A problem encapsulated: the rare peritoneal encapsulation syndrome

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ABSTRACT

Peritoneal encapsulation (PE) is a rare condition that has been described interchangeably with sclerosing encapsulated peritonitis as well as abdominal cocoon. We report an otherwise well 38-year-old man who presented with two episodes of intestinal obstruction about two months apart. Computed tomography (CT) of the abdomen showed characteristic features of the PE syndrome. Exploratory laparotomy was performed and small bowel was freed of adhesions and the encapsulation. Post-operative recovery was uneventful. PE is an extremely rare congenital condition in which there is abnormal return of the midgut loop to the abdominal cavity in the early stages of development. The small intestine is thus covered by the original dorsal mesentery, forming the characteristic accessory peritoneal sac. Management of cases is difficult as CT findings may not be characteristic and may only be diagnosed at laparotomy.

Keywords: intestinal obstruction, peritoneal encapsulation syndrome, peritonitis

Singapore Med J 2006; 47(9):808-810

INTRODUCTION

Peritoneal encapsulation (PE) is a rare condition that has been described interchangeably with sclerosing encapsulated peritonitis (SEP) as well as abdominal cocoon. This is however a distinctive pathology and was first described in 1868[1]. We present a case with this condition that we believe to be the first one reported in Singapore, as well as in the region.

CASE REPORT

A 38-year-old Chinese man presented with right groin pain and swelling of two months' duration. He had no past medical history, except for previous elective right inguinal hernia mesh repair performed eight months before in another hospital. Clinical examination revealed a reducible hernia with positive cough impulse. The surgery was performed with no complications and he was well when discharged.

He subsequently presented to our institution with a five-day history of abdominal distension, colicky abdominal pain and non-bilious vomiting. Bowel sounds were normal. Blood investigations were unremarkable. Abdominal radiograph showed mildly dilated small and large bowel loops. A colonoscopy that was performed was normal. The patient was managed conservatively and he was able to pass stools spontaneously the next day, with resolution of symptoms. An outpatient small bowel series was performed. This showed normal contrast passage until the distal jejunum, where it was dilated and exhibited air-fluid interfaces when the patient was erect, consistent with subacute small intestinal obstruction at the level of the distal jejunum. The patient however defaulted follow-up from the specialist outpatient clinic.

The patient was readmitted about two months later with complaints of intermittent non-bilious vomiting of ten days' duration. He also had complaints of no bowel movement for five days and progressive abdominal distension for the past two months. The patient was dehydrated and abdominal examination revealed an asymmetrical, vague central abdominal fullness and sluggish bowel sounds. Investigations performed revealed a hypochloraemic, hypokalaemic metabolic alkalosis secondary to vomiting. Abdominal radiograph showed dilated small bowel loops and multiple air-fluid levels. Computed tomography (CT) of the abdomen revealed dilatation of the small intestine with marked dilatation of the duodenum and the stomach, with a large loculated fluid collection encasing the small bowel from the fourth part of the duodenum to the terminal ileum. This layer was thin-walled with some enhancement, but the fluid was otherwise homogeneous with no debris or layering. The rest of the abdomen was otherwise normal (Fig. 1).

Exploratory laparotomy was performed as

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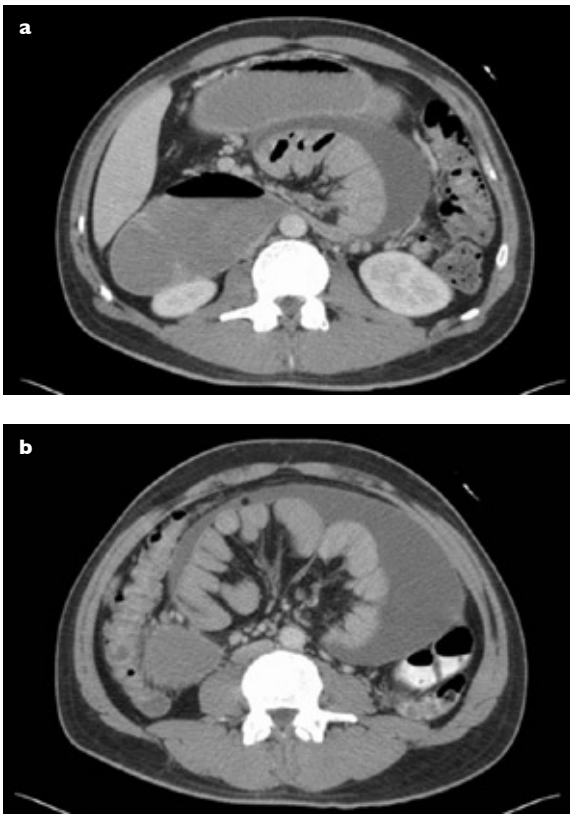

Fig. 1 Axial CT images (a,b) show encapsulated small bowel loops within a large fluid loculation and the colon.

