction (c,d) showing marked small bowel dilation, bowel wall thickening (black arrows), and calcifications (white arrows). |