Abdominal cocoon syndrome: Preoperative diagnostic criteria, good clinical outcome with medical treatment and review of the literature

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Sclerosing encapsulating peritonitis, or abdominal cocoon syndrome, was described firstly in young adolescent girls. It is characterized by a thick fibrotic peritoneum that wraps the bowel in a concertina-like fashion with some adhesions. We report a man with intermittent intestinal obstruction and an abdominal cocoon encasing the small bowel. Our patient had no history of peritonitis or tuberculosis. We think he had primary abdominal cocoon syndrome. To the best of our knowledge, very few male patients have been reported in the medical literature as developing this condition. We treated the patient with drug therapy, including steroid and mycophenolate mofetil, and a liquid diet program, without surgical operation. He was symptom-free on follow-up over a period of 11 months.

Key words: Intestinal obstruction, abdominal cocoon, sclerosing peritonitis, computerized tomography

Abdominal koza sendromu: Preoperatif tanı kriterleri, medikal tedaviye olumlu yanıt ve literatürün gözden geçirilmesi


Anahtar kelimeler: Intestinal obstrüksiyon, abdominal koza, sclerozan peritonit, bilgisayarlı tomografi

INTRODUCTION

Abdominal cocoon syndrome (ACS) or sclerosing encapsulating peritonitis is a relatively rare cause of intestinal obstruction characterized by fibrotic encapsulation of the bowel (1-3). It was first described by Owtschinnikow in 1907 as ‘peritonitis chronica fibrosa incapsulata’ and termed ‘abdominal cocoon’ by Foo in 1978. The primary or idiopathic form was found in adolescent girls from tropical or subtropical countries. The acquired form is more common than the primary type. Early diagnosis can result in proper management, and may prevent the need for bowel resection (4-7). Because of the non-specific symptoms and clinical picture, the clinical diagnosis is difficult. Computed tomography (CT) findings are useful for the diagnosis because CT can show intestinal lumen, wall and extra-intestinal tissue planes (8). We report a case who presented with mild intestinal obstruction and showed typical CT findings of an AC.
CASE REPORT
A 58-year-old male presented with colicky pain in the periumbilical region and abdominal distention. He reported having had several such episodes over the previous year, but had received no specific treatment because of spontaneous symptomatic relief. There was no history of constipation, nausea or vomiting. He had undergone laparotomy for intestinal obstruction about two years before. A thick fibrous membrane encasing the small intestine was found during the operation. During the dissection of the intestinal wall from the surrounding thick membrane, the ileum was injured and ileostomy was performed. In a second surgery two months later, the ileostomy was closed. His symptoms did not disappear and continued to worsen. Examination of the abdomen revealed distention and hyperactive bowel sounds. Findings of laboratory studies (including complete blood count (CBC), electrolytes, liver and renal function tests, etc.) were within normal limits. Plain abdominal X-ray showed a few small air-fluid levels, with no free gas under the diaphragm. Ultrasound showed multiple dilated hyperperistaltic intestinal segments and a little free fluid around the umbilicus. A hypoechoic band-like structure enclosed these segments (Figure 1A, B). Contrast-enhanced multidetector computerized tomography (MDCT) scan of the abdomen revealed dilated small bowel loops congested to the center of the abdomen. A mild hyperdense membrane-like sac surrounded the bowel loops, and minimal ascites was seen between the intestinal loops (Figure 2A, B, C). Because of the patient’s intermittent obstructive symptoms and the lack of complete blockage signs, surgical treatment was not considered. A liquid diet was administered while the patient remained in the hospital. The abdominal pain decreased with conservative care over three days. Oral steroid therapy with 40 mg daily prednisone and mycophenolate mofetil (MMF) 720 mg/day were started. Prednisone was gradually decreased after 30 days of treatment. The patient’s complaints gradually decreased and disappeared after treatment. He was symptom-free on follow-up over a period of 11 months.

