CASE REPORT

ABDOMINAL COCOON: A RARE CAUSE OF INTESTINAL OBSTRUCTION

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ABSTRACT: INTRODUCTION: Abdominal cocoon is a rare acquired benign condition causing intestinal obstruction due to thick fibrotic peritoneum encasing the small bowel partially or completely. We present a case of a patient who presented to our institution with abdominal distension and guarding. Computed tomography was obtained which revealed findings concerning for sclerosing encapsulating peritonitis. Sonographic imaging was also obtained and provides correlative imaging.

KEYWORDS: Abdominal cocoon, intestinal obstruction Sclerosing Encapsulating Peritonitis; Cocoon bowel; Peritonitis; Peritoneal Dialysis.

CASE REPORT: A 30 years old female presented with vague complaints of intermittent abdominal pain, nausea, vomiting and abdominal distension and irregular bowel habits.

The patient had no previous history of practolol use, hepatic disease, abdominal surgery, peritoneal dialysis, ventriculoperitoneal and peritoneovenous shunting. Additionally, no clinical features of connective tissue disease or sarcoidosis could be identified.

Plain radiograph of the abdomen in supine posture revealed mildly dilated small bowel loops in the mid-abdominal region.

Abdominal ultrasonography revealed clustering of the small bowel loops in the mid-abdominal region, which showed numerous septations and loculations throughout the abdomen as shown below.

Fig. 1  Fig. 2
Computed tomography findings seen in sclerosing encapsulating peritonitis include peritoneal calcification and a thick fibrous membrane that encases several small bowel loops or the entire small bowel. Ascites is often seen as well and can be diffuse or loculated.
PARACENTESIS YIELDED AROUND 1 LITER OF REDDISH ASCITIC FLUID:

DISCUSSION: Sclerosing encapsulating peritonitis, previously termed cocoon bowel, is characterized by an abnormally thickened, fibrotic peritoneum that encapsulates the small bowel loops resulting in obstruction.

Although many risk factors exist for sclerosing encapsulating peritonitis, the exact etiology and underlying pathophysiology of this disease process are not well understood. One proposed mechanism pertains to patients with cirrhosis who have peritoneovenous shunts.[1]

Since these patients tend to have a faster ascitic fluid circulation, it is believed that the release of fibrogenic cytokines causes increased deposition of fibrin on the peritoneum, which converts fibrinous adhesions to generalized peritoneal fibrosis.[2]

Histologically, mononuclear inflammatory cells infiltrate the peritoneal membrane.[3]

The incidence of sclerosing encapsulating peritonitis ranges from 0.5–4. 4%, and the incidence increases with the duration of peritoneal dialysis. Furthermore, there appears to be a direct relationship between duration of peritoneal dialysis, mortality, and the incidence of sclerosing encapsulating peritonitis.[4]

A prospective, multicenter study in Japan performed from 1999 to 2003 by Kawanishi et al. involved 1,958 peritoneal dialysis patients across 57 facilities.[4] Results of this study showed...
both an incidence and mortality of 0% at 3 years.\textsuperscript{[4]} Incidence and mortality rose to 2.1% and 8.3%, respectively, at 8 years.\textsuperscript{[4]} After 15 years of peritoneal dialysis, the incidence and mortality were 17.2% and 100%, respectively.\textsuperscript{[5,6]} The overall mortality of sclerosing encapsulating peritonitis is high at 56% with a range of 38-83%. Currently, no established age predilection or gender ratio exists for sclerosing encapsulating peritonitis.

