

Cocoon carcinomatosa: An unusual cause of intestinal obstruction

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Summary

Abdominal cocoon, also termed sclerosing encapsulating peritonitis, is an uncommon entity associated with formation of a fibro-collagenous membrane around intestinal loops resulting in intestinal obstruction. Most reported cases are idiopathic, but many other causes have been implicated in cocoon formation, including use of continuous ambulatory peritoneal dialysis, peritoneal tuberculosis, and connective tissue disease. However, peritoneal carcinomatosis is a rarely reported entity that causes this condition. Reported here are two cases of abdominal cocoon secondary to peritoneal carcinomatosis. Both patients presented with intestinal obstruction; one underwent surgery but the other refused surgery.

Keywords: Peritoneum, peritonitis, intestinal obstruction, abdominal pain, sclerosis

1. Introduction

Abdominal cocoon, also referred to as sclerosing encapsulating peritonitis (SEP), is related to small bowel encasement by a cocoon like fibro-collagenous sac resulting in intestinal obstruction. It can be either idiopathic or secondary. The pathogenesis of abdominal cocoon is theorized to be related to an increase in the release of fibrogenic cytokines resulting in deposition of fibrin-like material on the peritoneum (1). Abdominal cocoon may present with intestinal obstruction or with nonspecific symptoms like nausea, vomiting, and malnutrition. Ultrasound or computed tomography can be used for diagnosis, and scans will reveal a thick membrane-like covering around loops of the small intestine (1,2). Surgical intervention with resection of the membrane and adhesiolysis is the usual treatment. Peritoneal carcinomatosis occurs due to peritoneal seeding by various neoplastic diseases. In very rare cases, such peritoneal seeding can lead to formation of a membranous cocoon surrounding the bowel that causes intestinal obstruction. Such cocoon formation in peritoneal carcinomatosis is a rarely reported entity.

Reported here are two cases of cocoon formation resulting from peritoneal carcinomatosis.

2. Case Report

2.1. Case 1

An 18-year-old male presented with gradually increasing abdominal pain of one month's duration that was generalized and colicky in nature. The patient had features of acute intestinal obstruction (recurrent vomiting, abdominal distension, and obstipation) for 3 days before he was seen. The patient was started on anti-tubercular therapy at another facility based on a low serum-ascites albumin gradient (SAAG) and an adenosine deaminase (ADA) level of 48 U/L in ascitic fluid. On examination, the abdomen was tender and distended with the presence of nodules on palpation. An abdominal x-ray revealed dilated bowel loops with multiple air-fluid levels, and a contrast-enhanced computed tomography (CECT) scan of the abdomen revealed the presence of gross ascites with nodular peritoneal thickening, omental caking, and scalloping of the liver (Figure 1). The patient underwent an urgent exploratory laparotomy that revealed the presence of multiple nodular deposits over the peritoneum with adherent small bowel and a surrounding membranous "cocoon". The cocoon was excised and a loop ileostomy was created around 30 cm proximal to the IC junction. Histopathological examination of biopsy samples from omental deposits and the peritoneal cocoon revealed the

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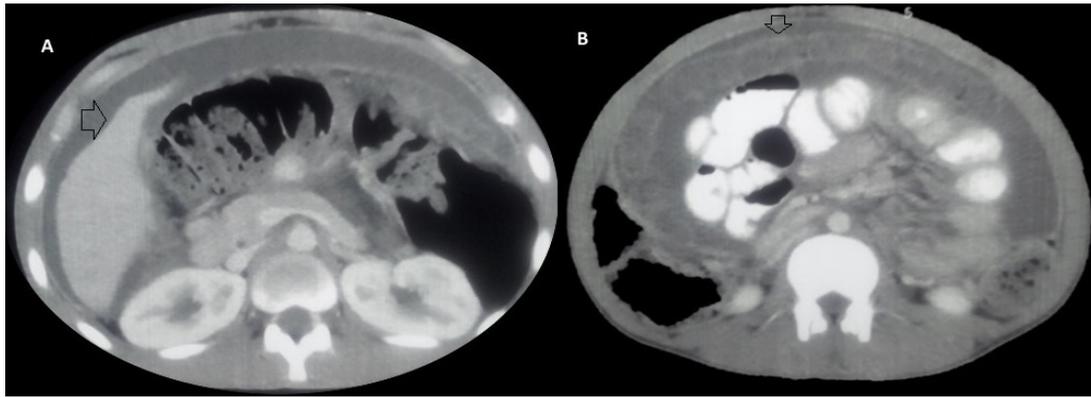


Figure 1. CT scan showing ascites, liver scalloping (A), and clumping of bowel loops with membrane (B). The arrow indicates liver scalloping (A) and a membrane surrounding the bowel loops (B). CT, computed tomography.

