Thrombotic Thrombocytopenic Purpura (TTP)



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Inhibition or deficiency of ADAMTS 13
Inhibiting-chains on Broken ADAM and Friday the 13th Guy
ADAMTS13 is a protease that acts to cleave von Willebrand factor (vWF) into smaller monomers so they can be degraded. In TTP, the function of ADAMTS13 is decreased due to inhibition, deficiency or reduced activity. Defects can be hereditary or acquired with the formation of an ADAMTS13 autoantibody.

vWF Multimers are Not Degraded
Van Willy-brand
Due to decreased activity of ADAMTS13, vWF multimers are not degraded, and instead activate platelet adhesion. This leads to several downstream effects, including platelet consumption and activation of the complement system.

Microthrombi (and Emboli)
Micro-trombones
vWF multimers combine with platelets to form thrombi within the microvasculature of various organ systems. Formation of microthrombi consumes platelets and thrombocytopenia develops. These microthrombi may also cause mechanical shearing of red blood cells as they pass by, resulting in a condition called microangiopathic hemolytic anemia (MAHA).

Symptoms

FAT RN Pentad
Fat RN with (5) Hand
The pentad of symptoms seen in TTP can be remembered with the acronym "FAT RN". F is for fever, A is for anemia, and T is for thrombocytopenia. The R in RN is for renal dysfunction, while the N is for neurologic abnormalities.

Fever
Fever-beaver
Fever is one of the first presenting symptoms of TTP, and occurs due to excessive activity of the complement system.

Anemia
Anemone
Microthrombi may shear red blood cells as they pass by and cause mechanical fragmentation of red blood cells in a condition known as microangiopathic hemolytic anemia (MAHA). In this condition, a peripheral blood smear will show schistocytes and labs will show an increased lactate dehydrogenase (LDH) due to cell lysis.

Thrombocytopenia
Trombone-side-toe-peanut
Platelets are consumed during the formation of microthrombi leading to severe thrombocytopenia. Patients may manifest with petechiae, ecchymosis and purpura. Labs will show a normal PT and PTT with an increased bleeding time.

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Renal Dysfunction

Kidney Dysfunctioning

Renal insufficiency occurs in TTP secondary to complement-mediated damage and microthrombi formation within renal vessels. Acute kidney injury (AKI) may develop in severe cases.

Neurologic Abnormalities (Altered Mental status)

Nerve-guy with Altered-brain

A wide range of neurologic symptoms can be seen with TTP when microthrombi form within the cerebral vasculature, with headache and confusion being the most common. Severe cases may present with transient focal neurologic deficits, seizures, stroke or coma.

Treatment

Plasmapheresis

Plasma-fairy

Due to the high mortality of TTP, plasma exchange therapy (PEX) is indicated for empiric treatment of patients with presumed TTP. PEX prevents further microthrombi formation by removing ADAMTS13 autoantibodies and excess vWF multimers; this procedure can reverse the symptoms of early organ system damage.

Corticosteroids

Quarter-on-steroids

Glucocorticoids are used in addition to PEX to reduce inhibitor propagation and prevent the need for emergent plasma exchange therapy.

Splenectomy

Chopped-off Spleen

In patients with refractory disease or multiple relapses, splenectomy may be a treatment option. In some patients, splenectomy is effective as it removes the source of lymphocytes producing the ADAMTS13 autoantibody.

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