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Case Report

Bowel obstruction complicating an encapsulating peritoneal sclerosis: A case report ☆

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ABSTRACT

Encapsulating peritoneal sclerosis (EPS) is a rare pathological entity that can be classified into an idiopathic or secondary affection. Bowel obstruction is a frequent complication that has a characteristic radiological finding, with a cocoon appearance. We present the case of a 40-year-old man who suffered an absolute constipation. He underwent a CT scan that was suggestive of encapsulating peritoneal sclerosis or abdominal cocoon responsible for small bowel obstruction.

Introduction

Encapsulating peritoneal sclerosis (EPS), also known as peritonitis chronica fibrosa incapsulata, was first identified by Owtschinnikow in 1907. This rare condition is a significant cause of bowel obstruction characterized by the encasement of the small bowel within a fibrocollagenic cocoon-like sac. It can be categorized as either secondary or idiopathic [1]. Peritoneal dialysis represents the most common cause in the secondary category, while instances without identifiable risk factors fall under the idiopathic classification, often termed abdominal cocoon, as observed in this case [2].

Case report

A 40-year-old patient, with no previous medical conditions except for postbulbar ulceration, presented at the emergency room with a 3-day history of absolute constipation, acute abdominal pain, and nausea. On physical examination, the patient exhibited abdominal distension, diffuse tenderness, and a tachycardic state (106 beats per minute). Laboratory test results showed an elevated white blood cell count (WBC) of 15,320/mm^3 and a C-reactive protein of 33.5 mg/dL.

Suspecting bowel obstruction, a contrast-enhanced CT scan was conducted, revealing mildly distended and thickened ileal loops in the hypogastric quadrant. These loops were

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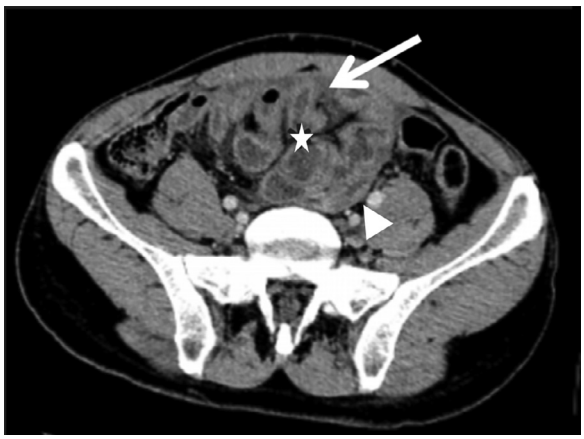

Fig. 1 – Axial contrast-enhanced CT image showing a cluster of encased bowel loops (asterisk), and peritoneal fluid (arrow) within a thickened peritoneal membrane with contrast uptake (head arrow).

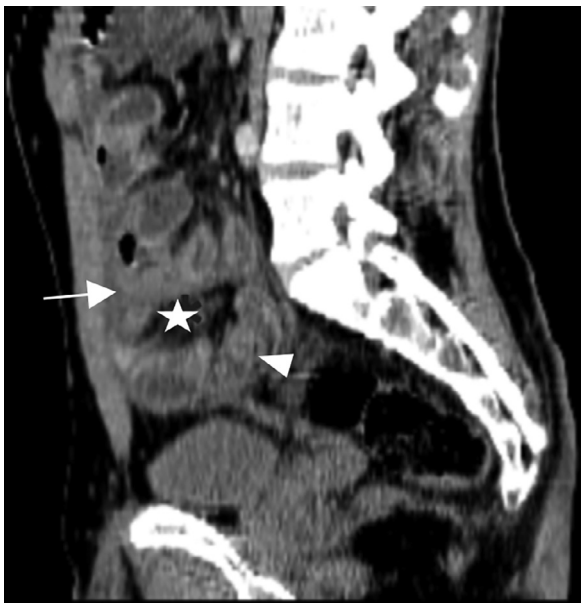

Fig. 2 – Sagittal contrast-enhanced CT image showing a cluster of encased bowel loops (asterisk), and peritoneal fluid (arrow) within a thickened peritoneal membrane with contrast uptake (head arrow).

enclosed within a sac formed by a thickened peritoneal membrane exhibiting a "cocoon-like appearance," contributing to upstream bowel dilatation with air-fluid levels (Figs. 1, 2 and 3). The presence of peritoneal fluid within the sac was also noted (Fig. 1). Additionally, perihepatic calcifications were observed within the thickened peritoneal membrane (Fig. 4). These CT findings suggested the possibility of encapsulating peritoneal sclerosis or an abdominal cocoon causing small bowel obstruction. Despite considering an internal hernia, this was ruled out due to the distinctive presence of the peritoneal membrane encasing the ileal loops.

Upon admission to the operating room, dense fibrous tissue resembling a cocoon enveloping the ileal bowel was dis-

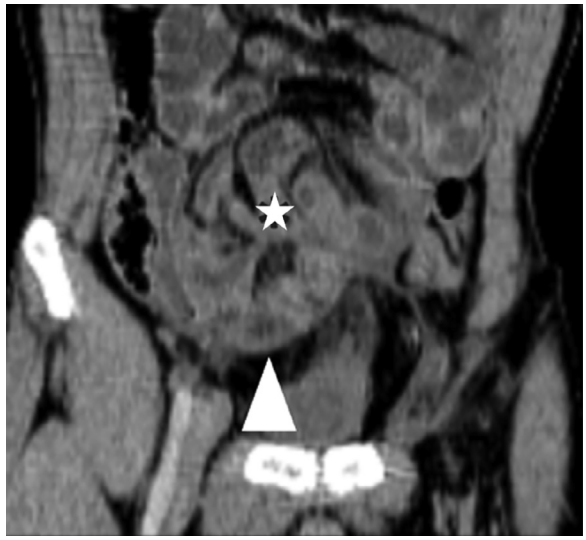

Fig. 3 – Coronal contrast-enhanced CT image showing a cluster of encased bowel loops (asterisk) within a thickened peritoneal membrane with contrast uptake (head arrow).

