Recurrent Atypical Hemolytic Uremic Syndrome in a Kidney Transplantation Recipient

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Background: Atypical hemolytic uremic syndrome (aHUS) is a rare disorder characterized by dysregulation of the complement pathway, presenting thrombotic microangiopathy (TMA), leading to end-stage renal disease (ESRD). Importantly, kidney transplantation (KT) in patients with a genetic predisposition to aHUS brings about recurrent disease leading to graft loss. Here we describe a kidney transplant recipient who developed recurrent hemolytic uremic syndrome one month after KT.

Case Report: A 45-aged male kidney transplant recipient was admitted due to rapid deterioration of graft function after 1 month from KT. Serum creatinine level was elevated from 1.43 mg/dl to 2.72mg/dl within 2 weeks. Hemolytic anemia (serum Hb 6.8g/dl), thrombocytopenia (platelet 72 x 109/L), elevated LDH (458mg/dl), depleted haptoglobin (<10mg/dl), and microangiopathic hemolytic anemia (MAHA) were found, suggesting TMA. He kept on deflazacort, tacrolimus, and mycophenolate mofetil, so tacrolimus was switched to sirolimus considering drug-induced TMA. However, there was no improvement. Shiga toxin was negative and ADAMTS13 activity was 63.6%. Plasmapheresis was undergone 7 times, but there was no clinical response for TMA. Graft biopsy pathology was reported as postinfectious glomerulonephritis, which was inconsistent with the clinical feature. Therefore, we reviewed his chart retrospectively. On his first visit to the hospital 9-years ago, he had malignant hypertension with severely decreased renal function (serum Cr 23.35mg/dl), anemia (Hb 5.7g/dl), elevated LDL (304mg/dl), and decreased complement 3 (56mg/dl). And most of all, the native kidney biopsy revealed TMA, but we did not evaluate for underlying other causes at that time. Therefore, we performed a genetic study for aHUS, and the next-generation sequence (NGS) revealed heterozygous mutation for complement factor I (CFI) (c.119A>C), which is pathogenic for aHUS, so recurrent aHUS after KT was confirmed.

Discussion: This report emphasizes the importance of clarifying primary renal disease of kidney transplant candidates with TMA before KT, especially screening for genetic study for aHUS, not to induce recurrence of aHUS after KT.

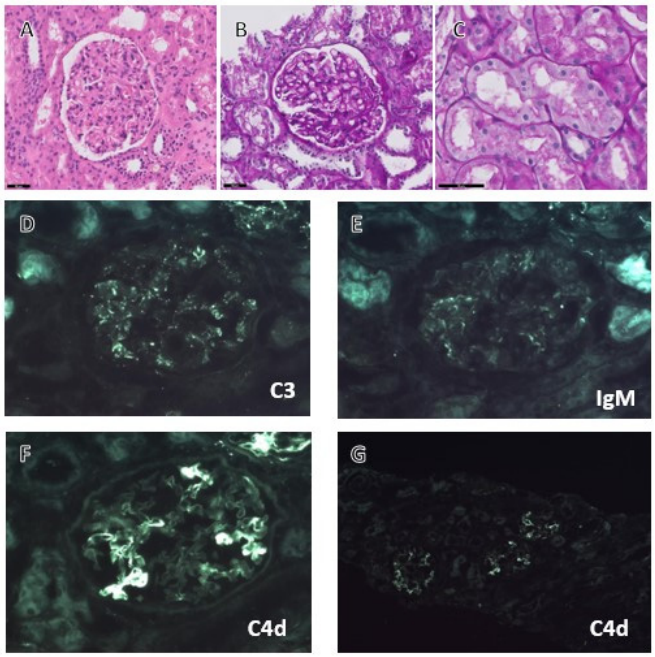

Figure 1. Microscopy and immunofluorescence of kidney biopsy in case patient. Hypercellularity of the glomeruli (A, B) with vacuolation of tubules is shown (C). An immunofluorescence study showed diffuse mesangial C3 (D), IgM (E), and C4d (F, G) deposits in glomeruli. Such findings are consistent with an early finding for TMA recurrence, regarding the clinical index.