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Encapsulating peritoneal sclerosis: A case report describing the surgical management of such a dilemma

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ABSTRACT

Abdominal Cocoon syndrome is a rare entity of disease that is characterized by an encapsulation of the small bowel resulting in the development of tissue fibrosis of the peritoneal surface. This is a 27 years old male vitally stable with a background of end-stage renal disease (ESRD) who presented to the Emergency department (ED) multiple times with ileus-like symptoms, leukocytosis and critically elevated creatinine soon after, we performed a Computed tomography (CT) that concluded Loculated fluid collections with thickening and enhancement of the peritoneal reflections. The patient was taken to the Operating room (OR) for a laparoscopic adhesiolysis and resection of the cocoon; Encapsulating peritoneal sclerosis (EPS) is a serious dilemma, imaging can raise suspicion of diagnosis and surgery is a crucial management for such an entity with nonspecific clinical and radiological features.

Keywords: Encapsulating peritoneal sclerosis, Sub acute intestinal obstruction, acute abdomen, general surgery, acute care surgery, nephrology, peritoneal dialysis

1. INTRODUCTION

Abdominal Cocoon syndrome is a rare entity of disease that is characterized by a complete or partial encapsulation of the small bowel resulting in the development of tissue adhesion, fibrosis and vasculopathy across the affected peritoneal surface exaggerating a presenting status of chronic ileus (Kawanishi et al., 2006; Vagholkar and Doctor-Ganju, 2018). As the prevalence of renal failure needing peritoneal dialysis has grown over time, the surgeon now frequently encounters this situation (Leeoloy et al., 2021). Knowledge of this condition is crucial for an early diagnosis and the best possible care (Vagholkar and Doctor-Ganju, 2018). In this case report, we will present clinical and radiological findings and complicated surgical management of this disease. As there is a scarcity of case reports describing the diagnosis,

surgical management of such an entity (Leeoloy et al., 2021; Wei et al., 2009).

2. CASE PRESENTATION

A 27-year-old Saudi male with a background of ESRD since he was 15 years old resulted from focal segmental glomerulosclerosis (FSGS) since the age of 9 years old, maintained on peritoneal dialysis (PD) since he was 19 years old but removed PD catheter six months before his presentation due to multiple episodes of resistant peritonitis since he started PD, he had several other complications including Deep venous thrombosis (DVT) after right femoral temporary line insertion, childhood-onset seizure disorder all associated with hypothyroidism and hypoparathyroidism and Dilated cardiomyopathy with moderate left ventricular dysfunction.

Presented to the ED complaining of left upper quadrant abdominal pain, colicky in nature, not radiating and partially relieved by analgesia started just after he completed his dialysis session, associated with nausea and vomiting, diarrhea without abdominal distention or fever, melena or hematochezia, he was vitally stable, we took the patient for abdominal X-ray and it showed air-fluid level we followed it with CT and it concluded signs of adhesive bowel obstruction with a large pelvic collection, the patient was discharged after he was responded to conservative management and drainage of the collection, nine days later the patient presented to the ED again with a similar image however on physical examination, his abdomen becomes distended and tender on palpation with digital rectal examination being unremarkable, his blood investigation revealed an elevation of white blood cell count with neutrophilic predominance, hemoglobin was reaching as low as 9.2 g/dL, platelets count, potassium and blood urea nitrogen level all were elevated and serum creatinine was critically high, reaching 1090 mmol/L with the serum lactic acid level being within the normal range. CT scan was done at a time it showed what represent a thickening and enhancement of the peritoneal reflections. Minimal traces of pelvic free fluid are visualized with a huge right-side abdominopelvic collection (Figure 1A).

