Abdominal Cocoon Syndrome as a cause of Intestinal Obstruction: A Case Report

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Abstract: Abdominal cocoon syndrome is also known as sclerosing encapsulating peritonitis, characterized by small bowel encapsulation by a fibro-collagenous membrane or “cocoon”. It is a rare cause of intestinal obstruction and has been reported predominantly in adolescent girls living in tropical/subtropical region. The cause and pathogenesis of the condition have not been elucidated. Prolonged administration of practolol, meconium peritonitis, and tuberculous infection of the female genital tract have been incriminated as possible causes. Timely and accurate imaging and diagnosis is important to avoid morbidity and mortality. Preoperative diagnosis is difficult. It is usually diagnosed during surgery. Simple excision of the membrane and lysis of the adhesions produces optimal results. Breaking of adhesions needs to be done carefully; to prevent damage to serosal surface and perforation. This case report is of a 38yr old lady who presented with sub-acute intestinal obstruction that was secondary to an abdominal cocoon and was managed by Laparoscopic surgery in our hospital.

Keywords: Abdomen, Cocoon, Intestinal Obstruction, Sclerosing Encapsulating Peritonitis, Subacute Intestinal Obstruction

1. Introduction

Abdominal cocoon refers to total or partial encapsulation of the small intestine by a fibro-collagenous membrane leading to acute or partial small bowel obstruction. It has been described by various names including ‘peritonitis chronica fibrosa incapsulata’ and sclerosing encapsulating peritonitis [1].

Two varieties of this condition are reported: primary or idiopathic and a more common secondary variety. Various hypotheses have been suggested for pathogenesis of the primary variety, first described by Foo et al., in 1978 [2]. The secondary form has been reported to be associated with use of β blockers (Practolol), abdominal Tuberculosis, chronic ambulatory peritoneal dialysis (CAPD), sarcoidosis, Systemic Lupus Erythematosus, cirrhosis and ventriculoperitoneal (VP) and peritoneovenous shunts [3].

Accurate diagnosis of this uncommon cause of a common surgical emergency is challenging and requires a high index of suspicion.

2. Case Report

A 38yr old lady presented with nausea, vomiting, and abdominal pain which started a few days before. She had no similar episodes before. No history of previous abdominal operations or medication. Patient was clinically stable. Local examination of the abdomen revealed a soft abdomen with normal bowel sounds and minimal tenderness in the left upper quadrant. There was no guarding or rigidity. A vague
mass was palpable left subcostal region. Findings of laboratory studies were within normal limits. CECT Abdomen revealed a long segment of proximal jejunum mildly dilated and densely adherent to each other in the left side of the abdomen antero-medial to the descending colon, the adherent bowel loops are seen to form a oval shaped well defined mass, there is evidence of fecal stasis – findings are suggestive of either cocoon abdomen or internal hernia “Figure 1”.

Figure 1. Mildly dilated proximal jejunum encapsulated in a fibro-collagenous membrane “cocoon”.

Laparoscopy and proceed was decided upon with a pre-operative diagnosis of acute mechanical intestinal obstruction. Per operatively, the internal herniated jejunum was found encased in a cocoon-like fibrotic tissue. Intraoperatively the fibrous membrane was excised and the small bowel was released. Rest of the abdominal visera appears to be normal. The fibrous membrane was sent for histopathological examination “Figure 2”.

Figure 2. (A) jejunum encapsulation by a fibro-collagenous membrane, (B) fibrous membrane excision.

Circulation of the bowel segment was intact; therefore, no resection was needed during the operation. On the fourth post-operative day the patient was discharged. Histopathological report – Mesothelial tissue with no histopathological abnormality, membranous layer. No evidence of granuloma or malignancy.

3. Discussion

Abdominal cocoon has been classified as primary and secondary based on whether it has a definite cause or not [1]. The various possible actiologies of the more common secondary type has been discussed in the Introduction. In view of the fact that tuberculosis is highly prevalent in our part of the world, it is likely to have a role in the formation of cocoon due to retrograde infection via fallopian tubes from subclinical pelvic inflammatory disease/genito urinary tuberculosis [4].

The idiopathic or primary variety is probably caused by a subclinical primary viral peritonitis leading to the formation of a cocoon or as an immunological reaction to gynaecological infections, or due to retrograde menstruation. It has been reported to primarily affect young females from tropical and subtropical regions. However, reports of this condition in adult males, premenopausal women and children from temperate zones are found in the literature [5].

The fibrocollagenous membrane can extend to involve other organs like the large intestine, liver and stomach [6].

Clinical presentation is nonspecific and can present with recurrent episodes of acute, subacute or chronic small bowel obstruction, weight loss, nausea and anorexia, palpable abdominal mass, vague abdominal pain though some patients may be asymptomatic [5]. Abdominal or pelvic masses may appear due to an encapsulated cluster of dilated small bowel loops. Recurrent acute or chronic small bowel obstruction is usually secondary to kinking and/or compression of the intestines within the constricting cocoon [3].

Radiological features are non specific but a better awareness of this entity and imaging techniques may facilitate pre-operative diagnosis [1]. Plain radiographs and oral contrast studies may show partial or complete small bowel obstruction, with a circumscribed conglomerated mass of bowel loops with delayed passage of contrast. Ultrasonography and computed tomographic (CT) studies are of more specific diagnostic value, and may show a mass of tightly bound small bowel loops surrounded by a thick rim of echo-poor tissue. Loculated ascitic fluid, thickened peritoneal membranes, and adherent bowel segments of various diameters are commonly seen. The imaging features are, however, not pathognomonic [7]. Similar CT findings have been reported in some cases of paraduodenal hernias [3].

Due to nonspecific clinical presentation and radiological findings, most cases are diagnosed incidentally at laparotomy [8].

Radiological findings, especially CT findings, may suggest the diagnosis but final diagnosis is generally by surgery, as confirmed by the ACS literature and summarized in (Table 1).
Management of abdominal cocoon is controversial. But in most cases surgical intervention is required. During surgery, a careful dissection and excision of the covering membrane and lysis of interloop adhesions is needed to free the entire length of small gut. The membrane often needs to be stripped from the small bowel until the loops lie free. Every effort must be made to avoid iatrogenic bowel injury during this process. Resection of bowel is indicated only in cases of nonviability to avoid complications of postoperative intestinal leakage and short-bowel syndrome [5]. Use of laparoscopic approach has also been suggested in the literature [9] but one must be careful while establishing a pneumoperitoneum to avoid injury to the bowel loops. An open approach to establishing pneumoperitoneum should be preferred [10]. In our case, underwent laparoscopic surgery and pneumoperitoneum created by Hasson’s technique (an open approach to establish pneumoperitoneum). Manifestations of abdominal tuberculosis such as mesenteric abscesses, enlarged and caseating mesenteric lymph nodes, and tubercles over the bowel serosa must be looked for during surgery, especially in countries where prevalence of abdominal TB is high [6]. Such features were found in 5/9 patients ultimately diagnosed to have TB.

No surgical treatment is required in asymptomatic cases.

The long-term prognosis of cocoon abdomen after lysis of adhesions is usually excellent [9]. In the presence of histological evidence of tuberculosis, anti-tubercular medications should be started and the outcome in terms of symptomatic relief is generally good.

4. Conclusion

Abdominal cocoon is a rare condition causing intestinal obstruction and diagnosis requires a high index of suspicion because of the nonspecific clinical picture. CECT of the abdomen is a useful radiological tool to aid in preoperative diagnosis. Peritoneal sac excision and adhesiolysis is the treatment and the outcome is usually excellent.

References


