Case Report

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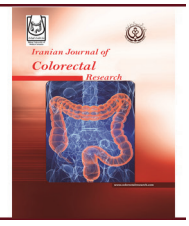

Encapsulating Peritoneal Sclerosis: A Rare Cause of Acute and Recurrent Abdominal Pain; A Case Report of Abdominal Cocoon Syndrome

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Abstract

Encapsulating peritoneal sclerosis (EPS), also known as abdominal cocoon syndrome, is a rare condition that may present with abdominal pain or intestinal obstruction due to the encasement of bowel loops within a fibrocollagenous membrane. We report the case of a 27-year-old woman who presented with severe generalized abdominal pain, nausea, and vomiting that did not respond to conservative treatment. Physical examination revealed right lower quadrant tenderness, and subsequent exploratory laparotomy demonstrated abdominal cocoon syndrome associated with an appendiceal mucocele. This rare entity typically manifests as recurrent abdominal pain and obstructive symptoms, and while conservative therapy may provide transient relief, it often delays diagnosis. Surgical excision of the fibrous membrane with adhesionolysis remains the definitive treatment in severe cases. Abdominal cocoon syndrome should be considered in the differential diagnosis of unexplained abdominal pain and obstruction, as diagnosis is frequently established only intraoperatively.

Keywords: Encapsulating peritoneal sclerosis, Abdominal Cocoon, Peritoneal Diseases, Intestinal Obstruction, Case Reportsn

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Introduction

Encapsulating peritoneal sclerosis (EPS), also known as abdominal cocoon syndrome, is a rare condition and an uncommon cause of abdominal pain and intestinal obstruction. It typically presents with recurrent episodes of intestinal obstruction, with or without an associated abdominal mass (1). The diagnosis is most often established intraoperatively (2). Although first described by Owtschinnikow in 1907 as peritonitis chronica fibrosa incapsulata, the term "abdominal cocoon" was introduced by Foo et al. in 1978 (3) mostly among the young adolescent girls suffered from retrograde menstruation (4, 5). It primarily affects adolescent girls of the tropical and subtropical areas (6); however, it occurs in different ethnically and geographically locations (7). A fibrous membrane with inflammatory infiltrates partially or completely encases the small intestine, leading to varying degrees of obstruction. The syndrome is classified into two types: primary (idiopathic), which represents an exceptionally rare form and secondary.

The secondary form is most commonly associated with conditions such as tuberculosis, previous abdominal surgery, neoplasms, beta-blocker use, and peritoneal dialysis (8).

The clinical features are nonspecific and diagnosis is often made intraoperatively, with imaging—particularly CT—playing an adjunctive role. Differential diagnoses include congenital peritoneal encapsulation, peritoneal carcinomatosis, and internal hernias. Internal hernias are an important differential diagnosis, as they may display similar CT features to abdominal cocoon syndrome; however, they typically do not present with a membrane-like sac on imaging (9). Reported mortality can be high in severe cases, particularly in patients on long-term peritoneal dialysis (10).

This case is noteworthy because it highlights an unusual presentation of EPS in a young female who was initially misdiagnosed with acute appendicitis. The intraoperative discovery of a fibrotic membrane encasing the small intestine underscores the diagnostic challenges and the need for awareness of this rare entity in patients presenting with unexplained intestinal obstruction.

Case Presentation

A 27-year-old female with a past medical history significant for epigastric pain was previously diagnosed with *Helicobacter pylori* infection via endoscopy, as well as ovarian cysts and endometriosis diagnosed several years ago. She presented with severe abdominal pain and nausea, but maintained normal intestinal transit for the past seven months. The patient reported multiple prior emergency department admissions with similar symptoms, which improved with conservative treatment.

However, on her most recent presentation, she developed severe generalized abdominal pain, nausea, and vomiting that were unresponsive to conservative measures. During the workup, abdominopelvic ultrasonography revealed mild free fluid in the lower peritoneal cavity and a tubular cystic structure measuring 45 × 15 mm adjacent to bowel loops on the left side, without significant perimesenteric inflammation or edema.

