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THROMBOTIC MICROANGIOPATHIES: THROMBOTIC THROMBOCYTOPENIC PURPURA VS ATYPICAL HEMOLYTIC UREMIC SYNDROME

Thrombotic microangiopathies (TMAs) are rare and life-threatening diseases. In recent years, there have been major advances in understanding the pathophysiology, classification, and treatment of these disorders, including the introduction of new drugs. Early differential diagnosis is crucial for prompt treatment to reduce high mortality rates and late organ damage. The classic forms of thrombotic microangiopathy are thrombotic thrombocytopenic purpura (TTP), typical hemolytic uremic syndrome (HUS) associated with toxin-producing bacteria, atypical HUS (aHUS) due to dysregulation of alternative complement C3 convertase attributable to genetic causes, and HUS secondary to coexisting disease. The hallmark of TMA is the association of microangiopathic hemolytic anemia, detected by positive schistocytes in peripheral blood smears, with thrombocytopenia and organ changes, that may vary and be more or less pronounced depending on the TMA subtype. TTP is caused by an acquired or congenital deficiency of the von Willebrand factor-cleaving protease ADAMTS-13. Determination of ADAMTS13 activity and ADAMTS13 inhibitors are critical for differentiating TTP from other TMAs. This presentation will specifically focus on the differential diagnosis between TTP and aHUS, their clinical management and the different therapeutic approaches.