Small Bowel Perforation: a Late and Fatal Complication of Encapsulating Peritoneal Sclerosis

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Abstract

Encapsulating peritoneal sclerosis (EPS) is a rare and serious complication in patients undergoing peritoneal dialysis (PD). The incidence of EPS increases with duration of PD. The rate of EPS-associated mortality rate is usually high. We encountered a 51-year-old uremic female patient who received PD therapy initially but was shifted to hemodialysis 9 years later due to dialysis inadequacy and ultrafiltration failure. There were three episodes of PD-related peritonitis during her PD therapy. EPS was diagnosed according to image and operation findings during a peritonitis episode. Enterolysis was performed and short-term steroid therapy was also administered. However, she experienced another episode of rapid progressive abdominal pain with compromised hemodynamic status. Abdominal computed tomography demonstrated perforation of small bowel with pneumoperitoneum. Despite emergency laparotomy and antibacterial therapy, the patient died 2 days later. We conclude that EPS can still progress in uremic patients despite suspension of PD. Our patients died of small bowel perforation with overwhelming sepsis.

KEY WORDS: encapsulating peritoneal sclerosis, bowel perforation, peritoneal dialysis

Introduction

Encapsulating peritoneal sclerosis (EPS), a serious and potentially devastating complication of peritoneal dialysis, results from peritoneal and systemic inflammation, followed by progressive bowel obstruction and severe malnutrition. The incidence of EPS in large, multi-center studies was estimated to be 0.8-3.3% (1). The incidence increased with duration of peritoneal dialysis (PD), which was 1.9%, 6.4%, 10.8% and 19.4% in patients on PD for more than 2, 5, 6 and 8 years, respectively (2). Other risk factors of EPS include bioincompatible dialysate, icodextrin use, chlorhexidine disinfectant, povidone-iodine, dialysate contamination with plasticizers and particulate matter, PD catheters, beta-blockers, and severe peritonitis (1). One previous study revealed that 33 (72%) out of 46 patients were diagnosed as EPS within 2 years after PD had stopped (3). Furthermore, it has also been reported that 5 (45%) out of 11 patients were diagnosed with EPS after switching to hemodialysis (HD) or receiving a transplant (4). Clinical features of EPS include ascites, abdominal mass, impaired peritoneal ultrafiltration, or partial or complete small bowel obstruction which results in anorexia, nausea, vomiting, abdominal fullness, abdominal pain, absent bowel sounds, and constipation (5). The mortality of EPS is high with most patients attributed to severe infection (6). Among all causes of death, small bowel perforation has rarely been reported.

Case Report

A 51-year-old female uremic patient had received PD therapy since 1999. The underlying renal disease was chronic glomerulonephritis. Her PD re-
gimen included three 1.5% dextrose (Dianeal® 2.5L) and two 2.5% Dextrose (Dianeal® 2.5L) exchanges per day. Without any remarkable event, her residual kidney function became negligible 4 years later. There were three episodes of PD-related peritonitis which occurred 1, 4 and 8 years respectively after the commencement of PD and eventually she was shifted to HD in 2008 due to dialysis inadequacy and ultrafiltration failure. The responsible pathogens of PD-related peritonitis were *Staphylococcus epidermidis* and *Serratia marcescens* for two episodes and no definite microorganism was identified for one episode. Peritoneal function as assessed by peritoneal equilibration test showed low average initially but progressed to high average after the second peritonitis. The course of PD treatment was quite smooth and she was in good nutritional status with no obvious body weight loss and the serum albumin level ranging from 3.3 gm/dL to 4.2 gm/dL. The total peritoneal clearance estimated by Kt/V was 1.91, 2.23 and 1.65, respectively after resolution of peritonitis episodes. After the switch to HD, the treatment course was rather smooth. Her HD schedule was thrice weekly with four hours per session. The latest Kt/V was 1.28 and URR was 72.5%. The PD catheter was removed later under the consideration of permanent treatment shift from PD to HD. One week after the removal of PD catheter, she presented to our emergency department with progressive diffuse abdominal pain. No evidence of hollow organ perforation was recorded. Abdominal computed tomography showed loculated fluid collection, peritoneal calcification, focal small bowel dilatation and thickened peritoneum. EPS was thus diagnosed according to clinical manifestations and image study. Exploratory laparotomy with enterolysis was performed in July, 2008. The operation record remarked pelvic abscess, extensive adhesion at terminal ileum, and calcification along with abscess wall. The culture of pelvic abscess did not show any identified microorganisms. Afterwards, to prevent disease progression, a short-term steroid therapy with prednisolone 20 mg per day was also prescribed but was soon withheld due to intolerance. In October, 2009, she was sent to our emergency department with abrupt diffuse abdominal pain. Upon arrival, profound hypotension (blood pressure: 77/42 mmHg), abdominal distention with rebounding pain were noted. Laboratory tests revealed leukocytosis (WBC: 10300/mm³) and elevated CRP (12.5 mg/L) and lactate (32 mg/dL). Biochemical data of blood sample were as follows: blood urea nitrogen 82 mg/dL, creatinine 9.0 mg/dL, sodium 133 meq/L, and potassium 6.5 meq/L. Abdominal computed tomography demonstrated ascites, multiple perforations of small bowel loops with pneumoperitoneum and multiple peritoneal calcification (Fig. 1). Despite emergency laparotomy and antibacterial therapy, the patient died 2 days later. Marked distended abdomen, multiple perforations and ischemic change of the whole small bowel were noted during operation. The culture of ascites yielded multiple microorganisms: including *E. coli*, *Enterococcus faecalis*, *Enterobacter cloacae*, and *Clostridium perfringens*.

