Idiopathic Sclerosing Peritonitis – A Rare Cause of Sub-acute Intestinal Obstruction Presenting as Massive Ovarian Edema and Ascites – a Case Report

A Nadkarni, B Rao, S Bhandary, P Shetty, R Jain

Citation

Abstract
Introduction: Idiopathic encapsulating sclerosing peritonitis (or abdominal cocoon) is a rare cause of small bowel obstruction characterized by fibrotic encapsulation of the bowel. Diagnosis is usually during laparotomy. Case report: A 21-year-old female patient was admitted with symptoms of subacute intestinal obstruction. She had a history of bilateral ovariotomy for bilateral ovarian enlargement with ascites and right hydrothorax one month back, which had revealed massive edema of the ovaries with stromal hyperplasia and hyperthecosis. After a conservative management trial for few days, she underwent an exploratory laparotomy: Mildly dilated loops of small bowel were found with a thin flimsy layer of fibrous tissue over the loops with interloopal adhesions and three strictures in the region of the ileum. The serosa of the sigmoid colon was studded with multiple nodules which were soft and friable. With a suspicion of tuberculosis or malignancy, limited resection of the involved ileum with caecum and ascending colon and an end-to-end ileo-transverse anastomosis was done and multiple biopsies were taken from the surface of the nodules on the sigmoid colon. The histological diagnosis revealed sclerosing peritonitis with serositis. The patient had an uneventful recovery. Discussion: The presentation of subacute intestinal obstruction due to sclerosing peritonitis in a young female with massive ovarian masses due to ovarian edema and hyperthecosis with stromal hyperplasia makes this case a rare one. The fibrosed capsule that usually is seen in patients with abdominal cocoon was flimsy and not well-formed in this case and did not draw attention to the diagnosis even at the time of laparotomy. Clinicians must rigorously pursue a preoperative diagnosis, as it may prevent a "surprise" upon laparotomy.

INTRODUCTION
Idiopathic encapsulating sclerosing peritonitis (or abdominal cocoon) is a rare cause of small bowel obstruction characterized by fibrotic encapsulation of the bowel, first described and named by Foo et al. in 1978 (1). Diagnosis is usually during laparotomy. It is categorized into primary and secondary forms (2). The idiopathic (primary) form is rare. The exact etiology of the disease remains obscure (3). Meigs’ syndrome is characterized by ovarian fibromas with ascites and usually a right-sided hydrothorax. It is termed as Pseudo-Meigs’, if the ovarian tumor is malignant.

We present a rare case of a 21-year-old female patient with an ovarian tumor with ascites suggestive of Meigs’ or Pseudo-Meigs’ syndrome initially, who underwent surgery and subsequently developed features of subacute intestinal obstruction one month later, which turned out to be caused by multiple ileal strictures due to sclerosing peritonitis. The ovarian pathology was massive edema of ovary with hyperthecosis and stromal hyperplasia and had no features of malignancy. We also discuss the various clinical conditions that can be thought of in such a clinical scenario with a review of the literature.

CASE REPORT
A 21-year-old female patient presented with a 15-day history of dull aching lower abdominal pain with slowly progressive distension of abdomen associated with few episodes of non-bilious vomiting for 2 days. She did not give any history of irregular bowel habits, jaundice, loss of appetite or weight, or generalized swelling of limbs. She experienced shortness of breath especially on lying down. She gave a past history of having irregular menses for the past one year for which she had been taking oral contraceptive pills (estrogen+progestrone) for the past 3 months. She did not have any past history of abdominal surgeries or tuberculosis. There was no family history of any ovarian, breast or bowel cancer. Examination revealed a pale but comfortable girl.
Her vital parameters were stable and the positive findings were diffuse abdominal distension with fluid thrill and horse-shoe shaped dullness suggestive of gross ascites. There was no palpable mass, visible peristalsis or organomegaly. Vaginal and rectal examinations did not reveal any mass but fullness in the pouch of Douglas.

Routine blood investigations including liver function tests and renal parameters were normal except for mild hypoproteinemina. Chest x-ray suggested mild right-sided pleural effusion, abdominal x-ray (erect) was normal. USG of the abdomen suggested gross ascites with bilateral ovarian enlargement with multiple thin-walled cysts of varying sizes (figure 1). Abdominal CT with contrast also showed massive ascites with bilateral enlarged ovaries with multiple enlarged follicles and fat planes maintained with adjacent structures (figure 2).

