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THROMBOTIC MICROANGIOPATHIES

Thrombotic microangiopathies (TMAs) are a wide spectrum of diseases characterized by thrombocytopenia, microangiopathic hemolytic anemia and widespread ischemic damage due to microvascular thrombosis. TMAs could be life-threatening unless promptly recognized and treated, hence demand a rapid differential diagnosis and initiation of proper therapy, especially in the light of the new targeted therapies available nowadays. Thrombotic thrombocytopenic purpura (TTP) is a rare and severe primary TMA, caused by the congenital or acquired deficiency of ADAMTS13, the von Willebrand factor-cleaving protease. The severity of TTP, the significant recent developments in treatment and the heterogeneity of management worldwide, among other reasons, prompted the International Society of Thrombosis and Hemostasis (ISTH) to promote the draw up of evidence-based guidelines for the diagnosis and treatment of TTP. The new ISTH TTP guidelines were developed using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach, which provides a framework for rating quality of evidence and grading strength of recommendations in a structured, transparent and comprehensive way. Very recently, the Italian TTP guidelines have been adapted from the ISTH TTP guidelines by the Italian Society of Hematology (SIE). This lecture will provide a brief overview of TMAs and their differential diagnosis and summarize the epidemiology, pathogenesis, diagnosis and management of TTP, focusing on the new ISTH and SIE guidelines.