Atypical Hemolytic Uremic Syndrome (aHUS) Successfully Treated With Eculizumab

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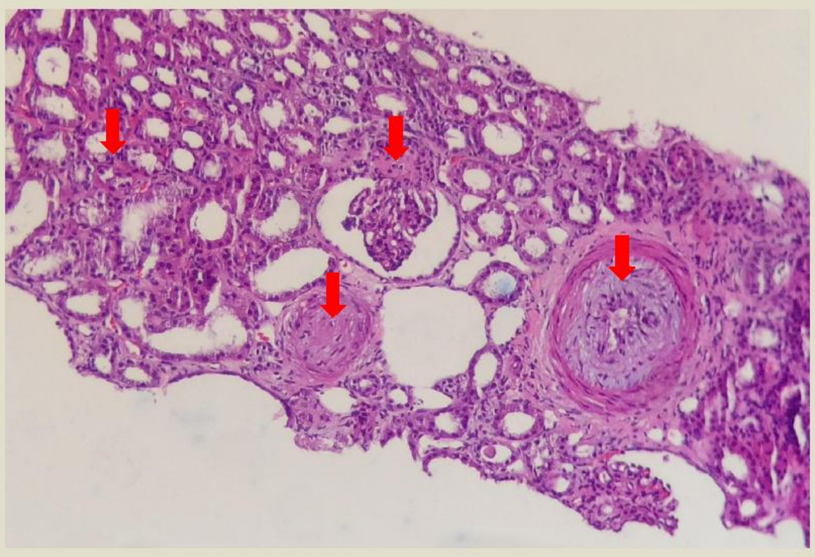
Introduction and Objectives

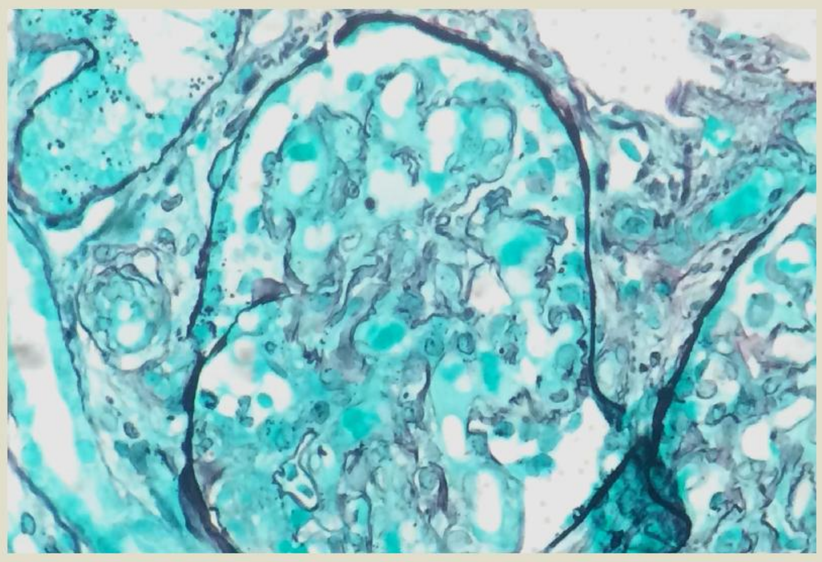
aHUS is a rare clinical syndrome characterized by hemolysis, thrombocytopenia, and acute kidney injury. In 90% of the patients, aHUS is triggered by enteric infections which are caused by the Shiga toxin–producing bacteria. The remaining aHUS cases are related to the dysregulation of the alternative pathway of the complement system. They are related to various triggers, such as infection by the human immunodeficiency virus, cancer, organ transplantation, pregnancy, use of certain anticancer drugs, immunotherapeutic agents and antiplatelet. The case below reports an aHUS with acute kidney injury (AKI) which promptly recovered the kidney function with eculizumab (Soliris®).

