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Atypical hemolytic uremic syndrome in South Korea

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Thrombotic microangiopathy (TMA) is defined by specific clinical characteristics, including microangiopathic hemolytic anemia, thrombocytopenia, and pathologic evidence of endothelial cell damage, as well as the resulting ischemic end-organ injuries. A variety of clinical scenarios have features of TMA, including infection, pregnancy, malignancy, autoimmune disease, and medications. These overlapping manifestations hamper differential diagnosis of the underlying pathogenesis, despite recent advances in understanding the mechanisms of several types of TMA syndrome. Atypical hemolytic uremic syndrome (aHUS) is caused by a genetic or acquired defect in regulation of the alternative complement pathway. It is important to consider the possibility of aHUS in all patients who exhibit TMA with triggering conditions because of the incomplete genetic penetrance of aHUS. Therapeutic strategies for aHUS are based on functional restoration of the complement system. Eculizumab, a monoclonal antibody against the terminal complement component 5 inhibitor, yields good outcomes that include prevention of organ damage and premature death. However, there remain unresolved challenges in terms of treatment duration, cost, and infectious complications. After the introduction of eculizumab in the treatment of aHUS in Korea, about 90 patients who were suspicious of aHUS and were applied to KFDA's approval for using the novel drug, although only about one-third of them were approved ultimately. Recently, South Korean experts, including hematologists, adult and pediatric nephrologists, transplantation surgeons, pathologists, and genetic laboratory medicine specialists have published a consensus report regarding the diagnosis and management of TMA syndromes. Sharing the consensus report and recent cases receiving KFDA's approval to eculizumab treatment may be helpful to reflect on the last years and to find out the future direction on the diagnosis and management of Korean aHUS.