

## CASE REPORT

# ENCAPSULATING PERITONEAL SCLEROSIS SECONDARY TO PERITONEAL DIALYSIS

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### Abstract

*Encapsulating peritoneal sclerosis (EPS) is a rare and morbid complication of peritoneal dialysis, characterized by peritoneal thickening, fibrosis, and encasement of the bowel. Here, we report a 37-year-old man who presented with encapsulating peritoneal sclerosis complicated by a significantly large, loculated fluid collection several months after discontinuation of peritoneal dialysis. The case, overall, highlights a lesser-known complication of peritoneal dialysis and provides clinical pearls for clinicians to optimize patient care.*

**Keywords:** Encapsulating peritoneal sclerosis, EPS, Peritoneal dialysis, Peritoneal fibrosis, Thickened peritoneum, Bowel cocooning, Bowel encapsulation, Peritoneal calcification

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### Introduction:

Encapsulating peritoneal sclerosis (EPS) is a poorly understood entity that can lead to encasement of the bowel leading to bowel obstruction and malnutrition. It is a life-threatening complication of long-term peritoneal dialysis (PD). Clinicians should remain vigilant to ensure a timely diagnosis, as the rarity of this disease and variable symptoms often contribute to delays in diagnosis. To describe a case of encapsulating peritoneal sclerosis which is a rare but fatal complication of peritoneal dialysis.

### Case report:

A 37-year-old man, with a past medical history of hypertension, asthma, and end-stage renal disease (ESRD) secondary to congenital obstructive uropathy, underwent a living donor renal transplant in 2004. Unfortunately, the transplant failed in 2012, leading to the reinitiation of hemodialysis (HD). He transitioned to peritoneal dialysis (PD) from 2018 to 2023 but returned to HD six months before presentation due to tumoral calcinosis of his left thigh. He presented to

our emergency department (ED) due to two weeks of anorexia, non-bloody diarrhea, non-bloody non-bilious emesis, abdominal distention, and right upper quadrant abdominal pain.

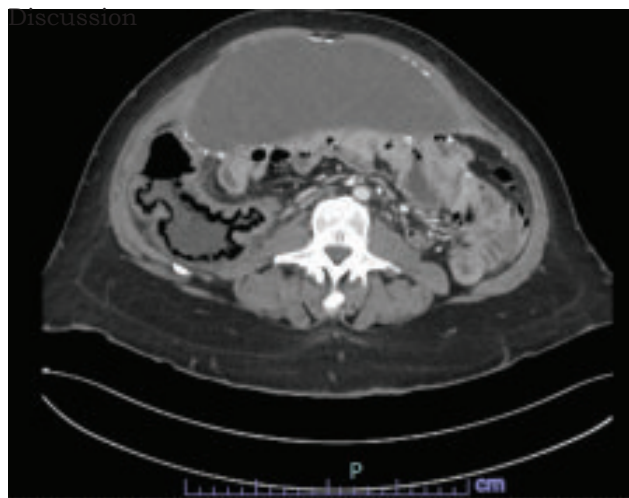
In the ED, the patient's temperature was 37°C, heart rate 110/min, and blood pressure was 88/45. Laboratory test results showed leucocytes  $17 \times 10^9 / L$  ( $4.1-11.1 \times 10^9 / L$ ), lactic acid 2.9 mmol/L (less than 2 mmol/L), creatinine 16 mg/dL (0.7-1.3 mg/dL), calcium 10.0 mg/dL (8.6-10.3 mg/dL), and phosphorus 7.2 mg/dL (3-4.5 mg/dL). A CT scan of the chest, abdomen, and pelvis was significant for calcified peritoneum, a large loculated peritoneal fluid collection with rim calcification, mild colonic wall thickening/fat stranding with pseudo pneumatosis (air surrounding fecal content), no evidence of acute bowel obstruction (Figures 1-3). A CT scan of the chest, abdomen, and pelvis 1 year ago revealed mild peritoneal calcification. Given symptoms, abdominal findings, and imaging, encapsulating peritoneal sclerosis (EPS) was higher in our differential.

