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Atypical Hemolytic Uremic Syndrome Occurring During Anti-GBM Glomerulonephritis Treatment

Hyejin Jeon, Hyun-Jung Kim, Seunghye Lee, Sehyun Jung, Se-Ho Chang
Department of Internal Medicine-Nephrology, Gyeongsang National University Hospital, Korea, Republic of

Case Study : Anti-glomerular basement membrane (anti-GBM) glomerulonephritis is a unique autoimmune disease affecting the kidneys, causing acute renal damage. Atypical hemolytic uremic syndrome (aHUS), a life-threatening condition, is characterized by microangiopathic hemolytic anemia, thrombocytopenia, and renal injury due to complement system dysregulation. We report a rare case of aHUS occurring during anti-GBM glomerulonephritis treatment in South Korea. A 49-year-old woman presented with gross hematuria, headache, nausea, and vomiting. Initial findings were serum creatinine 6.68mg/dL, urine protein to creatinine ratio (mg/mg) 5.8, urine blood 3+, urine RBC >100, with 10 to 30% dysmorphic RBC, and positive anti-GBM antibody test. She received hemodialysis and a renal biopsy revealed anti-GBM glomerulonephritis. Despite treatment with steroids and cyclophosphamide, renal function deteriorated. The anti-GBM antibody titer was elevated to 85, prompting 5 sessions of plasmapheresis, which decreased to 19. Nine days after discharge with maintenance hemodialysis, she had abrupt pancytopenia with high LDH and schistocytes, suggesting thrombotic microangiopathy. ADAMTS13 activity was normal at 56.1%, and aHUS was suspected as there was no history of diarrhea or exposure to other drugs. Eculizumab was considered, but the HIRA rejected it as the eGFR decline was less than 20% from the previous value, and the ADAMTS13 test was conducted post-plasmapheresis. After 6 plasmapheresis sessions, the platelet count increased to 66,000/mm^3. Despite the improvement, the patient declined additional plasmapheresis. About 6 weeks later, the platelet count exceeded 100,000/mm^3, and schistocyte disappeared. Suspected lung cancer on the initial chest CT, she underwent RUL lobectomy after 6 months; it was adenocarcinoma. Genetic test did not reveal an apparent genetic mutation of the aHUS. Some reports of anti-GBM disease with TMA showed that improvement typically occurs within 12weeks and our case aligns with this observation. We believe that rapid diagnosis may be useful to establishing the appropriate therapy and prognosis in these cases