CASE REPORT

COCOONING OF THE INTESTINE: A CASE REPORT AND A BRIEF REVIEW OF THE LITERATURE

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Abstract

Sclerosing encapsulating peritonitis (SEP) is a rare entity. There are well established secondary causes for this condition, but primary SEP is rare. This is a case report of SEP occurring in a 52- year-old female who is diabetic. She did not have any other risk factors for the development of this condition. Therefore she was diagnosed to have primary sclerosing encapsulating peritonitis.

Keywords: Sclerosing encapsulating peritonitis, SEP, Cocooning of small intestine

Introduction

Cocooning of the small intestine and omentum is a rare entity. This is known as sclerosing encapsulating peritonitis (SEP). In this condition, there is intra peritoneal fibrosis that creates a fibrous sheath, which encapsulates the small intestine, omentum and transverse colon into one large cocoon. It is a chronic condition where the pathophysiology is poorly elucidated.

The most common presentation of SEP is usually with small bowel obstruction[1]. During surgery, the encasing fibrous layer can be easily stripped off from the serosal surface. In addition, there are very loose adhesive tissues in between the bowel loops. Usually this does not damage the glistening serosal surface of the bowel.

There are two forms of this disease, namely primary idiopathic SEP and the secondary SEP. Of the two, secondary SEP is common and is usually associated with chronic ambulatory peritoneal dialysis[2], VP and PVshunts[3], beta blockers[4], and systemic diseases such as sarcoidosis and SLE[5]. Sometimes a similar picture may manifest in patients with Fitz-Hugh Curtis syndrome, due to chronic pelvic inflammatory disease. In idiopathic SEP, the aeitiology is unknown and it raises a challenge in diagnosis and management.

This is a case report of idiopathic SEP of an elderly female.

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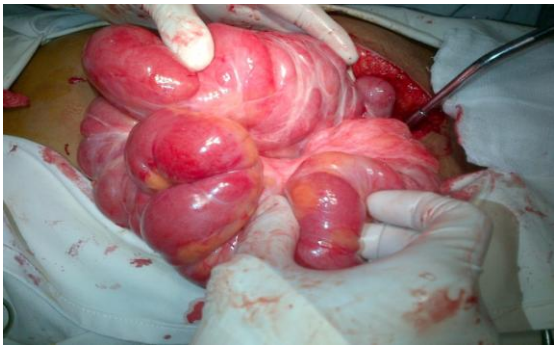
Case report

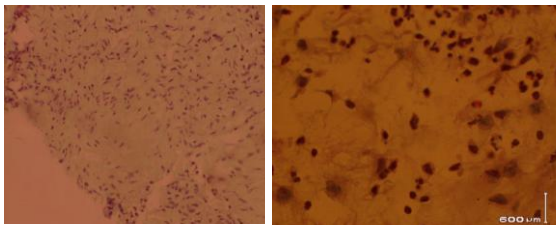
A 52-year old lady presented with sudden onset, diffuse colicky abdominal pain with abdominal distension and repeated vomiting. She had been prescribed atorvastatin for hypercholesterolemia for nearly a year and was on hormone replacement for perimenaupausal symptoms. Other than the above medication, she did not have significant medical or past surgical history. Abdominal examination revealed gaseous distension and a soft mass in the right lower quadrant of the abdomen. The abdominal mass was resonant to percussion, non-tender and bowel sounds were exaggerated. The rectum was found to be empty on examination.

Laboratory findings showed mild leukocytosis with normal biochemistry. The abdominal X-ray revealed multiple dilated bowel loops specially concentrated in the lower abdomen. Ultrasonagraphy showed dilated bowel loops with a small amount of free fluid. A clinical diagnosis of intestinal obstruction was made and laparotomy was performed.

The findings of the laparotomy were quite remarkable. The entire small bowel was wrapped in a thick shiny fibrous layer, incorporating the omentum and the transverse colon. This was like a cocoon encasing the viscera. The fibrous layer was not tightly adherent to the serosa; therefore careful release of the intestine was possible. Part of the membranous layer was sent for histological examination and the ascitic fluid was sent for microbiological examination. Even though there was dilatation of the small intestine, it was possible to primarily close the abdomen.



The histology revealed fibro adipose tissue, with areas of acute and chronic inflammation and fibrosis. There was no granuloma formation or any evidence of malignancy. The histological findings were compatible with sclerosing peritonitis.



Microbiology was negative with no evidence of tuberculosis. The patient made an uneventful recovery. She was followed up for a period of 6 months without any complications.

