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Case report

Encapsulating peritoneal sclerosis as a late complication of peritoneal dialysis

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HIGHLIGHTS

- We presented the patient experienced surgery because of encapsulating peritoneal sclerosis (EPS).
- EPS should be born in mind as a complication of the long term peritoneal dialysis.
- Diagnosis is primarily based on clinical finding, then generally confirmed by CT.
- Early identification of EPS is important in order to achieve better prognosis.

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ABSTRACT

Introduction: Encapsulating peritoneal sclerosis (EPS) is a rare cause of intestinal obstruction which is characterized by fibrotic encapsulation of the bowel. Although its pathogenesis is still not clear, many etiological factors have been stated.
Presentation of case: In this report, we present a 26-year old woman with peritoneal dialysis related EPS. Because of the unresolving intestinal obstructive symptoms, she underwent surgical intervention in which the thick dense whitish membranous sac was excised from the surrounding intestine along with adhesiolysis. She recovered uneventfully. She is symptom-free on the eight months of follow-up.
Discussion: EPS should be born in mind as a complication of the long term peritoneal dialysis in patients with progressive obstructive ileus and recurrent peritonitis. Its treatment either medically or surgically varies depending on the stage of this entity.
Conclusion: Early identification of EPS is important in order to achieve better prognosis.

1. Introduction

Encapsulating peritoneal sclerosis (EPS) is a rare cause of intestinal obstruction which is characterized by fibrotic encapsulation of the bowel due to a progressive intraabdominal inflammatory process [1]. Firstly Owtschinnikow [2] described this condition as 'peritonitis chronica fibrosa incapsulata' in 1907. Then, different terms such as sclerosing encapsulated peritonitis, sclerotic thickening of the peritoneal membrane, abdominal cocoon, peritoneal fibrosis, peritoneal sclerosis have been used but encapsulating peritoneal sclerosis is more descriptive and appropriate term because of the frequently absence of inflammatory component in the marred peritoneum [1].

EPS could be classified as primary (idiopathic) or secondary type. The former is usually seen in adolescent girls from tropical countries and peritonitis evoked by retrograde menstruation may be a causative factor in this form [3]. The latter is more common. EPS is commonly referred to as being multifactorial, genetic susceptibility may also be an etiological agent [1]. Which predisposing factor prompt the patient to proceed to EPS has not exactly known, however plenty of etiologies in the secondary form including abdominal surgery, generalized peritonitis, intraabdominal malignancy, intraperitoneal chemotherapy, ventriculoperitoneal shunt, β-blockers, peritoneal lavage using certain disinfectants such as chlorhexidine, and peritoneal dialysis have been stated as related to EPS [1–4].

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Although the prevalence of EPS among subjects under peritoneal dialysis (PD) is reported between 0.5% and 4.4%, it leads to devastating outcomes with the mortality rate varying from 20% to 93% (1,4). Longer duration of PD, acetate-buffered or hypertonic solutions, recurrent episodes of peritonitis which especially result from *Staphylococcus aureus*, fungi and Pseudomonas sp. are contributing factors to its development [1].

In this report, we present a case of EPS in a patient on peritoneal dialysis who underwent surgical management for her condition. A critical review of the relevant literature on this subject is also presented to determine the best approach.

2. Presentation of case

A 26-year old female suffering from hemolytic uremic syndrome related to end-stage renal disease had received automated peritoneal dialysis during ten years. She experienced renal transplantation 6 years ago, which resulted in early rejection of the transplant. The patient had 3 previous histories of peritonitis and 2 catheter exit-site infections. She presented with fever, abdominal pain, and cloudy peritoneal effluent. Her vital signs were as follows: the respiratory rate was 40 breaths/min, heart rate 110 beats/min and blood pressure 110/70 mm Hg.

She was cachectic and on physical examination mild abdominal tenderness and distension with hyperactive bowel sounds were detected as well as peritoneal dialysis catheter on the right lower quadrant. There was neither muscular defense nor rebound on her abdominal palpation. The blood chemistry was compatible with chronic renal failure. Laboratory tests were as following: creatinine 5.4 mg/dL, albumin 2.3 g/dL; hemoglobin 7, 8 g/dL; leukocytosis 13,000 mm [3]; CRP 86 mg/dL. *E. coli* was isolated from the peritoneal fluid, and antibiotic treatment was initiated.