Computed tomography (CT) scan identified a tubular structure on the left side of the abdominal cavity, approximately 12 mm in diameter, likely representing a left-sided appendix that was fluid-distended but demonstrated normal wall thickness and no signs of inflammation. The possibility of an appendiceal mucocele was included in the differential diagnosis (Figure 1).

Therefore, laparoscopic exploration was performed, revealing severe encasement of the abdominal

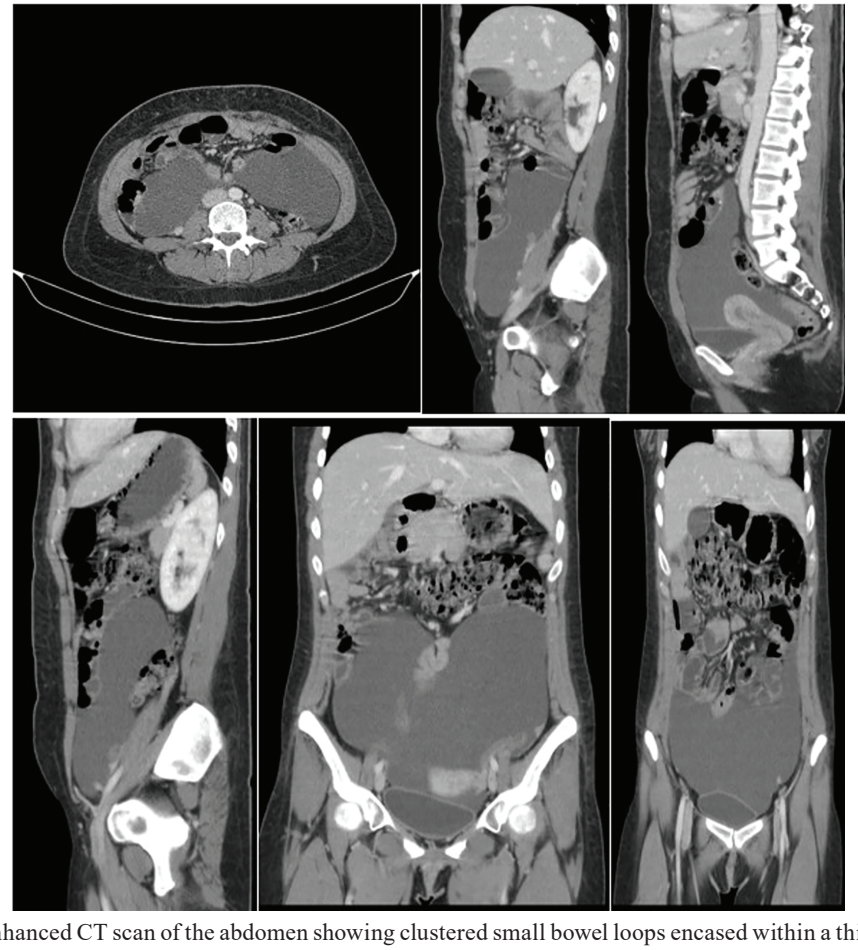

Figure 1: Contrast-enhanced CT scan of the abdomen showing clustered small bowel loops encased within a thin, fibrous membrane, consistent with abdominal cocoon. The sagittal, axial, and coronal thin-cut portovenous phase of a 16-slice CT scan (Siemens, Emotion) revealed that the small bowel loops are enveloped in a layer of peritoneum. The patient has ascites accumulating in the pelvic cavity, not distributed in the abdominal cavity due to the aforementioned peritoneal layer.