Figure 1
Figure 1: USG of the abdomen: Gross ascites with bilateral ovarian enlargement with multiple thin-walled cysts of varying sizes

Ascitic fluid analysis showed the transudative nature of the fluid with no malignant cells or tubercle bacilli. CEA (carcinoembryonic antigen), serum testosterone, thyroid function tests and CA-125 levels were normal. Even tests for collagen vascular diseases like SLE (Systemic Lupus Erythematosus) were negative. With a strong suspicion of ovarian malignancy, she underwent exploratory staging laparotomy with frozen section biopsies from omentum, peritoneum and ovaries, which showed massive edema with hyperthecosis and stromal hyperplasia, peritoneal and omental sections being normal. She underwent bilateral ovariectomy with drainage of ascitic fluid of about 1 liter. The post-operative period was uneventful; the patient went home on the 7th post-operative day and was tolerating normal diet.

The patient came back one month later with complaints of abdominal pain and vomiting for ten days. The pain was colicky in nature, radiating from upper to lower abdomen usually few hours after meals and relieved after vomiting. These symptoms used to occur once in 2 days and intermittent. There was no associated constipation or abdominal distension. On examination, there was no visible peristalsis or distension and the abdomen was soft with no guarding or rigidity. All her investigations including
abdominal x-ray (erect) were inconclusive. So she was given a trial of conservative management with nil per-orally, intravenous fluids and analgesics. The patient’s symptoms used to subside with this but as soon as she was started on oral feeding, the symptoms would reappear. The examination findings were the same and did not change over a period of ten days after admission. USG of the abdomen revealed mild ascites. Abdominal x-ray (erect) taken 10 days after conservative management revealed few air-fluid levels (figure 3). Barium meal follow-through was attempted but the patient could not tolerate the procedure. The only important finding was the presence of barium on the erect abdominal x-ray even 5 days after the attempted barium test, suggesting a delayed transit time. She underwent exploratory laparotomy in view of her non-resolving symptoms and suspected post-operative adhesions as the cause of her symptoms.

**Figure 3**
Figure 3: Abdominal x-ray (erect) taken 10 days after conservative management revealed few air-fluid levels

At laparotomy, mildly dilated loops of small bowel were found with a thin flimsy layer of fibrous tissue over the loops and inter-loopal adhesions. Mild ascites was noted. On removing the flimsy adhesions and tracing the bowel distally, three strictures in the region of the ileum were noticed – 3cm, 11cm and 36cm from the ileocaecal junction, respectively (figure 4). The serosa of the sigmoid colon was studded with multiple nodules (figure 5) which were soft and friable. Few mesenteric lymph nodes were found to be enlarged. The rest of the bowel was normal. Limited resection of the involved ileum with caecum and ascending colon and an end-to-end ileo-transverse anastomosis was done and multiple biopsies were taken from the surface of the nodules on the sigmoid colon.

**Figure 4**
Figure 4: Stricture seen

**Figure 5**
Figure 5: Multiple serosal nodules on the sigmoid colon
The ascitic fluid analysis revealed a transudative fluid with no acid-fast bacilli or malignant cells. The resected segment (figure 6) showed a thickened serosa throughout the length of the ileum resected. Section showed three ileal strictures at 3, 11 and 36cm from the ileocaecal junction. Microscopic examination of the sections from the strictures revealed dense lymphoplasmacytic infiltration of lamina propria and the serosa was markedly thickened showing dense fibroblastic and myofibroblastic proliferation with plump vesicular fusiform nuclei showing atypia interspersed with collagen deposition, thin capillaries and sparse lymphoplasmacytic infiltration (figure 7).

