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Case Report

Calcified encapsulating peritoneal sclerosis associated with peritoneal dialysis: A case report☆,☆☆,★

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ABSTRACT

Encapsulating peritoneal sclerosis (EPS) is a rare, but sometimes fatal, complication of peritoneal dialysis characterized by diffuse thickening and encapsulation of the bowel and peritoneum. In more advanced cases, the peritoneum will gradually calcify. EPS usually presents as partial small bowel obstruction and diagnosed on imaging studies. We present a case of a 19-year-old female on long-term peritoneal dialysis with EPS and diffuse peritoneal calcifications.

Introduction

Encapsulating peritoneal sclerosis (EPS) is an uncommon, however often fatal, condition characterized by diffuse thickening and encapsulation of the bowel and peritoneum [1]. While idiopathic cases have been described, secondary EPS has been associated with peritoneal dialysis or abdominal tuberculosis, among other etiologies resulting in peritoneal inflammation [2].

Case report

We present a case of a 19-year-old female with complex medical history presenting to the emergency department with abdominal pain and emesis. The patient has Turner syndrome and a remote history of hypoplastic left heart syndrome, for which she underwent heart transplantation and was placed on calcineurin inhibitor. Her treatment course was complicated by immunosuppressant nephrotoxicity resulting in

Abbreviations: EPS, Encapsulating peritoneal sclerosis.

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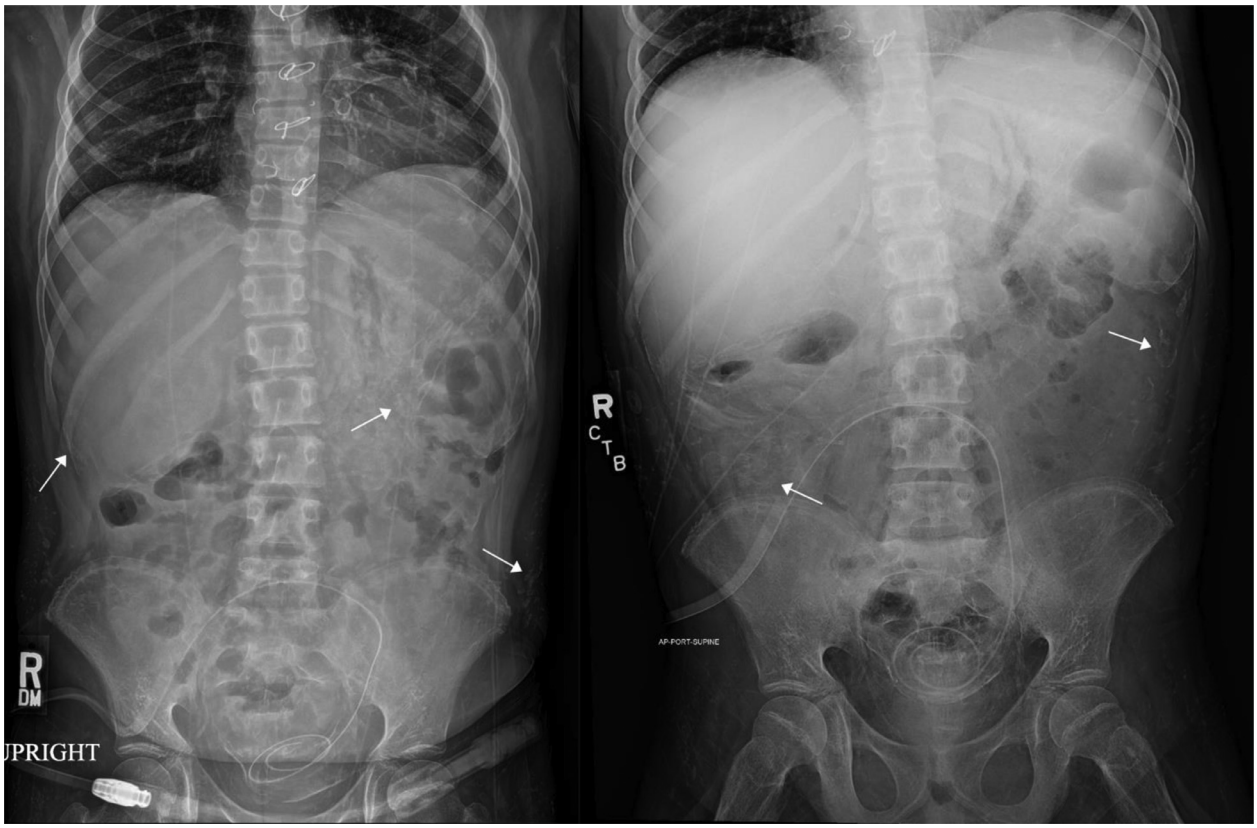

