Cocoon abdomen after chronic peritoneal dialysis

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ABSTRACT

A 66-year-old man presented with nausea, vomiting, and ascites. At laparotomy, an extensive fibrosis of the peritoneum was found to encase the bowel in a cocoon-like fashion. A meticulous dissection was undertaken to strip this encapsulating membrane from the bowel. Histologic study of the excised tissue disclosed extensive hyalinized fibroconnective tissue.

KEYWORDS Ascites; bowel obstruction; encapsulating peritoneal sclerosis; fibrosis; hyalinized fibroconnective tissue; peritoneal dialysis

Encapsulating peritoneal sclerosis (EPS) is a rare complication of peritoneal dialysis (PD) characterized by progressive fibrosis of the peritoneum with subsequent encapsulation of the viscera and resulting partial bowel obstruction.¹ This case highlights the clinical presentation, pathologic findings, and complicated surgical management of this disease.

CASE REPORT

A 66-year-old Vietnamese man with a history of end-stage renal disease presented to his nephrologist with chronic nausea, vomiting, weight loss, and pedal edema that began 3 months after his second kidney transplant. He had previously been on PD for 12 years without complication. His PD catheter had been removed 2 months prior to presentation.

In addition to his primary complaints, his initial evaluation revealed new-onset ascites in the setting of normal hepatic and renal function and normal electrolytes. He underwent an upper endoscopy, colonoscopy, liver biopsy, hepatic venogram with portal vein pressures, cardiac catheterization, positron emission tomography scan, and paracenteses—all of which were nondiagnostic. A small bowel series and abdominal computed tomography (CT) scan were concerning for a partial small bowel obstruction with omental and peritoneal thickening combined with loculated ascites (*Figure 1a*).

The patient was referred for surgical evaluation regarding the partial small bowel obstruction and ascites of unknown

origin. A diagnosis of encapsulating peritoneal sclerosis was made based on the classic CT findings and history of PD. He was offered an exploratory laparotomy with lysis of adhesions and stripping of the encapsulating membrane. Total parental nutrition was initiated prior to surgery due to feeding intolerance, posttransplant immunosuppressant therapy, and severe malnutrition.

At laparotomy, a thick capsule was found to encase the bowel in a cocoon-like fashion. In a 9-hour operation this was dissected from the peritoneum and bowel, which had the appearance of tanned leather (*Figures 1 and 2*). The distal small bowel was collapsed and portions of the mid- and proximal bowel were dilated without any specific point of obstruction. Pathologic examination demonstrated extensive hyalinized fibroconnective tissue with areas of hemosiderosis. At discharge, the patient was tolerating oral intake without total parental nutrition and had normal bowel function.

DISCUSSION

EPS is a rare and frequently fatal complication of PD, with mortality rates as high as 50%.^{2–4} It was initially reported in 1980 in several patients with thickening of the peritoneal membrane months to years after the initiation of PD.⁵ Since that time, more than 200 scientific articles have been published describing the disease process.⁶ There is limited consensus, however, on its pathologic origin or treatment, in part due to the rarity of the condition, with a reported incidence of about 2% among patients undergoing PD.⁶ Most reports come from Japan where PD is used more

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Figure 1. (a) CT scan showing thickened peritoneum and calcifications. (b) Entrance to the abdomen with a cocoon-like membrane. (c) Loops of bowel visible underneath the encapsulating membrane. (d) Stripping of the encapsulating membrane. Note the tanned leather appearance of the viscera.

Figure 2. (a) Bowel free from encapsulating sclerosis. (b) Representative samples of specimen sent to pathology.

commonly than in the USA. A large multicenter study of EPS in Japan identified 6923 patients undergoing PD, with only 62 (0.009%) documented EPS cases between 1980 and 1994.⁷

Risk factors for the development of EPS include recurrent episodes of peritonitis, the use of acetate-based dialysis

solutions, the use of chlorhexidine antiseptics, as well as long-term PD therapy.⁸ There is little consensus regarding the contributions of these risk factors to the development of the disease. PD duration appears to be the strongest risk factor for the development of EPS.⁹

Management recommendations include tamoxifen, corticosteroids, immunosuppressant therapy, discontinuation of PD, and surgical stripping.^{10,11} The recent experience of Kawanishi et al has demonstrated excellent outcomes with a surgical approach.¹²

1. Chin AI, Yeun JY. Encapsulating peritoneal sclerosis: an unpredictable and devastating complication of peritoneal dialysis. *Am J Kidney Dis.* 2006;47(4):697–712. doi:10.1053/j.ajkd.2005.12.049.
2. Brown MC, Simpson K, Kerssens JJ, Mactier RA. Encapsulating peritoneal sclerosis in the new millennium: a national cohort study. *Clin J Am Soc Nephrol.* 2009;4(7):1222–1229. doi:10.2215/CJN.01260209.
3. Korte MR, Boeschoten EW, Betjes MG. The Dutch EPS registry: increasing the knowledge of encapsulating peritoneal sclerosis. *Neth J Med.* 2009;67(8):359–362.
4. Maruyama Y, Nakayama M. Encapsulating peritoneal sclerosis in Japan. *Perit Dial Int.* 2008;28(Suppl 3):S201–S204.
5. Gandhi VC, Humayun HM, Ing TS, et al. Sclerotic thickening of the peritoneal membrane in maintenance peritoneal dialysis patients. *Arch Intern Med.* 1980;140(9):1201–1203. doi:10.1001/archinte.1980.00330200077024.
6. Kawaguchi Y, Tranaeus A. A historical review of encapsulating peritoneal sclerosis. *Perit Dial Int.* 2005;25(Suppl 4):S7–S13.
7. Nomoto Y, Kawaguchi Y, Kubo H, Hirano H, Sakai S, Kurokawa K. Sclerosing encapsulating peritonitis in patients undergoing continuous ambulatory peritoneal dialysis: a report of the Japanese Sclerosing Encapsulating Peritonitis Study Group. *Am J Kidney Dis.* 1996;28(3):420–427. doi:10.1016/S0272-6386(96)90501-6.
8. Yamamoto Y, Otsuka Y, Nakayama M, et al. Risk factors for encapsulating peritoneal sclerosis in patients who have experienced peritoneal dialysis treatment. *Clin Exp Nephrol.* 2005;9(2):148–152. doi:10.1007/s10157-005-0349-8.
9. Johnson DW, Cho Y, Livingston BE, et al. Encapsulating peritoneal sclerosis: incidence, predictors, and outcomes. *Kidney Int.* 2010;77(10):904–912. doi:10.1038/ki.2010.16.
10. Mori Y, Matsuo S, Sutoh H, Toriyama T, Kawahara H, Hotta N. A case of a dialysis patient with sclerosing peritonitis successfully treated with corticosteroid therapy alone. *Am J Kidney Dis.* 1997;30(2):275–278. doi:10.1016/S0272-6386(97)90064-0.
11. Kawaguchi Y, Saito A, Kawanishi H, et al. Recommendations on the management of encapsulating peritoneal sclerosis in Japan, 2005: diagnosis, predictive markers, treatment, and preventive measures. *Perit Dial Int.* 2005;25(Suppl 4):S83–S95.
12. Kawanishi H, Watanabe H, Moriishi M, Tsuchiya S. Successful surgical management of encapsulating peritoneal sclerosis. *Perit Dial Int.* 2005;25(Suppl 4):S39–S47.