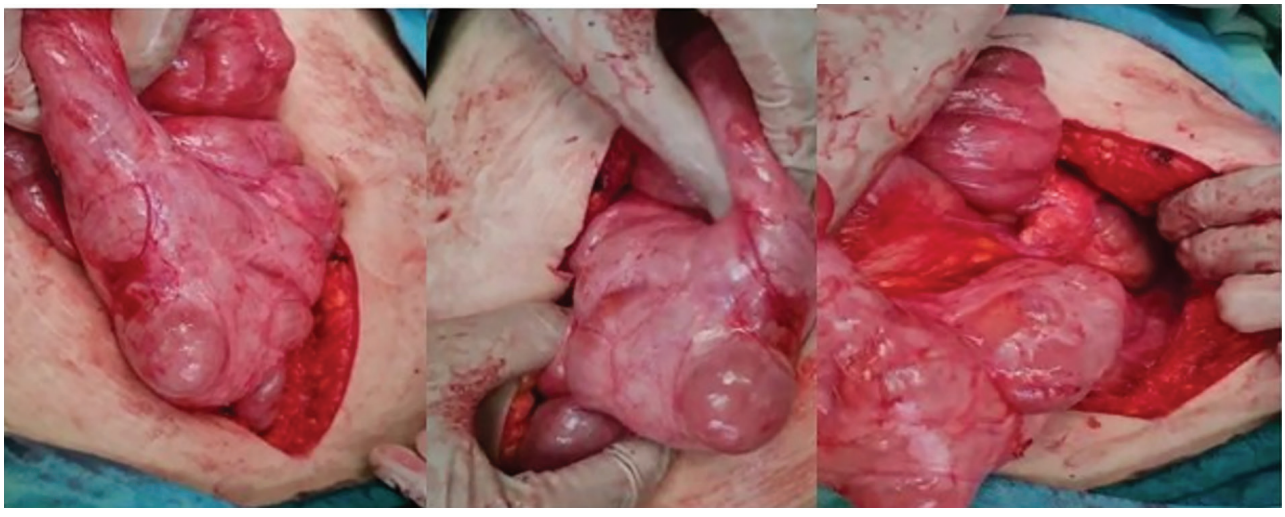

Figure 2: Intraoperative findings demonstrating small bowel loops enveloped by a dense fibrous sac.

viscera within a fibro-collagenous membrane. Due to the extent of involvement, the procedure was converted to an open laparotomy, during which a characteristic "cocoon"-like fibrous membrane was identified (Figure 2).

An appendectomy along with resection of the surrounding fibrous membrane was performed. Intraoperative findings supported the diagnosis of abdominal cocoon syndrome accompanied by an appendiceal mucocele. Histopathological examination revealed a well-differentiated (G1), low-grade appendiceal mucinous neoplasm diffusely involving the appendix. The tumor measured approximately 5 × 2 cm and was confined to the muscularis propria. Additionally, severe serositis of the fibrous membrane was observed, with involvement by endometriosis confirmed through immunohistochemical analysis (Figures 3 and 4).

Two days after surgery, the patient was discharged in stable condition. She remained relatively well during follow-up until six months later, when she developed colicky abdominal pain and abdominal distension. Subsequent evaluation included abdominopelvic ultrasonography, which revealed severe loculated anechoic fluid throughout the left side of the peritoneal cavity, displacing small bowel loops, raising suspicion for pseudomyxoma peritonei.

Abdominopelvic CT scan confirmed a large fluid collection of approximately 2500 cc in the lower abdomen and pelvic cavity without thick septations or solid components, causing anterior and superior displacement of bowel loops. Due to the significant ascites, the patient underwent ultrasound-guided abdominal paracentesis, during which approximately 120 cc of yellowish, clear fluid was aspirated and sent for laboratory analysis.

Cytopathological examination showed no malignant cells, with only a few lymphocytes, histiocytes, and polymorphonuclear leukocytes present. Gram stain and culture were negative for microorganisms and showed no growth. Other biochemical parameters indicated a transudative nature of the fluid. Tumor markers CA-125 and CEA were within normal limits.

