

Abdominal cocoon: a rare cause of intestinal obstruction

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ABSTRACT

The abdominal cocoon was described as a rare condition characterized by small bowel encapsulation leading to intestinal obstruction. In it a part or whole of small bowel is encapsulated within a dense fibrous membrane. It is also called as idiopathic sclerosing encapsulating peritonitis (SEP) or encapsulating peritoneal sclerosis (EPS). We report a young female who presented with intestinal obstruction with localized mass palpable on abdominal examination. At surgery laparotomy was performed and found partial encasement of small bowel within a dense fibrous sac and causing obstruction and the diagnosis of abdominal cocoon with acute intestinal obstruction was made. The sac was excised and the gut interloop adhesions were lysed. Outcome was satisfactory.

Keywords : Abdominal cocoon, sclerosing encapsulating peritonitis, intestinal obstruction, dense fibrous adhesions

INTRODUCTION

Abdominal cocoon is a rare condition that refers to total or partial encapsulation of small bowel by fibrocollagenous membrane with local inflammatory infiltrate leading to acute or chronic bowel obstruction. Abdominal cocoon is otherwise termed “encapsulating peritoneal sclerosis”(EPS) concisely describes the essential pathological features of that condition: intestinal encapsulation resembling a cocoon, and peritoneal membrane damage (peritoneal sclerosis). Previously, EPS had been called sclerosing encapsulating peritonitis (SEP). However, because inflammatory features are not necessarily apparent in advanced EPS cases, “encapsulating peritoneal sclerosis” has been proposed as a more accurate description of the disease entity. Abdominal cocoon was first described by Owtshimatous in 1907 ‘as Peritonitis Chronic Fibrosa Encapsulata’ and termed ‘Abdominal cocoon’ by Foo in 1978¹. It is also called as ‘abdominal cocoon syndrome’. The condition is acquired and the cause is usually unknown.

CASE HISTORY

A 35 year old female presented to the surgery emergency with complaints of abdominal pain and distension for the past 5 days. Patient gave history of similar complaints

six months back which was managed conservatively. Patient gave history of constipation since five days, patient was afebrile, there was a history of 4 to 5 episodes of vomiting. There was no history of any medical disease or cough in the family. On admission, pulse rate was 80 pm; BP- 140/90mmHg; Sr Bilirubin 2mg/dl. Systemic examination revealed no abnormality.

Examination of abdomen revealed distension, diffuse pain and sluggish bowel sounds. A vague mass was palpable in the periumbilical region with mild tenderness. X ray of erect abdomen showed 3 to 4 multiple dilated gut loops and no gas under the diaphragm. On contrast enhanced computer tomography (CECT) abdomen showed multiple dilated small bowels adherent to each other forming a mass with free fluid, suggestive of acute intestinal obstruction [Figure 1].

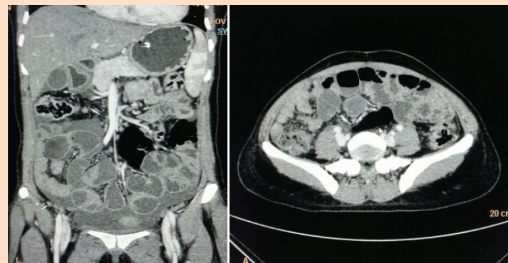


Figure 1: CECT showing multiple dilated small bowel loops.

Patient was provisionally diagnosed as acute intestinal obstruction. Exploratory laparotomy was planned and performed through midline incision, it revealed a dense white fibrous membrane encasing distal jejunal and ileal loops as a mass/cocoon along with 500ml of straw coloured fluid [Figure 2].



Figure 2: Figure showing dense white fibrous membrane encasing distal jejunal and ileal loops as a mass/cocoon.

On opening the membrane, small gut loops showing obstruction at multiple levels were noted. Adhesionlysis was performed with sharp dissection to release the gut loops [Figure 3]. The intestinal content was drawn off into the colon.

