

Case Report

Encapsulating Peritoneal Sclerosis: Case Report and Short Review of the Literature

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Abstract

Encapsulating peritoneal sclerosis (EPS) is an uncommon complication of peritoneal dialysis. Diagnosis can be made with a clinical history and radiological or surgical findings. We present the case of a 39-year-old female presenting with small-bowel obstruction secondary to EPS, who was treated conservatively and had an adequate resolution. We show the main radiological findings in this patient and offer a synthesis of the literature regarding the main characteristics of these findings.

ABBREVIATIONS

EPS: Encapsulating Peritoneal Sclerosis; PD: Peritoneal Dialysis; ESRD: End Stage Renal Disease

INTRODUCTION

End stage renal disease (ESRD) is one of the most common diseases worldwide, and sometimes, peritoneal dialysis (PD) is the only treatment available in many patients. Encapsulating Peritoneal Sclerosis (EPS) is a rare complication of this treatment, but its associated high morbidity and mortality rates are worrisome. Thus, we present the following case to raise awareness of this complication and to highlight the main radiological characteristics.

CASE PRESENTATION

A 39-year-old female with ESRD secondary to systemic lupus erythematosus, treated with PD since she was 21 years old during which she developed three episodes of malfunction and replacement of peritoneal catheters and two episodes of peritonitis. The last episode of peritonitis occurred in 2015 and required catheter removal, consequently she started hemodialysis treatment (three times per week, 3 hour, 10 minute session, one liter of ultrafiltration, no uresis). Currently, she is waiting for a transplant.

Two years after the removal of the peritoneal catheter, she started having multiple episodes of diffuse abdominal pain which were first treated as irritable bowel syndrome. In the last episode, she presented in the emergency room with four days of abdominal pain, food intolerance, nausea, vomiting and diarrhea. Vital signs were normal, and laboratory tests showed mild anemia (hemoglobin 10.5 g/dL), evidence of an adequate dialysis scheme (BUN 33.7 md/dL creatinine 7.7 mg/dl), and a negative lipase (93

U/L), the rest of the exams were found to be within normal range. A plain abdominal film in the upright position showed small-bowel dilatation, multiple air-fluid levels and small calcifications at the pelvis, no evidence of pneumoperitoneum (Figure 1). An abdominal ultrasound reported an abdominal mass, however, the origin was not clear. An abdominal contrast-enhanced CT confirmed small-bowel tethering, dilatation and air-fluid levels, associated to smooth peritoneal thickening abnormalities and ascites and focal calcifications in the peritoneum (Figure 2). Current and a prior abdominal CTs done three years before were compared showing no previous peritoneal abnormalities and further development of calcification and peritoneal thickening along the course of the dialysis catheter (Figure 3). Surgery interconsultation was requested, and they decided to treat the patient conservatively with close monitoring, IV hydration, nasogastric tube, and a follow up re-evaluation 7 days after. No steroids or tamoxifen were given. She had a favorable clinical course. The patient was discharged and no further episodes of small-bowel obstruction up to 6 months of follow-up have been documented.

DISCUSSION

Encapsulating peritoneal sclerosis (EPS) is a rare complication of peritoneal dialysis. First described in 1980 by Gandhi et al., it is also known as peritoneal fibrosis, peritoneal sclerosis, sclerosing obstructive peritonitis, sclerosing encapsulating peritonitis, calcific peritonitis, abdominal cocoon, and sclerosing peritonitis [1]. Nevertheless, the term “encapsulating peritoneal sclerosis” is preferred because it describes more accurately the morphologic changes [2-4].

The International Society for Peritoneal Dialysis (ISPD) defines EPS as a clinical syndrome with persistent or recurrent



Figure 1 Plain abdominal films in upright position (a) and decubitus (b and c) show small bowel dilatation, fluid levels, multiple vascular calcifications (arrowheads) and a non-vascular calcification (arrow). Abdominal CT correlation of the calcification is shown in panel d (arrow).

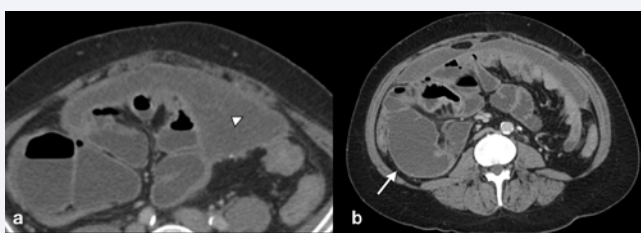


Figure 2 Contrast-enhanced abdominal CT scan showing loculated and septated ascites (arrowhead), peritoneal thickening, small bowel tethering and focal small-bowel dilatation (arrow).