The patient was closely followed, with gradual improvement in symptoms and resolution of the ascites, and her condition remained stable.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Discussion

As a rare fibroinflammatory disorder, Encapsulating peritoneal sclerosis (EPS), also known as abdominal cocoon syndrome, is characterized by a dense fibrous membrane formation encasing the small

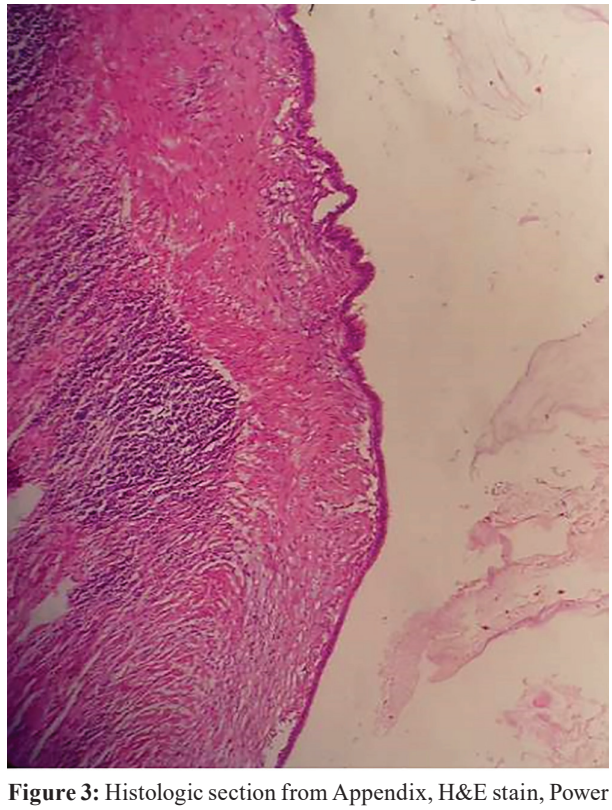

Figure 3: Histologic section from Appendix, H&E stain, Power 100, single layer of mucinous epithelium with low grade atypia without invasion and intraluminal mucin consistent with low grade appendiceal mucinous neoplasm (LAMN)