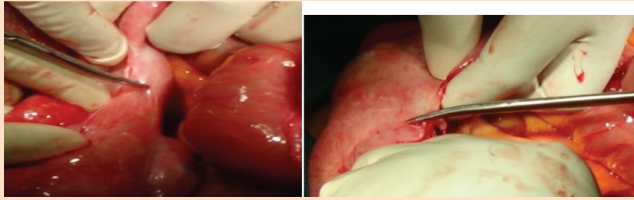


Figure 3: Figure showing pseudosac encasing small bowel along with fibrous membrane. Sharp dissection is given (adhesionlysis)

The white fibrous membrane pieces were sent for histopathological examination (HPE) which revealed pseudosac externally lined by flattened and at places by low cuboidal epithelium underneath showed moderate to focal dense collection of spindle to elongated cells in loose collagen tissue [Figure 4]. Foci of congested vessels and sparse collection of lymphocytes were seen, the features suggestive of chronic non specific inflammation. Post operative period was uneventful.

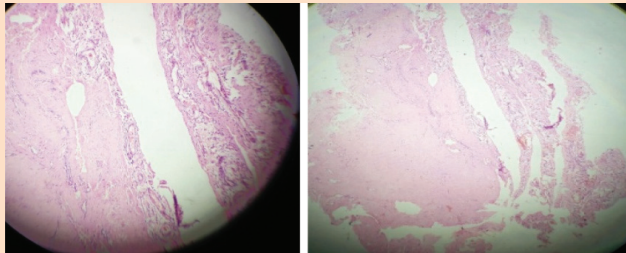


Figure 4: Section showing lining flattened to cuboidal epithelium with collection of inflammatory cells, collagen and fibrous tissue. [H&E,x10]

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DISCUSSION

The definition of SEP is associated with confusion and lack of information. The concepts of primary and secondary SEP are erroneously used interchangeably in many previously

published articles on SEP. Thus, we aimed to emphasize the correct use of the definitions of peritoneal encapsulation (PE), abdominal cocoon, idiopathic SEP, and secondary SEP in the present review.

PE was first described by Cleland in 1868. It is a developmental anomaly characterized by the congenital presence of an accessory peritoneal membrane, which is believed to be derived from the yolk sac peritoneum in the early stages of fetal life^{2,3}. This peritoneal membrane is classically found between the mesocolon and omentum, and most of the small intestines lie posterior to this membrane. In other words, PE is an anatomical anomaly unrelated to any inflammatory process.

Abdominal cocoon may be classified into primary or idiopathic and secondary forms depending on its underlying cause and the pathogenetic properties of fibrocollagenous membrane. Abdominal cocoon is categorized into three types according to the extent of the encasing membrane that covers the intestine⁴. Encasement of part of the intestine by a fibrocollagenous membrane is called type 1 cocoon syndrome, complete coverage of the intestine by the membrane is called type 2 abdominal cocoon syndrome, type 3 cocoon syndrome refers to encasement of the whole intestine, as well as other intra-abdominal organs, such as the appendix, caecum, ascending colon, and ovaries^{5,6}. Primary abdominal cocoon occurs mainly in young females from tropical and subtropical zones. Although retrograde menstruation with or without viral infection of the fallopian tubes has been suggested as a possible cause.

Secondary abdominal cocoon is apparently associated with predisposing factors such as -recurrent peritonitis, peritoneal dialysis, abdominal koch's, intake of intra peritoneal antibiotics or calcium channel blockers, sarcoidosis, carcinoid syndrome exposure to asbestos, tubercular pelvic inflammatory diseases. The clinical presentation of abdominal cocoon vary with the severity and duration of the disease, underlying causes, and affected person's immunological status. The clinical presentation includes acute, sub acute, chronic small bowel obstruction, abdominal distension, nausea and vomiting and life threatening complications in severe cases⁷.

An accurate diagnosis is difficult to make preoperatively, because findings on biochemical investigations are normal and imaging findings are non specific. Most cases are diagnosed at laparotomy. In the cases of abdominal cocoon described in the literature till date, the diagnosis was either made during surgery for unrelated reasons, or at exploratory laparotomy in patients presenting with obstruction. Treatment in present case is usually an midline laparotomy along with excision of sac with lysis of interloop bowel adhesions; bowel resection is done if a non viable segment is found.

CONCLUSION

A high index of suspicion is required in the absence of other possible causes of the symptoms of abdominal obstruction. Radiology may not provide accurate diagnosis. Diagnosis of abdominal cocoon is best made intraoperatively and the treatment consists of excision of sac and lysis of interloop adhesions with good outcome.

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Please cite this article as: Arun, Rajendra, Ravindar rao. Abdominal cocoon: a rare cause of intestinal obstruction. *Perspectives in medical research* 2016;4:1:57-59.

Sources of Support: Nil, Conflict of interest: None declared