Case Report

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Abdominal Cocoon Syndrome

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ABSTRACT

Abdominal cocoon syndrome is rare cause of intestinal obstruction characterized by small bowel encapsulation by a fibro-collagenous membrane or "cocoon".A 30 yearman presented in emergency department with abdominal pain. Preoperatively contrast enhanced computed tomography of abdomen revealed encapsulated cluster of mildly dilated and edematous small bowel loops with multiple air fluid levels with thin membrane and crowding of mesenteric vessels in left upper quadrant.Intra-operatively, the entire small bowel was found to be encapsulated in a dense fibrous sac. The peritoneal sac was excised, followed by lysis of the inter-loop adhesionswith smooth postoperative recovery. High index of suspicion is required in patient presenting with features of recurrent acute or chronic small bowel obstruction for diagnosis of abdominal cocoon syndrome. Contrast enhanced Computed Tomography of abdomen is a useful radiological to aid in preoperative diagnosis of syndrome.

Keywords: Abdomen; abdominal cocoon; CECT; encapsulated cluster.

INTRODUCTION

Abdominal cocoon syndrome is a rare condition that refers to total or partial encapsulation of the small bowel by a fibro-collagenous membrane with local inflammatory infiltrate leading to acute or chronic bowel obstruction.[1,2] The condition has been described by various synonyms including 'peritonitis chronica fibrosaincapsulata' and by sclerosing encapsulating peritonitis.[2] Encapsulating peritoneal sclerosis can occur at any age, with reports ranging from 2-day neonate to 82 years.[3] Since the time this disease was first described, approximately 35 cases have been reported .[4] We present the case of 30 years young male presenting in the emergency department of Civil Service Hospital presenting with acute abdomen. Patient was diagnosed of abdominal coccon syndrome by Contrast Enhanced Computed Tomography(CECT) which was confirmed intraoperatively. Patient had unremarkable perioperative period with good postoperative recovery.

CASE REPORT

Our patient, 30 year young male from Sindhupalchwok presented to Emergency department of Civil service hospital with epigastric pain for 5 days. The pain was gradual onset dull aching and located in epigastrium radiating to right ilac fossa.No vomiting; decreased appetite or loose stool noted. Similar history of pain abdomen was noted 2 years back. No past history of diabetes mellitus; hypertension or tuberculosis noted. At physical examination, vitals were stable; no pallor; jaundice; no edema; no clubbing noted, On superficial palpaltion; abdomen was not tender, but the umbilical area was distended. No evidence of organomegaly or rebound tenderness. On deep palpation, a soft mass approximately 6-8 cm in diameter was found in the umbilical regionextending up to right iliac fossa. Some firm areas were palpated in the mass. It was mobile atright angles to the axis of the mesentery. Bowel sounds were hyperperistaltic. The results of a rectal examination were unremarkable. CT revealed encapsulated cluster of mild dilated and edematous small bowel loops with maximum diameter measuring up to 3.5cm in AP diameter with multiple air fluid levels with thin membrane measuring up to 6mm in thickness. Crowding of mesenteric vessels with along with stretching of mesenteric veins noted in left upper quadrant. The clustered bowel loops is abutting anterior abdominal wall, inferiorly abutting and displacing caecum superiorly. (Fig.1 and Fig.2). Possible differentials were Abdominal cocoon (Sclerosing encapsulating peritonitis) with features of partial small bowel obstruction and transmesenteric internal hernia.

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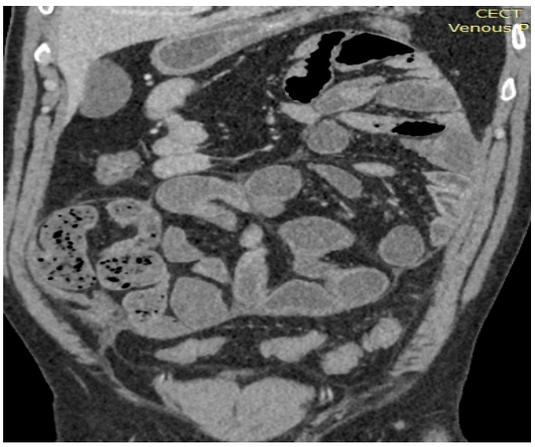

Figure 1. CT coronal contrast images showing encapsulated cluster of bowel loops with mild dilated small bowel loops.

