

Case Report:

Abdominal Cocoon Syndrome in a Child With Intestinal Obstruction: A Case Report and Literature Review



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ABSTRACT

Abdominal cocoon syndrome or idiopathic Sclerosing Encapsulating Peritonitis (SEP) is an extremely uncommon cause of intestinal obstruction. Its etiology is explainable through numerous theories.

An eleven-year-old girl referred to the pediatric surgery OPD with complaints of abdominal pain for the past two weeks and vomiting for two days. Her family history for tuberculosis was positive. On examination, the abdomen was distended and slightly firm on palpation. The X-ray of her abdomen revealed multiple air-fluid levels. The CECT of the abdomen indicated dilated abdomen, duodenum, and proximal bowel loops. Some trapped inter bowel free fluid was also observed. The clinical presentation of subacute intestinal obstruction and the radiological features suggested a differential diagnosis of tubercular peritonitis versus pseudomyxoma peritonei. The obstructive symptoms demonstrated that the patient underwent an explorative laparotomy. Preoperatively, a thin membranous sac was identified enclosing multiple dilated small bowel loops. The sac was released by blunt dissection and part of the sac was provided for histopathological examination. Based on clinical, histopathological, and radiological findings, a diagnosis of abdominal cocoon syndrome was determined. The postoperative follow-up period of 6 months was uneventful.

We presented a rare case of primary sclerosing encapsulating peritonitis, also reported as abdominal cocoon syndrome. It is among the rare potentially devastating causes of intestinal obstruction in children. A very high index of suspicion is imperative to arrive at its pre-operative diagnosis alone by clinical and radiological findings.

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1. Introduction

The abdominal cocoon syndrome is an extremely rare cause of intestinal obstruction. It is also recognized as Sclerosing Encapsulating Peritonitis (SEP), chronic fibrosa encystica, and idiopathic encapsulating sclerosing peritonitis. The condition is often reported in adolescent females of tropical and subtropical countries; however, elderly and male cases are also reported. The etiology of this condition remains unclear; however, idiopathic and cases secondary to localized or systemic processes have been reported in this regard (1). We reported a rare case of idiopathic SEP in a young girl who presented with recurrent spasmodic abdominal pain. Furthermore, this case highlights a rare cause of intestinal obstruction, to preoperatively diagnose it, and its further management. Informed consent was provided by the parents of the patient for reporting their case.

2. Case Presentation

An eleven-year-old girl presented to the paediatric surgery outpatient ward. She had complaints of abdominal pain for the past two weeks and vomiting for two days. The pain was generalized, spasmodic, and progressively increasing in intensity. She also reported that pain was associated with the passage of hard stools, constipation, and abdominal distension. Her family history for tuberculosis was positive. There was no significant history of the

loss of appetite or weight. On examination, the patient was afebrile and pale, but hemodynamically stable with a blood pressure of 114.78mmHg and a pulse rate of 90/min. No lymphadenopathy was seen. On auscultation, the chest was clear. The abdomen was distended and slightly firm on palpation. Hemogram revealed mild anaemia (hemoglobin: 9.5 g/dL), and mild neutrophilic leucocytosis (total leucocyte count: 15,000/m³). Moreover, C-reactive protein was raised and Mantoux read 6 mm. The patient's chest X-ray was clear. The x-ray of the abdomen revealed multiple air-fluid levels. The Contrast-Enhanced Computed Tomography (CECT) of the abdomen indicated dilated stomach, duodenum, and proximal bowel loops.

Some trapped inter bowel free fluid was also observed in the patient. The clinical presentation of subacute intestinal obstruction and the radiological features suggested a differential diagnosis of tubercular peritonitis versus pseudomyxoma peritonei. The patient's condition aggravated with severe pain. The obstructive symptoms revealed that the patient underwent an explorative laparotomy. Preoperatively, a fluid-filled thin membranous sac was identified enclosing multiple dilated small bowel loops (Figure 1a). The sac was released by blunt dissection and the bowel loops (Figure 1b) were decompressed by adhesiolysis along the avascular planes.

