

Case Report

Idiopathic encapsulating peritoneal sclerosis: a case report

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ABSTRACT

Idiopathic encapsulating peritonitis sclerosis is a rare cause of small bowel obstruction. It is diagnosed by the presence of dense white fibrotic membrane encasing a part or the entire small bowel. Diagnosis is usually made intraoperatively and the treatment is usually surgical. We report a case of 16 years old female with idiopathic encapsulating peritoneal sclerosis who presented with the features of recurrent intestinal obstruction. Surgical excision of the fibrotic sac and adhesiolysis was done.

Keywords: Encapsulating peritoneal sclerosis, Small bowel obstruction

INTRODUCTION

Encapsulating peritoneal sclerosis (EPS) is a rare cause of intestinal obstruction. It is characterised by partial and complete encapsulation of small bowel with a dense white fibrotic membrane.¹⁻⁵ Depending upon the cause, it is further divided as idiopathic (primary) and secondary. Primary EPS was first observed by Owtschinnikow in 1907 and was called *chronica fibrosa encapsulate*.⁶ The presentation is nonspecific and the diagnosis is usually made intraoperatively. Medical treatment includes antiinflammatory or antifibrogenic drugs such as tamoxifen, steroids, colchicine, azathioprine and mycophenolate mofetil.⁷ Surgical management includes membrane resection and adhesiolysis with or without bowel resection.

CASE REPORT

A 16-year-old female, presented with the complaint of pain in abdomen, abdominal distension for 5 days; nausea and vomiting with non-passage of faeces and flatus for 2 days. There was history of similar episodes in past and was managed conservatively. There was no history of tuberculosis or Koch's contact and no other significant medical history. On examination abdomen was distended

and diffuse tenderness with no guarding and rigidity was present. X-ray abdomen showed dilated jejunal loops with multiple air fluid levels.

Contrast enhanced computed tomography (CECT) abdomen showed dilated proximal jejunal loops with clumped distal jejunal and ileal loops surrounded in a peritoneal sac. There was no evidence of mesenteric lymphadenopathy and ascites.

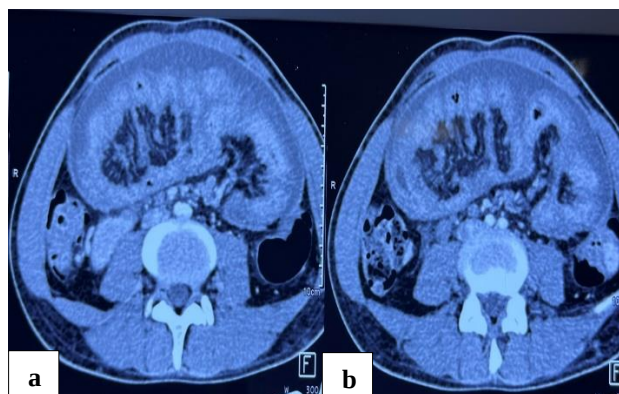


Figure 1: Axial CT images showing clumped small bowel enclosed within the thick peritoneal sac.

Exploratory laparotomy was done. Intraoperatively, proximal 70 cm of jejunum was found to be dilated with remaining jejunum and ileum encased in dense peritoneal sac that was firmly adhered to terminal ileum. There was no ascites or mesenteric lymphadenopathy. Peritoneal sac was excised and adhesiolysis was done. Approximately 180 cm bowel length was regained. Postoperative period was uneventful. Histopathology report showed nonspecific inflammatory changes, with no findings suggestive of tuberculosis. Patient was orally allowed on post-operative day (POD-3) and was discharged on POD-7. On follow up there was no evidence of any post op complication or recurrent obstruction.

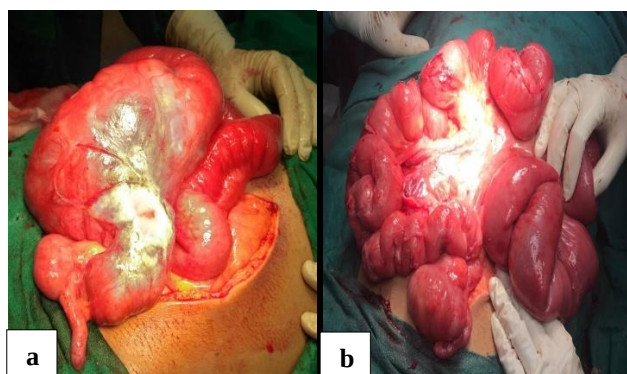


Figure 2: (a) Peritoneal sac enclosing small bowel with proximally dilated jejunum, and (b) post op image after resection of peritoneal sac and adhesiolysis.

DISCUSSION

EPS is rare with a mortality rate of 60-93%.² Depending upon the cause it is divided as primary (idiopathic) and secondary. Incidence of idiopathic EPS is 0.4-5.5%.² It is mostly observed in adolescent girls living in tropical and subtropical countries, although may be seen in children living in temperate regions or adults of advanced age.¹ Exact cause of idiopathic EPS is not known, however many theories to explain the primary aetiology, includes viral peritonitis due to infection from retrograde menstruation, developmental malformations, greater omentum hypoplasia, and malformation of mesenteric vessels.^{2,3} Most common secondary cause worldwide is chronic peritoneal dialysis (2). 3% of the patients on peritoneal dialysis develop EPS. Abdominal tuberculosis is one of the most common causes of secondary EPS in underdeveloped countries.¹¹ Other causes includes prior abdominal surgeries, prolonged beta blocker therapy, radiation enteritis, sarcoidosis, SLE, ventriculoperitoneal shunts, and malignancy.⁸⁻¹⁰

Clinical presentation includes asymptomatic course, acute or recurrent subacute or chronic intestinal obstruction.^{1,2,6} X-ray abdomen shows dilated bowel loops with air fluid levels, consistent with intestinal obstruction. CT scan is the most useful radiological investigation for its diagnosis.² Since most of the patients presents to emergency

department with acute abdomen, diagnosis is usually made intraoperatively.⁵

Asymptomatic patients are put on regular follow up.⁶ Mildly symptomatic patients are best managed by conservative management that includes intestinal rest, nasogastric decompression and nutritional support (enteral or parenteral).^{3,7} Cases with no regression of symptoms may be treated with anti-inflammatory or antifibrogenic drugs such as tamoxifen, steroids, colchicine, azathioprine and mycophenolate mofetil.⁷ Surgical management is the main stay of treatment for patients not responding to conservative management and those presenting with intestinal obstruction. It includes resection of peritoneal sac, enterolysis with or without bowel resection.

CONCLUSION

High degree of suspicion is fundamental for diagnosis of EPS. Surgical excision and adhesiolysis is usually curative.

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