CASE REPORT

ABDOMINAL COCOON

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Abstract

Background

The abdominal cocoon syndrome was first described as a rare condition where part of or the whole small bowel is encased within a fibrous membrane. Idiopathic sclerosing encapsulating peritonitis (or abdominal cocoon) is a rare cause of small bowel obstruction, especially in adult population.

Diagnosis is usually incidental at laparotomy. But preoperative diagnosis can be made.

Case presentation

This report is of a 27-year-old Bangladeshi male who presented with increasing abdominal pain and features of subacute intestinal obstruction. He had a history appendicectomy 2 months back through grid iron incision in a peripheral hospital. Pre-operative work-up did not reveal a sac like structure encasing small intestinal loops. At computed tomography of the abdomen and pelvis, a huge cystic structure was seen encasing loops of small bowel. At laparotomy, a fibrous capsule was revealed, in which small bowel loops were encased, with the presence of interloop adhesions. A diagnosis of primary abdominal cocoon was established and extensive adhesiolysis was performed. The patient had an uneventful recovery and follow-up. No evidence of Kochs noted in the abdomen or on histopathology of tissue sent for examination.

Conclusion

Abdominal cocoon is a rare cause of small bowel obstruction, but should be suspected especially in cases with attacks of non strangulating obstruction in the same individual. A high index of clinical suspicion may be generated by the recurrent character of small bowel obstruction. Clinicians must rigorously pursue a preoperative diagnosis. The overall prognosis is satisfactory.

Key Words

Fibrous membrane, peritonitis, sclerosing peritonitis, abdominal cocoon

Background

Abdominal cocoon (Sclerosing encapsulating peritonitis) is a rare cause of small bowel obstruction, and can be classified as idiopathic or secondary (most importantly and most frequently due to chronic ambulatory peritoneal dialysis) and abdominal Kochs. It was first described by Foo et al in 1978 in a patient with Celiac Disease.[1,2]

It affects mainly young females from tropical and subtropical regions, but adult case reports from temperate zones can be

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encountered in literature. It is characterized by a thick, fibrotic, cocoon-like membrane, partially or totally encasing the small bowel.

The entity is often misdiagnosed, and final diagnosis is often obtained late after laparotomy and peritoneal biopsy.[3] Although a preoperative diagnosis is feasible by computed tomography of the abdomen (small bowel loops congregated to the center of the abdomen encased by a soft-tissue density mantle).[3,4] However, preoperative diagnosis requires a high index of clinical suspicion.

Surgery (membrane dissection and extensive adhesiolysis) is the treatment of choice, and there is usually no need for bowel loop resection, especially when a preoperative diagnosis is feasible. An excellent long-term postoperative prognosis is most of the times expected.

Case Summary

In September 2012, a 27 year-old man presented in A&E with history of colicky abdominal pain and bilious vomiting. He mentioned 4 similar episodes before which were diagnosed as subacute small bowel obstruction in the past 2 years and required hospitalization and they resolved with conservative treatment. He also admitted to chronic constipation for the last 6 years, anorexia and 10 kg weight loss since his last admission. He had history of appendicectomy 2 months back in a peripheral hospital where the operating surgeon noticed a white sac in the abdomen. The surgeon took FNAC from it, which was reported as chronic inflammation. He had no other medical history.

On examination, he was anxious but afebrile and haemodynamically stable. His abdomen was distended but non-tender, with increased bowel sounds in pitch and frequency. No palpable abdominal mass or organomegaly and no external hernias were found on clinical examination. Laboratory blood analyses were within normal limits. Plain abdominal X-ray showed few air-fluid levels which were centrally located and without free intraperitoneal gas. Contrast enhanced abdomen computed tomography revealed a huge cystic mass containing crowded small bowel loops in the right side of abdomen. There was indentation of bladder at its dome as well.

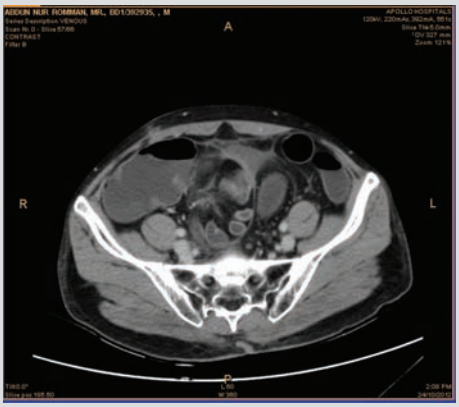

Figure 1: CT scan of abdomen showing a huge cystic mass encasing small bowel loops

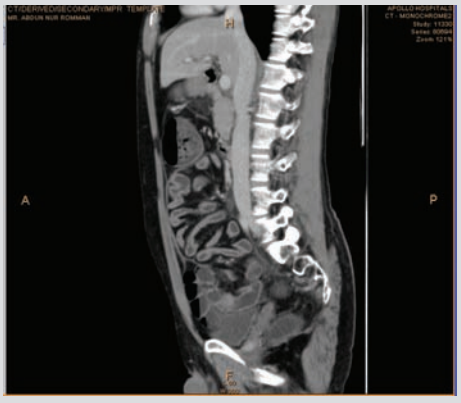

Figure 2: CT scan of abdomen showing a huge cystic mass encasing small bowel loops

On diagnostic laparoscopy, presence of the membrane was confirmed and exlporatory laparotomy was performed.

