Time Is Not Always the Matter: An Instance of Encapsulating Peritoneal Sclerosis Developing in a Patient on Peritoneal Dialysis for a Short Term

Radhames Ramos De Oleo,[1] Hugo Villanueva,[2] Lin Lwin,[2] Madhavi Katikaneni,[2] Jinil Yoo[2]

Encapsulating peritoneal sclerosis (EPS) is an infrequent but serious complication that is observed mostly in patients on long-term peritoneal dialysis (PD). However it can occur after short-term PD, in association with "second hit" risk factors such as peritonitis, acute cessation of PD, or kidney transplantation with the use of calcineurin inhibitors.

In our case, a young woman with second-hit risk factors presented with clinical and abdominal computed tomography findings consistent with EPS after short-term PD. She was treated conservatively with nutritional support and was discharged in improved and stable clinical status.

In general, the diagnosis of EPS requires clinical findings of bowel obstruction combined with typical computed tomography imaging features. However, the clinical manifestations can be very vague, and the diagnosis is often unclear. A recent study categorized EPS into 4 clinical stages, from pre-EPS to chronic ileus, with associated management from conservative treatment to surgical intervention.

In association with second-hit risk factors, EPS can occur after short-term PD. Severity is variable, and the outcome is often devastating. Timely recognition and expert management of EPS can change the outcome very favorably.

Key words
Encapsulating peritoneal sclerosis, peritonitis

From: [1]New York Medical College, Metropolitan Hospital Center, New York, and [2]Montefiore Medical Center, Wakefield Campus, Bronx, New York, U.S.A.

Introduction

Encapsulating peritoneal sclerosis (EPS) is an infrequent but a serious complication that arises primarily in patients on peritoneal dialysis (PD). Its strongest risk factor is the duration of PD, with the risk becoming significant after 5 years, and even more so after 10 years (1). Other risk factors include onset of PD at a young age, sudden cessation of PD, episodes of peritonitis, and kidney transplantation after PD (2). Very few cases of EPS have been observed after a short period of PD (3). Our case highlights one of the rare instances in which EPS develops after a short period of PD complicated by episodes of peritonitis, transition to hemodialysis (HD), and then to kidney transplantation.

Case presentation

A 24-year-old African American woman presented to the hospital complaining of abdominal discomfort for 3 months, followed by 3 days of constipation and poor oral intake. At age 7, she had undergone left nephrectomy for a primitive neuroendocrine tumor (malignant rhabdoid), with subsequent chemotherapy. At age 14, she was initiated on PD for end-stage kidney disease, which was twice complicated by peritonitis; after 9 months of PD, she was switched to HD. One year later, she received a cadaveric donor kidney graft that was rejected after 6 years. She then restarted HD.

The patient was feeling well on HD until 3 months before the current admission, when she noted abdominal discomfort, bloating, and episodic diarrhea or constipation. She denied fever or chills.

On physical examination at admission, her blood pressure was 96/67 mmHg; pulse, 110 and regular;

temperature, 97.8°F; respiration, 18 per minute, with no distress; weight, 55 kg; and height, 157 cm. Other findings included clear lungs; regular heart sounds, with a transmitted murmur from her arteriovenous fistula (HD access); a mildly distended abdomen, with positive fluid wave on percussion and slightly hypoactive bowel sounds; no leg edema; and a well-functioning arteriovenous fistula in her right arm.

Initial serum chemistry showed sodium 141 mEq/L, potassium 3.4 mEq/L, chloride 99 mEq/L, HCO_3 19 mEq/L, blood urea nitrogen 54 mg/dL, creatinine 8.9 mg/dL, glucose 101 mg/dL, calcium 9.1 mg/dL, and albumin 3.5 g/dL (total protein 5.6 g/dL); a complete blood count showed hemoglobin 12.4 g/dL, white blood cells $9.4\times10^3/\mu L$, platelets $194\times10^3/\mu L$, serum amylase and lipase within normal limits, and erythrocyte sedimentation rate 5 (normal: 0 – 20).

