Abdominal Cocoon Syndrome or Idiopathic Sclerosing Peritonitis

An Extremely Rare Cause of Intestinal Obstruction

A Case Report

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ABSTRACT

Background: Abdominal cocoon syndrome is an extremely rare cause of intestinal obstruction in which loops of small bowel encapsulated by a fibro-collagenous membrane . Condition is also known in the literature as sclerosing peritonitis and in the majority of cases, it has unknown cause. Although the majority of patients exhibit long-standing signs and symptoms of partial intestinal obstruction.

Case Summary: A 15-year old female presented with complete intestinal obstruction with unknown co-morbidity and no history of previous laparotomy. Intra-operatively, the entire small bowel was found to be encapsulated by a dense fibrous sac. The peritoneal sac was excised, followed by lysis of the inter-loop adhesions with appendicectomy done at the same time. Most patients with abdominal cocoon syndrome present with features of recurrent acute or chronic small bowel obstruction secondary to kinking and/or compression of the intestines within the constricting cocoon. An abdominal mass may also be present due to an encapsulated cluster of dilated small bowel loops.

Abdominal cocoon syndrome (ACS) is an inflammatory disease, characterized by a thick fibrocollagenous membrane encapsulating the small bowel. It is a rare cause of small bowel obstruction and is divided in idiopathic and secondary syndrome. It should be taken into consideration in differential diagnosis of small bowel obstruction especially in cases of repeated episodes of bowel obstruction. The gold standard for the diagnosis of SEP is laparoscopy or laparotomy, although CT has gained ground in preoperative diagnosis of SEP. Surgical treatment seems to be the effective therapeutic option in cases of conservative treatment's failure or in severe cases of SEP, and the outcome is usually satisfactory.

Keywords: Small bowel obstruction, Cocoon abdomen, Sclerosing encapsulating peritonitis, Emergency laparotomy.

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Abdominal cocoon syndrome (ACS); is a rare condition that refers to total or partial encapsulation of the small bowel by a fibro-collagenous membrane with local inflammatory infiltrate leading to acute or chronic bowel obstruction[1,2].

Abdominal cocoon syndrome is also known as sclerosing encapsulating peritonitis, primary sclerosing peritonitis, and idiopathic sclerosing peritonitis[3]. Usually occurs in young females, it is categorized into two classes, namely, primary (idiopathic) and secondary.

If no etiology is identified, the entity is labeled as primary or idiopathic sclerosing encapsulating peritonitis (SEP), also known as the abdominal cocoon syndrome. The secondary type is seen in patients with peritoneal dialysis, peritonitis, previous abdominal surgery, sarcoidosis, and tuberculosis[4,5].

Treatment is with beta blockers, ventriculoperitoneal shunts, etc. Sclerosing encapsulating peritonitis has also been known to occur in patients with a history of liver transplants[6]. Some of the causes of secondary SEP are listed in table 1.

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Table 1 : Some of the causes of secondary sclerosing encapsulating peritonitis.

Interventional or local causes.
1- Peritoneal dialysis.
2 - Intraperitoneal chemotherapy.
3 - Liver transplant.
Systemic causes like infection, drugs, and medical disease.
1- Abdominal tuberculosis.
2 - Recurrent peritonitis.
3 - Granulomatous peritonitis.
4 - Beta-blocker.
5 - Chemotherapy.
6 - Asbestos exposure.
7 - Endometriosis.
8 - Liver cirrhosis.
9 - Gastrointestinal malignancy.

In primary or idiopathic abdominal cocoons, diverse hypotheses have been proposed to explain the possible aetiology, including retrograde menstruation, viral infection, or cell-mediated immunological response. There is even a possible congenital aetiology[7], as in some cases it is accompanied by hypoplasia of the greater omentum or altered development due to the continuous growth of the intestinal loops contained in an accessory peritoneal membrane.

Abdominal cocoon, or "primary sclerosing encapsulating peritonitis", is a rare disease of unknown aetiology reported in subtropical and tropical countries, in which the intestine is covered by membranes that cause intestinal obstruction[8]. It was first described in 1908 by Owtschinnikow and it was defined in 1978 by Foo et al[8]. In the medical literature a total of 50 cases have been reported[8,9].

