

ENCAPSULATING PERITONEAL SCLEROSIS PRESENTING WITH HAEMORRHAGIC ASCITES AFTER TRANSFER FROM PERITONEAL DIALYSIS TO HAEMODIALYSIS

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ABSTRACT

A patient with end-stage kidney disease due to chronic glomerulonephritis was initiated on continuous ambulatory peritoneal dialysis. After three years he was transferred to haemodialysis following recurrent episodes of peritonitis. After the commencement of haemodialysis the patient developed progressive abdominal distension; paracentesis revealed bloody ascites. Radiographic imaging revealed features of small bowel obstruction with bowel loops matted to the posterior abdominal wall. A diagnosis of encapsulating peritoneal sclerosis was made. Treatment with prednisone was initiated but the patient's condition steadily worsened and he demised a year later due to severe malnutrition and sepsis.

KEY WORDS

Peritoneal dialysis, bloody ascites, encapsulating peritoneal sclerosis, prednisone.

BACKGROUND

Encapsulating peritoneal sclerosis (EPS) is a rare but devastating complication of peritoneal dialysis. A recent study reported an incidence rate of 1.8/1000 patient-years [12]. It usually presents insidiously with non-specific abdominal symptoms and has a very poor prognosis, with most cases proving resistant to therapy. The disease progresses relentlessly until death occurs from malnutrition and sepsis. We report a case of EPS presenting with bloody ascites.

CASE

A 40-year-old male with end-stage kidney disease secondary to chronic glomerulonephritis was commenced on continuous ambulatory peritoneal dialysis (CAPD) in December 2005. He did not have hypertension. His peritoneal dialysis prescription consisted of five exchanges daily (4 x 2L of 2.3% dextrose and 1 x 2L of 4.25% dextrose solution).

In 2006 he had two episodes of peritonitis secondary to *Staphylococcus epidermidis*, each episode being successfully treated with vancomycin. However, in February 2009, he developed non-resolving peritonitis, the Tenckhoff catheter was removed, and he was transferred to haemodialysis.

At the end of May 2009 he presented with weight loss, poor appetite and vague abdominal symptoms and was found to have abdominal distension with shifting dullness. CRP was raised at 135mg/L. Abdominal radiograph revealed multiple air fluid levels (Figure 1, panel A) and an ultrasound revealed a large volume of free intra-abdominal fluid. A paracentesis revealed that the ascitic fluid was bloody. In view of his previous history of tuberculous (TB) pericarditis, multiple samples of fluid were sent for TB culture. These were all negative. Cytological examination of the ascitic fluid did not reveal any malignant cells.

By June 2009, his symptoms had progressed. His CRP remained elevated at 128mg/L. An abdominal computed tomography (CT) scan revealed loculated ascites anteriorly with the bowels matted to the posterior abdominal wall

(Figure 1, panel B). A diagnosis of encapsulating peritoneal sclerosis was made and he was given a therapeutic trial of oral prednisone 40mg daily for six weeks. This was stopped after he failed to show any clinical response. He had progressive deterioration with wasting and malnutrition. In July 2010 he developed overwhelming Pseudomonas and Klebsiella sepsis which resulted in his demise.

DISCUSSION

EPS often has a multi-factorial aetiology [2] and our patient demonstrated several of these.

Our patient was relatively young, a risk factor in some studies [10], and had recurrent and non-resolving peritonitis. Peritonitis has been regarded as a risk for the future development of EPS [2]; however, some recent evidence suggests that peritonitis may not be a risk factor [10]. The duration on CAPD is the most important risk factor for the development of EPS [5]. The respective cumulative incidences in a recent study at three, five and eight years were 0.3%, 0.8% and 3.9% [10]. Our patient's duration on CAPD was only 38 months.

This case demonstrated two of the key features of EPS: symptoms of partial bowel obstruction, and typical radiologic findings. The diagnosis can usually be made on the basis of the clinical and radiologic evidence [2]. However, a definitive diagnosis may require histological evidence [2, 5]. We felt that a peritoneal biopsy was not needed due to the clear clinical and radiographic evidence.