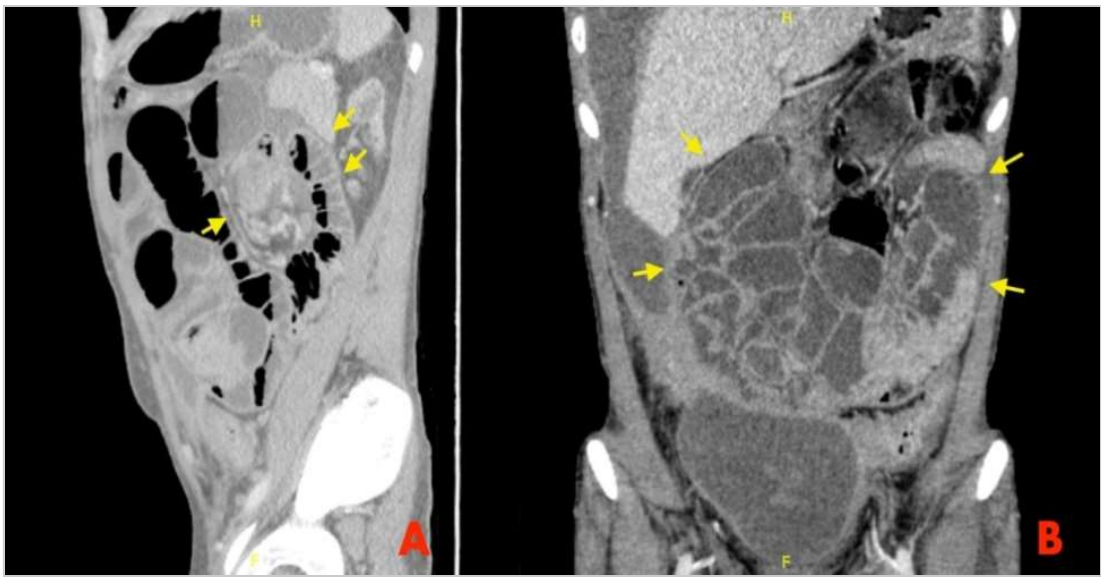

Figure 1 A: Axial CT with no contrast reveals air fluid levels consistent with blockage and dilated bowel loops (arrows). B: Coronal CT scan with no contrast with any contrast reveals an abdomen filled with fluid that is encircled by an enhanced, thickened peritoneum

He was taken to the OR for diagnostic laparoscopy abdomen was accessed initially by the closed technique using a Veress needle in palmer's point. However, we could not safely enter the abdomen after the saline test failed, so we entered the supra-umbilical area using the open technique. The skin incision was made and we dissected through subcutaneous tissue until reaching the fascia. The fascia was opened using a blade and the peritoneum was opened bluntly. We inserted a 12-millimeter (mm) port .and after abdomen was insufflated; we inspected the entry site for any injury or bleeding. The port was inserted into the cyst cavity, as visualized earlier in the CT scan Pigtail catheter tip was seen in the upper abdomen. Intra operatively we found an Extensive adhesion with a picture of a frozen abdomen (Figure 2A).

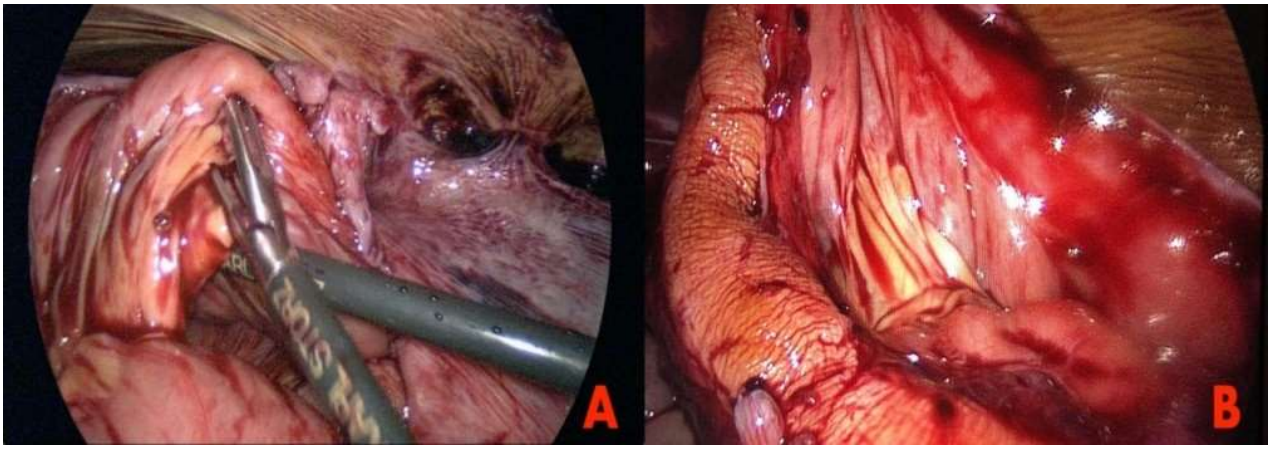

Figure 2 Intraoperative laparoscopic findings A: Inlet of the cocoon with hematomas; B: Outer vision of the cocoon, with cobbles toning of the small bowel