Fig. 1 – Abdominal radiographs demonstrate a nonobstructive bowel gas pattern with diffuse curvilinear hyperdensities in the abdomen and pelvis compatible with calcifications (arrows). A peritoneal dialysis catheter is seen coiling in the lower abdomen.

end-stage renal disease diagnosed at the age of 9 years. She started hemodialysis at 12 years old and was converted to peritoneal dialysis the following year.

Over the several few months prior to presentation, the patient had similar recurring issues with constipation, feculent emesis, and bloody peritoneal effluent, for which she previously received empiric antibiotic and antifungal therapy. The patient reported intermittent hypotension, but denied fever. She was admitted for further evaluation and treatment.

Physical examination was notable for abdominal tenderness predominantly in the epigastric region and right abdomen without rebound tenderness or guarding. There was no palpable abdominal mass, and her PD catheter site appeared clean without signs of inflammation or infection.

Relevant labs demonstrated leukocytosis, which was attributed to her weekly Neupogen injections. Blood and peritoneal fluid cultures did not demonstrate any growth. Peritoneal fluid was notable for both significant red blood cells and a few white blood cells.

She underwent upper and lower endoscopy which demonstrated nodularity and ulceration at the gastric antrum, external hemorrhoids, and patchy erythema within the colon.

Abdominal radiographs demonstrated a nonobstructive bowel gas pattern with diffuse curvilinear hyperdensities in the abdomen and pelvis concerning for calcifications (Fig. 1). No free air was identified. The peritoneal dialysis catheter was seen coiling in the lower abdomen.

Abdominal ultrasound (US) was performed which showed scattered curvilinear hyperechoic areas with acoustic shadowing compatible with calcifications, for example along the posterior hepatic margin and overlying several bowel loops (Fig. 2).

Computed tomography (CT) was subsequently obtained and confirmed the presence of significant visceral and parietal peritoneal calcifications (Fig. 3). There was diffuse bowel wall thickening with regions of clustered small bowel loops. The kidneys appeared multicystic with bilateral medullary nephrocalcinosis. Small volume peritoneal fluid with peritoneal dialysis catheter tubing in the lower abdominal were noted.

Comparison CT images from 2.5 years prior to presentation did not show any significant bowel wall thickening or peritoneal calcifications (Fig. 4).

A diagnosis of EPS was made based off the imaging findings. The patient was started on conservative therapy with systemic steroids and tamoxifen. She was initially kept on total parenteral nutrition with efforts to transition to PO diet. Peritoneal dialysis was terminated, and she was converted to hemodialysis via subclavian venous access.