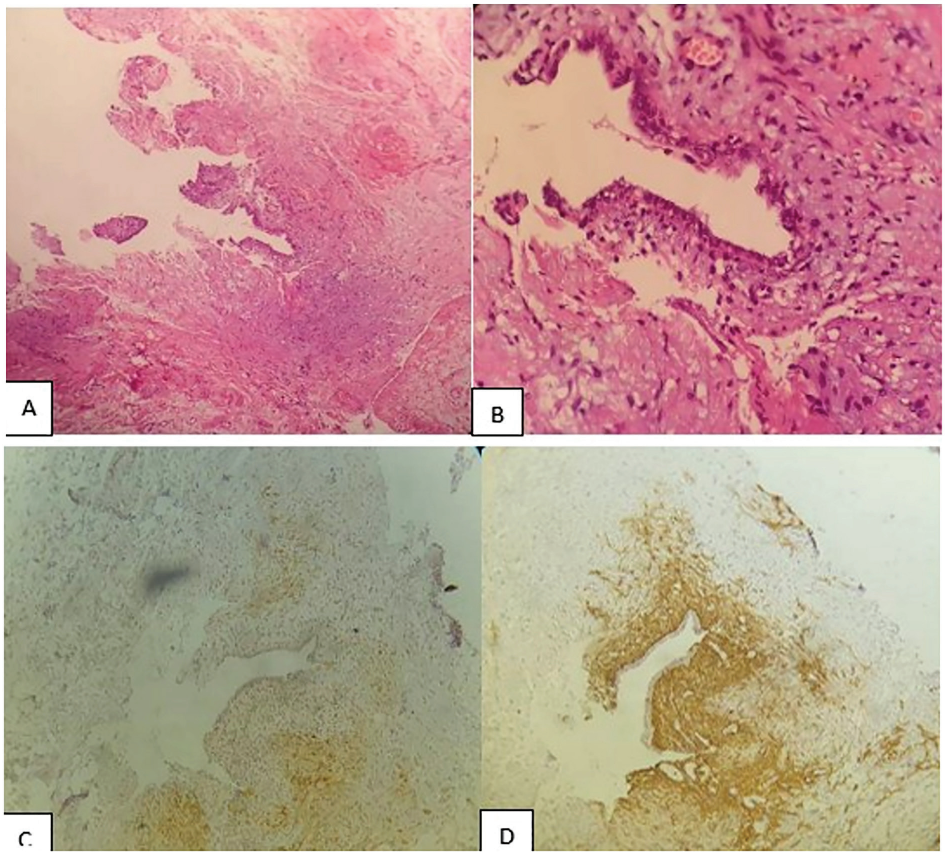

Figure 4: Histopathological image showing dense fibrocollagenous tissue with chronic inflammatory cell infiltration, confirming the diagnosis of abdominal cocoon. A) H&E stain, power 100, a focus with epithelial lining and stroma like endometriosis, B) power 400 of the same area, C) IHC stain for calretinin that is negative in epithelial lining, D) IHC stain for CD10 that is positive in stroma consistent with diagnosis of endometriosis.

bowel while involving other organs occasionally. Whilst most reported cases present with resurrent abdominal pain or subacute intestinal obstruction, diagnosis is frequently made intraoperatively due to the nonspecific nature of clinical and radiologic findings (10).

Contrary to the typical presentation, our patient developed with an acute clinical picture in favor of appendicitis, a presentation which is rarely introduced in the literature. It underscores the diagnostic challenges, especially when it pretends as common surgical emergencies. According to the reported cases, the intermittent pattern of symptoms leads to delayed recognition. Our case emphasizes on maintaining a broad differential diagnosis, particularly in young females exhibiting right lower quadrant pain. In most cases, like our study, the exact etiology and pathogenesis remain unclear; however, mesothelial cells, inflammatory cells, fibroblasts, and cytokines are believed to contribute to the fibrotic process and neoangiogenesis (11).

In our case radiologic assessment revealed nonspecific findings in accordance with previous studies that CT imaging are suggestive of obstruction; however, rarely confirms EPS preoperatively. Therefore, imaging is insufficient for diagnosis and surgical exploration is the gold standard.

The idiopathic EPS has an unknown pathogenesis. However, these are some hypotheses including mesothelial cell injury, chronic inflammation, fibroblast activities and cytokine- mediated fibrosis and neoangiogenesis. Our findingsare consistent with this framework. The absenceof prior peritoneal dialysis, TB, or abdominal surgery in our patient supports the idiopathic nature of the condition.

Considering similar case reports, comparative analysis reveals both concordance and divergence. Chien et al. described that nutritional status and early surgical intervention had a great effect on EPS patients' outcomes (10) which aligns with our patient's favorable postoperative course. Conversely, Al- Lawati et al. emphasized the diagnostic complexity in long-term dialysis patients (12), which is not applicable to our case. Mudarres et al. study revealed a similar idiopathic pattern of presentation with delayed diagnosis, highlithing the need for surgical awareness (13). These differences explain the heterogeneity of EPS and the need for individualized diagnostic and therapeutic strategies.

Our case contributes to the growing recognition of EPS in population outside the common geographic and demographic distributions. It also signifies the importance of surgical awareness and intraoperative vigilance, while encountering unexplained fibrotic encapsulation.

Future research should focus on improving noninvasive diagnostic modalities, including advanced imaging techniques and biomarkers profiling.

Investigating hormonal, genetic, and immunologic contributors may help identify at-risk individuals and guide preventive strategies. Furthermore, multicenter registries and larger case series are essential to establish standardized management protocols and assess long-term outcomes.

Conclusion

Encapsulating peritoneal sclerosis (EPS) is a rare and diagnostically challenging cause of intestinal obstruction. This case demonstrates an atypical presentation mimicking acute appendicitis, underscoring the importance of considering EPS in patients with unexplained abdominal pain or obstruction. Intraoperative recognition and careful excision of the fibrotic membrane were critical for definitive diagnosis and management. Awareness of such unusual presentations can aid timely diagnosis, guide appropriate surgical intervention, and improve patient outcomes.

Authors' Contribution

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work. Concept and design: S.M.K.T and M.M.S Acquisition, analysis, or interpretation of data: M.M.S. and A.A.F and M.S Drafting of the manuscript: M.M.S Critical review of the manuscript for important intellectual content: S.M.K.T and M.M.S Supervision: S.M.K.T and M.M.S.

Conflict of interest: No conflict of interest.

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