Sclerosing Encapsulating Peritonitis in a Case on Continuous Cyclic Peritoneal Dialysis

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Abstract

Sclerosing encapsulating peritonitis (SEP) is a rare but serious and devastating complication with a very high mortality rate in patients undergoing peritoneal dialysis. We hereby report a 63-year-old female patient diagnosed to have end-stage renal disease (ESRD) on continuous cyclic peritoneal dialysis (CCPD) for 14 years. Owing to ultrafiltration failure, aside from CCPD and daytime dwell, she had twice monthly hemodialysis for 9 years. She presented with persistent bloody dialysate effluent associated with anorexia, poor appetite, nausea, vomiting and progressive body weight loss. Peritoneal dialysis was discontinued and patient was shifted to thrice weekly regular hemodialysis. However, despite nutritional support, patient died eight months later because of severe malnutrition and cachexia. (Acta Nephrologica 2011; 25: 77-80)

KEY WORDS: sclerosing encapsulating peritonitis (SEP), peritoneal dialysis, ultrafiltration failure

Introduction

Sclerosing encapsulating peritonitis (SEP) is characterized by progressive intraabdominal inflammatory process, resulting in formation of thick fibrous sheath, encasement of small bowel, often with calcification of small bowel and peritoneum, leading to adhesion, sometimes bleeding, bowel obstruction, malnutrition and eventually death in 24-83% of this disorder (1).

Owing to rarity and relatively long period of development of this disorder, diagnosis is usually made late in the disease with a very poor outcome, after extensive fibrosis of peritoneal membrane has occurred, a stage where treatment has been shown to have a very limited success.

Case Report

The patient is a 63-year-old female, diagnosed to have end stage renal disease (ESRD) on continuous cyclic peritoneal dialysis (CCPD) for 14 years. Her peritoneal equilibration test (PET) showed a high transporter with D/D0 glucose of 0.25 and D/P creatinine of 0.9. Dialysates include 1.5% and 2.5% Dianeal peritoneal dialysis solution, 5000 mL each as continuous cyclic peritoneal dialysis at night with additional daytime dwell of Icodextrin 2000 mL and 2.5% Dianeal solution 2000 mL. An ultrafiltration of less than 1000 mL was achieved in a day. Owing to ultrafiltration failure, aside from CCPD and daytime dwell, she had twice monthly hemodialysis for 9 years. During the past 14 years, she had no episodes of peritonitis. Her hypertension was controlled with Labetalol 200 mg BID. Eight months ago, she had a tunnel infection. The peritoneal dialysis (PD) catheter was removed and a new PD catheter was reimplemented. Since then, the patient complained of vague abdominal discomfort and bloody dialysate effluent. Ascites routine study showed a bloody turbid fluid with specific gravity 1.018, glucose 91 mg/dL, lactate dehydrogenase (LDH) 185/iu/L, protein 2.0 mg/dL, red blood cell (RBC) 20000/µL, white blood cell (WBC) 110/µL, polymorphonuclear cell (PMN) 15%, and negative ascites.

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bacterial culture, acid fast stain and TB culture. Ultrasound revealed septated complicated peritoneal fluid (Fig. 1) while computerized tomography (CT) scan revealed curvilinear plaque of calcification outlining the peritoneum and small bowel loops (Fig. 2). Two months later, because of persistent intraabdominal bleeding, laparoscopy was done which revealed chronic inflammation, diffuse edema of peritoneum and all intraabdominal organs with thick fibrin, adhesion and minor oozing between small bowel and peritoneum (Fig. 3). Peritoneal biopsy revealed fibrous and fibrinoid tissue (Fig. 4). Aside from the above symptoms, the patient also complained of anorexia, poor appetite, nausea, vomiting, and body weight loss. Three months later, the patient was convinced to have the PD catheter removed and shifted to regular hemodialysis three times a week. However, the above symptoms did not improve. She had persistent intraabdominal bleeding, requiring weekly abdominal paracentesis of around 700-1000 mL of bloody ascites and frequent blood transfusion. Despite nutritional support with albumin and intradialysis parenteral nutrition, the patient died eight months later because of severe malnutrition and cachexia.
Discussion

SEP was first described by Gandhi et al., (2) in 1980. It is a rare but a life-threatening complication in patients undergoing peritoneal dialysis. The etiology remains elusive; peritonitis might be the most important risk factor; however, our patient did not have any episodes of peritonitis during the past 14 years of peritoneal dialysis. Other factors including peritoneal dialysis duration, exposure to hypertonic glucose, advanced glycation end products (AGE) (3), poor biocompatibility of dialysis solutions, chlorhexidine as disinfectant, acetate-based dialysis solutions, use of beta blocker and genetic predisposition have also been implicated as causative agents.