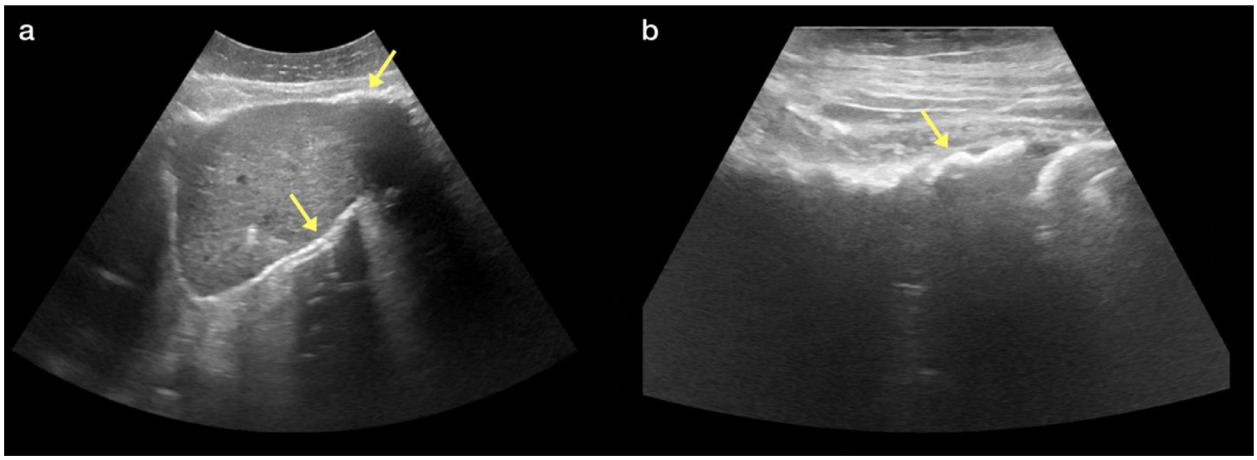

Fig. 2 – Abdominal ultrasound (US) shows scattered curvilinear hyperechoic areas with acoustic shadowing (arrows), for example along the posterior hepatic margin (a) and overlying several bowel loops (b).