DISCUSSION
Sclerosing encapsulated peritonitis or ACS is a rare cause of intestinal obstruction. It is characterized by a thick fibrotic membrane that encases the bowel, forming a sac or cocoon along with some internal adhesions. At times, the surrounding membrane may be thin and difficult to identify on a CT scan (3,4,8).

The exact etiology of ACS is obscure, but it can be classified as primary (idiopathic) type and secondary type. The primary or idiopathic form is found in adolescent girls from tropical or subtropical countries. Early menstruation and primary peritonitis caused by retrograde menstruation may be the most important etiological factors in this type (9,10). Rarely, idiopathic cocoon syndrome occurs in males, and an infective etiology has been suggested to account for this (11). The more common
secondary form is associated with prolonged beta blocker therapy (practolol), local irritation of the peritoneum by trauma or surgical operation, intraperitoneal medical therapy (sterilizing chemicals like povidone, intraperitoneal chemotherapeutic agents), peritoneal dialysis, peritoneal–venous shunting, ventriculoperitoneal shunt, and infectious peritonitis. These conditions may predispose patients to peritoneal irritation and inflammation, which as a final effect, leads to peritoneal fibrinogenesis (12,13). Tuberculous infection causing similar findings has been documented and described in some cases as well; however, it should be noted that more commonly, tuberculosis of the abdomen produces intense contrast-enhanced, dense and thick fibrotic adhesions, unlike those seen in this case (14,15). Our patient had no history of peritonitis, predisposing drug treatment or tuberculosis. Because of the presence of a membranous structure encasing the intestinal loops in the first operation, we think our patient had primary cocoon syndrome. This is an extremely rare form in males.

The two commonest clinical presentations are acute small bowel obstructions and subacute, chronic obstruction. In the latter type, patients have episodes of intermittent colicky pain, as in the patient presented in this report. Preoperative diagnosis is important because surgery is not needed in this form (16,17). Imaging plays the most important role in disease management. Plain abdominal X-ray may show air-fluid levels or normal findings. Ultrasonography shows dilated intestinal segments, free fluid and peritoneal membrane, if it is thick enough. In our case, the patient had a typical sac-like structure surrounding multiple dilated small bowel loops. These features were consistent with typical AC. Sometimes, a definitive diagnosis of this condition cannot be made with ultrasonography and/or plain films. CT is widely considered as the gold standard to image AC. Classic CT findings include ascites with small bowel loops congregated in a single area or the concentration of the small bowel to midline encased by a soft-tissue-density envelope. Beyond helical CT, MDCT is advantageous, not only in showing the extent of the disease and subtle radiologic findings, but also in planning for surgery, with the contribution of coronal and sagittal reformatted images (8,18,19). In our patient, typical findings on ultrasound and MDCT helped in making an exact diagnosis.

Surgical removal of the membrane and adhesions is the most appropriate choice of therapy. Recently, many cases in the worldwide literature have been successfully treated laparoscopically (20-22). Conservative treatment, including total parenteral nutrition, nasogastric decompression and peritoneal rest, is the first choice for those patients with mild symptoms. Drug treatment including colchicine, steroids, immunosuppressive agents (MMF, azathioprine) is the second step in the management of mild disease. Colchicine may be a promising treatment option for ACS due to its fewer side effects, cost effectiveness, and clinical benefits with respect to various fibrotic processes.
Lafrance et al. (23) reported two cases of ACS caused by peritoneal dialysis who were successfully treated with cortisone and MMF. We administered cortisone and MMF to our patient. Our patient is asymptomatic now, but if symptoms recur, drug treatment combination with total parenteral nutrition can be applied again. If complete intestinal obstruction occurs, surgical treatment is inevitable.

Here, we reported a man with intermittent intestinal obstruction and an idiopathic AC encasing the small bowel that was diagnosed preoperatively by radiology. To the best of our knowledge, there are few male patients reported in the medical literature as developing this condition. Recognition of this entity and awareness of the typical radiological findings result in proper management and can prevent an unnecessary surgical procedure.

REFERENCES


