Tuberculous Abdominal Cocoon With Internal Hernia - A Case Report And Review Of Literature

M Ajitha, V Abhishekh, U Mohan

Abstract
A 14-year-old girl was presented with features suggestive of intestinal obstruction. Past history revealed diagnosis of abdominal tuberculosis and default of anti-tuberculous therapy. Clinically, a globular mass was felt in epigastrium and umbilical region and free fluid was appreciated in the abdomen. An erect X-ray of the abdomen showed multiple air-fluid levels in the upper half of the abdomen. CECT of the abdomen revealed abnormally positioned crowded small-bowel loops with mild dilatation and multiple air-fluid levels. A provisional diagnosis of intestinal obstruction was made and the patient was taken up for explorative laparotomy which showed a well-formed thick capsule encasing the whole small bowel, the stomach and the large intestine except descending and sigmoid colon. On opening the capsule and releasing the bowel there were features suggestive of internal hernia with displaced superior mesenteric vessels. The histopathology of the capsule wall showed chronic non-specific inflammatory reaction and mature fibrous tissue. A diagnosis of abdominal cocoon with internal hernia secondary to abdominal tuberculosis was made.

INTRODUCTION
Abdominal cocoon presenting with intestinal obstruction is a rare finding with most cases of abdominal cocoon being primary or idiopathic. Cases of abdominal cocoon secondary to tuberculosis have been reported in literature and are on the rise in countries like India with increasing cases of tuberculosis. Internal hernia within the secondary abdominal cocoon is a rare finding and has not been reported in literature.

CASE REPORT
A 14-year-old girl was admitted to the emergency department with history of abdominal pain and distension since 10 days. Past history revealed similar episodes twice in the last year which resolved spontaneously. Past records showed workup for abdominal tuberculosis with a serum adenosine deaminase level of 49.1 U/L and ascitic fluid analysis of the exudative type with 95% lymphocytes. The patient had been started on anti-tuberculosis treatment with steroids based on the findings. The patient had defaulted on the treatment after taking it for 2 months.

Clinical examination of the patient revealed an anxious look and mild dehydration. Pulse was 110/minute, temperature 37.8°C, blood pressure 110/70 mmHg. There was no cyanosis or jaundice. No abnormalities of the chest or cardiovascular system were found. Local examination of the abdomen revealed a distended abdomen, hypoactive bowel sounds, and mild tenderness with free fluid in the abdomen. A tender lump was palpated in the umbilical region with mild guarding. There was no hepatomegaly or splenomegaly and rectal examination was unremarkable.

Routine laboratory workup revealed a total leukocyte count of 9300 cells/ml, hemoglobin of 10.9 g %, and normal serum chemistry and normal urine analysis. PA X-ray of the chest was normal but plain standing X-ray of the abdomen showed multiple air-fluid levels with no free air under the diaphragm. Contrast enhanced CT revealed abnormally positioned, concentric, clumped, dilated bowel loops in the upper abdomen, two below the level of the superior mesenteric artery and one left of the duodenojejunal junction. A thin capsule encasing the small and large bowel was seen. Moderate ascites was noted in lower abdomen and pelvis (figure 1-4).

Under a provisional clinical diagnosis of mechanical small-bowel obstruction, emergency laparotomy was performed through a midline incision. After opening the peritoneum, there was an encapsulated mass in the upper abdomen with brownish fluid occupying the lower abdomen and pelvis. The whole small bowel and part of the large bowel was
covered by a dense whitish and approximately 4mm thick membrane which gave the appearance of a cocoon (figure 5). At regions the capsule was densely adherent to the anterior abdominal wall. The membrane enveloping the small bowel was incised carefully and separated from the intestinal serosa by sharp and blunt dissection (figure 6). There appeared a clump of bowel enclosed in a thin sac which displaced the colon downward and the mesenteric vessels appeared twisted with displaced duodenojejunal junction (figure 7, 8). The sac was dissected and the whole small bowel was freed and followed from Treitz to ileocecal junction. The whole freed small bowel was viable and the colon was normal (figure 9); thus no further surgical procedure was deemed necessary. The cocoon wall and part of omentum were sent for histopathology examination which revealed necrotic material with chronic inflammation reaction and predominantly dense fibrocollagenous tissue.

