Abdominal Cocoon - An Interesting Case of Intestinal Obstruction

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Abstract: Introduction: Abdominal cocoon syndrome is characterized by small bowel encapsulation by a fibro-collagenous membrane forming a “cocoon”. It is a rare cause of intestinal obstruction. Presentation: A 18 year old female with no prior surgical history presented with acute onset pain in abdomen at night, with a palpable lump in right iliac fossa and hypogastric quadrants, after initial investigations exploratory laparotomy revealed terminal ileum in a fibrous encapsulated sac. The peritoneal sac was excised with adhesiolysis. The patient responded well to the treatment. Discussion: Most patients with abdominal cocoon syndrome present as a case of intestinal obstruction. Abdominal cocoon is a rare cause of the intestinal obstruction though the cause for cocoon formation is not evident in most cases. Conclusion: Abdominal cocoon is a rare cause of intestinal obstruction and proper diagnosis requires high index of suspicion. Peritoneal sac excision and adhesiolysis is treatment of choice.

1. Introduction

Abdominal cocoon syndrome refers to total or partial encapsulation of small intestine by fibro-collagenous membrane with local inflammatory infiltrate leading to acute or chronic intestinal obstruction. It is also described as ‘peritonitis chronicafibrosaincapsulata’. We found a abdominal cocoon causing intestinal obstruction in a 18 year old female with 2 months of amenorrhea which was treated surgically. It is first described by Foo et al. in 1978

2. Case Report

A 18 year old female presented to emergency room in the night with acute onset (1 hour) pain in abdomen with vomiting. There was no prior history of hospitalization or any surgical intervention. The patient was pregnant for 2 months at the time of presentation. Only positive history was of intermittent pain in abdomen which relied on itself.

On examination, patient had palpable mass in right iliac fossa and hypogastric region. There was asymmetrical distension of abdomen which was the unusual finding.

After ruling out the obstetric cause for pain in abdomen, ultrasonography revealed clumped up, encapsulated bowel loops in right iliac fossa and hypogastric region. X-ray and CT abdomen was not done considering first trimester of pregnancy.

Patient underwent laparotomy which revealed encapsulation of terminal ileum with proximal colon in a fibrous sac. The sac was excised and adhesiolysis relieved the obstruction and patient tolerated the procedure well and recovered successfully. The histopathological report of the excised peritoneal sac was chronic nonspecific inflammation.

3. Discussion

Abdominal cocoon or Sclerosing encapsulating peritonitis has been classified as primary and secondary based on whether it is idiopathic or has adefinite cause. The aetiology of the primary form is uncertain with various hypotheses, although it is probably caused by a sub-clinical peritonitis leading to the formation of a cocoon. History of recurrent pain in abdomen episodes in this case can be due to subclinical peritonitis.

Secondary causes include theplacement of Le Veenshunts for refractory ascites, continuous ambulatory peritoneal dialysis, systemic lupus erythematosis, peritonitis, previous abdominal surgery, sarcoidosis, and tuberculosis and the use of povidone iodine for abdominal wash-out.

Clinically, 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. An abdominal mass may also be present due to an encapsulated cluster of dilated small bowel loops.

Abdominal X-ray findings are usually non-specific. CECT is a useful tool for preoperative diagnosis of abdominal cocoon. The imaging features are, however, not pathognomonic. CT findings of a membrane enveloping loops of small bowel were seen in some paraduodenal hernias, abdominal cocoon, and in peritoneal encapsulation. However, the clinical and pathological features of these entities are different.

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, consists of excision of the accessory peritoneal sac with lysis of the inter-loop adhesions. Bowel resection is unnecessary unless a nonviable segment is found.

Retroperitoneal fibrosis (also known as Ormond disease) is a rare syndrome featuring fibrosclerosis of the tissue in the retroperitoneum, often leading to encasement of the ureters, such as encasement of the intestines in cocoon syndrome. The difference between Ormond disease and Abdominal Cocoon Syndrome is the location. Retroperitoneal fibrosis is treated with glucocorticoids. If there is ureteral obstruction, stent or surgery is indicated.

Intra-operative photograph of abdominal cocoon

References