It is characterized by a thick grayish-white fibrotic membrane that partially or completely encases the small bowel in a concertina-like manner. It may extend into other organs such as the large bowel, liver, or stomach, and usually presents with symptoms of recurrent intestinal obstructions[10].

This condition is usually diagnosed intra-operatively due to the difficulty of the diagnosis via laboratory and imaging modalities[11]. There are no clear guidelines regarding the management of SEP. However, surgical intervention including excision of the sac, adhesiolysis, and prophylactic appendectomy gives a good outcome[12].

Abdominal cocoon syndrome is classified into three types according to the extent of membrane encasement, and its involvement of various organs[13]. In type I, only a part of the small intestine is covered. In type II, the small intestine is covered completely. In type III, the fibro-collagenous membrane extends to cover various organs such as the stomach, appendix, cecum, ascending colon, and/or ovaries.

Symptoms are non-specific and preoperative diagnosis is extremely difficult. Computed tomography with

intravenous contrast is considered the most useful radiological examination for the diagnosis of this abnormality as far as for the decision-making[14]. In most cases, the definitive diagnosis is made during surgery[15,16].

Treatment is essentially surgical and is key for favourable patient progress. Intestinal resection of the mass is not necessary. Surgical treatment should be limited to releasing the trapped bowel by dividing and resecting the fibro-collagenous membrane, which are easily separated from the normal intestinal wall.

Thick fibrotic peritoneum encasing the small bowel partially or completely is a pathognomonic feature, and the correct diagnosis is not often made preoperatively.

Case Report

A 15-year-old female presented to the emergency department in Al-Shaheed Muhammed Baqir Al-Hakeem Hospital complaining for three days of severe colicky abdominal pain, repeated vomiting and absolute constipation with loss of weight more than five kilogram in the last three months.

She complained from similar episodes in the past with many hospital admissions, which were resolved on conservative management. The patient had no history of tuberculosis or past positive contacts. There was no history of previous surgery, and no family history of similar presentation.

Clinical examination revealed a moderately ill-looking slightly dehydrated young girl, temperature 38°C; BP 100/60 mmHg; pulse 90/min; RR 26/min.

Abdomen was tender and moderately distended on the left side of abdomen. A palpable fixed mass occupied the entire left side of abdomen. Bowel sounds were sluggish.

Plain abdominal x-rays in erect position revealed mildly dilated, gas-filled small and large bowel loops with few air-fluid levels, (Figure 1).

Because of the presence of a palpable mass on abdominal examination with a possible tumour origin, ultrasound examination and abdominal-pelvic CT scan were ordered. CT scan of abdomen and pelvis showed clustered, dilated small bowel loops in the same location as the mass, and adjacent structures had been displaced with occasional gaseous shadows, but does not confirm the diagnosis of primary sclerosing encapsulating peritonitis (PSEP) and was diagnosed as complicated intussusception of small-bowel, (Figure 2).

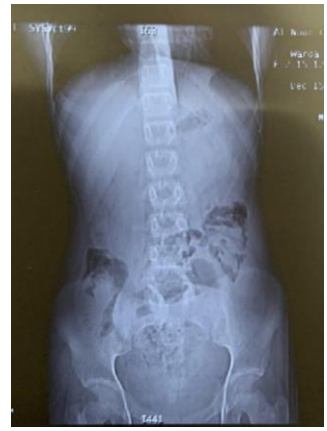

Figure 1: Plain abdominal x-ray in erect position; revealed mildly dilated, gas-filled small and large bowel loops with few air-fluid levels.

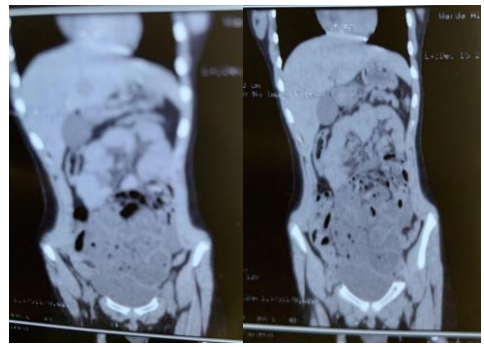

Figure 2A: CT scan of abdomen (Sagittal section) showed clustered, dilated small bowel loops.