On plain abdominal graphy there were few air-fluid levels. Contrast-enhanced abdominal computed tomography scans revealed diffuse mild thickening and contrast enhancement of the peritoneum along with massive ascites in the peritoneal cavity. Retroperitoneal calcified mass lesion compatible with ex-renal transplant was also seen in the left lower quadrant (Fig. 1). Jejunal and ileal loop walls appeared as thickened and edematous (Fig. 2). After 5 days of hospitalization, total parenteral nutrition was initiated due to intolerance of enteral route. Her catheter was removed 6 days later because of the resistance to treatment. Despite her transfer to hemodialysis therapy and adequate antibiotic treatment, abdominal symptoms and fever were persisted. She lost 17 kg during the clinical course of hospitalization. After surgical consultation, we decided to operate her because of the unresolving intestinal obstructive symptoms.

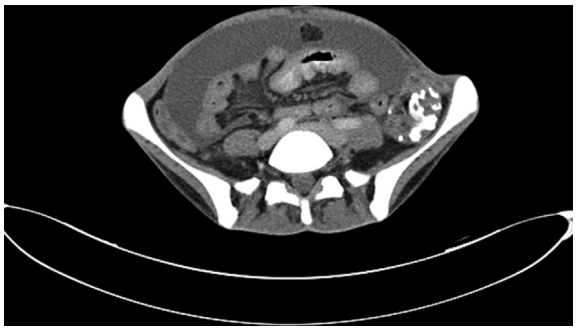

Fig. 1. Abdominal tomography scans reveal diffuse mild thickening and contrast enhancement of the peritoneum along with massive ascites. Retroperitoneal calcified mass lesion compatible with ex-renal transplant is seen in the left lower quadrant.

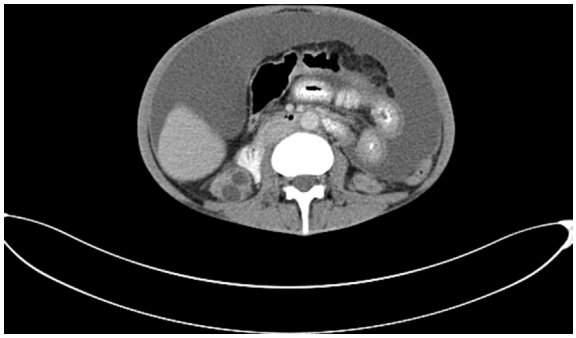

Fig. 2. Intestinal loops localized in the center of the abdomen due to massive ascites have thickened and edematous walls.

At the explorative laparotomy, ascites and a thick dense whitish membrane encasing small bowel and extending laterally to involve right colon was found. The membranous sac was excised from the surrounding intestine along with adhesiolysis. We repaired two ileal serosal injuries which had developed during the dissection of releasing of loops.

The histopathological evaluation of the membrane revealed fibrous tissue with absence of mesothelium and accompanied by massive collagen deposition, scattered mononuclear inflammatory cells and fibroblast proliferation, which reported as consistent with a diagnosis of EPS (Fig. 3). The specimen was evaluated by PAS and Giemsa stains which detected no microorganism.

The patient showed a significant recovery postoperatively and was discharged from the hospital uneventfully. She is symptom-free on the eight months of follow-up.

3. Discussion

EPS is a rare but life threatening complication of peritoneal dialysis. Moreover, the incidence is reached up to 17% for patients with history of PD longer than 15 years [5]. Its pathogenesis is unclear. Peritoneal inflammation and fibrosis occur as a response to peritoneal irritation. Ultimately, it results in encasement of the whole or part of the bowel in a thick shiny membrane which is a characteristic finding of this condition [5].

Symptoms of EPS depend on the disturbance of intestinal function due to obstructive and paralytic ileus, which account for progressive impairment in the nutritional status of the patient.

