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Idiopathic sclerosing encapsulating peritonitis (abdominal cocoon) in adult male. A case report



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ABSTRACT

INTRODUCTION: Abdominal cocoon (sclerosing encapsulating peritonitis) (SEP) is a rare condition, mostly affecting adolescent girls living in tropical/subtropical region. Its etiology is unknown. It may cause acute or sub-acute intestinal obstruction.

PRESENTATION OF CASE: We report here a 39 year old male who complained of long standing colicky abdominal pain, with significant weight loss. Abdomen CT scan showed clumping of ileal loops at the level of umbilicus, with a thin capsule surrounding it. Laparoscopy revealed abdominal cocoon, biopsy of which showed dense hypocellular fibro-collagenous tissue with no neoplastic or granulomatous process. Excision of fibrous tissue and release of adhesions was done. Patient was symptoms free after five months follow up.

DISCUSSION: Abdominal exploration is usually needed for the diagnosis and treatment of abdominal cocoon. A thick fibrotic peritoneal wrapping of the bowel is usually found. Complete recovery is the result in majority of cases after surgical removal of the wrap causing the cocoon.

CONCLUSION: Primary sclerosing encapsulating peritonitis (cocoon abdomen) diagnosis needs a high index of suspicion, as signs and symptoms are nonspecific and imaging findings are not always conclusive. Careful excision of the accessory peritoneal sac and lysis of adhesions between bowels is the best treatment. Prognosis is generally good.

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1. Introduction

Abdominal cocoon is a rare condition,¹ which refers to partial or total encapsulation of abdominal viscera within a dense fibrous membrane.¹ It has been referred to as “peritonitis chronica fibrosa incapsulata” by Owtschinnikow.² And was first described in details by Foo et al.¹ Abdominal cocoon is predominantly reported among females from the tropical and subtropical regions; however, cases of adult males were also reported.^{1,3} It can be classified as primary (idiopathic) or secondary.^{4,5} The correct diagnosis is not often made preoperatively,^{6,7} as CT and MRI finding are nonspecific.⁸

2. Case report

A 39 year old male was admitted to our hospital with complaints of abdominal pain for years and weight loss of 10 kg within the last year. The pain was colicky, started in center of the abdomen, radiated to the periphery, aggravated by eating. He had no nausea or vomiting, constipation or diarrhea. His appetite was good, but he was afraid to eat. He had no fever. There was no history of illnesses like: familial Mediterranean fever, tuberculosis, sarcoidosis, systemic lupus erythematosus, protein S deficiency, malignancy, ascites or peritoneal dialysis. There was no history of previous abdominal wash (e.g. by povidone iodine). Patient was not on regular medication (e.g. beta-adrenergic blocker). There is no history of travel to tropical or subtropical countries.

On examination he was thin built, with distended non tender abdomen. No organomegaly was observed. Bowel sounds were exaggerated. Digital rectal examination was normal. All his laboratory blood tests were normal. Abdomen CT scan showed clumping of ileal loops at the level of umbilicus, with a thin capsule surrounding it. Small amount of fluid is seen around these bowel loops.

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Fig. 1. Axial contrast enhanced CT scan. Note the clump of small bowel loops with minimal fluid around it inside the cocoon. Capsule of cocoon (white arrow). Loop entering through opening (black thin arrow).



Fig. 2. Para sagittal reconstructed CT image. The cocoon (white thick arrow). Proximal dilated loop (black thin arrow).

The proximal loops appear mildly dilated and distal ileal loops collapsed. No evidence of vascular axis rotation (Figs. 1 and 2).

Upper and lower gastrointestinal endoscopy were unremarkable. Diagnostic laparoscopy showed that most of the small bowel was wrapped up in a grayish-pink membrane forming a cocoon (Fig. 3). The procedure was changed to laparotomy to avoid injury as it was very difficult to release the encased bowel laparoscopically. It seems from the diagnostic laparoscopy and laparotomy view that the membrane is originated from the mesothelial covering of the root of the mesentery. Excision of the whole membrane was done carefully and was sent for pathology examination. Encased small bowel loops were separated by sharp dissection (Fig. 4).

All loops of small bowel were viable. Small bowel full thickness biopsy was taken. Two cm in diameter firm central mesenteric lymph node was excised. Appendectomy was done as the appendix was embedded in the membrane (Fig. 5). All other abdominal organs were normal. Histopathology of the membrane, showed dense poorly cellular collagenised fibrous tissue, features consistent with sclerosing encapsulating peritonitis (abdominal cocoon; Fig. 6).