Computed tomography (CT) imaging of abdomen and pelvis disclosed a moderate amount of low-density fluid, mostly surrounding the spleen and stomach, with loculated areas adherent to the descending colon and left paracolic gutter. Nodularity of the peritoneum in the left quadrant and near the transverse colon was also evident, and several bowel loops (the transverse colon and small-bowel loops) demonstrated focal wall thickening; however, no significant retroperitoneal lymphadenopathy was observed (Figure 1).

The patient refused to undergo CT-guided paracentesis, and the work-up for malignancy was negative, except for a slightly elevated cancer antigen 125. She was treated conservatively, without any interventional procedures. Her oral intake and nutrition status improved. She was discharged in stable condition and has since been lost to follow-up.

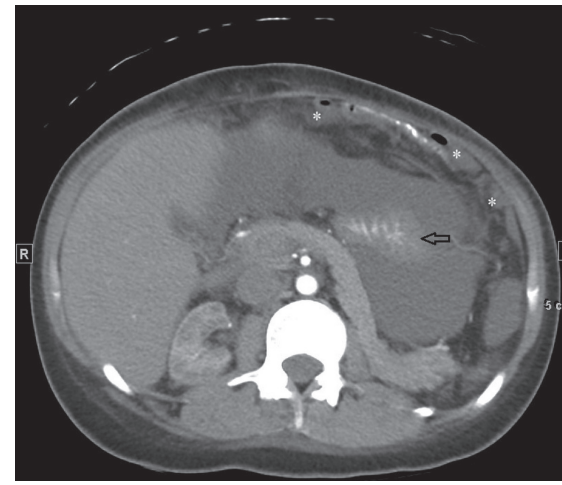

FIGURE 1 Computed tomography image of abdomen with oral and intravenous contrast. Asterisks mark thickened peritoneal membrane with nodularities. Thickened bowel wall is surrounded by encapsulated ascitic fluid (arrow).

Discussion

A rare clinical entity, EPS occurs primarily in patients on a long-term PD therapy; however, it can, for various reasons, develop in patients not on PD (4) and in patients on short-term PD (3). The strongest risk factor for EPS is duration of PD therapy; other risk factors include episodes of peritonitis, acute cessation of PD upon transition to HD, kidney transplantation (with the use of calcineurin inhibitors), onset of PD at a young age, or an underlying genetic predisposition (4).

A proposed pathogenetic mechanism for EPS is an initial injury to the peritoneal membrane by PD-related factors, resulting in peritoneal inflammation and a simple sclerosis process (the "first hit"), followed by second-hit factors such as acute cessation of PD, transplantation, peritonitis episodes, or genetic predisposition triggering a fibrotic and encapsulating process of the peritoneal membrane (5). Acute cessation of PD because of transition to HD or transplantation might be prevented with continuous lavage of the peritoneal cavity to remove accumulated EPS-promoting factors (6). An increased incidence of EPS after kidney transplantation has been reported. The suggested reason is the profibrotic effect of immunosuppressive agents (calcineurin inhibitors) or the acute cessation of PD, or both (7).

In general, a diagnosis of EPS requires both clinical signs and symptoms of bowel obstruction, combined with either typical features on CT imaging or confirmation of fibrous cocooning at laparotomy (8). However, clinical manifestations can be very vague, and the diagnosis is often not clear. Based on clinical presentation, one study (9) categorized EPS into 4 clinical stages:

- Stage1: Pre-EPS (asymptomatic with mild ascites and no inflammation)
- Stage 2: Inflammatory stage (symptomatic with partial encapsulation of bowel and intestinal wall swelling)

- Stage 3: Encapsulation (symptoms of bowel obstruction)
- Stage 4: Chronic ileus (absolute bowel obstruction, with encapsulating fibrous cocoon)

Imaging by CT is helpful in diagnosing the encapsulating stage, where cocooning of the bowel is seen; however, thickening of the peritoneal membrane is, by itself, not diagnostic of EPS because such thickening is seen in any long-term PD patient, and regular CT surveillance of long-term patients has not been shown to be helpful (1).

