THROMBOTIC THROMBOCYTOPENIC PURPURA

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TTP consists of the pentad of:

- Microangiopathic haemolytic anaemia
- Thrombocytopenic purpura
- Neurologic abnormalities
- Fever
- Renal disease

Untreated mortality = 90%

Treatment with plasma exchange = 10-20%

Lack of plasma protease – A Disintegrinlike And Metalloprotease with ThromboSpondin type 1 motif 13 (ADAMTS13) – responsible for the breakdown of ultralarge vWF multimers.

Platelet-vWF complexes form small blood clots which circulate in the blood vessels and cause shearing of red blood cells, resulting in their rupture – Haemolytic anaemia and Schistocyte formation.

Similar to HUS but differentiated by coagulation screen results.

In TTP, the organ most obviously affected is the brain; in HUS, the kidney.

Relapses are not uncommon, occurring in 13-36% of patients.

Severe bleeding from thrombocytopenia is unusual, although petechiae are common

Laboratory Investigations:

- Measuring ADAMTS13 activity level may aid in diagnosis.
- FBC – ? Hb + Platelets
- Bilirubin + LDH
- Blood film
- Coagulation screen

- U+E

Tx of choice for TTP is plasma exchange with fresh frozen plasma

Transfusion is contraindicated in thrombotic TTP, as it fuels the coagulopathy.

Corticosteroids may be used in refractory patients

Rituximab, a monoclonal antibody aimed at the CD20 molecule on B lymphocytes – destruction of B cells reduces the production of the protease inhibitor.