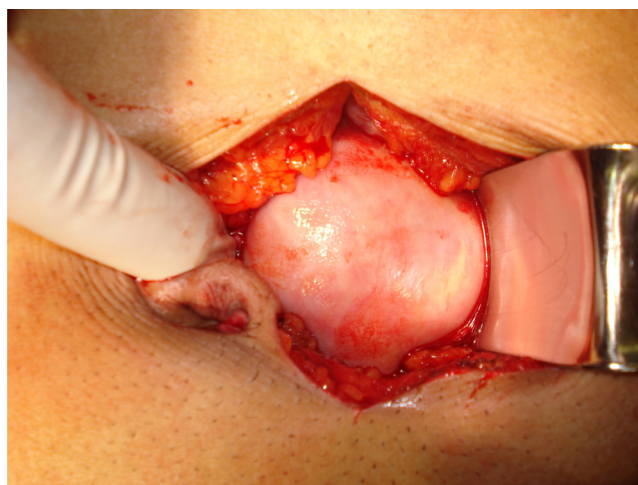


Fig. 3. Grayish-pink membrane covering the small bowel forming a cocoon. (For interpretation of the references to color in figure legend, the reader is referred to the web version of the article.)

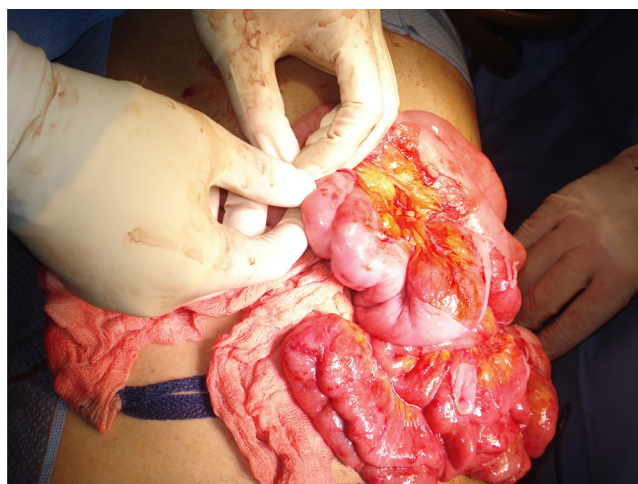


Fig. 4. Encased small bowel loops were separated with excision of the whole membrane.

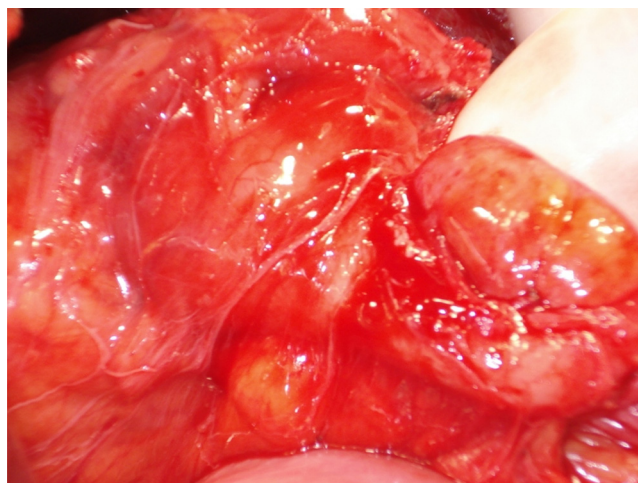


Fig. 5. The appendix was embedded in the membrane.

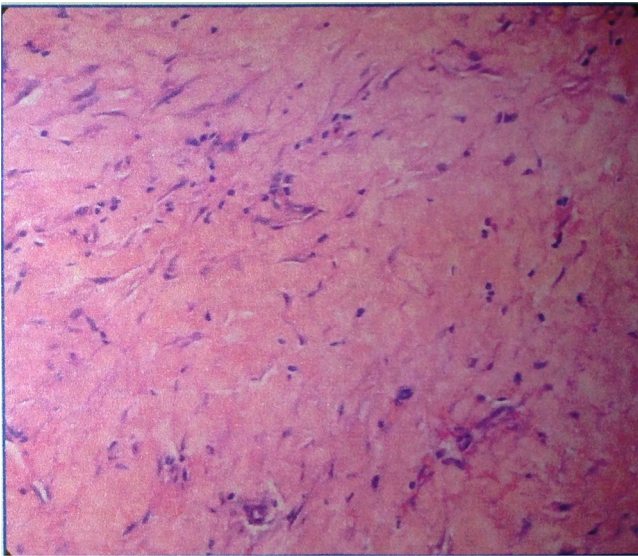


Fig. 6. High power view displaying dense poorly cellular collagenised fibrous tissue.

