Management of Scleroderma-related End-stage Renal Disease With Automated Peritoneal Dialysis

Malleshappa Pavan, Ashwinikumar Aiyangar, Anup Chaudhari, Prashanth Rajputh, Ravi Ranganath, Shah Arunkumar Babulal

Department of Nephrology, Lilavati Hospital and Research Centre, Mumbai, India

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INTRODUCTION

Kidney failure in scleroderma is a serious complication and a major cause of death. Hemodialysis, peritoneal dialysis, and kidney transplantation are established treatment options for end-stage renal disease (ESRD).1 Difficulty in creating a vascular access makes hemodialysis a difficult option for patients with scleroderma. Survival up to 18 months has been reported in 1 patient on continuous ambulatory peritoneal dialysis (PD). We describe our experience with ESRD in a patient with scleroderma, who had difficulty in making a vascular access and was treated with automated PD.

CASE REPORT

A 62-year-old woman with stage 2 chronic kidney disease due to sclerodermal kidney disease presented with sudden-onset breathlessness and oliguria following ingestion of ayurvedic medication for her skin ailment. This admission brought her in uremia with fluid overload, needing initiation of dialysis. Hemodialysis was initiated, but she repeatedly developed intradialytic hypotension and recurrent pulmonary edema after hemodialysis settings. Arteriovenous fistula was deferred in view of severe narrowing of upper limb veins on Doppler ultrasonography. Also, the permanent catheter insertion was deferred in view of her severely taught skin. Decision to insert a PD catheter was made as she had difficulty in obtaining a vascular access.

In spite of several case studies reporting poor outcomes of scleroderma on PD, we had no other option but to initiate PD. She was initially advised 3 daytime exchanges (2 L of 2.5% dextrose solution) and 1 overnight dwell of 2 L of 2.5% solution. She used to receive an ultrafiltration volume of 900 mL/d. Peritoneal equilibration test done at the end of 6 weeks revealed a high-average transporter status. Her weight was 40 kg, and she complained of frequent abdominal discomfort and nausea with 2000 mL of PD fluid.

Since then, she was on automated PD prescription with 5 cycles of 1600 mL of 1.5% solution with a therapy time of 8 hours. With this prescription, she received an ultrafiltration volume of 900 mL/d. Her Kt/V was 2.08/wk and creatinine clearance was 57 L/wk. She was having a minimal sieving effect of the peritoneal membrane on sodium and her serum sodium was around 135 mEq/L (Table).

In spite of being a high-average transporter, she was having minimum protein leakage and her
Blood pressure was 110/70 mm Hg with a good preservation of residual kidney function. Also, there was a marked improvement in her left ventricular ejection fraction from 25% to 50% on echocardiography along with subjective loosening of her skin. Over the past 18 months, her morale was high and there were no psychological changes while on this regimen.

**DISCUSSION**

Management of scleroderma-related ESRD requires some form of renal replacement therapy. Hemodialysis may be complicated by the difficulty in fashioning and maintaining a suitable vascular access and the inherent hemodynamic instability that can be present in these patients. Like hemodialysis, continuous ambulatory PD also has some potentially adverse features in patients with scleroderma. These include sclerosis of the peritoneal membrane, diminished blood flow due to Raynaud phenomenon of the peritoneal vasculature, and exit site problems (poor healing and infection). Patients undergoing kidney transplantation may have a poorer outcome, given the gravity of their underlying condition compared with other groups.

The role of PD in scleroderma patients with ESRD has not been fully evaluated because of the paucity of cases. Robson and colleagues reported that the decrease in peritoneal clearance was minimal and that control of uremia was satisfactory in 2 scleroderma patients studied for up to 10 months.

Supporting this, our patient also showed a minimal decrease in peritoneal clearance, with good control of uremia at 18 months of follow-up. In summary, automated PD helped maintain a reasonable quality of life in a scleroderma patient and could be continued for many months. Automated PD even may be preferable to kidney transplantation in certain patients with ongoing multisystem disease. Further studies are required in this regard to use automated PD as a treatment modality of choice in scleroderma patients with ESRD.

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**CONFLICT OF INTEREST**

None declared.

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Correspondence to:
Malleshappa Pavan, MD
Department of Nephrology, A-791, Lilavati Hospital and Research Centre, Bandra Reclamation, Bandra West, Mumbai – 400050, Maharashtra, India
Tel: +91 22 2655 2222, ext: 2628
Fax: +91 22 2640 7655
E-mail: dr_pavanm@yahoo.co.in

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