Because most patients with EPS show malnutrition, appropriate nutritional support is necessary (4), and patients in the inflammatory phase of EPS (early stage) can be treated with moderate doses of corticosteroids if infectious causes are ruled out (10). Tamoxifen, which has antifibrotic properties, has also been used for treatment (11). In patients with an established abdominal cocooning and recurrent bowel obstruction, a surgical approach is required, especially by an experienced surgical team for a successful outcome (1).

Our patient's clinical presentation without bowel obstruction and with CT findings of partial encapsulation of the bowel, intestinal swelling, and loculated ascites all pointed to inflammatory-stage (early-stage) EPS, and she was treated conservatively with nutritional support, reaching an improved and stabilized state. However, she refused further evaluation and was lost to follow-up management.

Conclusions

Encapsulating peritoneal sclerosis is observed mostly in patients after long-term PD therapy; however, it can be encountered after short-term PD in association with second-hit risk factors. Severity is variable, but outcomes are often devastating. Timely recognition of EPS and expert management can lead to a more favorable outcome.

Disclosures

The authors have no conflicts of interest to disclose.

References

1 Bargman JM. Hernias, leaks, and encapsulating peritoneal sclerosis. In: Daugirdas JT, Blake PG, Ing TS, eds. Handbook of dialysis. 5th ed. Boston, MA: Wolters Kluwer Publisher; 2015: 519–20.
2 Korte MR, Sampimon DE, Lingsma HF, *et al.* Risk factors associated with encapsulating peritoneal sclerosis in Dutch EPS study. Perit Dial Int 2011:31:269–78.
3 Mutoh S, Machida J, Ueda S, Kitamoto Y, Uemura S. Sclerosing encapsulating peritonitis occurring after very short-term intermittent peritoneal dialysis. Nephron 1992;62:119–20.
4 Moinddin Z, Summers A, Van Dellen D, Augustine T, Herrick SE. Encapsulating peritoneal sclerosis—a rare but devastating peritoneal disease. Front Physiol 2015;5:470.
5 Brown MC, Simpson K, Kerssens JJ, Mactier RA on behalf of the Scottish Renal Registry. Encapsulating peritoneal sclerosis in the new millennium: a national cohort study. Clin J Am Soc Nephrol 2009;4:1222–9.
6 Yamamoto T, Nagasue K, Okuno S, Yamakawa T. The role of peritoneal lavage and the prognostic significance of mesothelial cell area in preventing encapsulating peritoneal sclerosis. Perit Dial Int 2010;30:343–52.
7 Korte MR, Habib SM, Lingsma H, Weimar W, Betjes MG. Posttransplantation encapsulating peritoneal sclerosis contributes significantly to mortality after kidney transplantation. Am J Transplant 2011;11:599–605.
8 Davis SJ, Wilkie M. Complications of peritoneal dialysis. In: Johnson RJ, Feehally J, Floege J. Comprehensive clinical nephrology. 5th ed. Amsterdam, Netherlands: Elsevier; 2015: 1114.
9 Nakamoto H. Encapsulating peritoneal sclerosis—a clinician's approach to diagnosis and medical treatment. Perit Dial Int 2005;25(suppl 4):S30–8.
10 Kawanishi H. Surgical and medical treatments of encapsulating peritoneal sclerosis. Contrib Nephrol 2012;177:38–47.
11 Cornelis P, Oreopoulos DG. Update on potential medical treatments for encapsulating peritoneal sclerosis; human and experimental data. Int Urol Nephrol 2011;43:147–56.

Corresponding author:
Jinil Yoo, MD, Renal Unit, Montefiore Medical Center–Wakefield Campus, 4141 Carpenter Avenue, Bronx, New York 10466 U.S.A.
E-mail:
jiyoo@montefiore.org