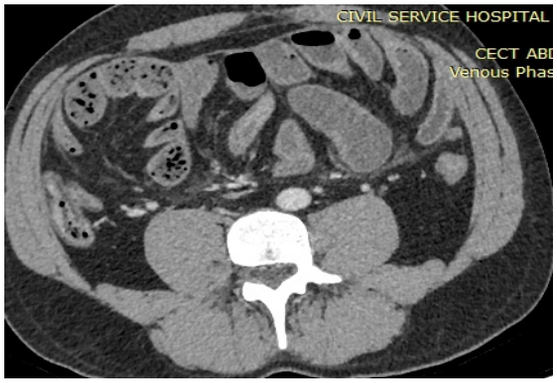

Figure 2. CT contrast axial images showing mild dilated small bowel loops with thin surrounding capsule.

SURGICAL FINDINGS

Emergencyexploratory laparotomy and adhesioloysis was done on 2075-11-6;there is evidence of almost whole of small bowel approximately 10 cm from DJ flexure to IC junction covered by membrane layer forming small bowel coccon. Minimal ascites seen. Peritoenal loose body was found.

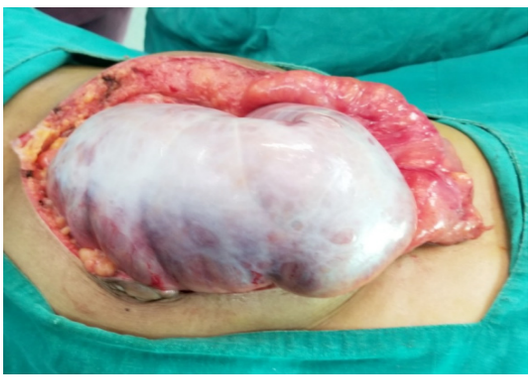

Figure 3. small bowel covered by membrane layer forming small bowel coccon.

DISCUSSION

Sclerorising encapsulating peritonitis has been classified as primary and secondary based on whether it is idiopathic or has a definite cause.[2] It can be secondary due to continuous ambulatory peritoneal dialysis (prevalence ~0.7%),[5] tuberculosis,[6] peritoneovenous or ventriculoperitoneal shunts, Various abdominal disorders such as sarcodosis, familial Mediterranean fever, gastrointestinal malignancy, protein S deficiency, liver transplantation, fibrogenic foreign material, and luteinised ovarian thecomas are the other rare causes.

Abdominal X-ray findings are non-specific. CECT is a useful tool for preoperative diagnosis of abdominal cocoon. The imaging features are, however, not pathognomic. CT findings of a membrane enveloping loops of small bowel were seen in some paraduodenal hernias, abdominal cocoon, and in peritoneal encapsulation.[7-8]

The final diagnosis of abdominal cocoon is usually based on intra-operative and histopathology findings, with a significant number presenting for emergency treatment without any imaging being performed. In all the reported patients, portions of the small bowel were encased within a fibrous cocoon.

Differential diagnosis includes peritoneal encapsulation, which was described as a developmental anomaly where the whole of the small bowel is encased in a thin accessory membrane. The clinical symptoms of this condition differ from those of the abdominal cocoon syndrome, in that the patients are mostly asymptomatic and the findings are incidental and late in life. Treatment, as in the present case, consist of excision of the thin accessory membrane with lysis of the inter-loop adhesions. Bowel resection is unnecessary unless a nonviable segment is found.The purpose of our case report is to make all the radiologist colleagues especially in the country like ours aware of the abdominal cocoon as the possible cause of epigastric pain and subacute intestinal obstruction. Currently, abdominal CT scans are requested commonly by clinicians, and the radiologist must be aware of this entity to make an appropriate CT diagnosis. Compared with other imaging techniques, CT gives a more complete picture of this entity as well as any associated complications, if any, and may also help to exclude other causes of intestinal obstruction. Use of computed tomography as a modality for assessment of subacute bowel obstruction is still not completely practised in low income country like Nepal due to financial burden to the patient party. Similarily , radiologist can have tendency to misinterpret it as mesenteric hernia can make the syndrome diagnosis

a rare entity. High degree of suspicion is required in imaging to look for the conditions and correlation with the intraoperative findings is key in coming to diagnosis.

CONCLUSIONS

Abdominal coccon syndrome is a rare entity in patient presenting with subacute bowel obstructions. Radiologists should be aware of this relatively rare cause of ofsubacute intestinal obstruction and its imaging findings since CECT abdomen can most often clinch the diagnosis and helps in the treatment of the patient.

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