Pathogenesis in immune thrombocytopenia: new insights

Jill Johnsen

Abstract

Idiopathic (immune) thrombocytopenic purpura (ITP) is a common autoimmune disorder resulting in isolated thrombocytopenia. ITP can present either alone (primary) or in the setting of other conditions (secondary) such as infections or altered immune states. ITP is associated with a loss of tolerance to platelet antigens and a phenotype of accelerated platelet destruction and impaired platelet production. Although the etiology of ITP remains unknown, complex dysregulation of the immune system is observed in ITP patients. Antiplatelet antibodies mediate accelerated clearance from the circulation in large part via the reticuloendothelial (monocytic phagocytic) system. In addition, cellular immunity is perturbed and T-cell and cytokine profiles are significantly shifted toward a type 1 and Th17 proinflammatory immune response. Further clues into immune dysregulation in ITP may be gleaned from studies of secondary ITP. Some infections can induce antiplatelet Abs by molecular mimicry, and there may be common elements involved in breaking tolerance with other autoimmune disorders. There is also evidence for a genetic predisposition to both ITP and responsiveness to therapy, which may in part lie within immune-related genes. Lastly, treatment with immunomodulatory agents remains the mainstay of ITP therapies.
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