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The many faces of thrombotic microangiopathies

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The thrombotic microangiopathies (TMAs), are a complex group of disorders that typically present with a schistocytes hemolytic anemia and associated thrombocytopenia with ensuing microvascular occlusion leading to tissue ischemia and end organ damage. CNS, GI and cardiac microcirculations are frequent targets. Signs and symptoms related to organ dysfunction may evolve over weeks to months and may not be present simultaneously. LDH elevation due to microvascular ischemia is frequently disproportionate to elevation of bilirubin or reticulocyte count. The major thrombotic microangiopathies include TTP, DIC/sepsis and Hemolytic Uremic Syndrome. HUS may be further divided into "typical", related to Shiga toxin, "atypical", related to dysregulation or over-activation of complement and secondary, including disorders of pregnancy such as the HELLP syndrome or pre-eclampsia, certain other infections such as Strep Pneumoniae, auto-immune disorders such as Sjogren's syndrome, cancer, chemotherapy, or other medications, such as quinine and calcineurin inhibitors. These disorders can provoke direct microvascular damage and present as a thrombotic microangiopathy or act as a trigger for a microangiopathic syndrome in individuals with a genetic predisposition. The level of ADAM-TS 13, Von Willebrand Factor cleaving enzyme, is a key discriminator between TTP and HUS being severely reduced in TTP but not HUS. Plasma exchange with or without steroids is the mainstay of treatment for TTP. Anti C5 antibody therapy has evolved as an important treatment for a HUS. Although we have gained significant insight into the pathophysiology of many of these disorders, given the complex interplay between genetic factors, acquired factors, the roles of the humoral, cellular and innate immune systems, the inflammatory response and the coagulation system, TMAs remain clinically challenging. This review will focus on a summary of our current knowledge with regard to diagnosis and treatment of TTP and HUS and how they relate to each other and the broader family of TMAs. Three clinical cases will be used to illustrate key points.

Biography

James Granfortuna is a practicing Hematologist-Oncologist for 30 years. He is currently a full time Faculty Member at the Cone Health Internal Medicine Teaching program, affiliate hospital of the University of North Carolina Chapel Hill Medical Center. His special interests include platelet and clotting disorders in the general population and in pregnancy.

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