THE NEW ZEALAND MEDICAL JOURNAL
Journal of the New Zealand Medical Association

Encapsulating peritoneal sclerosis—a complication of peritoneal dialysis

Caroline L Chembo, Alastair Macdonald, Nicola Hay, Philip Matheson, Grant Pidgeon, Murray Leikis

Abstract

Encapsulating peritoneal sclerosis (EPS) is a rare complication of peritoneal dialysis. It often presents with non-specific symptoms, leading to a delay in diagnosis and a poor prognosis. Here we report a case of EPS in a patient treated with peritoneal dialysis and discuss the risk factors, diagnostic challenges and treatment options available.

Case report

We present a 66-year-old Caucasian man on haemodialysis three times a week at time of presentation with gastrointestinal symptoms. He had a history of end-stage renal failure secondary to IgA nephropathy. His other comorbidities included a 35-year history of hypertension treated with various beta-blockers and previous parathyroidectomy. The beta-blockers included atenolol in the earlier years later changed to controlled-release metoprolol.

He had been on peritoneal dialysis from 1986 to 1992 before receiving a deceased donor renal transplant. 13 years later his renal function deteriorated and biopsy showed chronic allograft nephropathy. In September 2005 he returned to dialysis, opting for peritoneal dialysis as this suited his lifestyle.

In April 2009 he started experiencing non-specific symptoms such as general malaise, anorexia and fatigue. These were initially attributed to under-dialysis the symptoms coincided with declining dialysis adequacy. Switching to haemodialysis was advised but did not happen until September of that year due to patient reluctance to switch and vascular access issues. In total, the patient had 10 years of peritoneal dialysis exposure and had two episodes of bacterial peritonitis, once in 1988 and again in 2007.

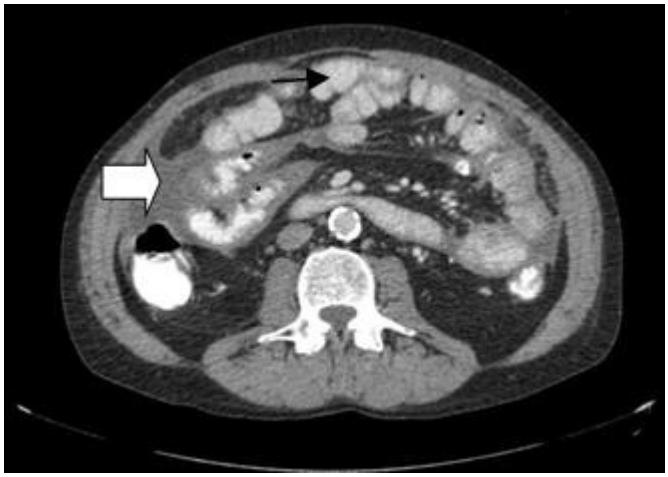
In November 2009 he presented to hospital with abdominal discomfort, early satiety and feeling generally unwell. As he still had his tenchkoff catheter *in situ*, he had a peritoneal flush. The fluid had 800×10^6/L mononuclear cells, Gram stain was negative and there were no acid fast bacilli. He was treated with intraperitoneal antibiotics (gentamicin and vancomycin).

The initial symptoms resolved but a month later he represented with similar symptoms. Gastroscopy showed mild gastritis for which he received omeprazole. The symptoms abated slightly but 2 weeks later he represented with abdominal pain and vomiting copious amounts of fluid about 2 hours after eating. His tenchkoff was removed and a peritoneal biopsy was done.

He had a CT scan (Figure 1) which showed loculated peritoneal fluid and slightly thick walled and enhancing small bowel. Given his symptoms, CT findings and peritoneal biopsy report (Figure 2), a diagnosis of encapsulating peritoneal sclerosis was made.