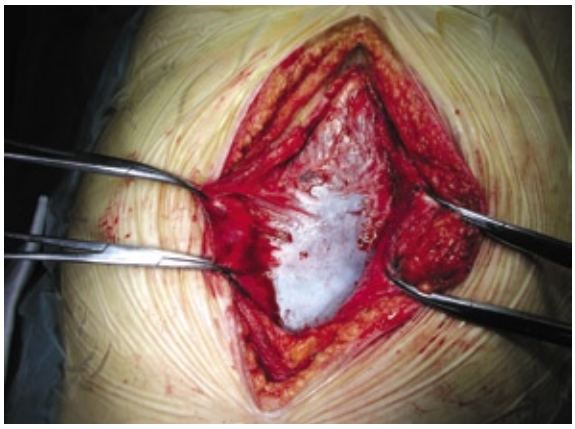

Fig. 2 Operative photograph shows the peritoneal capsule at laparotomy.

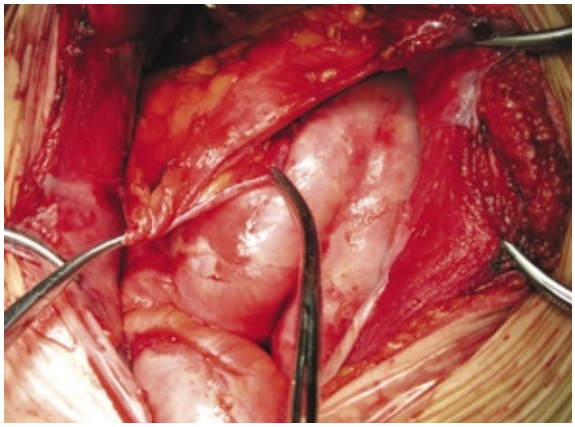

Fig. 3 Operative photograph shows the encapsulated small bowel with a dense fibrous layer. The peritoneal layer is reflected upwards.

there was no resolution of symptoms on conservative treatment. Intraoperatively, PE from the duodenal-jejunal flexure to the terminal ileum was noted, with distension of small bowel to the mid-jejunum. There was no gut malrotation. There were also 1.6 litres of haemoserous ascites within the PE. The stomach, duodenum and the large bowel were all extra-peritoneal (Figs. 2-4). The abnormal peritoneum overlying the small bowel was split along the anti-mesenteric border through the entire length of small bowel that was encapsulated and this procedure allowed greater bowel mobility. Concurrently, intestine adhesions were released. Bowel resection was not required. A layer of Sepra-film® adhesion barrier (Genzyme Corporation, Cambridge, USA) was placed over the small bowel in an attempt to prevent future adhesions from occurring. The patient had transient ileus post surgery and nutrition was maintained with parenteral nutrition for five days, until the patient could eat orally. He was discharged well on the tenth postoperative day.

Peritoneal cytology was negative for malignancy. It was also negative for acid-fast bacilli smears and cultures. Biopsy of the peritoneal lining was performed intraoperatively and histology revealed strips of dense fibrous tissue with focal areas of myxoid degeneration, haemorrhage and necrosis. In other areas, granulation tissue predominated. This was consistent with the PE syndrome.

DISCUSSION

PE is an extremely rare condition. This however is often used interchangeably with the entity of "abdominal cocoon" and "SEP". These are actually three different pathological entities and an understanding of the various aetiologies is necessary for this differentiation. SEP was first described by Owtschinnikow in 1907 under the name of "peritonitis chronic fibrosa incapsulata". This is characterised by a thick grayish-white fibrous membrane that covered the small intestinal wall. This condition is a rare complication of chronic ambulatory peritoneal dialysis, and the presence of acetate in the dialysate, as well as antiseptics used during bag exchanges, have been postulated as the factors that predispose to SEP. Beta-blocker practolol and recurrent peritonitis are additional causative factors. Other rare causes include ventriculoperitoneal and peritoneovenous shunting,

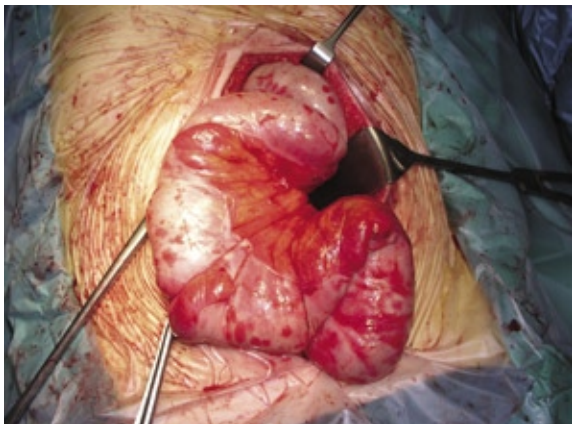

Fig. 4 Operative photograph shows abnormal peritoneum overlying the antimesenteric border of the small bowel.

