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Successful peritoneal dialysis application in a patient with catastrophic Behçet's disease

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Abstract

Behçet's disease (BD) is a type of systemic inflammatory vasculitis characterized by oral and genital ulcers, ocular lesions, skin lesions. Renal involvement is rare but may have serious consequences. In this report we present a patient diagnosed as BD 5 years ago and nowadays presented with a catastrophic course necessitating renal replacement therapy (RRT). We successfully applied acute peritoneal dialysis as RRT and, he relatively ameliorated.

Keywords: Henoch-Schonlein purpura, IgA nephropathy, migratory polyarthritis

Introduction

Behçet's disease (BD) is a vasculitic syndrome with systemic involvement such as oral-genital ulcers, ocular lesions, skin lesions, and some other manifestations such as renal pathologies. Immunity, genetic and environmental factors play role in the pathogenesis of BD. Although renal BD is not common, in some cases renal replacement therapy (RRT) may be necessary^[1]. Peritoneal dialysis is a feasible RRT option in BD patients considering their vascular involvement. In this paper we present a 41 year- old male patient diagnosed as Behçet's disease 5 years ago and nowadays presented with a catastrophic clinical course as multiple thrombosis, kidney failure and sepsis.

Case presentation

A 41-year-old man was transferred to our department of nephrology and intensive care with serious vascular, lung, skin and kidney involvement of BD. He had been diagnosed as BD 5 years ago. We learned from his relatives that he had irregular use of colchicine, steroids and anti-metabolite agents in his medical history. When he admitted to our hospital, his blood pressure was 95/50 mmHg, body temperature was 36.2°C, pulse rate was 80 beats per minute. On physical examination he was intubated, breath sounds were decreased and there were occasional rales in the lung parenchyma. Diffuse edema and, skin lesions secondary to circulatory disorder and venous pathologies were detected in the lower extremities. His family history was negative for BD and renal diseases as well. Laboratory values were as follows: urea: 353 mg/dl, creatinine: 6.2 mg/dl, uric acid: 10.1 mg/dl, pH: 7.1, HCO₃: 17 mmol/l, pCO₂: 29.6 mmHg, lactate 4 mmol/l, CRP> 200 mg/l.

Based on these clinical and laboratory features, the patient was diagnosed as catastrophic BD with systemic involvement and sepsis. Treatment with parenteral antibiotherapy and inotropic agents were started. Because of insufficient urine output, RRT was planned. However, dialysis catheter insertion efforts were failed due to disseminated venous thrombosis. Therefore we inserted a percutaneous Tenckhoff PD catheter and started continuous ambulatory peritoneal dialysis with low amount of dialysate. Four- 5 cycles/ day with 1- 1.5 liters of % 1.36 glucose containing PD solution were executed and 300- 400 ml ultrafiltration volumes were achieved. With successful acute PD application, his pulmonary and peripheral hypervolemic state was resolved and laboratory values were also started to ameliorate:

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Fig 1: Lower extremity skin lesions and PD application of the patient.

Discussion

Behçet's disease is a vasculitis syndrome with multisystemic involvement. In 1937 Dr. Behçet described the co-occurrence of recurrent oral ulcers, genital ulcers and uveitis as BD. In addition to oral, genital aphtha and uveitis, neurologic disease, arterial vasculitis/aneurysms, venous thrombosis, pulmonary disease, arthritis of lower extremity joints, gastrointestinal involvement and skin lesions such as erythema nodosum, acneiform nodules are other systemic presentations. The severity is generally greater in men. In patients with possible genetic predisposition, chemicals, metals and infections trigger abnormal immune activity which causes the disease. The laboratory tests are not pathognomonic in BD. Inflammation markers such as C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) and ferritin, white blood cells, monocyte-lymphocyte ratio, neutrophil-lymphocyte ratio, and platelet-lymphocyte ratio may elevate in active disease. Treatment aims to protect organs and systems as well as to control disease activity. For this purpose, corticosteroids, colchicine and some other immunosuppressive/immune modulatory agents are used. Clinical presentation of renal BD may be as hematuria, proteinuria and end stage renal disease (ESRD) as result of glomerular/vascular involvement or AA amyloidosis [2,3]. Although ESRD is not common in BD, RRT may be necessary in case of serious renal insufficiency. In haemodialysis, it is necessary to create a vascular access such as arteriovenous fistula or dialysis catheter. However these ways are likely to thrombose in Behçet's patients. In this case PD is feasible RRT modality. On the other hand, in case of transplantation, clotting of the transplanted renal blood vessels is an important problem [4,5]. In our case catastrophic BD with lung, skin, kidney and vascular involvement necessitated intensive care, inotropic and antibiotic treatment and RRT. Due to multiple venous thrombosis, HD catheter insertion efforts were failed and after insertion of PD catheter we executed acute PD successfully. Uremic toxins in the circulating blood pass to dialysate that filled in the peritoneal cavity, in a PD experience. Peritoneal membrane is used as a dialyser in PD physiology [6].

Conclusion

In cases of serious renal failure due to BD that necessitates RRT, PD may be logic, feasible and safe RRT option.

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