The post-operative period was uneventful. Part of the sac excision was provided for histopathological examination. Grossly, we received a flattened grey white soft tissue measuring 2x1.5x0.5 cm. The histopathology of the

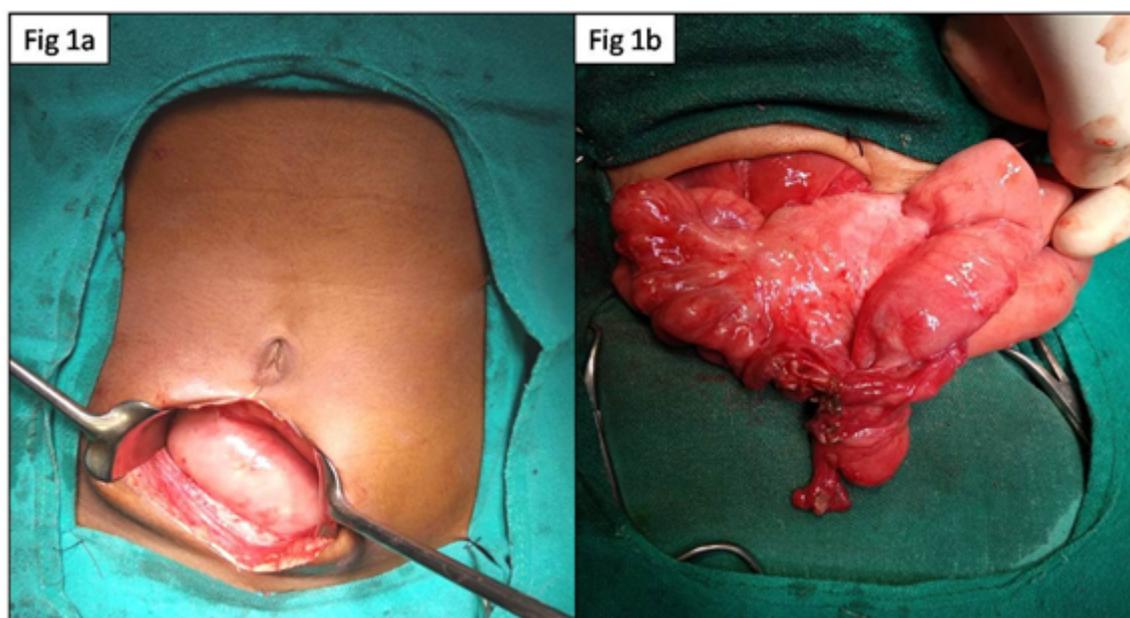


Figure 1. 1a: Intra-operative image showing a membranous sac.1b: showing healthy bowel after release from the sac

