

Abdominal Cocoon Syndrome: A Rare Cause of Intestinal Obstruction

Alvin Lyle Kim and Seung Hun Lee*

Department of Surgery, Kosin University Gospel Hospital, Kosin University College of Medicine, Busan, Korea

***Corresponding Author:** Seung Hun Lee, Department of Surgery, Kosin University Gospel Hospital, Kosin University College of Medicine, Busan, Korea.

Received: January 13, 2019; **Published:** February 26, 2019

Abstract

There are various causative factors which can lead to intestinal obstruction. One extremely rare cause is abdominal cocoon syndrome, also known as sclerosing encapsulating peritonitis (SEP). This disease is characterized by a thick fibrous membrane that wraps the abdominal organs, especially the small intestine. We report a SEP case, which presented as the cause of intermittent intestinal obstruction episodes. A 52-year-old male patient had several episodes of abdominal pain associated with dyspepsia, nausea and vomiting for 4 months. It was suspected to be SEP by preoperative contrast-enhanced computed tomography scan. Diagnostic laparotomy was performed. A thick fibrotic membrane covered the whole small intestine, especially the terminal ileum. Adhesiolysis and removal of the fibrotic membrane were done. The patient recovered successfully with no postoperative complications.

Keywords: *Intestinal Obstruction; Sclerosis; Peritonitis; Small Intestine*

Abbreviations

SEP: Sclerosing Encapsulating Peritonitis; CT: Computed Tomography; EGD: Esophagoduodenoscopy; IC: Ileocecal; CAPD: Continuous Ambulatory Peritoneal Dialysis

Introduction

SEP is a rare disease, which can cause an intestinal obstruction. The pathogenesis of this disease is unknown. It is characterized by a thick fibrous membrane wrapping the abdominal organs, especially the small intestine [1]. It is theorized to be related to an increased release of fibrogenic cytokines that transform the fibrin-like materials on the peritoneum. It can be categorized into two groups by pathogenesis: Primary, which is idiopathic in nature, and secondary, which is related to various underlying diseases [2,3]. Herein, we present a suspected SEP case by preoperative contrast-enhanced computed tomography scan (CT). This case allowed us to analyze SEP from various angles using radiological, surgical, and histopathological characteristics of idiopathic SEP in order to understand its pathophysiology.

Case Report

In our case, a 52-year-old male suffered from intermittent intestinal obstruction for 4 months. The patient first visited a local clinic and underwent esophagoduodenoscopy (EGD) and colonoscopy. As there were no significant lesions, he took digestive medicines for several weeks, which did not relieve his symptoms. He had no significant medical history. There was no evidence of peritonitis. Initial laboratory investigation showed an elevated white cell count of 11.30 x/L (reference range 4.7 - 9.6 x/L). Abdominal plain film was normal. Contrast enhanced CT demonstrated mild dilatation with encasement of the small bowel divided into multiple sacs by a thick membrane (Figure 1). The image showed no evidence of bowel ischemia or lymphadenopathy. SEP was suspected by preoperative CT scan. The patient underwent diagnostic laparotomy. The laparoscopic approach failed because of the severe adhesion between the peritoneum and the sac membrane, which was wrapping the small bowel. The whole bowel was covered by a thick fibrotic membrane (Figure 2). After removing the huge sac, multiple lesser sacs were seen. The fibrous membrane with small bowel adhesions extended from the Treitz ligament to the ileocecal (IC) junction forming 4 lesser sacs; each sac was formed by a length of dilated small bowel loops with fluid (Figure 3). The thickest sac was 80 - 100 cm proximal to the IC junction. The thickness of the membrane grew gradually thinner proximally; the membrane thickness covering the bowel and the mesentery was about 0.3 cm at the main sac and less than 0.1 cm around the Treitz ligament. Cystostomy of the sac and adhesiolysis were subsequently performed, and then the total length of the small bowel got longer, and peristaltic

movement was observed. Removing the whole fibrotic membrane was impossible. Incision of the membrane was done longitudinally at several small bowel lesions (Figure 4). Histology of the sac showed a dense fibrocollagenous wall and underlying loose connective tissue (Figure 5). There was no evidence of cellular atypia or malignancy. The patient recovered successfully without complication and was discharged on postoperative day 8.