Both transverse colon and liver were adherent to the anterior abdominal wall and the small intestine had an appearance of cobble stoning (Figure 2B) a large cavity found along the right side of the abdomen extending from the diaphragm to the right iliac fossa with thick-walled cyst afterword we inserted three additional ports with a size of 5 mm in the right upper quadrant, subxiphoid and left upper quadrants. Under direct vision with no injury or bleeding, cystic cavity content was suctioned; we followed that with resection of part of the cyst, then we extract the cyst through the supra-umbilical site using an Endo bag and sent it for pathology/culture. We performed adhesiolysis using a combination of sharp and blunt dissectors (Figure 2A, 2B), releasing all of the small bowels safely with no obvious enterotomies or injury. We ran the bowel from the ileocecal valve to the duodenojejunal flexure to find that multiple segments of the abnormal cobblestone appeared small bowel wall (Figure 3B).

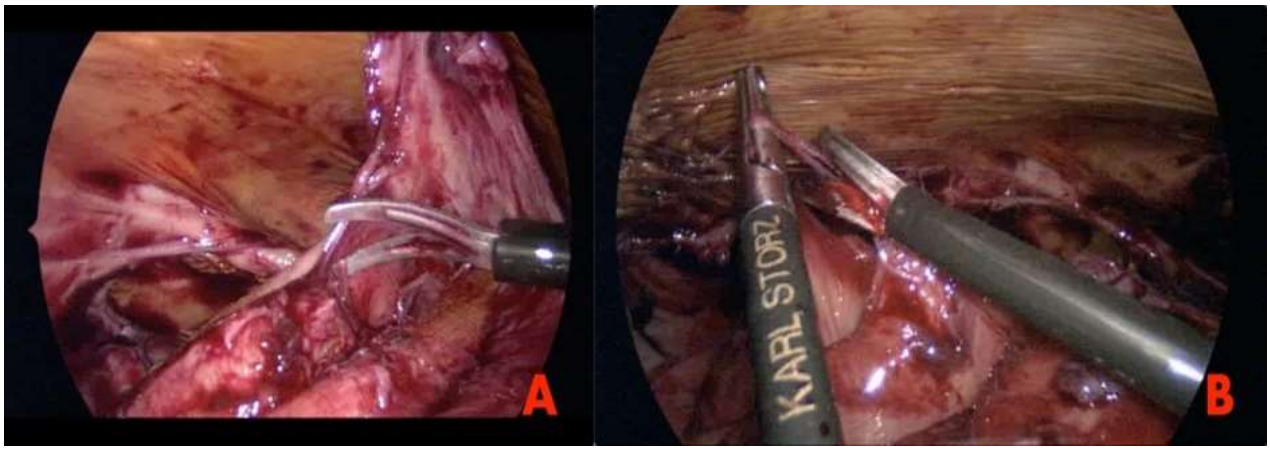

Figure 3 Intra operative laparoscopic views of adhesionlysis. A: Adhesions formation as a part of intestinal cocooning. B: The inter-enteric sclerotic membranes that surround the small intestine resemble cobwebs

Multiple small hematomas occurred along the small bowel wall (Figure 3A). We reexamined those hematomas prior to closure and we found them to be small with no expansion. Copious irrigation and suctioning were done for the abdomen. Two large 19 French Blake (FR) Blake Drains were introduced and brought out through the previously inserted port sites, fixed to the skin using silk suture right lateral drain was positioned along the right paracolic gutter within the cyst cavity, the subxiphoid right upper drain positioned the pelvis and along the left paracolic gutter and Hemostasis was ensured. 12 mm supra umbilical port closed with an enclosure device using polydioxanone I suture in a figure-of-eight manner. All ports were retrieved from the abdomen under direct vision with no bleeding. The wounds were closed with Monocryl 4-0 suture subcuticular in an interrupted fashion. Steristrips and simple dressing were applied to all wounds. Extubation was done and the patient tolerated the procedure without immediate complications. However, towards the end of the procedure, the patient had an acid-base and electrolyte disturbance, so he was shifted to the intensive care unit (ICU) for observation. We downgraded the patient to the ward the following day. The postoperative course went smoothly with no complications.

