Thrombotic thrombocytopenic purpura in 2022 - novel therapies and focus on long term outcomes

Shruti Chaturvedi

Johns Hopkins University School of Medicine, USA

Thrombotic thrombocytopenic purpura (TTP) is a rare thrombotic microangiopathy caused by ADAMTS13 deficiency. TTP used to be a universally fatal disorder; however, rapid diagnosis and treatment with plasma exchange and immunosuppression has improved survival of acute TTP episodes to > 90%. Recent insights into TTP pathogenesis have led to the development of novel therapies targeting pathogenic anti-ADAMTS13 antibody production, von Willebrand factor (VWF)-platelet interactions, and ADAMTS13 replacement. TTP is now understood as a chronic disease characterized by recurrent episodes of thrombotic microangiopathy. TTP survivors are also at increased risk for a number of long-term complications such as stroke, cognitive impairment, cardiovascular disease, poor quality of life and shortened survival. This talk will discuss current ideas on the pathophysiology, diagnosis, and management of TTP with a focus on targeted therapy therapies and emerging treatment paradigms. Strategies to optimize long term outcomes will also be discussed.