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Interesting AKI : Is atypical HUS a rare disease? case and literature review

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Primary atypical hemolytic uremic syndrome (aHUS) is caused by complement pathway dysregulation. Although there are few reports of aHUS in Korea, most patients were children; adult-onset aHUS confirmed by genetic testing is rarely reported yet. We recently treated several patients with adult-onset aHUS with mutations of the complement regulatory genes including factor H (CFH). In the first case, uncontrolled hypertension developed with microangiopathic hemolytic anemia and renal failure, which then progressed to ESRD. In the second case, upper respiratory infection occurred followed by renal failure with clinical features of aHUS. Renal function was recovered after treatment with plasmapheresis. In the third case, patient had recurred aHUS after 2nd kidney transplantation and finally genetic analysis revealed the gene mutation. Despite similar gene mutation, triggering factors, clinical presentations, outcomes, and treatments were variable. Even though aHUS is a rare disease, clinicians need to perform a genetic analysis in patients with ESRD of unknown origin, especially when considering renal transplantation.