The prevalence rate ranges from 0.5 to 7.3% according to literature review in patients undergoing peritoneal dialysis. The frequency and occurrence are related to duration of PD (4, 5). SEP usually occurs in patients receiving continuous ambulatory CAPD for more than 4-5 years (6). Rigby and Hawley (7) also emphasized the impact of CAPD duration on the development of SEP, reporting low frequency in patients on peritoneal dialysis for less than 2 years with prevalence rate of 1.9% and rising to 19.4% in patients on peritoneal dialysis for more than 8 years. In a multicenter experience reported by Kim et al. (6) in Korea, SEP was commonly noted in patients with CAPD duration of more than 4 years with a prevalence rate of 21%. The prognosis is poor with a reported mortality rate of 43.5% in a multicenter Japanese study 8 and 56% in an Australian multicenter study (7). In the prospective study conducted by Kawanishi in Japan (8), the prevalence and mortality rate were 2.5% and 38%, respectively.

The clinical features of SEP include abdominal pain, nausea, vomiting, sometimes with blood-stained dialysate and loss of ultrafiltration (9). Abdominal pain is common as disease progresses. However, due to rarity and relatively long period of development of this disorder with no specific clinical predictors of SEP, early diagnosis and treatment prior to development of symptoms is difficult.

Better care of our peritoneal dialysis patients and decreased incidence of peritonitis allow our patients to remain longer on peritoneal dialysis, as in our patient who underwent peritoneal dialysis for 14 years, a relatively long period. Loss of ultrafiltration and change in permeability characteristic of peritoneum (10) may be the only clues to the occurrence of SEP. However, image findings, including CT scan, are not sensitive enough to demonstrate early peritoneal changes of SEP. Hence, it is not useful as a screening tool. Direct peritoneal visualization by a well-trained laparoscopic surgeon may be helpful in highly suspected cases.

The above clinical features in conjunction with findings from imaging allow for a confident diagnosis. Radiologic findings include thickening and calcification of the bowel wall and calcification of peritoneum. Centrally located gas-filled and dilated loops of bowel wall may be seen in the presence of small bowel encapsulation. The earliest ultrasound findings are abnormal peristalsis of bowel loops and peritoneal fluid collection, with echogenic peripancreatic stranding that may progress to loculation (11). Our patient’s ultrasound revealed septation of peritoneal fluid. CT scan showed peritoneal thickening and calcification. Loculated fluid collection is seen in up to 90% of cases (12). The above findings are very typical in our patient; however, these radiologic findings are seen only in the late stage of the disease with a very poor outcome.

Eighteen F-fluorodeoxyglucose positron emission tomography (FDG-PET) enable us to identify areas of high metabolic activity within the body, with the tracer being taken up by tissues that are utilizing glucose. Tarzi et al. (13) used FDG-PET scan in 4 patients with suspected SEP and all showed positive scan results. The most important risk factor for SEP in our case was the duration of peritoneal dialysis (more than 14 years) with ultrafiltration failure. However, not all patients on longterm peritoneal dialysis developed this complication, so a screening test such as PET scan to detect patients at risk would be very useful. However, the validity of PET scan needs further investigation.

Treatment of SEP is largely supportive with cessation of peritoneal dialysis, shifting to other forms of renal replacement therapy and nutritional support. However, it takes us five months to finally convince the patient to have the PD catheter removed. In patients presenting with gastrointestinal symptoms and bloody dialysate, we should have a high index of suspicion of possible SEP and early discontinuation of peritoneal dialysis may prevent fatal disease pro-

Fig. 4. Peritoneal biopsy shows low cellularity and fibrinoid tissue. (Haematoxylin and Eosin stain × 100)
gression like our patient. There is paucity of evidence for determining the optimal treatment of sclerosing encapsulating peritonitis. Surgical treatment is reserved for intestinal obstruction with poor outcome and extremely high mortality rate of 60% due to post-operative complications such as sepsis caused by rupture of anastomosis or intestinal leakage, intraabdominal infection and enterocutaneous fistula. However, in recent years, several series have been published with new promising surgical outcomes (8, 10, 14). Enterolysis is a preferred surgical technique with avoidance of intestinal resection and anastomosis. Recent data indicate that surgical technique is of major importance for favorable outcome (15).

Kawanishi et al. (16) reported a dramatic improvement in outcome after surgical treatment. In reported series, the mortality was just 4% (2 out of 50 patients). These successful outcomes were achieved in a center with high level of expertise in EPS surgery, the procedure were conducted by an experienced and dedicated surgeon using technique with ablation of the capsules and intestinal adhesions with appropriate enterolysis, which constitute the key elements in resolving the state of ileus.

Immunosuppressants, including prednisolone and cyclosporine may be useful when active inflammation is present. The use of Tamoxifen (17) has been reported. The drug has antifibrotic effect and may reduce the expression of transforming growth factor β-1, which is related to the development of SEP. However, it is only conceptually attractive but proved disappointing in clinical use.

Conclusion

We need to have a high index of suspicion in peritoneal dialysis patients with abdominal pain and associated risk factors, especially in patients with long duration of CAPD and ultrafiltration failure. Diagnosis should be pursued aggressively through direct observation of peritoneal membrane by skilled laparoscopic surgeon. Radiologic diagnosis, like CT scan, is not sensitive enough to demonstrate early peritoneal changes of SEP, and is not useful as a screening tool. Early recognition of the above complications, followed by immediate cessation of peritoneal dialysis and initiation of an alternate form of dialysis, may prevent potentially fatal disease progression.

References