

中文標題：一位 65 歲長期腹膜透析的女性患有包囊性腹膜硬化症

英文標題：Encapsulating Peritoneal Sclerosis in a 65-Year-Old Woman under Peritoneal Dialysis

作者：黃柏諭¹、蔡任弼²

服務單位：¹大林慈濟醫院內科部；²大林慈濟醫院腎臟內科

Introduction

Encapsulating peritoneal sclerosis (EPS) is an uncommon but serious condition of peritoneal dialysis (PD), and its prevalence and incidence vary among centers and countries. The most significant risk factor is the duration on PD. The diagnosis of EPS requires both clinical features and evidence of encapsulation of bowel. The management of EPS includes supportive care (cessation of PD, nutrition support), medical therapy (immunosuppressants and antifibrotics) and surgical enterolysis [1]; however, there was limited evidence upon optimal dosing or duration for medical treatment. Here we reported a 65-year-old woman presenting with EPS which occurred about 13 years after initiation of PD.

Case Report

A 65-year-old female patient presented with insidious-onset, progressive abdominal fullness, anorexia, malnutrition and weight loss in January 2020. She had history of hypertension and end-stage renal disease, and started on peritoneal dialysis in 2007. Her stage IA endometrial cancer discovered in 2014 was managed with operation, and total parathyroidectomy with autoimplantation was done in 2017 due to renal hyperparathyroidism. The abdominal CT scans in April 2020 revealed calcification and thickening of peritoneum surrounding the small intestines (figure 1). She suffered no episodes of peritonitis previously. Her renal replacement modality was switched to hemodialysis, and she was treated with oral prednisolone 5 mg per day and tamoxifen 10 mg per day. There was gradual improvement in gastrointestinal symptoms after a 3-month treatment, and follow-up CT scans showed stationary changes regarding the peritoneum.

Discussion

The prevalence of encapsulating peritoneal sclerosis (EPS) among patients on peritoneal dialysis (PD) in Taiwan was about 1.8% [2]; around the world the prevalence based on observational cohort studies was 0.4-8.9% [3]. The "two-hit theory" explains the pathogenesis of EPS. The first hit is chronic inflammation and damage of peritoneal membrane by uremic toxins, glucose degradation products, low dialysate pH, among others; the second factor was associated with infection, autoimmunity, or neoplasms. In this case, the possible predisposing factors for EPS included long period of PD and abdominal malignancy and surgery.

It is challenging to treat patients with EPS. Transforming growth factor beta (TGF- β) plays a role in epithelial-mesenchymal transition of peritoneum, and tamoxifen induces TGF- β 1 suppression of fibroblasts [4]. However, the level of evidence about efficacy of tamoxifen is low, and the optimal dosing and treatment duration is unknown. A number of novel therapies for EPS have been mentioned [5], including colchicine, thalidomide, dissolved molecular hydrogen, and stem cell

treatment. To the best of our knowledge, currently there are no ongoing large-scale randomized trials about these treatments.

References

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Figure 1. (A) (B) Abdominal CT scans showing calcification and thickening of peritoneum along the bowel wall.

(A)



(B)