Small bowel, appendix and lymph node biopsy were unremarkable. Post operatively the patient showed improvement in his symptoms; with uneventful five months follow up.

3. Discussion

Sclerosing encapsulating peritonitis (Cocoon abdomen) is a rare condition.¹ With few cases described in literatures.^{1,2,7} Cocoon abdomen is mostly observed in young girls living in tropical and subtropical countries of the world, especially China, Malaysia, Singapore, Pakistan, India, Nigeria, Kenya, Saudi Arabia, Israel, and South Africa.^{1,3} Although few cases have been documented in males.⁶ It is classified as primary (idiopathic) or secondary.^{4,5} The etiology of the primary form is uncertain with various hypotheses proposed. These include retrograde menstruation with a superimposed viral infection,¹ retrograde peritonitis and cell-mediated immunological tissue damage incited by gynecological infection.^{1,6,9} However, since this condition has also been seen to affect males, premenopausal females and children, there seems to be little support for these theories.³ The secondary causes of abdominal cocoon include continuous ambulatory peritoneal dialysis, tuberculosis, systemic lupus erythematosus, sarcoidosis, familial Mediterranean fever, gastrointestinal malignancy, protein S deficiency, liver transplantation, fibrogenic foreign material, luteinized ovarian thecomas, the use of povidone iodine for abdominal washout, placement of LeVeen shunt for refractory ascites, as well as the beta-adrenergic blocker (practolol).^{4,5,8,10} In our patient, the etiology appears to be primary, as all the known causes of secondary cocoon abdomen were ruled out. Patients with cocoon abdomen may present in one or multiple episodes with symptoms of acute or sub acute small bowel obstruction (that could sometimes resolve spontaneously). In this case, the patient has recurrent attacks of colicky abdominal pain and distension, which was sometimes resolved spontaneously. Abdominal mass may also be present due to an encapsulated cluster of dilated small bowel loops.^{3,5-7} A pre-operative diagnosis is usually very difficult and requires a high index of clinical suspicion. Most cases are diagnosed intraoperatively during laparotomy performed for intestinal obstruction, or during diagnostic laparoscopy.^{6,7,9} Plain radiographs of the abdomen may suggest features of intestinal obstruction.^{6,8} Abdominal ultrasonography may be of help in the diagnosis of cocoon abdomen by revealing an echogenic mass of dilated small-bowel loops surrounded by a thick rim of hypoechoic

fibrous membrane.^{6,8} Barium follow-through may show delayed transit of contrast and clustering of the bowel loops in the pelvis.^{6,8} CT findings may include clumping of small bowel loops in the center of the abdomen encased by a soft-tissue density mantle, peritoneal thickening and calcification, clumping of small bowel loops, and lobulated fluid collections.⁸ In this study preoperative diagnosis was possible by abdominal CT scan which showed clumping of ileal loops, surrounded by a thin capsule and Small amount of fluid. The proximal loops were mildly dilated while the distal ileal loops were collapsed. Cocoon abdomen may be confused with congenital peritoneal encapsulation, which is characterized by a thin accessory peritoneal sac surrounding the small bowel, and is an incidental finding.³

Dissection (open or laparoscopic) of fibrotic membrane and adhesiolysis is usually sufficient to treat abdominal cocoon.^{3,7} Complications after surgery were reported; these include intra-abdominal infections, enterocutaneous fistula and perforated bowel.⁶

In this case laparotomy was decided as the membrane was dense and attached firmly to the bowel. The prognosis of primary Sclerosing encapsulating peritonitis is excellent. Follow-up has shown recurrence only in few cases, some of which resolved on conservative management.^{6,9}

4. Conclusion

Primary sclerosing encapsulating peritonitis (cocoon abdomen) is rare condition; with unclear etiology. It affects young patients and its diagnosis needs a high index of suspicion, as signs and symptoms are nonspecific and imaging findings are not always conclusive. Careful excision of the accessory peritoneal sac and lysis of adhesions between bowels is the best treatment. Prognosis is generally good.

Conflict of interest

The authors have no financial or personal relationships with other people or organisations to disclose.

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Ethical approval

This study had been approval by the ethics committee in Bahrain Specialist Hospital, Manama, Kingdom of Bahrain. The reference number for their judgement: 115/2014.

Author contributions

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Revision: Sunitha Jacob.

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