Basrah Journal Of Surgery

Original Article
Bas J Surg, September, 13, 2007

IDIOPATHIC SCLEROSING ENCAPSULATED PERITONITIS (ABDOMINAL COCOON) A CASE REPORT AND LITERATURE REVIEW

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Abstract

Sclerosing encapsulated peritonitis or abdominal cocoon is a rare disease that is characterized by a total or partial encasement of the small bowel by a thick and fibro tic membrane. Thirty eight cases were reported since it was first described. It occurs primarily in adolescent females in tropical and subtropical regions. Preoperative diagnosis is a matter of challenge and usually made at laparotomy. We report a patient with partial intestinal obstruction and abdominal cocoon that was diagnosed during surgery. We review the literature and discuss the etiology of this disease.

Introduction

Sclerosing encapsulating peritonitis (SEP) or cocoon abdomen is a rare cause of small bowel obstruction. In this condition, the bowel is partially or totally encased or wrapped by a thick fibrous membrane forming several compartments (cocoons) containing loops of small bowel. SEP usually presents with symptoms of intestinal obstruction, and pre-operative diagnosis is usually difficult. The pathogenesis of this condition remains unclear; however, it is a form of chronic irritation and inflammation, and may be summarized as primary (idiopathic) or secondarily induced[1].

Case report

A 33-year-old male, of middle-eastern ethnicity, presented at the emergency department with a history of recurrent generalized abdominal pain and vomiting of one week duration. He gave a history of a similar attack of pain one month prior. There was neither history of recent weight loss, nor loss of appetite. There was no significant past medical history or prolonged drug intake. His surgical past history is negative.

On evaluation the patient was not in distress, well nourished and hydrated. He was afebrile and heamodynamically stable. However, the abdominal examination revealed a small palpable mass with minimal tenderness in the lower right quadrant. There was no hepatomegaly or spleenomegaly. His hernial orifices were intact. Genital and rectal examination was normal. Bowel sounds were exaggerated on auscultation.

Routine laboratory workup was normal. An abdominal X-ray showed multiple air fluid levels in the central abdomen. An ultra sound examination of the abdomen and pelvis failed to identify any pathology. We proceeded with a CT scan and it revealed multiple dilated small bowel loops with air fluid levels, with clustering of small bowel loops in the right iliac fossa (figures 1&2).

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Figure 1A, B: CT scan abdomen of the abdomen and pelvis revealed a cluster of small bowel seen in the right iliac fossa encased by a fibrous sac (arrow).

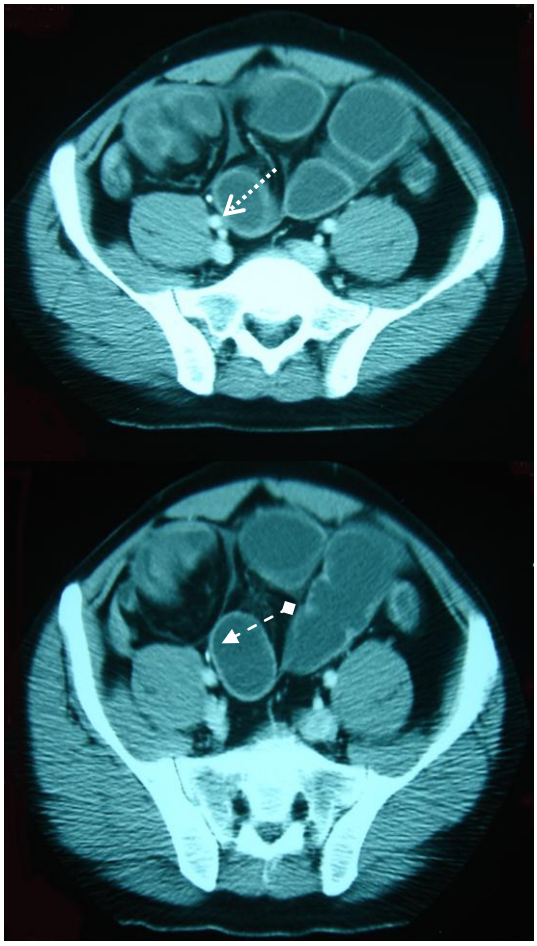
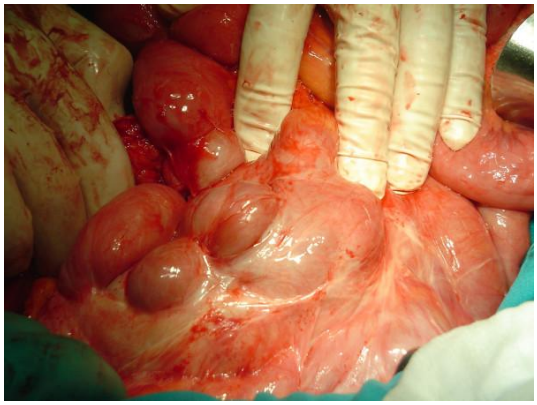