Many risk factors exist for sclerosing encapsulating peritonitis.\textsuperscript{[1,2,7-10]} Several case studies list long term peritoneal dialysis, prior abdominal surgery, retrograde peritonitis, practolol therapy, sarcoid, liver transplantation, peritoneal tuberculosis, ventriculoperitoneal shunts, and peritoneovenous shunts as potential risk factors.\textsuperscript{[9]}

Many cases are also idiopathic.\textsuperscript{[8,10]} The idiopathic form is commonly seen in adolescent females and is thought to result from retrograde menstruation that leads to a subclinical peritoneal infection.\textsuperscript{[11]}

Treatment options for sclerosing encapsulating peritonitis consist of surgical intervention or medical management depending on the clinical presentation.\textsuperscript{[11]} Medical options include supportive measures such as bowel rest and nutritional support.\textsuperscript{[11]} Medications that recently have been used to treat sclerosing encapsulating peritonitis include: corticosteroids, tamoxifen, and immunosuppressants. Each of these medications treat the condition by reducing the inflammation and fibrosis of sclerosing encapsulating peritonitis.\textsuperscript{[11]}

Surgical intervention consists of lysis of intestinal adhesions. Diagnosis of sclerosing encapsulating peritonitis is usually confirmed by laparotomy, which reveals a fibrous membrane surrounding multiple small bowel loops.\textsuperscript{[9,10]}

Key computed tomography findings seen in sclerosing encapsulating peritonitis include peritoneal calcification and a thick fibrous, enhancing membrane that encases several small bowel loops or the entire small bowel. Ascites is often seen as well and can be diffuse or loculated.\textsuperscript{[8]}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{Fig_7.png}
\caption{Fig. 7}
\end{figure}

Key sonographic features include internal septations, thick fibrous membranes, and loculated ascites surrounding the small bowel.[8] Additional ultrasound features include the small-bowel loops having the overall appearance of a cauliflower or concertina due to their arrangement within the fibrous membrane sac.

Sclerosing encapsulating peritonitis often has a vague clinical presentation that can lead to a delay in diagnosis and treatment. Physical exam and laboratory testing will not raise clinical suspicion for this rare disease process.[2] Symptoms of sclerosing encapsulating peritonitis are nonspecific and usually consist of anorexia, bloody ascites, nausea, diarrhea, and abdominal pain.[7] Sclerosing encapsulating peritonitis can lead to serious complications such as severe malnutrition, sepsis, and death if not promptly diagnosed and treated.

Abdominal plain films can often show dilated small bowel loops typical of obstruction; however, the fibrosing, encapsulating membrane typical of sclerosing encapsulating peritonitis will not be seen on this imaging modality. Since surgery is often needed to decompress the encased small bowel loops, CT and ultrasound imaging can lead to prompt diagnosis, treatment, and improved patient outcomes.[8,10]

The differential diagnosis for sclerosing encapsulating peritonitis includes disease processes that could result in similar imaging features such as peritoneal enhancement, peritoneal calcification, ascites, or possible small bowel obstruction.[8,10] Similar disease processes to consider in the differential diagnosis would include peritoneal carcinomatosis, pseudomyxoma peritonei, peritoneal mesothelioma, and tuberculous peritonitis. Clinical history and CT findings are critical in differentiating these respective disease entities, although overlap of CT features is commonly seen.[4]

Differentiating sclerosing encapsulating peritonitis from peritoneal carcinomatosis: the latter is commonly seen in patients with a history of ovarian cancer or gastrointestinal tract malignancy.[4] Presenting symptoms are nonspecific and usually consist of ascites, abdominal pain, nausea, and vomiting. Important CT features of peritoneal carcinomatosis include thickening, nodularity, and enhancement of the peritoneum and diffuse tumor infiltration of the mesentery producing a pleated or stellate pattern.[4] Small bowel obstruction is often seen in peritoneal carcinomatosis from colorectal carcinoma.[4] Omental caking is another key CT feature of peritoneal carcinomatosis due to replacement of the omental fat by tumor and fibrosis.[12] Sonographically, peritoneal carcinomatosis can present with hypoechoic nodules that are commonly seen in Morison's pouch, the pouch of Douglas, and the right subphrenic region.[12] Peritoneal nodules can also appear hyperechoic with acoustic shadowing from psammomatous calcification.[12] Septated ascites is often present as well.