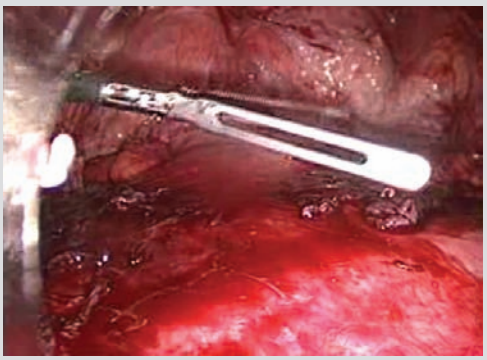

Figure 3: Diagnostic laparoscopy showing fibrous membrane beneath the bowel forceps

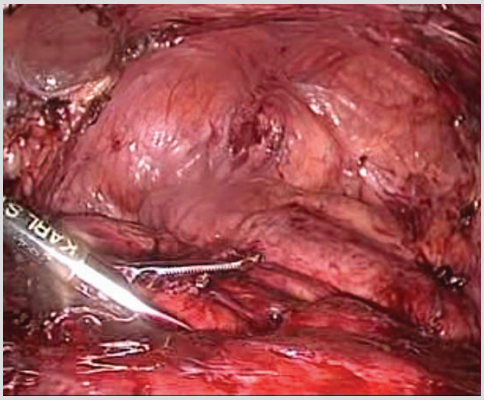

Figure 4: Diagnostic laparoscopy showing fibrous membrane beneath the bowel forceps

On laparotomy, a fibrous capsule covering the small intestines was revealed, with the presence of interloop adhesions. Encasement started 15 cm from DJ flexure to caecum. The thick membrane was excised and extensive adhesiolysis of small bowel loops were performed. Attention as paid to operate lightly and softly to avoid additional injury to serosa and thereby preventing postoperative adhesions.

Histology of the membrane showed thickened fibrocollagenous tissue with evidence of chronic inflammation.

A diagnosis of idiopathic sclerosing encapsulating peritonitis (abdominal cocoon) type II was established, due to intraoperative findings and by ruling-out any other condition that could explain the patient's pathology. Postoperative recovery was uneventful with additional nutritional support and 3 months after the operation he is in good health.

Discussion

Abdominal cocoon is a rare condition that refers to total or partial encapsulation of the small bowel by a fibrocollagenous membrane or cocoon with chronic inflammatory cells infiltrate leading to acute or chronic bowel obstruction characterized by thick fibrotic sac covering the small bowel partially or completely. It is mostly seen in adolescent girls.

It can be classified as primary and secondary. Secondary causes include chronic ambulatory peritoneal dialysis, Leveen shunt, TB PID, Orthotopic liver transplantation, sarcoidosis, SLE, practolol use. Most involved part is small intestine.But the cocoon membrane can encase stomach and liver also. According to extension it can be classified into 3 types - type I - the membrane encapsulated partial intestine; type II - the entire intestine was encapsulated by the membrane; and type III - the entire intestine and other organs (eg, appendix, cecum, ascending colon, ovary, etc).[5]

Most patients with abdominal cocoon syndrome present with features of recurrent acute or chronic small bowel obstruction secondary to kinking and/or compression of the intestines within the constricting cocoon. Patient may also have complaints of vague abdoiminal pain, weight loss, abdominal mass and sometimes jaundice.

Most of the time the diagnosis is made on laparotomy. But plain X-ray abdomen shows multiple air fluid level and sometimes calcification in the acute phase. Ultrasonogram of abdomen may show bowel surrounded by thick rim of hypo echoic tissue and tethering of bowel posteriorly or anteriorly. CT scan of abdomen and pelvis may reveal clustered small bowel loops, thick membrane like sac and proximal dilatation of small bowel. Barium follow-through study may reveal serpentine configuration of dilated small bowel within the cocoon.

Treatment options depend on patient's presentation. Surgical options range from excision of the accessory peritoneal sac with lysis of the interloop adhesions. Bowel resection is unnecessary unless a nonviable segment is found.Postsurgical recovery in most cases is smooth, and recurrence of the membrane is rare in long term follow up.[5]

Patient may present with recurrent intestinal obstruction due to adhesions. Surgical options for recurrent adhesion are adhesiolysis, intestinal intubation or plication of small intestinal loops according to Noble/ Charles Philips method.

Conclusion

The nonspecific clinical picture and benign imaging findings make diagnosis difficult.

A high index of suspicion is needed in the absence of other possible causes of the symptoms of abdominal obstruction.

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