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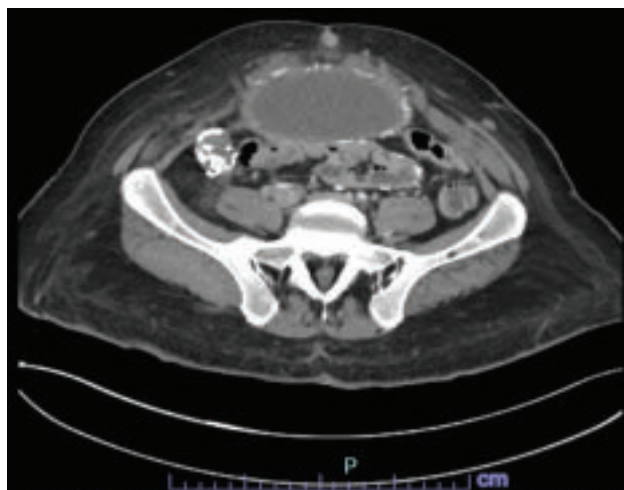
A paracentesis was performed draining approximately 1 Liter of serosanguinous fluid. He was started on empiric antibiotics (piperacillin-tazobactam) for presumed bacterial peritonitis. Hemodialysis was continued during hospitalization. Blood cultures and peritoneal fluid analysis were negative. The patient improved significantly with the resolution of symptoms. He was switched from piperacillin-tazobactam to 2 weeks of oral Augmentin at discharge.



**Fig.-1.** CT abdomen axial cut showing peritoneal calcification, large fluid collection, pseudo-pneumatosis of ascending colon, bowel loops drawn into center.



**Figure 3.** CT abdomen sagittal cut showing peritoneal calcification and large abdominal fluid collection.



**Fig.-2.** CT abdomen axial cut showing peritoneal and bowel wall calcification, large fluid collection, and calcified kidney graft.

Our patient developed encapsulating peritoneal sclerosis, a rare complication of long-term peritoneal dialysis.

#### Discussion:

Encapsulating peritoneal sclerosis (EPS) is a clinical syndrome within the spectrum of peritoneal fibrosis. It is characterized by the encasement of the bowel due to a diffusely thickened peritoneum and is associated with high morbidity and mortality (1). Risk factors for EPS include younger age at PD onset, increased duration of PD, prior episodes of peritonitis, and dialysate makeup (e.g. increased glucose concentration) (2). This patient's risk factors include younger age at PD onset, six years duration of PD, and previous peritonitis episodes. The pathogenesis of EPS is not well understood but inflammation leading to fibrosis and calcification is considered multifactorial because of prolonged contact between dialysate and peritoneum (3,4). Laparoscopic findings suggest that peritoneal degeneration is associated with the duration of peritoneal dialysis and frequent episodes of peritonitis. EPS is slowly progressive and often asymptomatic early in the disease course. It commonly presents after cessation of peritoneal dialysis and the time frame can vary (5). Our patient had PD six months before this presentation. Clinicians may notice serosanguinous dialysate or decreased ultrafiltrate if the patient still receives peritoneal dialysis. As EPS progresses to later stages, the disease can present with signs and symptoms of obstruction including abdominal pain, constipation, vomiting, palpable abdominal mass, or ascites. Diagnosis remains challenging due to diagnostic criteria that rely largely on clinical symptoms and variable radiographic

findings. CT scan of the abdomen is the most common modality used for imaging in EPS currently. Radiographic findings include peritoneal calcification, bowel and peritoneal thickening, bowel dilatation, loculated ascites, and bowel cocooning (bowel loops clustered in the central abdomen with tethering) (6).

EPS is a life-threatening complication of long-term PD. Prevention efforts have been primarily aimed at reducing glucose and glucose degradation product concentrations in dialysis fluid using icodextrin and neutral pH solutions (7,8). Length of peritonitis episodes have also been associated with EPS (9). Early recognition and treatment have been reported to improve outcomes. Nutritional status should be regularly assessed and optimized in all patients. Effective treatment with glucocorticoids, tamoxifen, or other immunosuppressive agents has been described, but no randomized controlled trials have been performed due to disease rarity (10,11). Tamoxifen is the most widely used anti-fibrotic treatment due to inhibition of TGF  $\alpha 1$ , which is thought to halt progression and lead to collagen degradation in EPS. Laparotomy is only recommended for surgical enterolysis in cases of complete bowel obstruction at presentation or cases of medical management failure.

In summary, we describe a rare case of encapsulating peritoneal sclerosis (EPS) following peritoneal dialysis. A high index of suspicion is warranted in patients experiencing symptoms such as abdominal pain, constipation, vomiting, palpable abdominal mass, or ascites, given the associated high morbidity and mortality.

### Conclusion:

Clinicians should be vigilant for signs of encapsulating peritoneal sclerosis (EPS) in long-term peritoneal dialysis patients. Recognizing prolonged dialysis, recurrent peritonitis, and high-glucose dialysate as risk factors is crucial for early detection. Management of EPS involves medical treatments like glucocorticoids and tamoxifen, with surgery for severe cases.

### Conflict of Interest:

The authors stated that there is no conflict of interest in this study

### Funding:

This research received no external funding.

### Consent:

For the purpose of publishing this case report and any related photos, the parents are written informed consent was acquired.

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