

aTypical HUS: Where do we Stand in Taiwan

Mai Szu-WU M.D., PhD.

Division of Nephrology, Taipei Medical University Hospital, Taipei, Taiwan;
Department of Internal Medicine, School of Medicine, Taipei Medical University, Taipei, Taiwan.

Abstract

Atypical hemolytic uremic syndrome (aHUS) is a genetic, life-threatening, systemic disease. When unrecognized or inappropriately treated, aHUS has a high degree of morbidity and mortality. aHUS results from chronic, uncontrolled activity of the alternative complement pathway, which damages the endothelium, activates platelets, and leads to organ damage. aHUS is clinically very similar to the other major TMAs: Shiga toxin-producing *Escherichia coli* (STEC)-HUS, thrombotic thrombocytopenic purpura (TTP), and disseminated intravascular coagulation (DIC). The signs and symptoms of all the TMAs overlap, complicating the differential diagnosis.

Nowadays, eculizumab, a terminal complement inhibitor is the only approved treatment for aHUS, were established in 4 prospective, multicenter clinical studies either in pediatric or adult populations. This year, eculizumab has got national funding in Taiwan since July. Here Dr. Wu will present the current clinical practices and moving forward the approach for advanced aHUS diagnosis and management under Taiwan health care system.