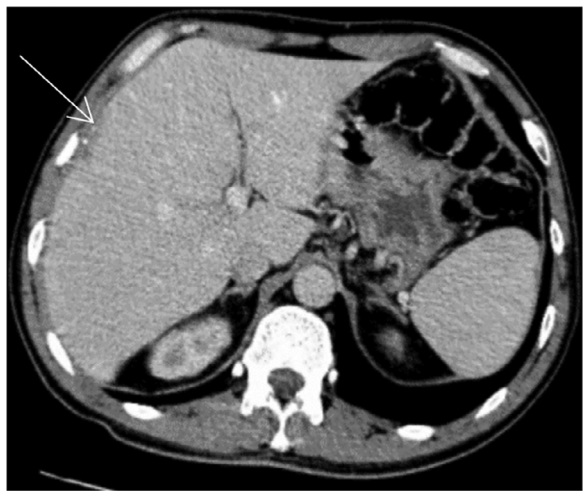

Fig. 4 – Contrast-enhanced CT axial image showing calcifications within the thickened peritoneal membrane in perihepatic.

covered, mirroring the sac identified in the contrast-enhanced CT scan. Excision of the fibrous tissue was performed to liberate the loops and alleviate the upstream bowel dilatation. Histopathological analysis revealed dense fibrosis with dystrophic calcifications and focal necrosis.

Following the surgical procedure, the patient's recovery was uneventful, allowing for a return to a regular diet, and discharge from the hospital occurred a week later.

Discussion

EPS represents a rare pathological entity that can be categorized as either idiopathic or secondary. While peritoneal dial-

ysis remains the primary cause [1], this condition can also manifest in patients with peritoneal tuberculosis, sarcoidosis, prior abdominal surgery, or gastrointestinal malignancies [2]. The exact pathophysiology remains unknown, but individuals undergoing long-term peritoneal dialysis have chronic peritoneal inflammation due to the dialysis fluid's properties, causing tissue fibrosis and mesothelium loss [3].

The "two-hit theory" describes EPS as a result of injury from long-term peritoneal dialysis causing chronic peritoneal membrane injury, followed by a second hit, such as peritonitis, genetic predisposition, or a sudden interruption of peritoneal dialysis. However, the exact pathogenesis is still unknown [3].

Early clinical symptoms of EPS are typically an early feeling of fullness, anorexia, nausea, and altered stool habits, such as constipation or diarrhea. It can be associated with ascites stained with blood or indicators of inflammation such as fever and elevated CRP [3]. In the late stage of EPS, patients usually present with abdominal pain, a sensation of fullness, abdominal mass, and possible bowel obstruction. According to Kawakuchi et al. [4], this is due to the formation of a fibrous cocoon that progressively encloses the intestines, causing various complications [3].

Nakamoto divided EPS into 4 groups according to the clinical manifestation [5]:

- Stage 1: Pre-EPS stage: no inflammation, minor ascites, and no symptoms.
- Stage 2: Inflammatory stage: Patients exhibit symptoms such as nausea and diarrhea in line with intestinal edema and partial encapsulation of the intestine.
- Stage 3: Encapsulation: Bowel blockage symptoms due to the fibrous cocoon.
- Stage 4: Chronic stage of ileus: Patients experience complete obstruction of their bowels due to a thickening of the fibrous cocoon.

Several studies have investigated possible biomarkers in peritoneal dialysis effluent to aid in early EPS identification. Low CA125 levels indicating mesothelial cell loss and high levels of Interleukin-6 (IL-6) were observed years before EPS diagnosis, suggesting potential early diagnostic markers. However, more research is needed for specific indicators [3].

EPS is a rare cause of bowel obstruction, recognized for its distinctive appearance on contrast-enhanced CT scans. These scans reveal dilated small bowel loops with air-fluid levels, suggesting encapsulating peritoneal sclerosis by showing bowel loops enclosed within a thickened peritoneal membrane, potential calcifications, and peritoneal fluid collections [3].

However, interpretation and diagnosis might be challenging due to the uncommon, complex nature of the illness and the non-specific character of most CT scan findings. Therefore, surgery with peritoneal biopsy might be necessary to confirm the diagnosis. Macroscopically, late stages of EPS usually show a thicker brownish peritoneum, with significant fibrosis and intestinal loops attached to one another [3].

The main differential diagnosis in congenital peritoneal encapsulation is a very rare disorder characterized by aberrant gastrointestinal development in embryos [6].

Therapeutic management of EPS includes discontinuation of peritoneal dialysis, nutritional support, immunosuppressive therapy, tamoxifen, and surgery for late stages or in cases of bowel obstruction [3].

Conclusion

Therapeutic approaches for encapsulating peritoneal sclerosis encompass pharmacological and surgical options. Treatment strategies often involve excision of the membrane or adhesiolysis to release affected bowel loops. Surgical resection may be necessary in cases where bowel integrity is compromised, albeit carrying a risk of recurrence following membrane excision [7].

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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