Intermittent Recurrent Renal Failure: Diagnosing Atypical Hemolytic Uremic Syndrome

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Introduction

Hemolytic uremic syndrome (HUS) is characterized by the clinical triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury. Atypical Hemolytic Uremic Syndrome (aHUS) refers to non-Shiga-toxin HUS and is a primary thrombotic microangiopathic disease due to chronic, uncontrolled activation of the complement system. Complement-mediated HUS is a relatively rare, life threatening disorder caused by mutations in the genes that encode complement proteins.

Case Report

A 68 year-old male with a past medical history of three previous hospitalizations for unexplained acute renal failure presented with fever, chills and a transient rash on his forearms. On admission his serum creatinine was 0.95mg/dl. He quickly developed anuric renal failure with a rapidly rising creatinine requiring initiation of hemodialysis. Peripheral smear demonstrated schistocytes. Labs revealed thrombocytopenia, anemia, and hypocomplementemia. A renal biopsy was performed which demonstrated features of membranoproliferative glomerulonephritis and thrombotic microangiopathy. Initial therapy included high dose steroids and the initiation of plasmapheresis, however he has since been started on Eclizumab.

Discussion

The diagnosis of complement-mediated HUS remains challenging and is based on the clinical presentation of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury, not meeting criteria for typical HUS and TTP.[1] Mutations in proteins that regulate the alternative complement pathway such as Factor H (CFH), membrane cofactor protein (MCP or CD46), and Factor I (IF) have been implicated in causing aHUS.[2] In the past, plasma exchange was first line treatment however Eculizumab, a humanized monoclonal antibody to C5, has now been shown to more effective resulting in decreased morbidity.[3]

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