Figure 2, the cocoon encasement of the distal small bowel.



As the origin of the underlying pathology was not defined, a decision for an exploratory laparotomy was made on hospital day 3. Upon surgery, a thick transparent fibrous capsule encasing the distal small bowel was revealed, with the presence of interloop adhesions. There was no obvious origin of the sac. The liver, stomach, appendix, right and left colon, as well as the sigmoid were not included in the fibrous sac. The greater omentum looked hypo plastic and adherent to the fibrous sac (figure 3). Proximal ileum was dilated with a collapsed colon. No sings of bowel ischemia were present. Incision of the thick membrane and extensive adhesiolysis of small bowel loops were performed, at some stages simple finger dissection was sufficient. The serosa of the bowel was intact. No bowel resection was required. Normal restoration of the bowel integrity was reestablished.

Histology of the excised membrane showed thickened fibrocollagenous tissue and infiltration of mononuclear cells around blood vessels. The diagnosis of idiopathic sclerosing encapsulating peritonitis (abdominal cocoon) was established, due to intraoperative findings and by ruling-out any other condition explaining the patient's pathology. Post operative recovery was uneventful, Patient was discharged home on post-operative day 4 and he was seen in the surgical outpatient department for follow up after 6 months, he was in good health.

Discussion

Abdominal cocoon is a rare entity first described and named by Foo et al. in 1978[2] This pathology is characterized by thick fibrous membrane wrapping the bowel in a concertina-like fashion. Terms such as sclerosing peritonitis[2,3], encapsulating peritonitis[4] and sclerosing encapsulating peritonitis (SEP)[5,6] have also been used to describe this condition. A total of 38 cases were reported since abdominal cocoon was first described in 1978. Twenty seven of them were females,

and eleven males. Four of the female patients were children. Most of the patients were from tropical and subtropical regions. Reported cases are from Singapore[7], India[8-12], Malaysia[13,14], Middle East[15-17], United Kingdom[5], Nigeria[18], Australia[19], Japan[2], America[20], and Turkey[21]. To our knowledge, this is the first case reported in Kuwait.

The etiology of this condition is obscure, but it can be classified as primary (idiopathic) SEP and secondary SEP, caused by local or systemic irritants. Several theories have been proposed as possible causes of primary SEP. Foo et al.[7] reported 10 abdominal cocoons within two years of menarche, and retrograde menstruation has been incriminated as a cause of primary peritonitis. Infection via the fallopian tubes was proposed also as a possible cause. The male patients, premenopausal sisters and children reported before, weaken both hypotheses. Some drugs, especially the beta-adrenergic blocker 'practolol' has been suggested as a possible cause of secondary SEP, because they may lead to enhanced collagen production and subsequent fibrosis. Elteringham et al.[3] reported 9 cases undergoing practolol therapy who developed peritonitis which caused sclerosing peritonitis. Brown et al.[22] and Windsoret al.[23] reported similar cases. Practolol is no longer in general use, but cases continue to emerge. LeVeen shunts also have been proposed as a possible cause of abdominal cocoon[21], but its direct relation to peritoneovenous shunt remains unproven. Holland[5] reported sclerosing peritonitis in patients on chronic ambulatory peritoneal dialysis. Dialysate solutions and bacterial peritonitis have been reported as etiological factors. But neither of these hypotheses has been proven.

Preoperative diagnosis of abdominal cocoon is difficult. Diagnosis is usually made at laparotomy. Suspicious sings and symptoms are; (1) nausea, vomiting, weight loss, distention and abdominal pain; (2) recurrent episodes of acute and sub acute small bowel obstruction, (3) the presence of a non tender mass on abdominal palpation; (3) typical appearance on radiological examination, and (4) negative history of previous abdominal operation. Four preoperative diagnostic features have been described by Yip and Lee[13]. The four clinical aspects are: a relative young girl without an obvious cause of intestinal obstruction, past history of similar episodes which resolved spontaneously, the presence of abdominal pain and vomiting but it is rarely to have the four cardinal symptoms of intestinal obstruction, and the presence of a non-tender mass on abdominal palpation. To some extent, the case presented here is matched to the above criteria. However, this case is in an adult healthy male with a history of sub acute intestinal obstruction.

The Pathological findings of SEP are characterized by a cocoon-like encapsulation of the entire intestine, which is accompanied by fibrin deposition, focal bleeding on the peritoneum and various quantities of bloody ascites[24]. Histologically, the membrane is composed mainly of organized fibrin, probably derived by plasma exudation from the peritoneal microvasculature[24].

The characteristic radiological findings of SEP have been sparsely described[25]. Ultrasound may show clumping of bowel loops with the bowel surrounded by a thick rim of hypo echoic tissue. Tethering of the bowel posteriorly, or the presence of a membrane anterior to the small bowel may be seen. Barium studies show varying length of small bowel tightly enclosed in a thickened peritoneum, proximal small bowel dilation, and an increase in transit time. The CT findings may also include an

encapsulated clump of bowel, peritoneal thickening, calcification, peritoneal enhancement, small bowel tethering, and locculated fluid collections. Peritoneal calcification and tethering of the small bowel loops are associated more specifically with SEP than the other CT findings.

Peritoneal encapsulation should not be confused with SEP, which may have a similar appearance on radiological studies. Peritoneal encapsulation is a congenital condition in which all or part of the small bowel is encased by an accessory peritoneal membrane. Usually, this is asymptomatic, but has been reported to cause bowel obstruction in a few cases. Pathologically, the encasing membrane is normal peritoneum rather than the thick fibrocollagenous tissue seen in abdominal cocoon, in addition the membrane is not adherent to the inner surface of the intestine.

Surgery is the required treatment, lysis of the membrane and adhesions being the main method. Bowel resection is unnecessary and it increases morbidity and mortality. Resection is indicated only if the bowel is non-viable. An excellent long-term postoperative prognosis is most of the times guaranteed. Recently, in a series of 5 patients, in addition to adhesolysis, small bowel intubation was performed with good results.

Conclusion

In the case presented, an intra-operative diagnosis of SEP was made in an adult male patient with a history of intestinal obstruction. Pre-operative findings were inconclusive in his current admission.

SEP or abdominal cocoon, although a rare cause of a common surgical emergency such as small bowel ileus, may be responsible, especially in cases with recurrent attacks of non-strangulating obstruction in the same individual. A high index of clinical suspicion may be generated by the recurrent presentation of small bowel ileus combined with relevant imaging findings and lack of other etiologies. Clinicians must rigorously pursue a preoperative diagnosis, as it may prevent a “surprise” upon laparotomy and unnecessary procedures for the patient, such as bowel resection.

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