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Perspective

Mechanisms and historical prognosis of atypical hemolytic uremic syndrome

Marina Joseph*

Department of Medicine and Nephrology, Taranaki Base Hospital, 8 David Street Westown, New Plymouth Taranaki, New Zealand.

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ABOUT THE STUDY

The highly rare, fatal, and progressive condition known as Atypical Hemolytic Uremic Syndrome (AHUS) frequently involves a genetic component. In most circumstances, stopping the complement cascade can successfully control it. Certain monoclonal antibodies, which are covered in more detail later in the article, have been shown to be effective. Kidney function is the main organ affected by the condition known as atypical hemolytic-uremic syndrome. This disorder results in the development of aberrant blood clots, which can happen at any age, in the tiny blood arteries of the kidneys. If these clots restrict or obstruct blood flow, they may result in major medical issues. Hemolytic anaemia, thrombocytopenia, and renal failure are the three main characteristics of atypical hemolytic-uremic syndrome that are linked to aberrant coagulation.

Usually, persistent, unchecked activation of the complement system, a portion of the body's immune system that eliminates and destroys foreign objects causes AHUS. The condition, which affects both children and adults, is characterised by systemic Thrombotic Micro Angiopathy (TMA), which causes blood clots to form in tiny blood vessels all over the body and can cause fatalities such heart attacks, strokes, and renal failure. Occasionally, acquired neutralising autoantibodies inhibitors of these complement system components, such as anti-factor H antibodies, or changes in the complement regulatory proteins might cause the complement system to become activated. Before eculizumab and ravulizumab were available, it was thought that 33%-40% of patients with the first clinical bout of AHUS would pass away or develop End-Stage Renal Disease (ESRD). Despite Plasma Exchange or Plasma Infusion (PE/PI), almost two-thirds (65%) of patients passed away, needed dialysis, or sustained permanent renal impairment within the first year of diagnosis.

*Corresponding author. Marina Joseph, E-mail: Marina99@yahoo.com.

Mechanisms

The complement system is tightly controlled to prevent it from harming healthy tissues and organs. In healthy people, complement is employed to destroy foreign chemicals. However, it has been shown that in the majority of individuals with AHUS, development of anti-factor H autoantibodies or genetic abnormalities in any of numerous complement regulatory proteins can cause chronic, unchecked, and excessive activation of complement. This causes platelet activation, harm to the endothelium cells, and white blood cell activation. This results in systemic TMA, which presents as hemolysis, damage to many organs, a decreased platelet count, and frequently, death.

Historical prognosis

Before the use of monoclonal antibodies, patients with AHUS had a very bad prognosis. The percentage of individuals who suffered poor outcomes within the first year increased to 70% among those with the most prevalent AHUS genetic variant. Nevertheless, regardless of a mutation's status, abrupt morbidity and mortality are still possible. More than 40% of AHUS cases are discovered after the age of 18; however it can occur at any age. As a result, the majority of untreated AHUS patients experience ESRD and chronic dialysis, which are linked to serious morbidities and a worsening prognosis. Patients with AHUS have had combined liver-kidney transplantation, although this high-risk treatment has a death rate close to 50%.

Patients with AHUS had very poor quality of life before the treatments were available and used; they suffered from weariness, renal problems, hypertension, neurological impairment, gastrointestinal distress, clotting at the site of venous access, and in the worst cases, death. Due to the need for extensive vascular access and frequent administration, PE/PI is also said to be highly disruptive to patients' lives and to be associated with major safety hazards. Patients with AHUS now have much better prognoses thanks to eculizumab approval. After stopping eculizumab treatment, there is a risk of relapse, necessitating continuous monitoring.