4. DISCUSSION

Encapsulating Peritoneal Sclerosis (EPS), EPS was first described in 1980 in a case series of five patients on peritoneal dialysis who had laparotomies for unrelated causes, during which a severe thickening and sclerosis of the whole peritoneal surface was appreciated intra operatively, creating a rim that bound the loops of the bowel, ever since a great variety of symptoms have been recorded by several institutes (Gandhi et al., 1980). Linked to what is now known as encapsulating peritoneal sclerosis (Gandhi et al., 1980; Machado, 2016), with a not fully understood pathology and multifactorial nature of such a disease (Kawanishi et al., 2006; Machado, 2016). EPS has persisted as a diagnostic dilemma with unclear risk factors that can predict such a disease; several studies, however, had described PD duration to be the strongest risk factor for the development of EPS; it is extremely uncommon during the first three years of PD therapy and then became more common after that as reported by Brown et al., (2009) and Leeoloy et al., (2021) we noticed this in our patient with a duration of dialysis exceeding eight years which is higher than the intervals observed in other cases described by those investigators. Our case had an early onset of seizures and we attribute this to hypertensive encephalopathy similarly noticed in some presented cases in the literature (Leeoloy et al., 2021). Signs and symptoms were not specific for this disease, with the most common symptoms being non-specific ileus-like symptoms such as nausea, vomiting, diarrhea, intermittent abdominal pain, anorexia and loss of appetite.

The physical exam can also be unremarkable. An abdominal mass with tenderness in light and deep palpation, hypoactive bowel sounds and potentially abdominal rigidity can all be appreciated in some cases (Brown et al., 2009; Kawanishi et al., 2006; Leeoloy et al., 2021; Machado, 2016). We cannot simply relay on a single specific test to diagnose this disease; bowel obstruction and dilated small bowel on imaging are among several factors had been noticed in many cases, including ascites, septation and peritoneal thickening (Brown et al., 2009; Ti et al., 2010). However, the uncertainty of those markings, a study described a sign that has been noticed in a patient with intra-abdominal cocoon, a "cocooning of the small bowel" in which the peritoneum encircles the colon as a defining characteristic of EPS (Ti et al., 2010). Cocooning was found to be a clear description of CT scan findings in our case. Although radiological characteristics are not diagnostic, a greater suspicion could be raised on this condition by imaging, as it can help with pre-operative diagnosis (George et al., 2007).

Contrary to many cases that managed this case with a laparotomy fashion surgery (Wei et al., 2009), we intervene in this patient in a laparoscopic fashion; the best approach for such a case is to perform adhesiolysis and to do an excision of the covering membrane to release thorough small bowel, regardless of the cause of forming adhesions, adhesiolysis continues to present the largest surgical obstacle to the operating surgeon. EPS has two folds and here, where its difficulty persists; attempts to cut through the adhering loops' membrane may harm the serosa and frequently result in bowel perforations (Vagholkar and Doctor-Ganju, 2018; Wei et al., 2009). When there is a vascular compromise, bowel resection may be required.

5. CONCLUSION

EPS is the most serious complication of peritoneal dialysis. Early diagnosis by imaging followed by appropriate surgical management is crucial in such an entity with nonspecific clinical features.

Author Contributions

Fayez and Hosam composed the manuscript and literature review; Ali and Salman provided imaging and case presentation; Khaled and Thamer, revising it critically for important intellectual content, final approval of the version to be published.

Informed consent

The written informed consent was obtained from the patient.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data collected during this study are available upon reasonable request from the corresponding author.

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