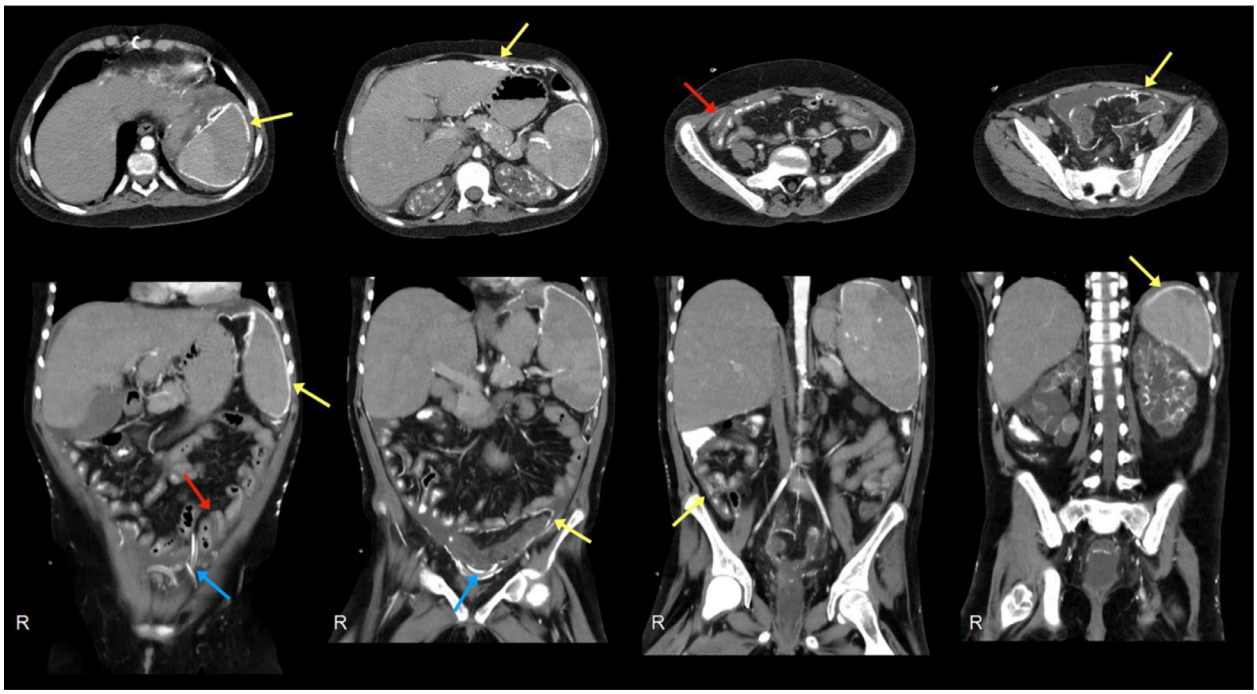

Fig. 3 – Axial and coronal computed tomography (CT) images of the abdomen and pelvis reveal scattered areas of visceral and parietal peritoneal calcification (yellow arrows). There is diffuse bowel wall thickening with regions of clustered small bowel loops (red arrows). There is no evidence to suggest acute bowel obstruction. The kidneys appear multicystic with bilateral medullary nephrocalcinosis. Small volume peritoneal fluid with peritoneal dialysis catheter tubing in the lower abdomen are noted (blue arrows).

Discussion

EPS is a rare abdominal condition, first described as "peritonitis chronica fibrosa incapsulata" by Owtschinnikow in 1907 [3] and later coined "abdominal cocoon" by Foo et al. in 1978 [4]. As its name implies, it is characterized by diffuse fibrocollagenous thickening of the peritoneum and encasement of the bowel and presents with intermittent episodes of nonspecific abdominal symptoms of small bowel obstruction such as diffuse pain and vomiting. Primary idiopathic EPS was initially described in young females from subtropical and tropical Asian countries [5]; however, it may occur in all age groups [1,6].

Its pathogenesis is not fully understood; however, current hypotheses propose that EPS develops secondary to peritoneal irritation. The most common cause of EPS is chronic peritoneal dialysis [7,8], followed by bacterial peritonitis including tubercular peritonitis, abdominal malignancies, and abdominal surgery, among others [2]. The overall prevalence of EPS in

