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Administration of eculizumab in atypical hemolytic uremic syndrome: Two case reports

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Case Study

The timing of eculizumab administration, maintenance period, and discontinuation time are still not clearly established. We compared and analyzed two cases in which eculizumab was administered after as a diagnosis of atypical hemolytic uremic syndrome (aHUS).

A 60 years old male with history of diabetic nephropathy performed kidney transplantation (KT) from a living unrelated donor at our hospital in April 2019. At three weeks later from KT, his serum creatinine was elevated and other abnormal laboratory findings (pancytopenia, schistocytes, decreased haptoglobin, and increased LDH) were newly developed. We suspected thrombotic microangiopathy (TMA). There were no abnormal findings on renal biopsy and the gene mutation test was not confirmed. Eculizumab could not be administered immediately. After 4 months, Genetic examination confirmed CFB and CD46 mutations. The patient's other abnormal lab findings improved while administering eculizumab, but creatinine gradually worsened. While maintaining hemodialysis, eculizumab was administered for 2 years, but there was no improvement.

A 35-year-old female patient with no underlying disease underwent D&C surgery at a local hospital. She came to our hospital with a sudden lower abdominal pain and was diagnosed with thrombocytopenia, renal failure. Initially we suspected DIC, but both PT and PTT were identified as normal range. Her platelet counts and renal dysfunction gradually worsened, and schistocytes were identified on the PB smear test. ADAMTS 13, autoantibody tests were confirmed to be in the normal range and we were diagnosed with aHUS. She was received eculizumab twice at intervals of a week. Most laboratory findings were rapidly improved after use of eclizumab. The additional eculizumab could not be administered due to insurance problem, but fortunately hemodialysis has stopped and serum creatinine levels, platelet count are gradually improving.

When comparing the two cases, clinicians should be able to consider aHUS in these situations and make quick decisions about treatment.