PERITONEAL DIALYSIS - 2

MP518  DISCONTINUATION OF PERITONEAL DIALYSIS (PD) AFTER LATE INITIATION OF ECULIZUMAB IN A PATIENT WITH FAMILIAL ATYPICAL HEMOLYTIC UREMIC SYNDROME (aHUS): A CASE REPORT

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Introduction and Aims: aHUS is a rare, genetically-related disease, in which complement-mediated thrombotic microangiopathy leads to chronic renal insufficiency. Its high mortality rate presents both a challenge and a dilemma for clinicians. A rapid identification of the disease and its etiology and initiation of therapy are crucial for successful management. Until recently, plasmapheresis (PPH) was the treatment of choice for aHUS, but eculizumab has shown promising efficacy both in de novo and dialysis patients. Here we present the case of an aHUS patient who started eculizumab treatment while on PD, 5 years after the debut of the disease.

Methods: A 33-year-old female was diagnosed with aHUS in July 2008. She had had no prior pregnancies and was not taking contraceptives or other regular medication. Due to rapid progressive deterioration of renal function, she underwent PPH and hemodialysis but showed no renal improvement. Genetic analysis identified a novel heterozygous mutation in complement factor H. In September 2008, she started nocturnal automated PD with 10 liters and wet day continuous cycling PD (CCPD) with 2 liters of extraneal. Dialysis was well managed and her residual renal function (RRF) was 3.2 mL/min with diuresis of 700 cc/day. The patient started eculizumab treatment in August 2013, at which time she was asymptomatic with good clinical status but her platelet count was low. She also needed five hypotensor drugs to control her hypertension.

Results: After 8.5 months of eculizumab treatment, the patient showed good tolerance of the drug and improved arterial hypertension. She was able to reduce the use of hypotensor drugs, and her PD exchange decreased by 50%. She stopped CCPD and started nocturnal intermittent PD with only 6 liters. Her diuresis was 2000 cc/day with RRF of 14 mL/min. Her platelet count, clinical status and quality of life also improved. In April 2015, PD was stopped. Nine months later, after 29 months of eculizumab treatment, her diuresis was 2500 cc/day, her creatinine count was 2.6 mg/dL, and her creatinine clearance was 38.7 mL/min.

Conclusions: Given the promising activity and good level of tolerance shown in our case and others, we recommend the use of eculizumab over standard PPH as first-line treatment, not only in newly diagnosed patients or after kidney transplantation but also in selected aHUS patients on dialysis.

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