Discussion

A possible cause for SEP may be due to chronic inflammation within the peritoneal cavity. This leads to fibrosis and membrane formation. Strangely this membrane formation encapsulates most of the small intestinal omentum and part of the transverse colon, thus forming a structure resembling a cocoon. This is why SEP is also known as cocooning of the intestine. A good example to explain the chronic inflammatory theory is that SEP is noted in patients with liver transplantation. After liver transplantation, there is usually ongoing peritonitis, which seems to contribute towards the formation of SEP [6].

This kind of chronic subclinical peritonitis is seen in patients undergoing peritoneal dialysis. SEP is seen in about 0.5- 7.3% of these patients [2].

SEP should also be suspected in patients with cirrhosis, especially with spontaneous subclinical bacterial peritonitis, as this can give rise to chronic inflammation, subsequent fibrosis and membrane formation. This is due to the chronic infection that occurs in these patients over a long period of time.

Beta blockers, particularly propanalol also have been implicated in SEP, as these drugs may enhance the collagen formation, subsequently leading to fibrosis[4]. This is also seen in patients who have undergone peritoneovenous shunting for refractory ascites[2, 3].

Primary or idiopathic SEP seen in young females may be due to the transvaginal spread of infection or retrograde menstruation. This is still a hypothesis and lacks supportive evidence.

Viral peritonitis and food contaminated by an ergot fungus (Clavipesfuciformis) is also thought to cause primary SEP[7, 8].

In adults, SEP is known to occur in chronic conditions such as sarcoidosis abdominal TB, familial Mediterranean fever and in protein C deficiency[7, 8].

Congenital SEP occurs due to a mesothelial layer encasing the intestine in a cocoon fashion rather than as a fibrocollegenous layer, that is usually seen in idiopathic or secondary SEP. Therefore this layer can be easily stripped off from the intestines as well [8].

In chronic encapsulating peritonitis, adhesions are seen both inside and outside the encapsulating fibrous membrane and also between the bowel loops. Strangely however, this condition is not common in patients with repeated adhesive small bowel obstruction or in cases of septic peritonitis.

The mesothelium is attenuated or absent in areas where the fibrocollagenous membrane covers the small intestine. Occasionally there is a nonspecific chronic inflammation of varying activity seen in this layer[8].

The stomach, duodenum, colon and liver can also get involved in SEP. The most frequently involved site is the small intestine. This process can take years to develop or it may have a rapid course of less than 12 weeks[9].

SEP is a recognized entity that may cause ascites and this may develop slowly as the disease progresses.

The clinical course usually includes episodes of intermittent and partial small bowel obstruction. This is caused by the kinking and compression of the bowel loops within the cocoon. Sometimes there can be torsion of the loops causing strangulation and peritonitis [9].

It can also present with chronic abdominal pain with malnutrition or as long standing abdominal masses. The clinical manifestations may sometimes be nonspecific, for example like anorexia and weight loss[9].

The preoperative diagnosis in SEP is difficult. Plain X rays and X rays with oral contrast will only demonstrate small bowel obstruction but will not determine the cause of the obstruction. Even though it is a difficult task to diagnose the etiology for obstruction, there are subtle changes that may give a clue to the diagnosis of SEP. Unlike in the other forms of intestinal obstruction, such as band adhesive obstruction or internal hernia, in SEP there

is no definitive transitional zone or dilated proximal bowel loops[10].
Computer tomography studies may be helpful in preoperative diagnosis. In SEP these studies will show tightly bound small bowel loops with thickened peritoneal membranes and ascites [7]. There will be thickened and dilated small bowel loops in various sizes inside the cocoon. There are occasional instances where SEP is diagnosed pre-operatively but these occasions are extremely rare[9].

It is usually diagnosed upon opening the abdomen. The appearance is quite unique. The other differential diagnoses for SEP include retractile mesoenteritis, sclerosing malignant lymphoma, malignant primary mesenteric tumour and other metastatic neoplasms.

It is difficult to determine appropriate therapeutic strategy in patients with SEP. The first few published data shows that, in primary or secondary SEP, surgical intervention is necessary as attempts of conservative measures usually fails when they present with intestinal obstruction [10]. However, there are isolated cases of successful conservative management of secondary SEP with immunosuppression, corticosteroids and tamoxifen. This has been reported only in patients who developed this condition secondary to chronic peritoneal dialysis [11].

Surgical adhesiolysis is the treatment for SEP. This is a tedious process and has to be performed with great care. There can be recurrences of the disease as well as recurrent attacks of intestinal obstruction following adhesiolysis. The combination of surgery with immunosuppression has been considered as an effective treatment option to prevent recurrences.

The excision of the whole membrane is also been done in addition to the adhesiolysis. This can be performed without much difficulty, as this fibrocollegenous membrane is not tightly adherent to the serosa of the small intestine. The need for bowel resection is rare, and is only performed in situations where a gangrenous bowel loops are detected. This is only seen in very late presentations.

The prognosis of this condition is usually excellent [1].

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