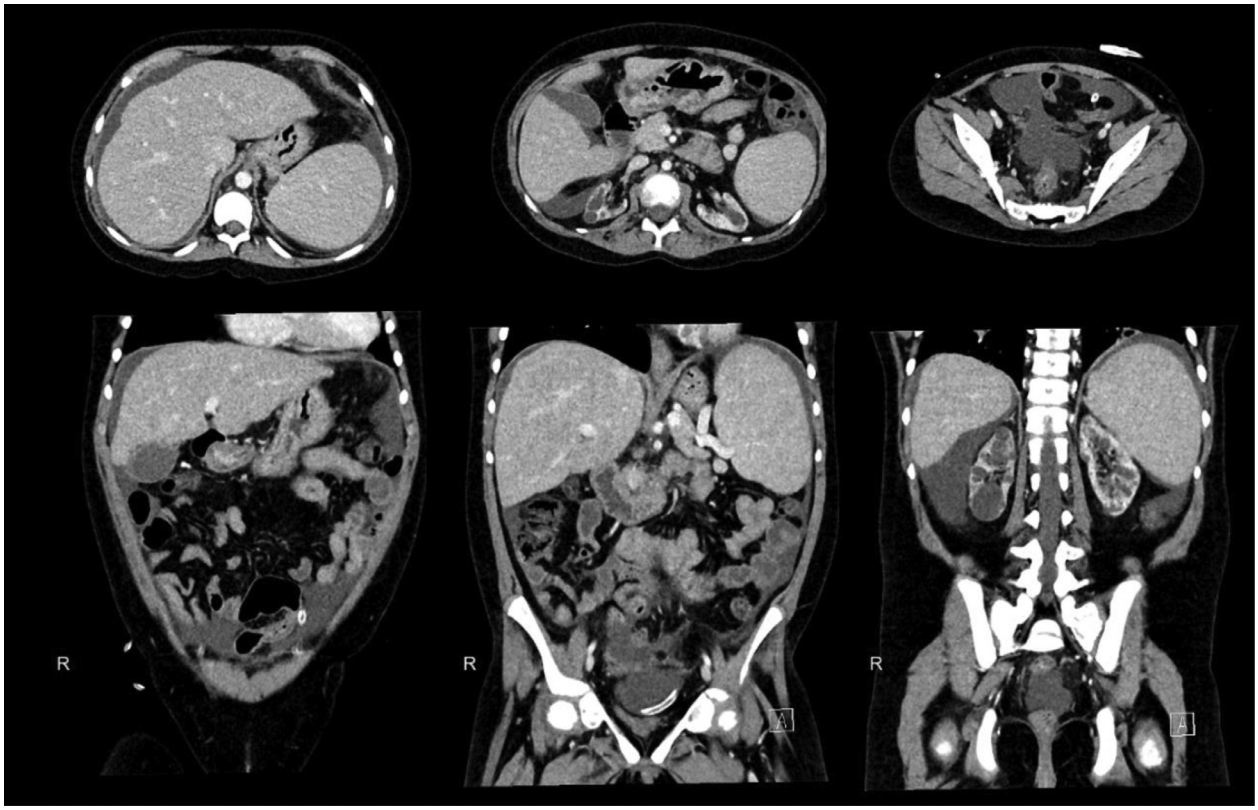

Fig. 4 – Axial and coronal computed tomography (CT) images of the abdomen and pelvis 2.5 years prior to presentation show normal spread-out bowel loops without bowel wall thickening or peritoneal calcifications. Small volume peritoneal fluid with peritoneal dialysis catheter tubing in the lower abdomen are again noted.

patients on chronic peritoneal dialysis is approximately 0.7% [9], with high mortality rate of on average 35% [2,10].

Imaging is an important part in the diagnostic evaluation of symptomatic EPS. Abdominal radiography is normal in most cases of EPS, however it may show diffuse peritoneal calcifications in more advanced cases [2,8]. Small bowel loops in EPS are often partially obstructed, but may not appear significantly dilated on radiographs. Small bowel follow-through studies may show a fixed cluster of mildly dilated bowel loops in a U-shaped configuration [2].

Ultrasound evaluation of the abdomen may show clustered bowel loops in a cauliflower-like configuration, hyperechoic wall thickening with a trilaminar appearance, shadowing areas of peritoneal calcifications, and ascites [2].

CT is the most useful modality in the diagnosis of EPS and can also identify complications of EPS such as bowel necrosis or perforation. The visceral and parietal peritoneum are usually involved; the thickened and enhancing peritoneum envelops and traps multiple small bowel loops into a cluster with surrounding “cocoon”, which can calcify over time [8]. Ascites is often seen, especially in patients on peritoneal dialysis. The bowel wall may also thicken and fibrose, resulting in luminal narrowing and proximal bowel obstruction without transition point [8]. Tethering and kinking of the bowel loops from adhesions indicate a poor prognosis [2]. Magnetic resonance imaging would show similar findings to that seen on CT.

Management of EPS includes conservative or surgical treatments. Conservative management options include transition from peritoneal dialysis to hemodialysis, total parenteral nutrition, renin-angiotensin-aldosterone system inhibitor therapy, immunosuppressant therapy, or tamoxifen, which may reverse findings of EPS [2]. Surgical management includes dissection of the peritoneal cocoon and adhesiolysis, during which the peritoneum will appear leathery and tan colored [2]. Small bowel resection should be avoided due to risk of fistulization [1].

EPS remains a serious complication of peritoneal dialysis with high mortality rate. Early diagnosis and treatment have been shown to provide better patient outcomes, and therefore should be considered in differential diagnoses in at-risk patients, not to be mistaken for congenital peritoneal encapsulation or internal herniation.

Patient consent

Appropriate patient consent has been obtained for this case study.

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