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COMPLEMENT-MEDIATED THROMBOTIC MICROANGIOPATHY IN A PATIENT WITH ANTI-SYNTHETASE SYNDROME - AN EXCEEDINGLY RARE ASSOCIATION

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BACKGROUND

Complement-mediated thrombotic microangiopathy (CM-TMA), previously denominated atypical hemolytic uremic syndrome, is a rare entity characterized by microvascular thrombosis associated with complement dearrangement, not completely understood. Classical clinical scenario comprises a life-threatening triad of microangiopathic hemolytic anemia (MAHA), thrombocytopenia and organ failure (typically renal failure) due to microvascular occlusions by platelet thrombi.

We present the first case to our knowledge of a spontaneous CM-TMA in a patient with anti-synthetase syndrome (AS)

CASE REPORT

A 76-year female with a medical past of hypertension and AS diagnosed in 2017's september, treating with cyclophosphamide due to pulmonary hypertension and non-specific interstitial pneumonia, searched our outpatient clinic because of worsening of shortness of breath and muscular weakness with elevated creatinokinase (CK) of >3000, when high-dose steroid therapy was initiated. She had been admitted in other hospital due to partial response, being detected with thrombocytopenia. She was discharged after one week without case elucidation, returning to our outpatient clinic informing reduced urine output.

Laboratory findings revealed severe acute kidney injury (creatinine 5,4 mg/dL from a basal value of 0,8 mg/dL), thrombocytopenia (17000 platelets) microscopic hematuria, direct antiglobulin test negative autoimmune hemolytic anemia (reticulocytosis, elevated indirect bilirrubin (IB) and lactate dehydrogenase (LDH) with low haptoglobin level) and >5% schizocytes in peripheral blood smear.

A rapidly progressive glomerulonephritis due to TMA diagnosis was suspected and ADAMTS13 activity result were normal. Plasmapheresis and hemodialysis were initiated with partial response (creatinine decreased to 2 mg/dL, platelets raised to 180000, reticulocyte count, IB, LDH, haptoglobin and schizocytes normalized). A kidney biopsy could not be performed because of respiratory discomfort due to positioning.

Mycophenolate mofetil was introduced and ecolizumab was requested, but kidney function did not recover.

After two months, our patient deceased from an at home cardiac arrest.

CONCLUSION

After MEDLINE database search, we found only one report of TMA associated with AS, which initiated only after steroid treatment (1mg/kg).

The pathogenesis underlying the relationship between AS and CM-TMA remain obscure. Thanks to elevated morbidity and mortality, TMA should be in the differential diagnosis of a AS patient that presents with cytopenias and acute kidney injury.