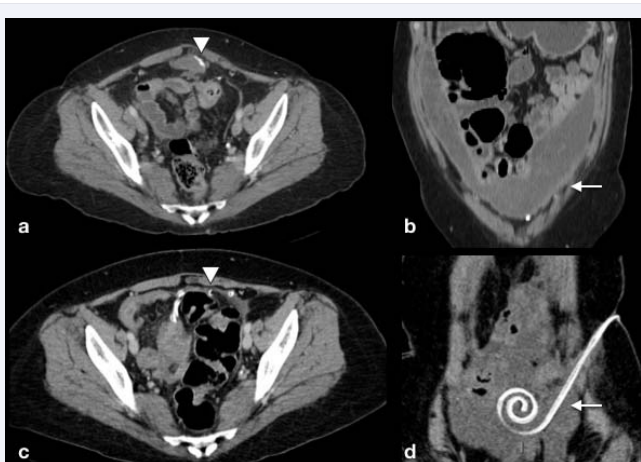


Figure 3 Axial (a and c) and coronal reformatted (b and d) abdominal CT scans. Current CT scan (a and b) show calcification (arrowhead) and peritoneal thickening (arrow) and loculated ascites. A three-year prior CT scan (c and d), shows the peritoneal catheter course which correspond to the sites of thickening (arrow) calcification (arrowhead) shown in the current abdominal CT.

episodes of intestinal obstruction with or without the existence of positive inflammation markers and the presence of peritoneal thickening, sclerosis, calcifications, and encapsulation confirmed by macroscopic inspection or radiologic findings [5,6]. The incidence rate varies from 0.7 to 13.6 per 1000 patient-years [2,7,8], and an increase in this rate has been documented [9,10]. The mortality rate ranges from 17 to 55% and is directly related to the duration of PD [7,8,11]. The major causes of mortality are related to bowel obstruction and complications of the associated surgical procedures such as malnutrition and septicemia, although lower rates have also been reported after surgical intervention [9,12].

It is believed that EPS is caused by a progressive damage to the peritoneum secondary to prolonged dialysis therapy, and a two-hit theory is now accepted. The first hit can trigger a fibrotic process, and a second hit (e.g. peritonitis and acute cessation of PD as in this case) triggers the development of EPS [6]. The high glucose concentrations and low pH of the dialysate fluid facilitates osmosis and diffusion to prevent glucose degradation products generated by heat sterilization of the peritoneal fluid. Multiple factors that predispose to the development of this complication have been identified and these include the number of episodes of peritonitis, duration and age of initiation of PD, use of hypertonic dextrose or icodextrin solutions, ultrafiltration failure associated with membrane fibrosis, cessation of PD [13] and the use of calcineurin inhibitors in renal transplant patients, which increase the expression of transforming growth factor β (TGF- β) which in turn induces peritoneal fibrosis [14-17].

EPS has an insidious onset. In the early stages, patients present with intermittent episodes of hyporexia, nausea, and vomiting. These non-specific manifestations explain why early diagnosis is so challenging. In later stages, constipation, an abdominal mass and abdominal pain (with or without intestinal occlusion) may appear [18]; the development of a fibrous "cocoon" can lead to malnutrition, weight loss, small-bowel obstruction and ischemia [6]. Other manifestations have been reported, such as ultrafiltration failure, diarrhea and peritonitis [12]. Elevated concentrations of inflammatory markers (C-reactive protein), hypoproteinemia, and hemorrhagic ascites can be also found in 7%-50% of cases [18].

In most patients, as in this case, there is a history of peritonitis which occurs 3.3 times more frequently as compared to patients without EPS [9,12]. However, EPS can occur without a clinical history of peritonitis [3,8,14,15].

Plain abdominal films are usually performed initially in the setting of suspected small-bowel occlusion, and they can show bowel dilatation, air-fluid levels, and calcifications in advance stages [18]. Abdominal ultrasound can detect loculated ascites with intraperitoneal echogenic strands and dilated loops [3], however, there is no evidence of its sensitivity and specificity to detect peritoneal thickening, adhesions, or dilated bowel in EPS [19].