Methods

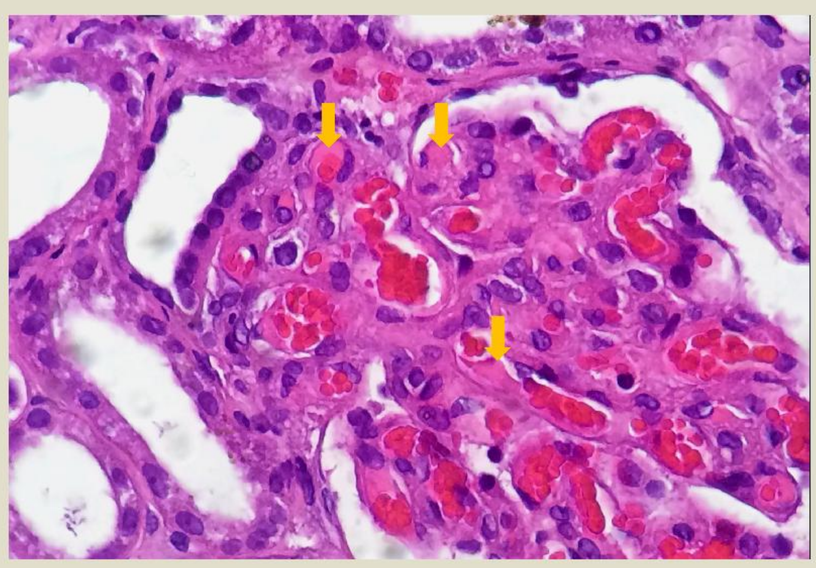
A 15 year-old boy was admitted with abdominal pain, edema and hematuria in October, 2014. After 1 week, he developed hypertension and oligoanuric AKI, hemolytic anemia, thrombocytopenia and increased LDH. Schizocytes were found in his blood smear, with high rates of reticulocytes, low C3 and haptoglobin. Direct Coombs, anticardiolipin antibody IgG and IgM, ANA, anti-DNAds and sorology for HBV, HCV and HIV were negative. ADAMTS 13 activity and C4 were normal. He underwent daily hemodialysis (HD). Treatment with eculizumab started 2 weeks after the first session of HD. He increased diuresis and recovered renal function not requiring HD a month after beginning the therapy. He underwent a 1-month induction therapy with eculizumab weekly and maintenance twice a month. Renal biopsy was done 3 weeks after admission.



Endothelial swelling and hyperplasia with narrowing of the lumen (red arrow)



Wrinkling of the capillary tuft (silver stain)

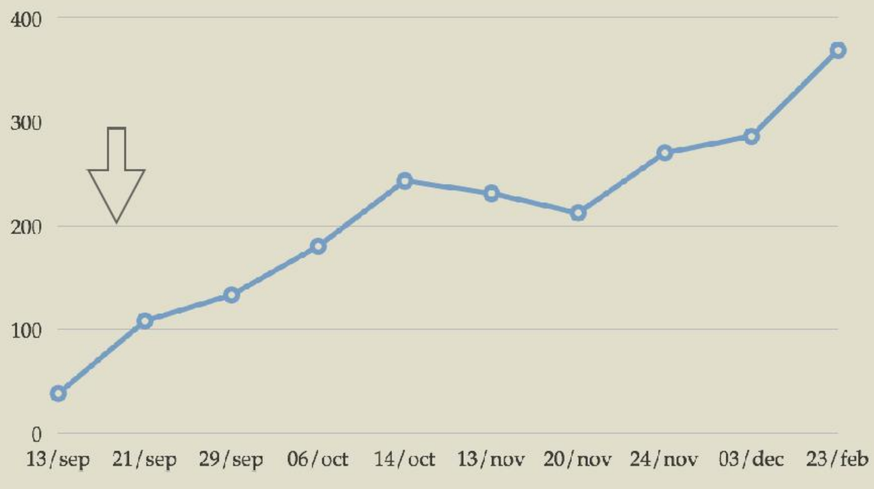


Capillary thrombosis, congestion, and thickening of the capillary tuft (yellow arrow)

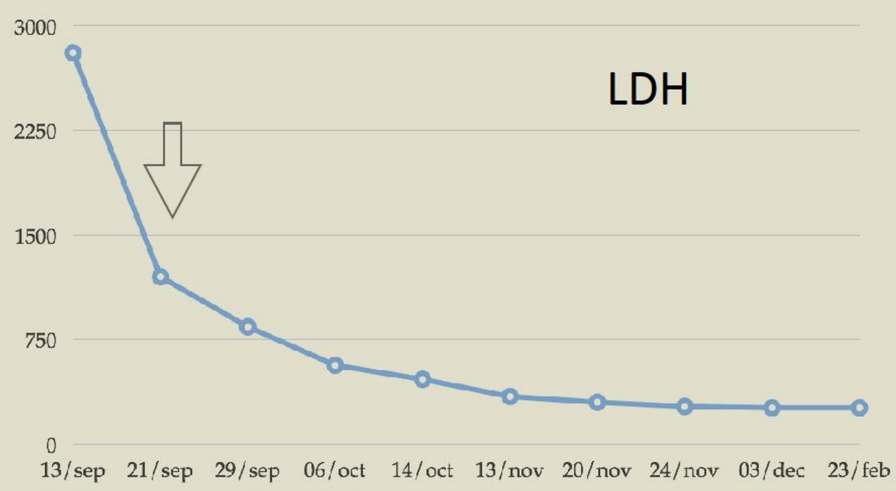
Results and Conclusions

He is still using eculizumab and, 4 months after the beginning of the treatment, his blood pressure was controlled with the use of two anti hypertensive medications. The patient is now asymptomatic, off dialysis and with normal renal function during follow-up.

Platelets



LDH



Unfortunately, aHUS has a poor prognosis, with death rates as high as 25% and progression to end-stage renal disease in half of the patients. Eculizumab is a monoclonal antibody that targets the complement's C5 to prevent the activation of the terminal membrane attack complex and thus slowing down the complement-mediated damage.
Considering the great risk of death by aHUS, as soon as it is highly suspected, the treatment with eculizumab should be considered

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H) Clinical Nephrology, primary and secondary glomerulonephritis.

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