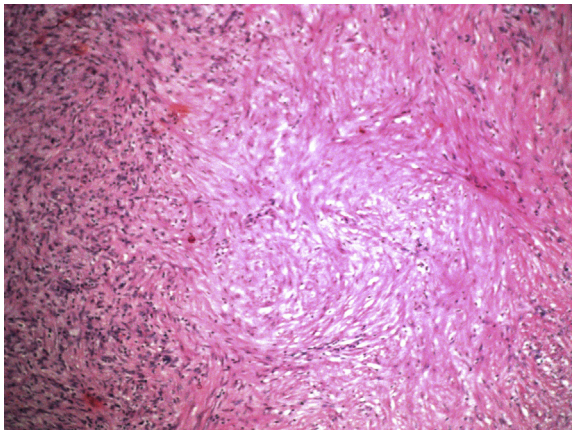

Fig. 3. Membrane composed of fibrous tissue and scattered mononuclear inflammatory cells on the pathologic exam with HEx200 are seen.

Because of the nonspecificity of its early clinical features, the diagnosis is very difficult before the patient develops bowel obstruction. The chronic course of the disease may help differentiating the EPS from other causes of ileus. A palpable abdominal mass constituted from encapsulated cluster of dilated small bowel loops can be another finding on clinical manifestation. Moreover, indolent peritonitis in the absence of positive peritoneal culture should alert the physician regarding the diagnosis of EPS. In some cases, bloody effluent may be observed. On the other hand, declining small solute clearance and failure of ultrafiltration capacity may be different signs of ESP. In addition, a history of peritonitis is not a prerequisite for its development [6]. In our case, the patient experienced multiple episodes of abdominal pain over the last two years.

There are no reliable predictive tests for EPS. Therefore, high index of suspicion is required in the diagnosis of this condition. Radiological examination also plays key role in the identification of the disease. Computed tomography (CT) is widely considered as the gold standard to image EPS [3]. CT demonstrates peritoneal and bowel wall thickening, peritoneal calcification, loculated fluid collection, small bowel tethering and dilatation [5].

Pathological exam establishes the diagnosis with complete loss of mesothelium accompanied by marked interstitial thickening composed of fibroblasts and collagen deposition within the peritoneal membrane. Inflammatory cells are variably present but leukocyte infiltration is not a crucial part of this diagnosis [5].

There is no uniformly accepted management of EPS. Conservative approach including nasogastric decompression, bowel rest and total parenteral nutrition is the initial treatment option for patients with mild symptoms. Transferring the patient to hemodialysis following the cessation of PD is an another choice. Whereas, EPS may even be accelerated after discontinuation of PD[6]. Medical treatment with steroids or immunosuppressive agents has been reported to be beneficial especially in the early inflammatory phase [6]. A recent study consisting of the large cohorts of 111 patients with EPS has showed that there was no difference on survival time among 54 patients treated with various medications [6]. Either by open or laparoscopic technique, surgical removal of the membrane from the peritoneal surface with releasing the intestine is an appropriate treatment option for patients who could not use gastrointestinal tract due to the end stage disease [3,7]. Achieving a clear cleavage plan between the membrane and bowel surface is extremely difficult [8]. Complications after surgical intervention include recurrent ileus, formation of intestinal fistula, and intraperitoneal abscesses. Mortality varies from 20% to 93% and increases with the duration on PD [1,4]. Death usually occurs in more than 60% of patients within 4 months of diagnosis [1]. On the other side, surgical mortality has been stated as low as 4% in experienced hands by She [9]. There is no obstacle for renal transplantation of patient with EPS. The overall median survival was 14 months (range 0–119) according to large cohorts study [6]. Moreover, better survival rates were obtained in transplanted patients compared to patients who were maintained on dialysis.

4. Conclusion

In which patient will develop EPS during the fate of the dialysis has not fully elucidated. Physicians should be familiar with symptoms, clinical and radiological signs suggestive of EPS. Diagnosis is primarily based on clinical finding, and then generally confirmed by CT. One of the goals of the management is early identification of the disease, which provides better prognosis.

Ethical approval

None.

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Author contribution

Ebru Oran: Study design and writing article.
Hakan Seyit: The acquisition of data.
Canan Besleyici: The acquisition of data.
Abdulkadir Ünsal: Revising article critically.
Halil Alış: Final approval of the version to be submitted.

Conflicts of interest

None.

Guarantor

Ebru Oran.

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