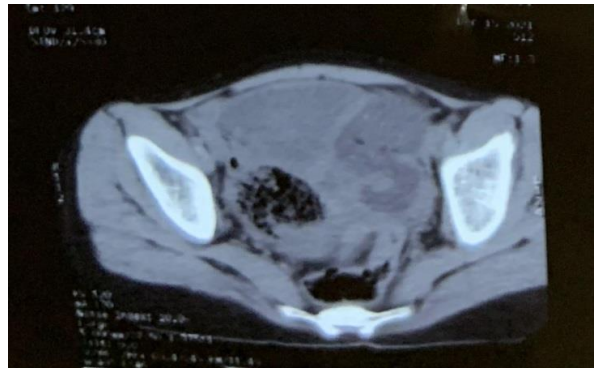

Figure 2B: A CT scan of the abdomen (axial section) showing clustered, dilated loops of small bowel.

The patient underwent exploratory midline laparotomy revealed the entire small bowel encapsulated in a thick, whitish fibrous sheath that was elastic, drawn back and adhered to itself from the duodenojejunal junction (Treitz angle) to the ileocecal valve, ascending colon and both ovaries , which is described in the literature as a “cocoon”, (Figure 3).

We dealt with the sheath, which was separated by blunt dissection from the intestinal wall, which was normal in appearance, complete excision of this membrane and adhesiolysis were carried out without any need for bowel resection from the duodenojejunal junction till the ileocecal junction, (Figure 4) with appendicectomy done at the same time, (Figure 5), which was inflamed and encapsulated by a fibro-collagenous membrane, and this led to relief of the obstruction. Biopsies were taken and samples of the membranes were sent for histopathology, (Figure 6).

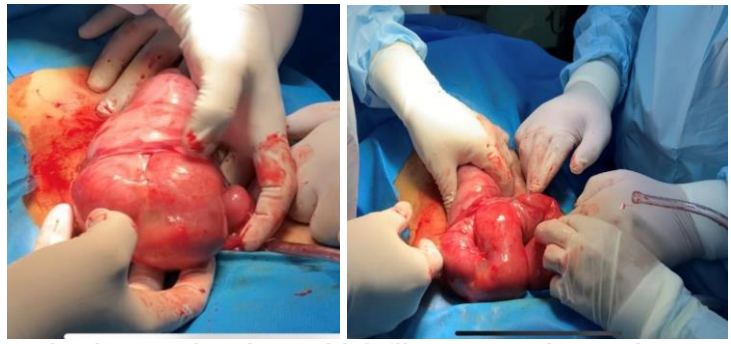

Figure 3: Intraoperative image showing a thick fibrous membrane that resembles a cocoon.

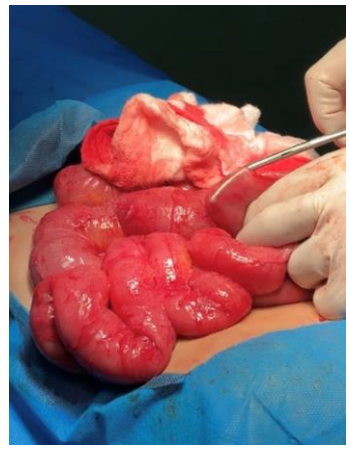

Figure 4: Intraoperative image showing complete excision of fibrotic membrane and adhesiolysis.

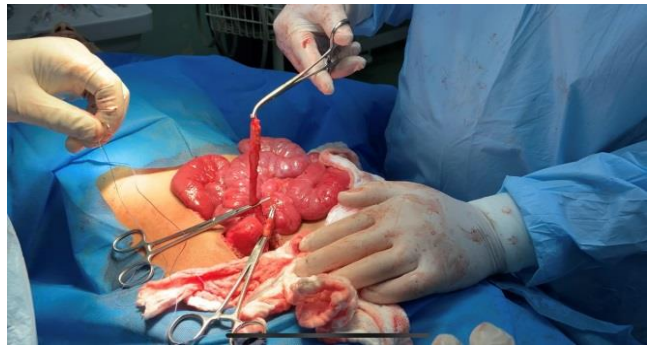

Figure 5: Intraoperative image showing appendicectomy.

Figure 6: The excised whitish fibrous sheath.

The postoperative progress was favourable, and the patient received parenteral nutrition for three days with a very good results, oral feeding started after three days.

She was discharged home after one week in a good condition. A two-week later follow-up visit was scheduled which was satisfactory with a good outcome.

