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Atypical hemolytic uremic syndrome : A Case report

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Case Study: Atypical hemolytic uremic syndrome (aHUS) is a life-threatening condition of thrombotic microangiopathy (TMA), caused by an uncontrolled complement dysregulation. In spite of its fatality however, aHUS is often masked by other complement activating conditions, making its diagnosis confounding. In this report, we present a case of patient with recurred TMA, finally diagnosed as aHUS, which is very rare.

On May of 2019, a 55-year-old woman without any medical history, was presented with general weakness. An initial laboratory test showed hemoglobin level of 5.2g/dL, with numerous schistocytes, high LDH level(1155U/L) and undetectable haptoglobin(<10mg/dl), indicating hemolytic anemia; the platelet count was low, with $44x10^6$/L. Along with massive proteinuria(over 10g/day), acute kidney injury was evident with serum creatinine level of 2.47mg/dL, whose level peaked to 3.53mg/dL. Other tests to exclude STEC-HUS, autoimmune disease, DIC, infection or malignancy were all negative. A renal biopsy was performed, which revealed 'thrombotic microangiopathy'. Therapeutic plasma exchange (TPE) was initiated soon after admission, but was discontinued after ADAMTS-13 activity was determined to be 49%. She was discharged after 7 times of daily TPE with hematologic and minimal renal improvement (Fig.1). Thus, she regularly returned to clinic to monitor her renal function. After a year later, TMA recurred. She developed thrombocytopenia $90x10^6$/L, microangiopathic hemolytic anemia 9.9g/dL; increased LDH level 370U/L ; undetectable haptoglobin(<10mg/dl), and acute kidney injury with serum creatinine level of 3.63mg/dL. No other complement activating condition was identified, and stool toxin assay as well as coagulation profile all turned out to be normal, with ADAMTS-13 activity of 94%, highly implicating atypical HUS. The patient was vaccinated against meningococcus with prophylactic antibiotics and Eculizumab therapy was initiated. Genetic test result to confirm aHUS are pending.

We report a case of eculizumab administration after diagnosing a recurrent TMA patient with nephrotic range proteinuria and acute kidney injury as aHUS.

hematologic parameters and renal function change in a patient with aHUS