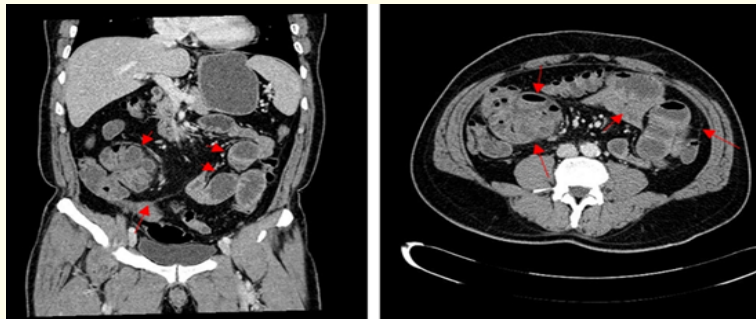


Figure 1: Contrast-enhanced CT in coronal and axial sections showing abdominal cocoon. Red arrows indicate the fibrous membrane around the bowel loop.

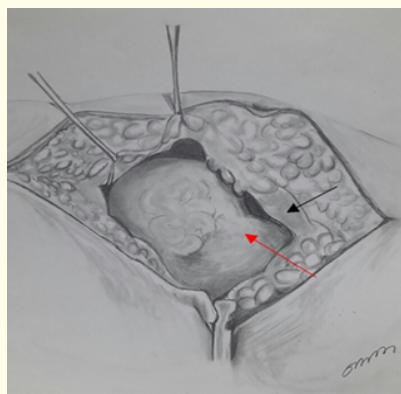


Figure 2: Illustration of the peritoneum. The red arrow is showing the fibrous membrane, which covers the whole bowel. The black arrow indicates the thickened peritoneum.

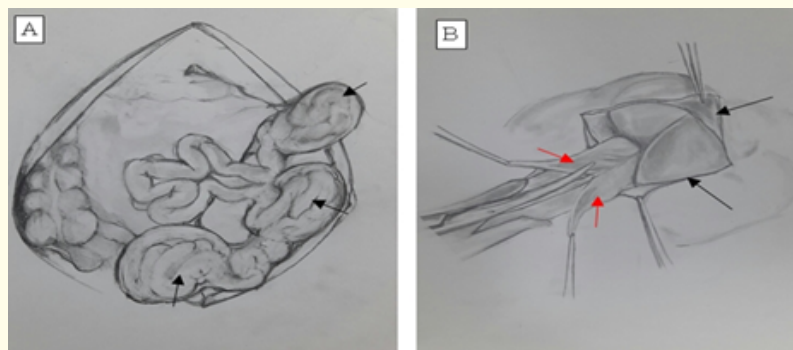


Figure 3: Illustration shows multiple lesser sacs (black arrows) formed by a certain length of dilated small bowel loops with fluid (A). Opening the lesser sac (black arrows), each small bowel was wrapped by a fibrous membrane (red arrows) (B).

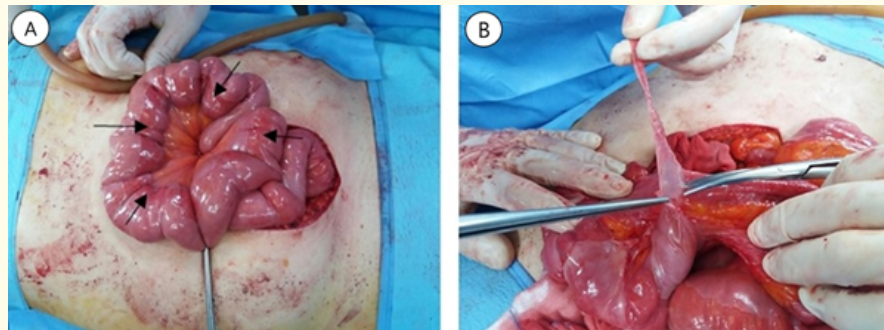


Figure 4: Observed surgical image of the small intestine. Black arrows showing incision sites at proximal jejunum, which was covered by a thin fibrous membrane (A) and a thick fibrous membrane at terminal ileum (B).

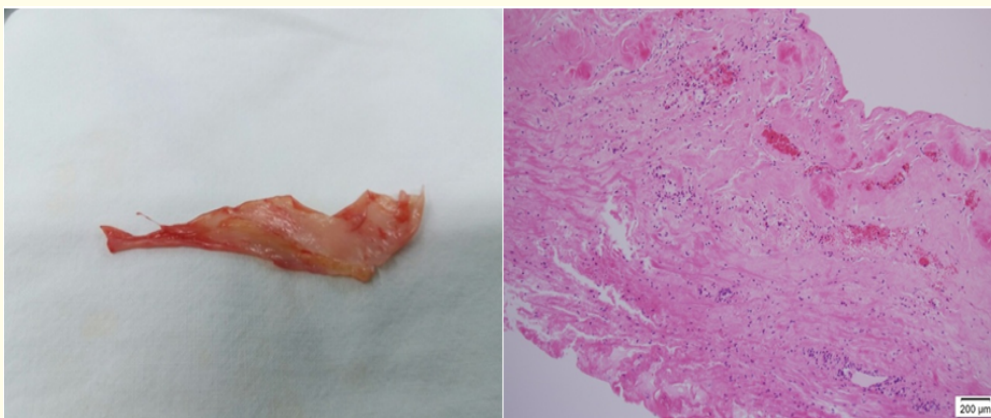


Figure 5: Fibrous membrane tissue and histological section; Histology of the cyst wall features a fibrocollagenous wall and underlying loose connective tissue.