sarcoidosis and systemic lupus erythematous, all of which were absent in our patient[2,3]. Lethality of this condition is extremely high and the mortality rate is close to 60%, and is associated with various surgical complications such as intestinal necrosis and frequent anastomosis breakdowns. Conservative management with success has been reported with corticosteroids, immunosuppressants and tamoxifen in SEP secondary to continuous ambulatory peritoneal dialysis (CAPD).

Abdominal cocoon is another entity that has to be differentiated. This was first described and named by Foo et al in 1978[4], and this condition was thought to be related to retrograde menstruation. However, sporadic cases have been reported in both children and men, hence the exact aetiology remains unknown. Characteristic features on barium follow-through examination are a characteristic serpentine configuration of the dilated distal small bowel within a cocoon-like structure. The small bowel is found totally or partially coiled up in a concertina-like fashion and encased in a dense white membrane on operation. It has been reported that removal of the membrane and release of intestine adhesions are adequate and bowel resection is usually not necessary[5]. PE was first described in 1868 by Cleland[1] and is characterised by the small intestine lying behind an accessory peritoneal membrane. In contrast to SEP which is an acquired pathology, PE is a congenital occurrence whereby there is abnormal return of the midgut loop to the abdominal cavity in the early stages of development. The small intestine is thus covered by the original dorsal mesentery which ordinarily forms the transverse mesocolon thus forming the characteristic accessory peritoneal sac[6].

Patients are usually asymptomatic and may present, as in our patient, with acute intestinal obstruction. Hence it is often only diagnosed at laparotomy. It has been suggested that two clinical signs may be elicited. Firstly, in view of the dense fibrous sac, only bowel proximal to and out of the fibrous layer can distend and the patient may have a fixed, asymmetrical distension of the abdomen that does not vary with peristalsis. In addition, the patient will have a difference in consistency of the abdominal wall to palpation. This is because the distended area is soft and the flat area, which is covered by the dense fibrous capsule, is firm[7]. In our patient, the entire small bowel was encased. However, there was also an adhesion band found in the mid-jejunum which caused marked distension proximal to this. Hence, asymmetry of the abdomen was evident. Patients with PE may not show characteristic radiological features. These are usually normal or show non-specific features of intestinal obstruction. Findings on CT may be suggestive of PE when small bowel is enveloped in a thin membrane, as in our patient.

Management of such cases requires urgent surgery. Our patient underwent laparotomy, with simple freeing of the adhesions as well as release of the trapped intestines. There was no resection of bowel required. In contrast to dialysis-induced SEP which reports surgical mortality of 60-80%, there is paucity of data on long-term follow-up for such patients in terms of survival, as well as disease recurrence. We remain cautious on the prognosis of this patient and plan to continue long-term follow-up for him. Our patient presented with suggestive signs as well as characteristic radiological features, enabling decisive management. Thus, recognition of this condition is important as it allows optimal management. So, although rare, clinicians should suspect this condition in patients with recurrent intestinal obstruction if no aetiological factors can be found.

REFERENCES

1. Cleland J. On an abnormal arrangement of the peritoneum with remarks on the developments of the mesocolon. J Anat Physiol 1868; 2:201-6.
2. Slingeneyer A. Preliminary report on a cooperative international study on sclerosing encapsulating peritonitis. Contrib Nephrol 1987; 57: 239-47.
3. Deeb LS, Mourad FH, El-Zein YR, Uthman SM. Abdominal cocoon in a man: preoperative diagnosis and literature review. J Clin Gastroenterol 1998: 26:148-50.
4. Foo KT, Ng KC, Rauff A, Foong WC, Sinniah R. Unusual small intestinal obstruction in adolescent girls: the abdominal cocoon. Br J Surg 1978; 65:427-30.
5. Macklin J, Hall C, Feldman MA. Unusual cause of small bowel obstruction in adolescent girls: the abdominal cocoon. J R Coll Surg Edinb 1991; 36:50-2. Comment in: J R Coll Surg Edinb 1992; 37:60.
6. Costantinides F, Di Nunno N, Bernasconi P, Jonjic N, Melato M. A new type of peritoneal encapsulation of the small bowel. Am J Surg Pathol 1998; 22:1297-8.
7. Naraynsingh V, Maharaj D, Singh M, Ramdass MJ. Peritoneal encapsulation: a preoperative diagnosis is possible. Postgrad Med J 2001; 77:725-6.