Symptoms usually have an insidious onset, as in our patient, and are due to thickening of the peritoneum with resultant intestinal obstruction [5]. Ultrafiltration failure is common and often develops well before a diagnosis of EPS is made [11]. At our centre, we routinely initiate our patients with a peritoneal dialysis prescription of four daily exchanges of 1.5% dialysate. Our patient required five daily exchanges including one 4.5% dialysate exchange.

Evidence of bowel obstruction can be obtained using radiographic techniques [5]. Our patient had an abdominal X-ray picture in keeping with bowel obstruction; however, this is not sufficient to make a diagnosis of EPS. An ultrasound of the abdomen only revealed free fluid. Ultrasound may demonstrate the so called "sandwich" appearance of the peritoneal membrane, trilaminar thickening of the membrane, tethering to the posterior abdominal wall and dilated, fixed and matted bowel loops [6]. CT of the abdomen revealed several of these features including loculated ascites, tethering of the bowels to the posterior abdominal wall and dilated bowel loops [5, 6].

TREATMENT AND PROGNOSIS

The only prospective, multicenter study [2] compared total parenteral nutrition (TPN), corticosteroids (in combination with TPN alone or TPN plus surgery) and surgery (with

all receiving TPN). Recovery was 0%, 38.5% and 58.3% respectively. Other drugs that have been used include tamoxifen, and as well as immunosuppressive drugs like cyclosporine and azathioprine [2]. A few studies showed some benefit with tamoxifen – in these studies the patients were receiving concomitant oral corticosteroids [7, 8, 9]. Data on immunosuppressive drugs other than corticosteroids are limited and therefore strong recommendations cannot be made on their efficacy. Our patient received approximately six weeks of oral prednisone without any improvement.

The prognosis of EPS is very poor once the patient develops symptoms, with a mortality rate of more than 60% within four months of being diagnosed. Our patient died exactly one year after diagnosis due to malnutrition with overwhelming sepsis. The usual causes of death are malnutrition with sepsis, or following surgery [2]. Preventive measures should be instituted in all patients commenced on CAPD. These include avoiding hypertonic dialysate solutions and preservation of residual renal function [2, 4]. Electively stopping CAPD after eight years and switching to haemodialysis is another consideration [2].

CONCLUSION

EPS is a rare but devastating complication of CAPD. It classically presents insidiously with clinico-radiographic features of bowel obstruction. Haemorrhagic ascites following transfer from peritoneal dialysis to haemodialysis may be the initial form of presentation. Clinicians should have a high index of suspicion for EPS in any patient on peritoneal dialysis with non-specific abdominal symptoms. Preventive strategies should be instituted in all patients commenced on CAPD as the response to treatment is poor.

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Figure 1, Panel A: Abdominal X-ray showing bowel obstruction.

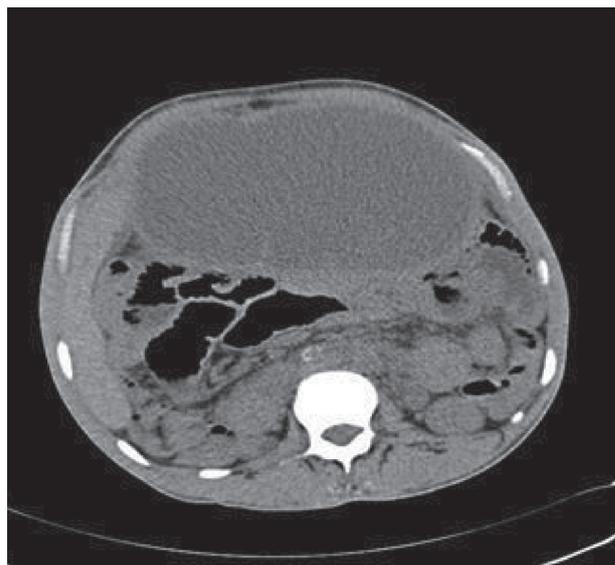


Figure 1, Panel B: Computed tomography of the abdomen showing loculated ascites anteriorly with tethering of bowel to the posterior abdominal wall and dilated loops of bowel.