Discussion

Idiopathic SEP is a chronic inflammatory process resulting in formation of a fibrocollagenous membrane, which can encase any organs in the abdomen [1]. It was first described by Owtschinnikow in 1907 and was called peritonitis chronica fibrosa encapsulate [3,4]. SEP can be classified into two forms. Primary (Idiopathic) SEP is known as abdominal cocoon syndrome. Secondary SEP is a more common form and is associated with continuous ambulatory peritoneal dialysis (CAPD), peritoneal-venous shunting, prolonged intake of beta-blockers (practolol), malignancy, penetrating abdominal injury, connective tissue diseases, and abdominal tuberculosis [2,3]. Although SEP is a rare disease, it is a highly treatable condition. Symptoms like intermittent vomiting, diarrhea, abdominal pain, and sudden abdominal distension with subacute bowel obstruction signs can act as clues. Preoperative imaging is important and contrast CT scan can be a good modality to make a diagnosis and to aid differential diagnoses. CT scan can be the most common requested imaging modality and helps guide SEP diagnosis [5,6]. The final diagnosis is made by operation [4]. Removing the fibrotic membrane via adhesiolysis and releasing the entrapped bowel can be the treatment of choice for SEP [7]. Bowel resection is required if there are signs of ischemic change to the bowel. Primary SEP has an excellent prognosis with low rates of complication while CAPD-associated SEP has a one-year mortality rate of 30 - 57% [8]. Histology often shows associated inflammatory cells with calcification. Its pathogenesis is not completely known. In our case, there was a bull's-eye pattern. As the wrapping was further from the main sac, the thickness of the fibrous layer which was entrapping the small bowel gradually became thinner.

Conclusion

Idiopathic SEP is rare, but benign and highly treatable. Secondary SEP, such as tubercular abdominal cocoon can be treated with medication initially. For that reason, identifying underlying diseases is important to prevent unnecessary operations. The recurrence rate is up to 20% after surgery, thus close observation with regular check-ups are required [9].

Conflicts of Interest

The authors have no conflicts of interest.

Bibliography

1. Rajagopal AS., *et al.* "Conundrum of the cocoon: report of a case and review of the literature". *Diseases of the Colon and Rectum* 46.8 (2003): 1141-1143.
2. Naidoo K., *et al.* "Small bowel injury in peritoneal encapsulation following penetrating abdominal trauma". *Case Reports in Surgery* (2013): 379464.
3. Tannoury JN., *et al.* "Idiopathic sclerosing encapsulating peritonitis: abdominal cocoon". *World Journal of Gastroenterology* 18.17 (2012): 1999-2004.
4. Akbulut S. "Accurate definition and management of idiopathic sclerosing encapsulating peritonitis". *World Journal of Gastroenterology* 21.2 (2015): 675-687.
5. Tarzi RM., *et al.* "Assessing the validity of an abdominal CT scoring system in the diagnosis of encapsulating peritoneal sclerosis". *Clinical Journal of the American Society of Nephrology* 3.6 (2008): 1702-1710.
6. Vlijm A., *et al.* "Computed tomographic findings characteristic for encapsulating peritoneal sclerosis: A Case-Control Study". *Peritoneal Dialysis International* 29.5 (2009): 517-522.
7. Qasaimeh GR., *et al.* "Laparoscopic diagnosis and management of an abdominal cocoon: a case report and literature review". *Surgical Laparoscopy Endoscopy and Percutaneous Techniques* 20.5 (2010): e169-e171.
8. Goodlad C., *et al.* "Encapsulating peritoneal sclerosis: what have we learned?" *Seminars in Nephrology* 31.2 (2011): 183-198.
9. Wei B., *et al.* "Diagnosis and treatment of abdominal cocoon: a report of 24 cases". *American Journal of Surgery* 198.3 (2009): 348-353.

Volume 6 Issue 3 March 2019

©All rights reserved by Alvin Lyle Kim and Seung Hun Lee.