

Abstract Submission No. : 2398

Eculizumab therapy on a patient with co-existent lupus nephritis and C3 mutation-related atypical haemolytic uremic syndrome: a case report

Mi Jung Kim¹, Haekyung Lee², Yon Hee Kim³, So Young Jin³, Jin Seok Jeon²

¹Department of Internal Medicine-Nephrology, Asan Yuri Hospital, Korea, Republic of

²Department of Internal Medicine-Nephrology, Soonchunhyang University College of Medicine, Korea, Republic of

³Department of Pathology, Soonchunhyang University College of Medicine, Korea, Republic of

Case Study: Thrombotic microangiopathy (TMA), a rare but serious complication of systemic lupus erythematosus (SLE), is associated with poor outcomes to conventional immunosuppressive therapy. Recently, eculizumab, a humanised monoclonal antibody that blocks the complement factor 5, has been known to effectively treat atypical haemolytic uremic syndrome (aHUS). Here, we report a case of aHUS co-existing with lupus nephritis that was successfully treated with eculizumab.

A 23-year-old man presented with abdominal pain and diarrhoea. Initial laboratory tests have shown thrombocytopenia, microangiopathic haemolytic anaemia, and acute kidney injury. Immunologic tests were consistent with SLE. Kidney biopsy have revealed lupus nephritis class IV-G with TMA. Genetic analysis have shown complement C3 gene mutations, which hints the co-existence of lupus nephritis with aHUS, a form of complement-mediated TMA. Although initial treatment with haemodialysis, plasma exchange, and conventional immunosuppressive therapy (steroid and cyclophosphamide) did not appreciably improve kidney function and thrombocytopenia, the patient was able to respond to eculizumab therapy.

Due to the strikingly similar features of TMA and SLE, clinical suspicion of aHUS in patients with lupus nephritis is important for early diagnosis and prompt management. Timely administration of eculizumab should be considered as a treatment option for aHUS in lupus nephritis patients to yield optimal therapeutic outcomes.