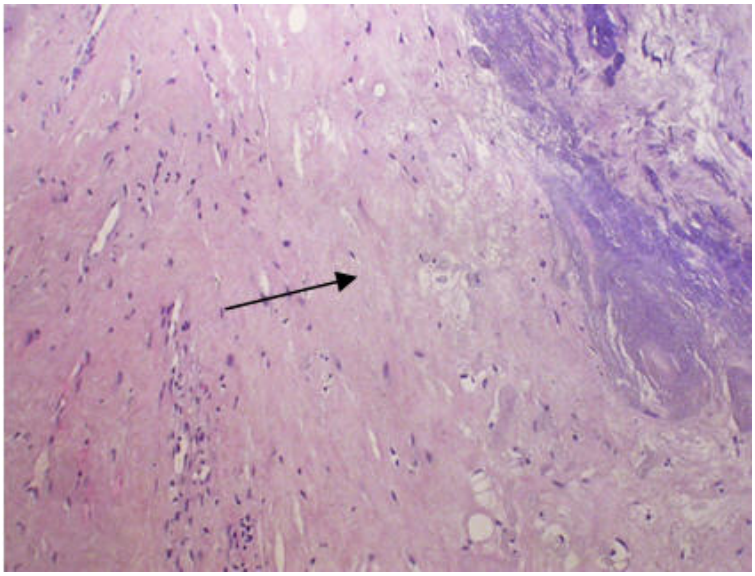
The bowel obstruction was managed conservatively with nasogastric (NG) tube on free drainage and antiemetics. He was commenced on intravenous hydrocortisone and parenteral nutrition. When the vomiting had settled he switched to oral prednisone at 60 mg and tamoxifen at 20 mg daily.

Figure 1. CT scan showing loculated peritoneal fluid and slightly thick walled and enhancing small bowel



White arrow=loculated effusion; black arrow=enhancing small bowel.

Figure 2. Peritoneal biopsy slide (×400) showing dense hyalinised fibrous tissue



Arrow indicates hyalinised fibrous tissue.

The vomiting abated for about 3 weeks but recurred leading to his readmission. His management was still conservative, he was kept nil by mouth and had an NG tube on free drainage. His steroids were change to parenteral.

The prognosis for encapsulating peritoneal sclerosis is quite poor, therefore a discussion with the patient and his family was arranged. After the family meeting and with the lack of patient improvement with conservative management, the patient decided to have all treatment withdrawn. Palliative care measures were instituted and the patient was discharged home with input from the Hospital Palliative Care Team. The patient died 2 days later at home.

Encapsulating peritoneal sclerosis (EPS)

EPS is a condition that is characterised by bowel loops being cocooned in thickened peritoneum. It is a rare but recognised complication of peritoneal dialysis.[1] Since the initial symptoms are non-specific and are usually attributed to failure of peritoneal dialysis, the patient is then prepared to switched to haemodialysis or transplanted. It has been known to occur immediately post bacterial peritonitis, however diagnosis is usually established after cessation of peritoneal dialysis.

Duration on peritoneal dialysis has been associated with EPS. It is rare if peritoneal exposure is less than 2 years but rises to around 20% with exposure over 8 years.[2] The peritoneal membrane transport status is also linked to EPS with high transport status being more commonly associated with EPS. History of recurrent episodes of peritoneal dialysis related peritonitis also increases the risk of developing EPS.[2] EPS has been associated with conditions such as SLE, malignancy, peritoneo-venous shunts and beta-blockers, especially practolol but idiopathic cases have been described.[3]

The pathophysiology of peritoneal sclerosis is not fully understood. When linked to peritoneal dialysis, the process of sclerosis is thought to be enhanced by advanced glycosylated end products. These products result from the glucose degradation in the PD fluid during the process of sterilisation.[4] However other mediators are thought to play a role. Vascular endothelial growth factor (VEGF) and transforming growth factor beta (TGF-β) are possible mediators of this process.[5] Angiotensin II levels may also have a role as levels are very elevated in PD effluent of patients with EPS.[6] Epithelial–mesenchymal transition has also been postulated as the potential mechanism of the peritoneal fibrosis.[6]

Clinical features are initially non-specific with low grade fever, raised inflammatory markers and malnutrition. Patients may also present with recurrent cloudy PD effluent without overt peritonitis. Dialysis adequacy may also be declining. As the condition progresses bowel obstruction can develop.[7] Mortality can be as high as 60% depending on the duration of peritoneal dialysis.[8]

Radiological features are non-specific and may not be diagnostic including features of bowel obstruction. CT may show calcification of peritoneum, loculated pockets of fluid and air fluid levels. PET scan may have a role especially in the early inflammatory stage.[9]

Histology reveals hyalinised fibrous connective tissue with chronic inflammatory cells and calcium deposition.[10]

As this condition is very rare, the treatment reports are based mostly on single case reports or a few case series. The initial management involves switching the patient to haemodialysis if they are on peritoneal dialysis. It will also involve treating the probable cause of the EPS. Management is usually supportive and involves resting the bowel with nasogastric aspiration. During this time nutrition is usually provided via total parenteral nutrition.