Preoperative diagnosis of this syndrome as the cause of her intestinal obstruction was not made until at laparotomy, when a thick fibrotic peritoneal wrapping of the bowel in a concertina-like fashion with some adhesions was found.

The pathology study reported whitish-grey membranes comprised of a fibrous tissue with a moderate amount of capillaries and chronic inflammatory infiltrate. There was no evidence of neoplasm or specific granuloma in the sample examined, which excluded abdominal tuberculosis, (Figure 7).

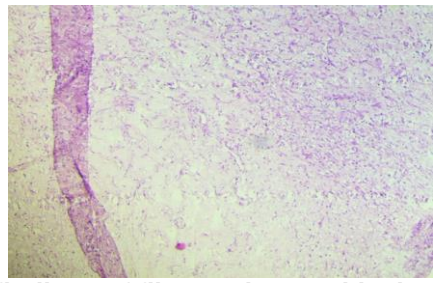

Figure 7: Histopathological findings of fibrous tissue with chronic inflammatory infiltration.

Discussion

Abdominal cocoon syndrome is an inflammatory disease, characterized by a thick fibrocollagenous membrane encapsulating the small bowel. It is a rare cause of small bowel obstruction and is divided in idiopathic and secondary syndrome. It should be taken into consideration in differential diagnosis of small bowel obstruction especially in cases of repeated episodes of bowel obstruction.

The gold standard for the diagnosis of SEP is laparoscopy or laparotomy, although CT has gained ground in preoperative diagnosis of SEP.

Surgical treatment seems to be the effective therapeutic option in cases of conservative treatment's failure or in severe cases of SEP, and the outcome is usually satisfactory.

Primary sclerosing encapsulating peritonitis or abdominal cocoon syndrome is a rare disease entity in which there is a thick, grayish-white membrane made of fibrous tissues and collagen enclosing the abdominal structures partially or totally, a cocoon-like sac[12]. There is no identifiable cause for primary sclerosing encapsulating peritonitis, although a role of cytokines, fibroblasts, and angiogenic factors has been suggested[17]. It classically presents in young females from tropical and subtropical countries[12].

The aetiology of the primary form is uncertain with various hypotheses, although it is probably caused by a subclinical peritonitis leading to the formation of a cocoon[1,8,14]. Foo et al detected the condition in 10 young girls with symptoms of bowel obstruction two years after menarche and postulated that a chemical peritonitis was caused by retrograde menstruation, leading to the formation of a cocoon[8].

Secondary SEP, where an identifiable etiology is present, is more common and associated with peritoneal dialysis, peritoneal venous shunting, beta-blocker use, penetrating abdominal injuries, tuberculosis, sarcoidosis, as well as liver transplantation[6,18,19]. The placement of Le Veenshunts for refractory ascites[19], continuous ambulatory peritoneal dialysis[20], systemic lupus erythematosis and the use of povidone iodine for abdominal wash-out[21] as well as the adrenergic blocker practolol[22]. Practolol has been withdrawn from use because it was noted to cause the formation of a peritoneal membrane.

In the current case, we believed she had primary sclerosing encapsulating peritonitis type 3, because the patient had no history of tuberculosis or any positive contacts. There was no history of previous abdominal surgery and no family history of similar presentation. Surgery revealed that the entire small bowel from the duodenojejunal junction to the ileocecal valve, ascending colon and both ovaries encapsulated by a thick, whitish fibro-collagenous membrane.

Clinically, most patients with ACS present with features of recurrent acute or chronic small bowel obstruction secondary to kinking and/or compression of the intestines within the constricting cocoon[1,8,14]. An abdominal mass may also

be present due to an encapsulated cluster of dilated small bowel loops.

Abdominal X-ray findings are non-specific. Contrast enhanced CT scan is a useful tool for preoperative diagnosis of abdominal cocoon. The imaging features are, however, not pathognomonic. CT findings of a membrane enveloping loops of small bowel were seen in some paraduodenal hernias, abdominal cocoon, and in peritoneal encapsulation[19]. However, the clinical and pathological features of these entities are different.

Thus, in idiopathic SEP cases, the diagnosis and treatment are based on surgical operation and histopathological examination of peritoneum[12].

Treatment, as in the present case, consists of excision of the accessory peritoneal sac with lysis of the inter-loop adhesions. Bowel resection is unnecessary unless a nonviable segment is found.

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