CLINICAL IMAGES

Encapsulating peritoneal sclerosis

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A 51-year-old man with acquired immunodeficiency syndrome and end-stage renal disease maintained on peritoneal dialysis (PD) for 8 years and on hemodialysis (HD) for the next 2 years, was admitted to the hospital with a 3-month history of recurrent abdominal pain, nausea, and progressive malnutrition. The physical examination revealed diffuse abdominal tenderness and ileus. Laboratory tests indicated an inflammatory state. Abdominal contrast-enhanced computed tomography showed marked peritoneum and bowel-wall thickening, presence of intestinal obstruction

and cocooning, with significant enlargement of the duodenum and jejunum, loculated ascites, and peritoneal calcifications (FIGURE 1A). Encapsulating peritoneal sclerosis (EPS) was diagnosed and the decision about surgical treatment was made. After 6 weeks of total parenteral nutrition and complete bowel rest, a laparotomy was performed. It revealed thickened, opaque sheath of sclerosed peritoneum (FIGURE 1B) encasing a mass of small-bowel loops tethered together in a kind of fibrous sac (FIGURE 1C). During a 5-hour surgery, a large portion of the fibrotic tissue was

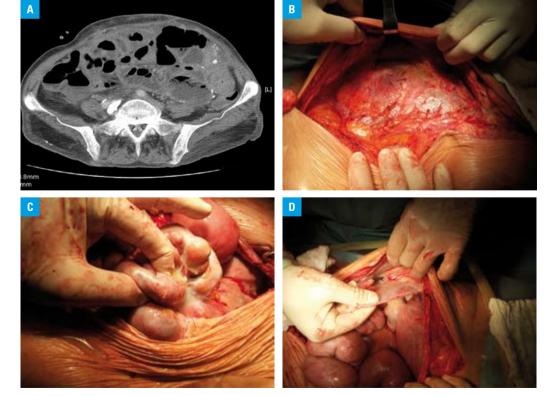


FIGURE 1 A – preoperative computed tomography scan; B – at laparotomy: small bowel loops covered with a fibrous sclerotic tissue; C – small bowel entrapped in cocoon; D – dissection of thickened parietal peritoneum with calcifications after removing its visceral part from bowel loops

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carefully dissected from the abdominal wall and bowel loops, and removed (FIGURE 1D). The patient was discharged in 2 weeks, with a complete resolution of all symptoms. His nutritional status improved and now – 2 years after the surgery – he is doing well on HD.

EPS is a rare but devastating complication of long-term PD associated with a significant mortality rate, reaching up to 55% at 1 year after the diagnosis.^{1,2} It is characterized by severe inflammation followed by peritoneal membrane fibrosis and intestinal encapsulation leading to recurrent small bowel obstruction. The etiology and pathogenesis of the syndrome remain unclear, and the most important risk factor seems to be the time on PD, possibly together with the recurrent episodes of bacterial peritonitis and peritoneal glucose exposure.^{1,2} In most cases, EPS occurs after the cessation of PD, and the transfer of patients to HD or kidney transplantation.^{2,3} Since the syndrome is relatively rare, there are no evidence-based treatment recommendations. The therapeutic strategies include corticosteroids and tamoxifen; however, in cases of bowel obstruction, the surgical management seems to be the most successful, provided it is performed by an experienced team.^{4,5}

REFERENCES

- 1 Brown MC, Simpson K, Kerssens J, et al. Encapsulating peritoneal sclerosis in the new millennium: a national cohort study. Clin J Am Soc Nephrol. 2009: 4: 1222-1229.
- 2 Latus J, Ulmer C, Fritz P, et al. Encapsulating peritoneal sclerosis: a rare, serious but potentially curable complication of peritoneal dialysis experience of a referral centre in Germany. Nephrol Dial Transplant. 2012 Jun 25. IEpub ahead of printl.
- 3 Brown EA, Van Biesen W, Finkelstein FO, et al. Length of time on peritoneal dialysis and encapsulating peritoneal sclerosis: position paper for ISPD. Perit Dial Int. 2009: 29: 595-600.
- 4 Kawanishi H, Ide K, Yamashita M, et al. Surgical techniques for prevention of recurrence after total enterolysis in encapsulating peritoneal sclerosis. Adv Perit Dial. 2008: 24: 51-55.
- 5 Augustine T, Brown PW, Davies SD, et al. Encapsulating peritoneal sclerosis: clinical significance and implications. Nephron Clin Pract. 2009; 111: c149-154.