Figure 6
Figure 6: Resected specimen with multiple strictures shown

These features suggested sclerosing serositis with peritonitis. The sections from appendix, colon and the lymph nodes were normal. Sections from the biopsies of the sigmoid colon nodules showed proliferating histiocytes, foam cells, giant cells and congested capillaries surrounded by fascicles of proliferating fibroblasts and myofibroblasts. There were no features suggestive of tuberculosis, Crohn’s disease or malignancy which had been suspected intraoperatively. A review of slides of the previous surgery on the ovaries revealed both ovaries showing normal parenchyma with loss of normal corticomedullary differentiation with ill-defined nodularity containing a thin subcapsular zone of condensed stromal cells overlying markedly edematous parenchyma with few primordial follicles surrounded by widely separate spindle and stellate plump stromal cells with foci of stromal hemorrhage with perivasular stromal cell condensation. Reticulin stain highlighted stromal edema with wide separation of collagen fibres. These features suggested massive edema of ovaries with stromal hyperplasia and hyperthecosis. The sections that were taken from the omentum and peritoneum, then, were normal.

The patient had an uneventful recovery from surgery. Six months after surgery, she remains asymptomatic.

DISCUSSION
Abdominal cocoon is a recognized cause of intestinal obstruction. It was first described in 1907 by Owtschinnikow as peritonitis chronica fibrosa incapsulata (5). Brown et al., in 1974, described the condition as a complication of long-term treatment with beta-adrenergic blocking agents (6). Foo, et al., in 1978, were the first to describe “cocoon abdomen” in an adolescent girl (1). It is a rare entity and its etiology remains unknown.

The main pathogenesis in this condition appears to be peritonitis leading to sclerosis with membrane formation (7). In primary or idiopathic SEP, the stimulus for this inflammatory reaction is open to speculation. Some suggest that it could be due to subclinical viral peritonitis which ascends vaginally with trans-fallopian migration or through hematogenous spread (8,9). Others postulate that retrograde menstruation may be the initial inflammatory stimulus (8). The argument against this is that the condition also afflicts premenarchal girls and males. In our case, the etiology of the peritonitis appears to be related to the ovarian edema which may have been induced by the oral contraceptive pills that the patient was taking, though this is just an assumption. In some patients, the congenital absence of greater omentum is postulated to be the predisposing factor (10,9). It is a known complication of chronic ambulatory peritoneal dialysis (11). In a study of 554 patients with cirrhosis, 69 patients had peritoneovenous shunts, and at autopsy, 38% of these patients were found to have generalized peritoneal fibrosis, with cocoon formation (22). Because these patients had
faster ascitic fluid circulation, it was hypothesized that increased deposition of fibrin on the peritoneum, after release of fibrogenic cytokines, converted fibrinous adhesions to generalized peritoneal fibrosis.

Peritoneal encapsulation is a separate entity, which may be confused with abdominal cocoon. This is a congenital malformation that usually presents in infancy or childhood as an acute intestinal obstruction and is characterized by a thin accessory peritoneal sac covering the small bowel with the loops lying freely within this membrane (12,7,13). This membrane is derived from the yolk sac as it is drawn into the embryonic abdominal cavity during the 12th week of gestation. It has been associated with midgut malrotation and vascular anomalies (14,15). Histologically, the membrane is similar to the normal peritoneum rather than the thick fibro-collagenous tissue seen in abdominal cocoon (16).

Abdominal cocoon usually presents with a variety of clinical presentations. Recurrent attacks of subacute intestinal obstruction with colicky abdominal pain, progressive nausea and vomiting (12), abdominal distention, and weight loss should raise suspicion regarding the diagnosis, as in this case. The symptoms may be quite confusing and the decision to operate may not be supported by many clinical signs or investigative procedures, as in this case. Occasionally, the patient may present with an abdominal or pelvic mass (4). On other occasions, it might be discovered accidentally during laparotomy for other surgical conditions (17).

Radiological findings may show various degrees of intestinal obstruction, especially in acute presentation. Abdominal ultrasound examination may show loops of bowel entrapped in a thick hyperemicogenic layer. The presence of a thick peritoneal layer anterior to the small bowel is characteristic (18,16). A plain x-ray of the abdomen only indicates multiple air-fluid levels like in any other case of small-bowel obstruction, as seen in this case. Computerized tomography findings may be more diagnostic, demonstrating clusters of small-bowel loops with mural wall thickening, thick peritoneal encapsulation, peritoneal enhancement, and loculated fluid collection (18,4). At times, the surrounding membrane may be thin and difficult to identify on a CT scan, making it difficult to diagnose the condition preoperatively on imaging (16). Barium small-bowel series occasionally show varying lengths of small-bowel aggregation suggestive of the characteristic ‘cauliflower sign’ (13,19) and a delayed transit. The “saw tooth” or “serpentine configuration” of the small-bowel loops is characteristic and may be seen in both computerized tomography and small-bowel series. Definitive diagnosis of abdominal cocoon is made at laparotomy. Characteristically, the small bowel is found compressed and coiled up within a dense whitish membrane. The membrane encasing the bowel loops consists predominantly of a proliferation of fibroblasts resulting in diffuse fibrous thickening without any or only mild inflammation (9).