Computed tomography is the most effective method to evaluate the peritoneum, and has a very good inter-rater agreement to diagnose EPS [20]. The main imaging findings are secondary to abnormalities of the peritoneum: peritoneal

thickening, peritoneal calcifications, small bowel manifestations: tethering of the small bowel, bowel wall thickening and loculated fluid collections [12,20,21]. Peritoneal thickening can be smooth, nodular or irregular and show enhancement after the administration of IV contrast media. Calcifications are present in more than half of the patients [12], they can manifest as fine linear, focal or as an extensive conglomerate and increase in size progressively involving the visceral and parietal peritoneum. Adherences and fibrosis generated from the thickened peritoneum displace the loops centrally, retracting the mesentery, this appearance is also known as “the gingerbread man” sign [22]. Small-bowel occlusion produces dilatation and air-fluid levels, this appearance has been described as a “cocoon” [8,13,19,21-24]. The severity of CT abnormalities correlate with clinical outcome [23].

Regarding abdominal CT, a retrospective 9.5 yr analysis showed that bowel tethering (in-drawing of the bowel to the center of the abdomen) and peritoneal calcifications are the most specific parameters to diagnose EPS, but they have low sensitivity, being abnormal only in 33% of the cases. On the other hand, loculation of ascites is the least reliable parameter. Furthermore, in the same study, almost 70% of the patients had a normal CT scan at least 4 months prior to the diagnosis of EPS, evidencing the rapid progression of the disease. Finally, the diagnostic performance of CT in the setting of asymptomatic patients appears to be lower [18]. Together, these data discourage using abdominal CT as a screening strategy [18,20,24].

Finally, in cases where the diagnosis is not definitive with a CT scan, surgery may be needed, which has the advantages of confirming the diagnosis and treating potential complications of this condition [6].

Another modality that has been used is dynamic cinematographic magnetic enteroresonance (cine-MR) which can demonstrate impaired peristalsis secondary to adherence or bowel tethering, with some reports claiming that cine-MRI can have similar or even better results than CT [25]. However, the efficacy of this modality is not well established [6,26].

It is important to differentiate this entity from other causes of peritoneal thickening, such as peritoneal carcinomatosis [4], extrapelvic endometriosis and causes leading to peritoneal calcification such as pseudomyxoma peritonei, tuberculosis, peritoneal mesothelioma and calcified peritoneal carcinomatosis [21,27]. Sheet like calcifications are more common in patients with benign causes (secondary to peritonitis or post-surgical) on the contrary, nodular calcifications are associated with a malignant origin (more commonly ovarian carcinomatosis) [28].

A few remarks on medical management include:

a) Discontinuation of peritoneal dialysis, which is crucial and should be done as soon as the diagnosis is made [11,19,29]. This strategy is controversial and contrasts with the second-hit theory which recognizes cessation of PD as a risk factor.

b) *Nil per os* management and total parenteral nutrition alone have not demonstrated to be effective. The latter has only been proposed as an adjuvant therapy in patients with malnutrition [11].

c) Medical treatment with corticosteroids is useful at early stages. However, only 38.5% of patients achieve symptomatic improvement [30].

d) Tamoxifen can prevent peritoneal deterioration because it stimulates metalloproteinase-9 to remove degenerated collagen, thereby preventing damage to mesothelial cells but its use is still controversial, because the observational studies that have been published have a limited number of patients. [11,29,31].

Other therapies have been used such as inhibitors of mammalian target of rapamycin (mTOR) [32].

Surgical management is reserved only for cases of high-grade small-bowel obstruction because the mortality has been mainly associated with surgical complications [8,18,29,31,33]. A Japanese study showed a high success rate with enterolysis and reported lower mortality rates compared with other studies (35.4% vs 52-55%) [11]. This procedure (sometimes combined with the Noble plication procedure) does not reverse peritoneal affection but can prevent recurrence of obstruction [11].

CONCLUSION

The insidious and unspecific clinical manifestations of EPS make the diagnosis challenging. This, combined with its rapid progressive nature and its high morbidity and mortality rate, indicates that we must increase awareness of this uncommon complication of PD. It is important to recognize typical radiological findings of this complication, especially on CT scans. This will allow early diagnosis and management, with the potential to improve patients' outcomes.

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