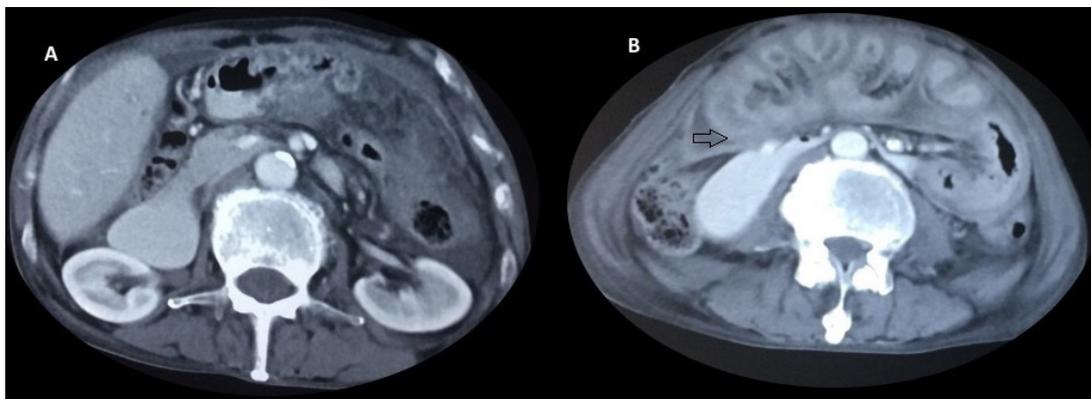


Figure 2. CT scan showing ascites (A) and clumped bowel loops encased by a membrane (B). The arrow indicates clumped bowel loops encased by a membrane (B). CT, computed tomography.

presence of metastatic mucinous adenocarcinoma. The patient was advised to follow up for treatment of the primary lesion and discharged.

2.2. Case 2

A 70-year-old male was evaluated for gradually increasing abdominal pain of two months' duration that was generalized and colicky in nature. The patient had a history of recurrent vomiting, abdominal distension, and constipation for 15-20 days. On examination, a vague lump approximately 22 × 20 cm in size was palpable in the periumbilical area. The lump was firm, non-tender, and non-mobile in nature with surface nodularity. On ultrasonography, the bowel loops appeared clumped in the periumbilical area with a surrounding sac-like structure. Contrast-enhanced CT of the abdomen revealed the presence of peritoneal thickening with clumped small bowel loops surrounded by a membrane "cocoon". Analysis of ascitic fluid revealed the presence of a low SAAG and ADA in ascitic fluid was 13 U/L. On further evaluation, analysis of the ascitic fluid revealed the presence of metastatic adenocarcinoma. The patient was counselled for the need of a further workup and various treatment

options, but the patient left treatment against medical advice.

3. Discussion

First described by Foo *et al.* in 1978, SEP is an uncommon entity (2). The term encompasses three components: "sclerosing", which refers to the growth of dense collagenous fibrotic tissue, "encapsulating", which refers to small bowel encasement and restricted motility due to this fibrotic process, and "peritonitis", which refers to the inflammatory changes in the peritoneal membrane, though these may not be universal (1,2). SEP may be idiopathic or secondary to chronic ambulatory peritoneal dialysis, prior abdominal surgery, beta blocker therapy, post-liver transplantation, abdominal tuberculosis, ventriculoperitoneal shunts, and peritoneovenous shunts (1-5). Patients may be entirely asymptomatic or have non-specific symptoms like nausea, anorexia, and diffuse abdominal pain or they can present with stark features of intestinal obstruction.

Plain films of the abdomen may reveal dilated small bowel loops due to obstruction and these loops may be restricted to one location due to encapsulation (6).

Barium studies may reveal a serpentine or concertina-like configuration of dilated small bowel loops fixed in a U-fashioned cluster often termed a "cauliflower" sign (Figure 2B). On CT, a fibrous membrane surrounding the bowel loops is usually seen. Other signs include fixation of intestinal loops, mural thickening of the bowel wall, ascites, and lymphadenopathy (7). On CT, peritoneal carcinomatosis can appear with thickening, nodularity, and enhancement of the peritoneum and diffuse tumor infiltration of the mesentery producing a pleated or stellate pattern. Omental caking is another key CT feature of peritoneal carcinomatosis due to replacement of the omental fat by the tumor and fibrosis. In rare cases, such seeding with fibrosis can lead to the formation of a thick membrane (cocoon) surrounding the bowel, resulting in obstructive symptoms. Peritoneal nodules can also appear hyperechoic with acoustic shadowing from psammomatous calcification (8).

There are only a few case reports of cocoon secondary to peritoneal carcinomatosis in the literature. Various malignancies like ovarian and colorectal carcinoma have been found to present with cocoon formation (9,10). In one diagnosed case of colorectal carcinoma with peritoneal carcinomatosis, cocoon formation occurred after chemotherapy and was treated surgically (10). Interestingly, abdominal cocoon formation in cases of metastatic gastroenteropancreatic neuroendocrine tumors (NET) has been reported in late stages and is associated with poor surgical outcomes and universal mortality. In fact, a NET-related cocoon has been reported to be a type 4 cocoon that completely encases the entire contents of the abdomen. Surgical outcomes are poor and therefore surgery is not recommended (11). In conclusion, abdominal cocoon may be caused by peritoneal carcinomatosis and it represents a late feature of that condition with limited therapeutic options.

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