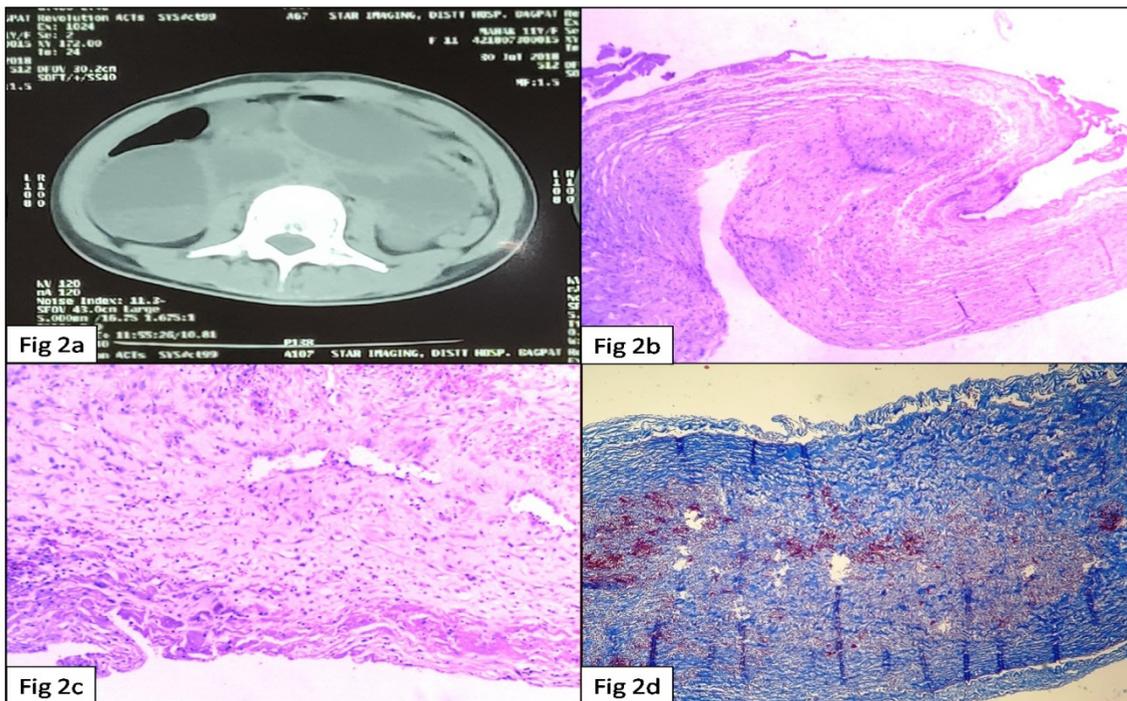


Figure 2. CT scan, Fibro collagenous, Hematoxylin & Eosin, and Masson's trichrome stain result

A: The axial section of CT scan showing an encasement of bowel in the center, B: Fibro collagenous wall observed on microscopy comprising fibroblastic proliferation (Hematoxylin & Eosin, 100X), C: High-power view showing mononuclear inflammatory infiltrate along with tiny vessels dispersed within the fibroblastic cells (Hematoxylin & Eosin, 400X), D: Masson's trichrome stain highlighting the fibroblastic nature of cells (100X)

sac indicated fibro-connective tissue proliferation as well as few dilated lymphatics, congested blood vessels, and inflammatory infiltrate comprising of lymphocytes, macrophages, and few neutrophils. No nuclear pleomorphism was detected (Figure 2b, 2c). No evidence of epithelioid cell granulomas (tubercular or foreign body), giant cells, or birefringent material was identified. Masson's trichrome stain highlighted the fibroblastic nature of the cells. (Figure 2d) The CECT highlighted the centering of the distal jejunal and ileal loops in the lower abdomen and pelvis by a thin fibrous capsule (Figure 1a). Accordingly, a diagnosis of idiopathic SEP was determined. The postoperative follow-up period of 6 months was uneventful.

3. Discussion

The term SEP was first addressed by Owtschinnikow in 1907 and later described by Foo et al. in 1978 (1). It was defined as the packing of the intestinal segments within a dense fibrotic membrane, leading to intestinal obstruction (2). The correct differentiation of abdominal cocoon or SEP from Peritoneal Encapsulation (PE) was identified by Akbulut S and associates. PE is a congenital anomaly and was first described by Cleland et al. in 1868; it is characterized by the presence of an accessory thin transpar-

ent peritoneal membrane. Its presence is unrelated to any inflammatory process and usually never leads to intestinal obstruction with the diagnosis made accidentally at laparotomy (3). In contrast, SEP is an acquired condition occurring secondary to an inflammatory process.

Two SEP types are detected; primary (idiopathic) and secondary (owing to any localized or systemic process). Secondary SEP could occur consequent to tuberculosis, sarcoidosis, SLE, continuous or automated peritoneal dialysis, liver cirrhosis, familial Mediterranean fever, protein S deficiency, gastrointestinal malignancies, fibrogenic foreign materials, endometriosis, ovarian thecomas, dermoid cyst rupture, liver transplant, penetrating abdominal injuries, and prolonged beta-blocker therapy (4, 5). It is essential to differentiate between these two, as the secondary forms are associated with a dense fibrous capsule formed due to recurrent inflammation. It is also associated with the presence of inter-bowel adhesions and is difficult to separate, due to the lack of an avascular plane.

Idiopathic SEP is more often observed in young adolescent females of tropical and subtropical countries. However, few Chinese studies have revealed male involvement both singly and in series (6). No gender pre-

dilection is observed in this condition. The etiology of it remains debatable; however, the most common theory behind the idiopathic forms is recurrent subclinical peritonitis causing sclerosis and membrane formation. Another hypothesis is the retrograde menstruation theory with superimposed viral infection; however, this theory fails to explain its occurrence in males and children (6). Some other hypotheses include cell-mediated immunological tissue damage secondary to gynecological infections. No useful biomarker has been found for predicting the occurrence of SEP. Based on the extent to which the intestine is covered, idiopathic SEP is categorized into three types. Type 1 indicates that only a part of the intestine is encased, type 2 suggests that complete intestine is covered, and type 3 occurs in intestine conditions along with the involvement of other body organs.