**Discussion**

EPS, an uncommon but catastrophic complication of long-term PD therapy, causes the thickened peritoneum and encasement of the small intestine. Of note, PD-associated peritonitis is a risk factor for EPS development (7) and most of the episodes of peritonitis developed between 4 and 14 years after the initiation of PD (6). However, EPS might occur even by mild bacterial peritonitis (8). A ‘two-hit’ hypothesis for EPS has been proposed in which disruption of normal peritoneal/mesothelial physiology like mesothelial denudation, interstitial fibrosis, fibrin deposition, and neovascularization as a consequence of exposure to PD with bioincompatible dialysis solutions that occurs generally over a period of years predisposes the individual to a second hit that triggers the process (9). This second hit, as an initiating factor, like an episode of peritonitis, discontinuing PD, or an acute intra-abdominal event is prone to trigger the extensive peritoneal lesions that are characteristic of EPS (10). Our patient experienced three episodes of peritonitis. Although not severe, the repetitive inflammation and destruction may provoke the development of EPS.

The mortality rate of EPS varies between 24 and 56% (11, 12). Overall the survival rate for patients with EPS at 1, 2, 3, and 5 years after diagnosis were 69, 62, 58, and 35%, respectively (1). The causes of mortality in patients with EPS are multiple. As demonstrated in recent studies, cardiac and respiratory arrest, bacte-
remia, sepsis or gastrointestinal bleeding constitutes the major causes of death (6, 13). Among all EPS-associated complications, small bowel perforation has rarely been reported (13, 14). All reported patients had catastrophic outcome as we presented here. The first case had received PD for 165 months but experienced perforation of small intestine with evolving pan-peritonitis and died of sepsis 17 months after EPS onset (14). Kawanishi et al. reported 48 EPS patients, among which 4 patients died of perforative peritonitis, though the locations were not mentioned (13). The pathophysiology of this rare complication has not been clarified yet. Mechanical obstruction secondary to encapsulation and vascular insufficiency may contribute to small bowel perforation. Of note, several perforations of small bowel were noted in our patient during operation. This finding indicates an extensive involvement and certainly our patient experienced a rapid deterioration course. Subsequent culture result proved to be multiple bacteria infection. The severe sepsis is difficult to control even though surgical intervention was not delayed.

Image study can help to make an early diagnosis of PD-related EPS and usually CT scan is of the greatest importance (15). Typical features of CT scanning in EPS include peritoneal abnormalities, small bowel abnormalities, and loculated fluid collections (16). The peritoneal abnormalities include peritoneal thickening which may progress to peritoneal encapsulation of small bowel loops, as cocooning of the small bowel, and peritoneal calcification (15). Peritoneal calcification can involve both visceral and parietal peritoneum (5). This visceral calcification may present as a small focal area, fine linear pattern or extensive conglomerate calcification (10). Small bowel abnormalities represent small bowel dilatation which is caused by the sclerosed, thickened peritoneum that surrounds the small bowel loops, while loculated fluid collections indicate loculated ascites that can be infectious (15). The CT scan finding in our case revealed peritoneal thickening, linear peritoneal calcification along several segments of small bowel, suggesting a generalized alteration. Currently, there are no randomized, controlled trials to guide the management of patients with EPS. Recommended treatments include use of immunosuppressant agents, predominantly the corticosteroids; antifibrotic agents such as tamoxifen, nutritional support, and surgery to remove the fibrotic material (17). In our patient, enterolysis had been performed in previous exploratory laparotomy for extensive adhesion. Additional short-term steroid therapy had also been provided. Even after PD was stopped, it appeared that EPS continued to progress and our patient died of sepsis due to multiple small bowel perforations.

We conclude that small bowel perforation is a rare complication of EPS. EPS can still progress even after withdrawal of PD and shift to HD. Our patient died of small bowel perforation with subsequent overwhelming sepsis in spite of emergency surgical intervention and antibiotics therapy.

References