The patient had an uneventful post-operative period and was discharged from the hospital on the 10th post-operative day after starting on anti-tuberculosis treatment. She has been regularly attending our follow-up clinics for the last six months and has remained free of bowel symptoms throughout this period.

**Figure 1**

Figure 1: CT scan showing bowel loops occupying the upper abdomen with ascitic fluid in lower abdomen and pelvis.

**Figure 2**

Figure 2: CT scan showing compacted bowel loops.

**Figure 3**

Figure 3: CT scan showing dilated bowel loops within the cocoon.

**Figure 4**

Figure 4: CT scan with an arrow showing areas of collapsed and compacted bowel.
Figure 5
Figure 5: Intra-operative findings with an arrow showing the abdominal cocoon in the upper half with empty lower abdomen.

Figure 6
Figure 6: Initial opening of the cocoon and dissecting of small bowel.

Figure 7
Figure 7: Internal hernia with sac dissected showing twisted mesenteric vessels.

Figure 8
Figure 8: Reduced internal hernia with defect.
DISCUSSION

Intestinal obstruction is a commonly encountered surgical emergency, and usually occurs secondary to intestinal adhesions, bands and obstructed hernias. However, at times rare causes of intestinal obstruction may be encountered such as the 'abdominal cocoon', also known as 'sclerosing encapsulating peritonitis' (SEP), which is a rare condition that is characterized by the encasement of the small bowel by a fibrocollagenic cocoon-like sac.

Abdominal cocoon is an infrequently reported clinical entity. Since the earliest description by Owtschinnikow in 1907, entitled “peritonitis chronica incapsulata”, the condition has been named differently by various authors. The term ‘abdominal cocoon’ was first applied by Foo et al. in 1978 [1].

Sclerosing encapsulating peritonitis (SEP) is an acquired condition that is often idiopathic.

It is of two forms – primary or idiopathic and secondary.

The primary form of the disease is commoner, and has been classically described in young adolescent females from the tropical and subtropical countries. Here, the exact stimulus for the inflammatory reaction is not known, but some suggest that it may arise due to a subclinical primary viral peritonitis, as an immunological reaction to gynecological infections, or due to retrograde menstruation. However, since this condition has also been seen to affect males, premenopausal females and children, there seems to be little support for these theories.

The secondary form of sclerosing peritonitis has been reported in association with practolol intake [2], chronic ambulatory peritoneal dialysis [3], ventriculoperitoneal [4] and peritoneovenous shunts [5], sarcoidosis [6], SLE [7], liver cirrhosis.

Constrictive pericarditis being treated with propranolol, intraperitoneal instillation of drugs, tuberculosis [8], familial Mediterranean fever, gastrointestinal malignancy, protein-S deficiency, liver transplantation [9], fibrogenic foreign material, leiomyomata of the uterus, endometriotic cysts [10] or tumours of the ovary and luteinized ovarian thecomas are the other rare causative factors.

Only occasional cases of SEP occurring secondarily to a tuberculous etiology have been reported in medical literature numbering about 12 cases. The largest case series is by Robin Kaushik et al. of 6 cases [11], 3 cases by Mohanty et al. [24] and one each by Foo et al. [1], Sahoo et al. [12] and Laloo et al. [8].

In 1992, Yip et al. [13] described four cardinal signs for suspected preoperative sclerosing encapsulated peritonitis: young woman without clear cause that justifies the intestinal obstruction, a history of similar episodes that resolve spontaneously, palpable abdominal mass with pain and clinical presentation of abdominal pain and vomiting without having a complete obstruction.

Histopathological examination of the encapsulating membrane constantly shows thickened vascular fibrocollagenous tissue, with or without areas of lymphocyte and plasma cell infiltrates. A covering of mesothelial cells may be found in focal areas. Mesenteric lymph nodes demonstrate non-specific reactive hyperplasia and organisms are typically absent.

Ascitic fluid analysis for ADA levels can be used to diagnose tuberculosis.