Steroids, because of their anti-inflammatory properties are commonly used with some success. They can be used as monotherapy but many investigators have used combined therapies with other immunosuppressants such as cyclosporine, azathioprine and mycophenolate.[1,11] A recent report of three cases has shown benefit from a combination of mycophenolate and steroids.[11]

Tamoxifen, in a dose of 20 mg, because of its anti-fibrotic properties, has been used in some cases with success.[12]

Surgery has previously been associated with poor outcome although some case reports of very good outcomes from surgery have been published. These cases have had meticulous adhesiolysis without enterostomy.[13]

There are currently other treatments that are under investigations e.g. intraperitoneal angiotensin converting inhibitors, angiotensin receptor blockers and COX-2 inhibitors and low glucose degradation product peritoneal fluids.[6]

We have presented a patient who developed encapsulating peritoneal sclerosis after having had 10 years of exposure to peritoneal dialysis fluid. During this time he had two episodes of culture proven bacterial peritonitis. He had also received beta-blockers for 32 years for treatment of his hypertension. Any of these factors may have increased his risk for developing EPS and the presence of multiple factors may have increased his risk further.

EPS, although rare, is a condition that has high mortality. More research is needed in understanding its pathophysiology and treatment options.

Author information: Caroline L Chembo, Advanced Trainee Nephrology, Renal Department, Palmerston North Hospital, MidCentral DHB, Palmerston North; Alastair Macdonald, Consultant Nephrologist, Renal Department Wellington Hospital, Wellington; Nicola Hay, Consultant Nephrologist, Renal Department Wellington Hospital, Wellington, Philip Matheson, Consultant Nephrologist, Renal Department Wellington Hospital, Wellington; Grant Pidgeon, Consultant Nephrologist, Renal Department Wellington Hospital, Wellington; Murray Leikis, Consultant Nephrologist, Renal Department Wellington Hospital, Wellington

Correspondence: Dr Caroline L Chembo, Palmerston North Hospital, MidCentral DHB, Ruahine Street, Palmerston North, New Zealand. Fax +64 (0)6 3508058; email: chemboc@doctors.net.uk

References

1. Kawanishi H, Kawaguchi Y, Fukui H, et al. Encapsulating peritoneal sclerosis in Japan: a prospective, controlled multicenter study. Am-J-Kidney-Dis. 2004;44:729-37.

2. Rigby RJ, Hawley CM. Sclerosing peritonitis The experience in Australia. Nephrol Dial Transplant. 1998;13:154-59.
3. Cancarini GC, Sandrini M, Vizzardi V, et al. Clinical aspects of peritoneal sclerosis. J Nephrol. 2001;14:(suppl 4) S39-47.
4. Krediet RT, Zweers MM, Van Westrhenen MM, et al. What can we do to preserve the peritoneum? Perit Dial Int. 2003;23:(suppl 2) S14-9.
5. Pecoits-Filho R, Araujo MR, Lindholm B, et al. Plasma and dialysate IL-6 and VEGF concentrations are associated with high peritoneal solute transfer rate. Nephrol. Dial Transplant. 2002;17:1480-6.
6. Jing S, Kezhou Y, Hong Z, et al. Effect of renin–angiotension system inhibitors on prevention of peritoneal fibrosis in peritoneal dialysis patients. Nephrology. 2010;15:27-32.
7. Hendriks MP, de Sévaux RGL, Hilbrands LB. Encapsulating peritoneal sclerosis in patients on peritoneal dialysis. The Netherlands Journal of Medicine. 2008; 7 July-August,.
8. Kawanishi H, Moriishi M. Encapsulating peritoneal sclerosis: prevention and treatment. suppl 2 S289-92, 2007, Vol. 27.
9. Tarzi RM, Frank JW, Ahmad, S, et al. Fluorodeoxyglucose positron emission tomography detectec the inflammatory phase of sclerosing peritonitis. Perit. Dial. Int. 2006;26:224-240.
10. Summers A, Clancy MJ, Syed F, et al. Single-center experience of encapsulating peritoneal sclerosis in patients on peritoneal dialysis for end-stage renal failure. Kidney International. 2005;68:2381-2388.
11. Lafrance J-P, Létourneau I, Ouimet D, et al. Successful Treatment of Encapsulating Peritoneal Sclerosis with Immunosuppresive Therapy. American Journal of Kidney Diseases. 2008;51(2):e7-e10.
12. Allaria PM, Giangrande A, Gandini E, Pisoni IB. Continous ambulatory peritoneal dialysis and sclerosing peritonitis; tamoxifen as new therapeutic agent? J Nephrol. 1999;12:395-7
13. Kawanishi H, Harada Y, Noriyuki T, et al. Treatment options for encapsulating peritoneal sclerosis based on progressive stage. Adv Perit Dial. 2001;17:200-4.