Differentiating sclerosing encapsulating peritonitis from pseudomyxoma peritonei: the latter is commonly seen with mucin-producing tumors, most commonly appendiceal, pancreas, gallbladder or ovarian in origin.[4] Pseudomyxoma peritonei is more commonly seen in women and has a mean age at diagnosis of 49.[4] Pseudomyxoma peritonei has nonspecific presenting
symptoms such as weight loss, abdominal pain, and increasing abdominal girth. Common CT features seen in pseudomyxoma peritonei include diffuse low attenuation mucinous ascites containing curvilinear or amorphous calcifications. Scalloped margins of abdominal visceral organs, mainly the liver and spleen, are commonly seen as well. Sonographically, pseudomyxoma peritonei can present with nonmobile echoes and ascites, suggesting gelatinous fluid. Septations are often present and appear echogenic. The gelatinous fluid appears hypoechoic and displaces the intestines centrally, leading to a starburst or star-like configuration due to echogenic bowel being surrounded by hypoechoic, gelatinous fluid.

Differentiating sclerosing encapsulating peritonitis from peritoneal mesothelioma: the latter is more commonly seen in males with an age range of 50-69 years and a history of prior asbestos exposure is seen in approximately 50% of peritoneal mesothelioma cases. Three distinct CT types of peritoneal mesothelioma have been described in the literature: dry-painful, wet, and mixed. The dry-painful type is most common and defined by a single large mass or multiple small peritoneal masses located in a single abdominal quadrant without ascites. The wet type presents with diffuse nodules and plaques along with ascites and intestinal distention. The mixed type contains features of both the dry-painful and wet types. Additional CT features of peritoneal mesothelioma include: omental masses, scalloped margins of abdominal visceral organs, and direct invasion of abdominal organs. Sonographically, peritoneal mesothelioma can present with hypoechoic, sheetlike or nodular masses that cause peritoneal thickening. Omental invasion can occur and presents as a hypoechoic mass with scattered hyperechoic areas suggesting entrapped omental fat. Ascites can be seen but is usually minimal compared to the amount tumor dissemination. Intestinal wall and solid organ invasion can also occur.

Differentiating sclerosing-encapsulating peritonitis from tuberculosis peritonitis: the latter is commonly seen in immune compromised individuals, usually with a history AIDS or prior transplant. Tuberculosis peritonitis commonly presents with abdominal pain, fever, night sweats, weight loss, and anorexia. Three CT types of tuberculosis peritonitis have been described: wet, fibrotic, and dry. The wet type is the most common occurring in 90% of cases and consists of diffuse ascites. The ascites in the wet type can be diffuse or loculated and is slightly hyper attenuating due to its high cellular or protein content. The fibrotic type is characterized by matted bowel loops along with low attenuating omental and mesenteric nodular masses. The dry type commonly presents with caseous nodules, mesentery thickening, or fibrous adhesions. Although three distinct subtypes of tuberculosis peritonitis have been described, the features of each subtype often overlap. Abdominal lymphadenopathy involving the mesenteric, retroperitoneal, periportal, and peripancreatic lymph nodes can also be seen in tuberculosis peritonitis. Sonographically, tuberculosis peritonitis often presents with ascites. The ascites can be free or loculated containing mobile septa that can produce a latticelike pattern. Sonographic findings of tuberculosis peritonitis are not pathognomonic but hypoechoic or nodular peritoneum thickening can be seen. Omental and mesenteric thickening with enlarged mesenteric lymph nodes can also be seen.
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CONCLUSION: Sclerosing encapsulating peritonitis, previously termed cocoon bowel, causes intestinal obstruction due to development of a fibrous membrane that encases multiple small bowel loops. Since surgery is often needed to decompress encased small bowel loops, CT and ultrasound imaging can lead to a prompt diagnose.

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REFERENCES:

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