CASE REPORT

A case of acute EPS with local cocoon formation in a patient on peritoneal dialysis

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Abstract Encapsulating peritoneal sclerosis is a rare and life-threatening complication of long-term peritoneal dialysis and until now there is no established medical treatment. Many factors have been incriminated in its pathogenesis but they do not explain all risk conditions. We report our experience and we investigate the predisposing factors. Probably unidentified factors make some patients more susceptible to developing encapsulating peritoneal sclerosis.

Keywords Peritoneal dialysis · Peritoneal histopathology · Peritoneal sclerosis

Introduction

Encapsulating peritoneal sclerosis (EPS) is a rare and life-threatening complication of long-term peritoneal dialysis (PD), characterized by rigidity of the peritoneum and dramatic fibrous thickening of small bowel [1, 2]. The incidence in case series of PD patients is approximately 1–3 % and increases with length of time on PD [3]. EPS is a challenge for clinical nephrology because there are no specific diagnostic effluent or blood markers for EPS. Until now, there is no established medical treatment for EPS and therapeutic guidelines have not yet been established [4].

Case report

A 71-year-old Caucasian female, for 7 years on automated PD, presented with recurrent partial small-bowel obstruction. PD has been her only dialysis treatment of end-stage renal disease due to left renal hypoplasia. Two years before she developed the episode of peritonitis, due to *Enterococcus faecalis*. We think that the route of infection was intrinsic because the patient was suffering from chronic colonic diverticular disease. The patient had no history of gastric or duodenal ulcer or previous bowel immobility episodes and no evidence of beta-blockers treatment, abdominal surgery, malignancy, ultrafiltration failure or hypertonic dialysis solutions use; no peritoneal deterioration features. The evaluation of PET (D/P creatinine 0.63, D/D0 glucose 0.39) was low-average transporter. The adequacy of PD treatment was always according to guidelines. The last K_t/V was 1.87. She was on therapy with calcium and calcium acetate 435 mg/magnesium carbonate 235 mg (4 tablets/day), calcitriol and epoietin. For 2 months she experienced abdominal discomfort, vomiting and recurrent abdominal pain. Since starting gastrointestinal symptoms, the dialysate leukocyte counts and the cultures were negative for peritoneal infection. We did not note a bloody drainage. C-reactive protein and procalcitonin levels and blood white cells counts were in the normal range. The abdominal CT scan demonstrated evidence of peritoneal sclerosis and absence of gas–fluid levels and calcifications. The patient refused the hospital admission and we could not start the steroid treatment; 2 weeks later she again experienced bowel immobility. Laboratory investigations for peritonitis were negative. Based on those radiologic and clinical findings, the patient was diagnosed with EPS and appropriate antibiotics and total parenteral nutrition were initiated. PD treatment was stopped and hemodialysis

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treatment was started. Two days later, after an initial improvement, the patient again developed a severe bowel obstruction. The new abdominal CT scan showed the same picture and air gas–fluid levels (Figs. 1, 2). The patient underwent surgical intervention. Following laparotomy, under a dense sclerotic membrane we found thickening of intestinal wall and entrapment of small bowel in fibrous tissue; before the ileocecal valve, a cocoon-like encapsulation was surrounding the last 80 cm of small-bowel loops. We detected a single intestinal perforation in the small-intestine last loop. We did not find lesions in the stomach, duodenum, liver, gallbladder, pancreas, spleen and large intestine. An ileocecal resection and an ileocolic anastomosis were performed but after 12 h the patient died for septic shock. There was no evidence of anastomotic insufficiency as a cause of death. Histopathology examination showed a massive alteration in peritoneal membrane morphology with a PD-induced chronic tissue damage, consisting of mesothelial denudation and interstitial fibrosis (Figs. 3, 4).

Discussion

Encapsulating peritoneal sclerosis is a serious complication of long-term PD; the mortality rate for EPS has been high, because of bowel obstruction and after-surgery complications [5]. There is a lack of well-defined diagnostic criteria, especially to distinguish early stages of EPS. Many factors have been incriminated in its pathogenesis, especially bio-incompatible PD fluids and peritoneal infections. EPS is significantly associated with PD duration, particularly more than 5 years. The underlying causal relationship between

Fig. 1 Axial image from a CT scan of abdomen demonstrating peritoneal thickening, tethering and dilatation of small-bowel loops (red arrow) with increased density of mesenteric fat; peritoneal catheter (green arrow) (color figure online)

Fig. 2 Coronal reformatted image from abdominal CT scan: aortic wall calcification (red arrow), ascites (white star), tethering and dilatation of small-bowel loops (white arrow), peritoneal catheter (green arrow) (color figure online)

Fig. 3 Peritoneal histological examination: the mesothelial cell layer is lost and replaced by a band of acellular collagen with a submesothelial thicknesses (H&E stain, $\times 10$)

long duration on PD and an increased risk of EPS is currently not known. Most of the patients on long-term PD have an advanced peritoneal deterioration (peritoneal sclerosis: mesothelial denudation, submesothelial interstitial fibrosis, vascular sclerosis) but not all patients with those morphologic changes will suffer from EPS. The “two-hit” hypothesis highlights the balance between two factors, PD-induced peritoneal damage and superimposed inflammation. This theory explains why EPS tends to occur rather easily in long-term PD patients, but the exact cause and pathogenesis are still not clear [5–7]. Nowadays the factors, PD-related (length of time on PD; solution constituents: plasticizers, chlorhexidine, acetate; peritonitis history; Increasing peritoneal transport rate) and non PD-

Fig. 4 Peritoneal histological examination: mesothelial denudation, interstitial fibrosis and interstitial mononuclear cell infiltration (H&E stain, $\times 10$)

related (abdominal surgery, hepatic ascites, tuberculous peritonitis, autoimmune disease, abdominal malignancy, beta-blockers therapy) incriminated in its pathogenesis, do not explain all risk conditions for the development of EPS.

Our patient did not show peritoneal deterioration clinical features neither chronic inflammatory state. She did not exhibit clinical findings (malnutrition, anasarca) nor persistent symptoms of gastrointestinal dysfunction (vomiting, constipation, early satiety) to suggest the EPS.

Regarding the “two-hit” hypothesis, in the present case among suggested predisposing factors of EPS we detected the duration of PD longer than 5 years and the episode of peritonitis. We cannot exclude some subclinical episodes of peritonitis that could explain a possible transition from simple sclerosis to EPS. Up to now, the natural history and the evolution of EPS are not quite known. Most of the reported cases show a slow and progressive course. In this patient, we report a fast course of her symptoms. The abdominal cocooning is a pathognomonic finding but requires further investigations.

Conclusion

Up to now, not all predisposing factors have been identified. Probably unidentified factors make some patients more susceptible to developing EPS. Future research needs to focus primarily on identifying risk factors and predictive markers of EPS that can be used in clinical practice [8].

Conflict of interest The authors declare that no conflict of interest exists.

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