Achieving a correct diagnosis of SEP preoperatively is extremely difficult, although not impossible. The clinical presentation of these cases could vary from asymptomatic to sub-acute or acute intestinal obstruction. Usually, the symptoms are nonspecific with multiple attacks of pain, constipation, vomiting, the loss of appetite, and weight. The presentation as an abdominal mass was first reported by Basu and colleagues (7).

Radiological investigations for diagnosis include abdominal x-ray, barium studies, ultrasound, CECT, and occasionally contrast-enhanced Magnetic Resonance Imaging (MRI). X-ray is non-specific; however, barium studies revealed conglomerated loops in the center of the abdomen, i.e., recognized as the 'cauliflower' sign (3, 4). This sign, along with delay in the transit time are essential clues to SEP. Barium studies may not be possible in patients presenting with acute abdomen; thus, CECT is more promising for the diagnosis. The latter presents the centering of the bowel loops in the midline and encasement by a dense fibrous capsule with a contrast free periphery. The histological features are not pathognomonic; however, when present in conjunction with radiological features and preoperational findings, they are diagnostic of idiopathic SEP (4).

The differential diagnosis of SEP includes congenital peritoneal encapsulation, and encapsulation secondary to the previously mentioned causes (4). A significant history of dialysis, prolonged medication, abdominal surgery, and tuberculosis could rule out secondary SEP. Sarcoidosis and endometriosis are associated with other systemic complaints. Radiological investigations may hint the diagnosis in these cases and histopathology significantly impacts delineating all the secondary causes of the abdominal cocoon. The reported patient had no

significant medical history, which would have acted as a trigger for peritoneal inflammation.

The significance of knowing the etiology cannot be undetermined and defines the correct management of the cases. The cases not presenting obstruction could be managed by conservative methods. Such techniques include nasogastric decompression, proper nutritional support, and medications, like corticosteroids, tamoxifen, and azathioprine (6, 8). Enterolysis surgery, however, remains the treatment of choice in acutely symptomatic cases and patients with failed medical management. The laparoscopic approach in these cases is beneficial, as it is both diagnostic and therapeutic. Bowel resection is usually not recommended until finding gross evidence of bowel ischemia. Complications are rare but include adhesions, infections, and the formation of enterocutaneous fistula. Idiopathic SEP has an excellent prognosis without reportable recurrences. The secondary forms of SEP, however, have a poorer outcome, compared to primary types (8). Recurrence is uncommon, although few patients present with adhesive intestinal obstruction in the view of the dissection, i.e., made during the surgical intervention (6). One study documented only a 6% recurrence rate (9).

The diagnosis of SEP usually surprises the clinicians on the operative table and is confirmed on biopsy; however, with radiological advances, like High-Resolution Computed Tomography (HRCT) scan, a pre-operative diagnosis is possible. Therefore, it directs the clinician to instigate conservative medical management for cases, not in obstruction, and avoid unnecessary surgical interventions.

4. Conclusion

The current case report aimed at increasing the awareness of this rare, potentially devastating but completely treatable entity as a cause of intestinal obstruction in children. A strong clinical and radiological suspicion could help reach a pre-operative definitive diagnosis. Careful dissection and excision of the sac with the release of the intestinal contents in primary SEP could lead to complete recovery.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article. The participants were informed about the purpose of the research and its implementation stages; they were also assured about the confidentiality of their information; moreover, they were free to leave the study whenever

they wished, and if desired, the research results would be available to them.

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Authors contributions

Conceptualization, design: Nidhi Mahajan; Manuscript preparation: Nidhi Mahajan, Mitali Agarwa, Arti Khatri; Literature search: Mitali Agarwal; Editing, and review of the manuscript: Arti Khatri; Manuscript review, treating surgeon: Yousuf Mohsin Bari Siddiqui, Mama Sengar.

Conflicts of interest

The authors declared no conflicts of interest.

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