The preoperative diagnosis of abdominal cocoon requires a high index of suspicion, supported by clinical data and imaging findings indicative of the condition.

The classic barium finding described by Sieck et al. [25] is a serpentine or concertina-like configuration of dilated small bowel loops in a wedged U-shaped cluster.

On sonography, encasement of most of the small-bowel loops in a thick fibrous membrane and arrangement of the loops in a concertina shape with a narrow posterior base,
having the overall appearance of a cauliflower [14], are characteristic features of an abdominal cocoon. Hollman et al. [15] described characteristic sonographic findings with changes of peristalsis, tethering of the bowel to the posterior abdominal wall, intraperitoneal echogenic strands, and membrane formation during the late stage of the disease.

The CT appearance of abdominal cocoon has rarely been reported. It may demonstrate adherent small-bowel loops encased within a thick enhancing peritoneal membrane. Other CT features of SEP include signs of obstruction, agglutination and resection of intestinal loops, mural thickening, ascites and localized fluid collections, peritoneal thickening and enhancement, peritoneal or intestinal mural calcifications, and reactive adenopathy [16]. Ascites is uncommon in the endemic form of abdominal cocoon [17].

According to Kawanishi et al., the SEP can be classified into three phases that would allow us to guide the diagnosis and treatment based on the ability of peritoneal response to this offending agent mode [18]:

In the first phase, the peritoneum undergoes a marked acute inflammatory reaction with increased acute phase reactants. At that time the use of steroids becomes important.

In the second phase begins the process of encapsulation, which is accompanied by a decrease in the inflammatory component and the onset of obstructive symptoms. The therapeutic approach in this phase should be directed to bowel rest with total parenteral nutrition. In a phase in which inflammation is not as important, the doses of corticosteroids should be reduced. However, in the midst of a bowel obstruction in its initial stage, you may find a positive response to corticosteroid therapy to decrease the intestinal wall edema associated.

The progression of the disease would lead to a third phase. In this one, the main feature would be recurrent episodes of intestinal obstruction. This is the time that would have to consider surgical treatment to resolve the process with the removal of the capsule and release of peritoneal adhesions.

The base of conservative treatment is bowel rest with total parenteral nutrition. The literature seems to focus on drug treatment with corticosteroids [19]. In this sense there are studies supporting the use of prednisone and methylprednisolone in the initial stages of SEP based on its immunosuppressive effect. The use of immunosuppressants such as azathioprine, cyclosporine and tamoxifen [20], has been reported. However, studies are required to confirm the usefulness of these drugs in the treatment.

In case of secondary SEP, the underlying cause is treated, like anti-tuberculosis therapy with steroids.

The role of surgery in this disease has been classically linked to the use of common surgical procedures in intestinal obstruction; however, one must consider the indication for surgery at the time when conservative treatment does not solve the obstructive picture. At present, knowledge of the pathogenesis of the disease and the results of recent studies [21, 22] suggest that surgical treatment has an important role in the resolution of the disease.

The unfavorable results are mainly related to bowel injury during surgery and attempt to repair it. There are publications in series with 45-69% operative mortality [23] in most cases associated with intestinal resection and primary anastomosis.

Peritoneal deterioration that accompanies these patients compels us to seek a surgical solution that minimizes the risk of perforation, and the ideal method is to excise the capsule and release the peritoneal adhesions before they reach the final stage of disease where there is usually calcification of membrane.

The prognosis of abdominal cocoon after surgery seems excellent and no recurrence has been described. Only one patient in literature who had presented with long-standing symptoms and weight loss died post-operatively after subclavian vein thrombosis due to intravenous hyperalimentation.

CONCLUSION
Abdominal cocoon is a rare cause of intestinal obstruction and its possibility must be considered in cases of subacute intestinal obstruction with palpable abdominal mass.

CECT is useful in clinching the diagnosis and planning elective surgery.

Meticulous dissection of the cocoon membrane from the gut to release the entrapped intestine and separation of the inter-loop adhesions is the treatment of choice.

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1477-1481.
Author Information

M B Ajitha
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute

V Abhishek
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute

Ullikashi Mohan
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute