Surg Today (2000) 30:950–953

SURGERY TODAY

Abdominal Cocoon: Report of a Case

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Abstract The abdominal cocoon is a rare cause of intestinal obstruction most often found in adolescent girls from tropical and subtropical countries. It is characterized by a thick fibrotic sac covering the small bowel partially or completely, the etiology of which is unknown. A correct diagnosis is not often made preoperatively; however, following simple surgical release of the entrapped bowel, these patients usually do well. We report herein our experience of a case of abdominal cocoon with a brief review of the medical literature on this unusual disease entity.

Key words Abdominal cocoon · Intestinal obstruction

Introduction

Abdominal cocoon is a rare condition of unknown etiology that primarily affects adolescent girls living in tropical and subtropical regions.[1–3] This condition is not often suspected preoperatively, and therefore the diagnosis is usually made at laparotomy. We present our experience of one such case of a young girl who presented as an emergency with features of intestinal obstruction.

Case Report

A 12-year-old girl presented to the Emergency Department of our hospital with a 10-h history of abdominal pain which had developed suddenly and had initially been colicky in nature. The pain later became constant during observation in hospital and was associated with two episodes of bilious vomiting. The patient had been constipated for 24 h and had not passed flatus for 12 h. There was no history of any similar episodes in the past, or of tuberculosis, diabetes mellitus, or hypertension. She had started menstruating only 4 months earlier, her last menstrual period having been 12 days prior to admission. On general examination the patient was conscious and oriented, well nourished, well hydrated, and afebrile, with a pulse rate of 120 beats/min and a blood pressure of 110/70 mmHg. No abnormalities of the chest or cardiovascular system were found (Fig. 1); however, an abdominal examination revealed fullness in the periumbilical and left hypochondrial region. A tender lump was palpated in the umbilical and left hypochondrial quadrants with mild guarding. It was able to be moved from side to side, but did not move with respiration. There was no hepatomegaly or splenomegaly, and no free fluid was evident on clinical examination. Bowel sounds were exaggerated on auscultation. A rectal examination was unremarkable.

Routine laboratory workup revealed a hemoglobin of 12.1 g%, a total leukocyte count of 5600 cells/ml, a blood sugar level of 92 mg%, and a blood urea of 32 mg%. An abdominal X-ray showed a few dilated loops of bowel in the left hypochondrial region, without any definite air fluid level (Figs. 2 and 3). An ultrasound examination of the abdomen showed a normal liver, gallbladder, pancreas, and kidneys. The uterus and left ovary appeared normal but the right ovary was slightly enlarged to 4 × 3 × 3.5 cm. There was no evidence of any cystic lesion in the abdomen, but a small amount of fluid was seen in the pelvis. Under a provisional clinical diagnosis of subacute intestinal obstruction associated with torsion of some benign cystic lesion in the abdomen, a laparotomy was performed through a midline incision. The entire small bowel and its mesentery was observed to be apparently only 2 feet (70 cm) in length from the duodenojejunal flexure to the ileocecal junction, and encased in a thick whitish membrane, the surface of which was covered in hemorrhagic spots, with a

Reprint requests to: R. Parshad
Received: May 26, 1999 / Accepted: May 30, 2000

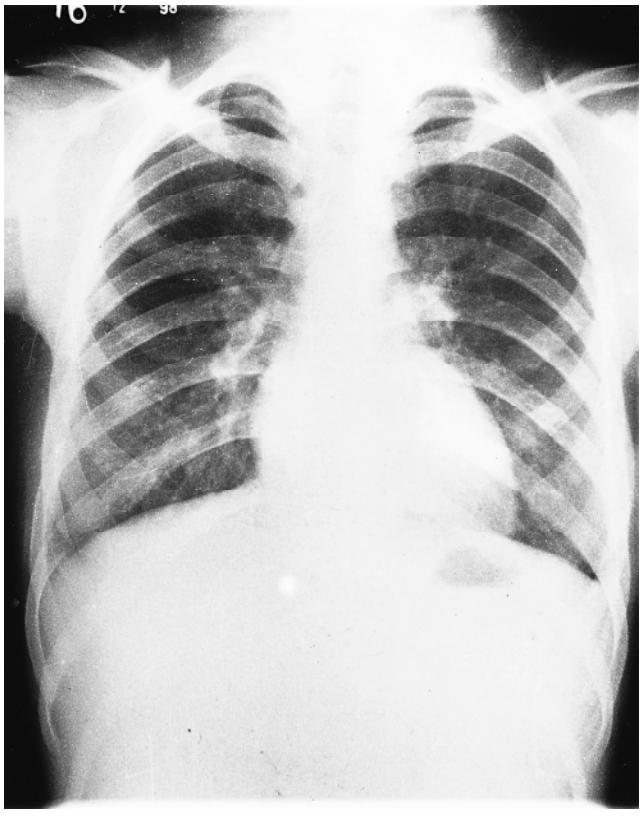

Fig. 1. Chest X-ray

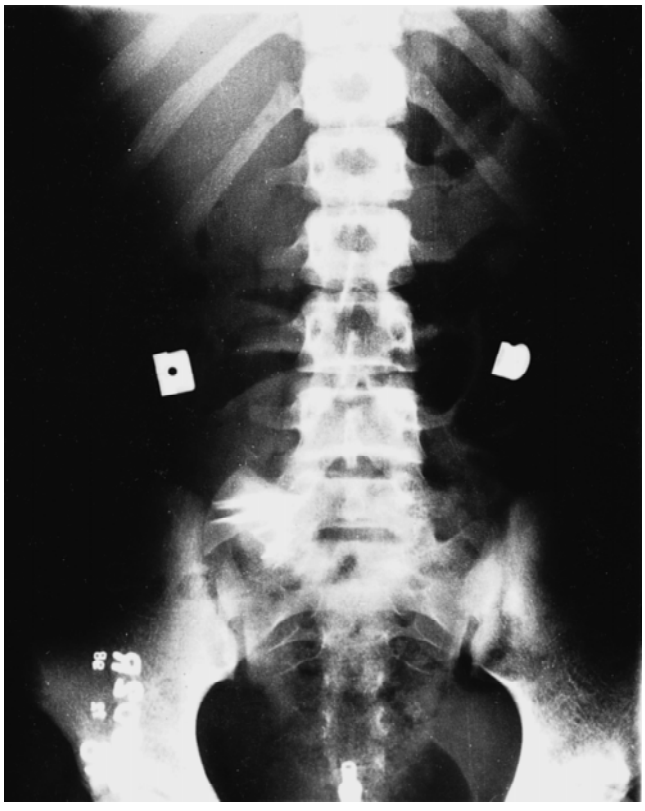

Fig. 2. Supine abdominal X-ray showing a few dilated bowel loops

narrow base. There was around 200 ml of straw-colored fluid in the pelvis. A relatively thin membrane was also found to be covering the liver, gallbladder, stomach omentum, and colon, that was separate from the cocoon.

On incising the over leaf of this thick membrane, the entire length of small bowel was found coiled in a serpiginous fashion within this 2 feet of cocoon. The bowel was otherwise normal, but with filmsy interloop adhesions. The mesentery was normal without any enlarged nodes. All of the flimsy adhesions were separated and the whole small bowel was freed. The patient had an uneventful postoperative recovery and was discharged on postoperative day 5. Histopathology showed thickened fibrocollagenous tissue with an area of local inflammatory infiltrate (Figs. 4 and 5). When last seen at her 3-month follow up, the patient was asymptomatic.

Discussion

Primary abdominal cocoon is a condition characterized by total or partial encasement of the small bowel by a fibrocollagenous cocoon-like sac. The abdominal cocoon was first described and so named by Foo et al. in 1978;[4] however, its etiology and pathogenesis remain obscure. A Medline search revealed that less than 50 cases have been reported up to now.

The affected individuals are usually from tropical and subtropical regions, with cases having been reported from India,[1,3–7] China,[8] Malaysia,[1,9] Singapore,[4] Japan,[10] Korea,[9] Pakistan,[10] Saudi Arabia,[12,13] and Lebanon.[14] The condition usually affects adolescent girls[4,5,7,13] ranging in age from 6 to 18 years; however, there have been anecdotal reports of a higher age range.[3,9] Regarding sex distribution, this condition is generally found exclusively in females, although cases in males have also been reported.[1,3,7,9,15] Clinically, these patients usually present with features of acute or subacute small bowel obstruction, symptoms of chronic obstruction and weight loss, and/or pain associated with an abdominal lump.[4,12,16] Our patient presented with features of subacute intestinal obstruction and an abdominal lump. Most cases are diagnosed when a laparotomy is performed for obstructive symptoms.[4,7,9,11,13]

This condition has been preoperatively diagnosed by barium meal follow-through and computed tomography (CT) scan of the abdomen in patients presenting with features of long-standing, worsening abdominal pain. A characteristic finding of coiling of the small bowel in a concertina-like fashion on barium examination has been described and termed as the "cauliflower" sign.[6] A

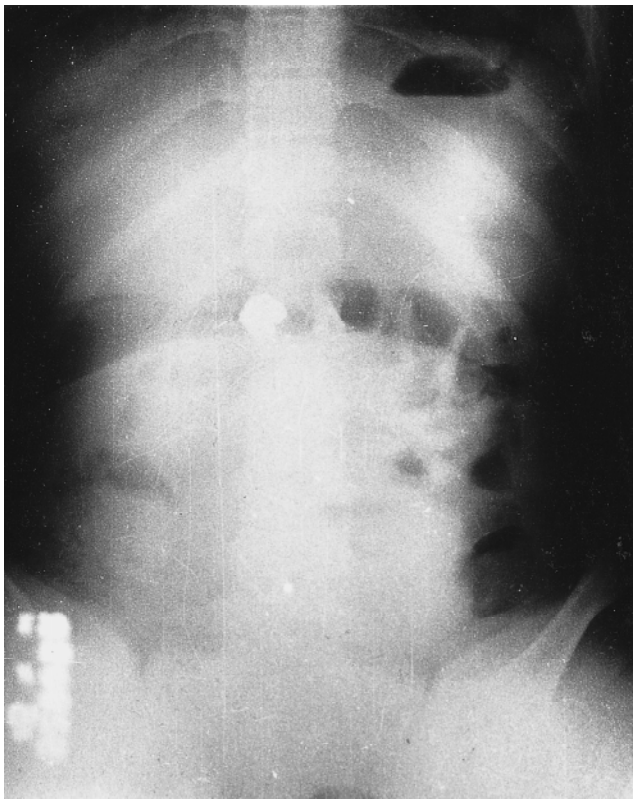

Fig. 3. Erect abdominal X-ray showing diffuse haziness without any significant air fluid levels

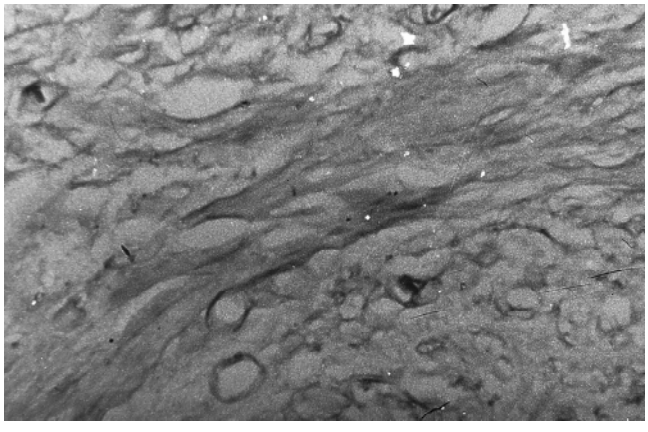

Fig. 4. Areas of diffuse fibrosis

preoperative CT scan of one patient suggested the clumping together of small bowel loops encased by a thick mantle of soft tissue infiltrating between the bowel loops and replacing mesenteric fat.[17] The degree of small bowel involvement by encaseation varies from a few feet to the entire length.[10,12,17] Reports of operative findings were consistent with the findings in our patient, in whom the whole small bowel from the duodenojejunal flexure to the ileocecal junction was encased. The histology in our patient was similar to that of previously reported cases, showing thickened fibrocollagenous tissue with an area of local inflammatory infiltrate. A neurofibromatous component in the histological section has also been reported.[5] According to one case report, ascitic fluid cytology revealed fragments of collagenous connective tissue.[10]

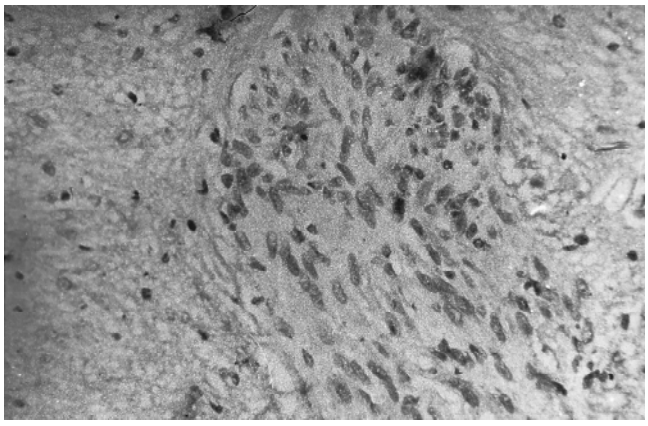

Fig. 5. Areas of fibrosis with focal inflammatory infiltrate

The etiology of this condition is still not well understood, although a number of hypotheses have been proposed to explain the formation of the membrane. These include: retrograde menstruation with a superimposed viral infection;[4] retrograde peritonitis via the fallopian tubes;[18] cell-mediated immunological tissue damage incited by gynecological infection;[18] practolol therapy;[19] and tuberculosis.[4] However, none of these hypotheses explain the characteristic age group, sex, and geographical distribution of this disease, and there is no objective evidence to substantiate them.

Peritoneal encapsulation is a related condition which differs[12,20] in that small or large bowel is found behind an accessory, but otherwise normal, peritoneal membrane. This condition has been reported in elderly males and as it is usually asymptomatic, it is generally discovered either at laparotomy for an unrelated condition or at autopsy.[21] No active management is required. Other conditions which can secondarily cause a similar clinical picture include those caused by practolol therapy,[19] continuous ambulatory peritoneal dialysis,[22] peritoneovenous shunt,[23,24] sarcoidiosis,[25] and systemic lupus erythematosus.[26]

Surgery remains the cornerstone in the management of abdominal cocoon. Careful dissection and excision of the thick sac with release of the small intestine leads to complete recovery. Resection of the bowel is indicated only if it is nonviable. The long-term prognosis of these patients is excellent if operative intervention is initiated without undue delay.

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