The interesting facts in this case are: (a) Appearance of obstructive symptoms few days after surgery for ovarian tumor - the first differential diagnosis for such a presentation is adhesive obstruction, due to previous surgery, but in this case, it was proved wrong. It also suggests a role of previous surgery or ovarian pathology as the stimulus for the, subsequently found, sclerosing peritonitis. (b) The membrane surrounding the intestines found during the second surgery was not well-formed – it may be considered as an early stage of pathogenesis. (c) Abdominal CT scan, done during the initial presentation of massive ovarian masses, showed normal bowel loops – this fact, too, suggests that the sclerosing peritonitis pathology may have been induced after the first surgery or as a part of the ovarian pathology, in this case being massive ovarian edema with hyperthecosis and stromal hyperplasia. (d) The presence of massive ascites – the evidence supporting this finding is that, in 1994, Clement et al. (21) reported six cases of sclerosing peritonitis with massive ovarian edema associated with luteinized thecoma. There seems to be a resemblance of the clinical feature in four of their cases with our patient whereby the dominating feature is the presence of the ovarian tumor. Furthermore, all the cases in their series had ascites and in some of them the ascitic fluid was voluminous as in our patient. It seems that the only pathology in gynecology that has been associated with abdominal cocoon is luteinized thecoma of ovary (21). As in this case, only the finding of sclerosing peritonitis, and not abdominal cocoon with encapsulating features, was specifically described in their cases. (e) The presence of strictures and peritoneal nodules – this finding makes this case a unique one as according to the authors’ knowledge from review of literature this has been the first case of sclerosing peritonitis with massive ovarian edema to present with peritoneal nodules and strictures, and least features of encapsulation. The various conditions presenting with multiple ileal strictures are tuberculosis, Crohn’s disease, pelvic inflammatory disease, ischemic bowel, radiation enteritis, carcinoid infiltration, lymphomas, diffuse enteropathies and, rarely, ganglioneuromatosis.
Conservative management in cases of sclerosing peritonitis often fails. Surgery includes freeing the bowel from the thickened encasing membrane to release the obstruction. Bowel resection is indicated if a segment is found to be non-viable. The most common complications of sclerosing peritonitis appear to be intestinal obstruction, small-bowel necrosis and entero-cutaneous fistulae. This condition necessitates surgical intervention, and the mortality rate is 60% within 4 months of diagnosis (20, 23). Surgery should be avoided if possible for asymptomatic patients, patients with co-morbid conditions such as liver cirrhosis and ascites (18, 12). Considering the presence of multiple strictures with peritoneal nodules, the age of the patient and the fact that tuberculosis and malignancy are the commonest causes of ileal strictures in India, the decision of resection and anastomosis was taken in this case.

Management of such a rare case presents a dilemma as it is impossible to decide on the best therapeutic approach on table. What intrigued the authors, including the pathologist involved, was the presence of two rare entities in the same patient, which are the voluminous ascitic fluid, a benign gynaecological disease and the abdominal sclerosing peritonitis.

CONCLUSIONS

Sclerosing peritonitis is a rare cause of subacute intestinal obstruction. The association of an ovarian pathology, too, is a rare finding. The presentation of this case with an ovarian pathology, multiple small-bowel strictures and peritoneal nodules with massive ascites and with least features of encapsulation, suggests a new dimension towards the etiology and pathogenesis of sclerosing peritonitis and thus adds a “feather to the cap” of confusing presentations of abdominal cocoon.

References

Author Information

Akshay Nadkarni
Department of General Surgery, Kasturba Medical College

B. H. Anand Rao
Department of General Surgery, Kasturba Medical College

Siddharth Bhandary
Department of General Surgery, Kasturba Medical College

Prashanth Shetty
Department of General Surgery, Kasturba Medical College

Rohit Jain
Department of General Surgery, Kasturba Medical College