Transitory and floating aphasic disease in a patience affected by Thrombotic Thrombocytopenic Purpura (Moschcowitz Syndrome)

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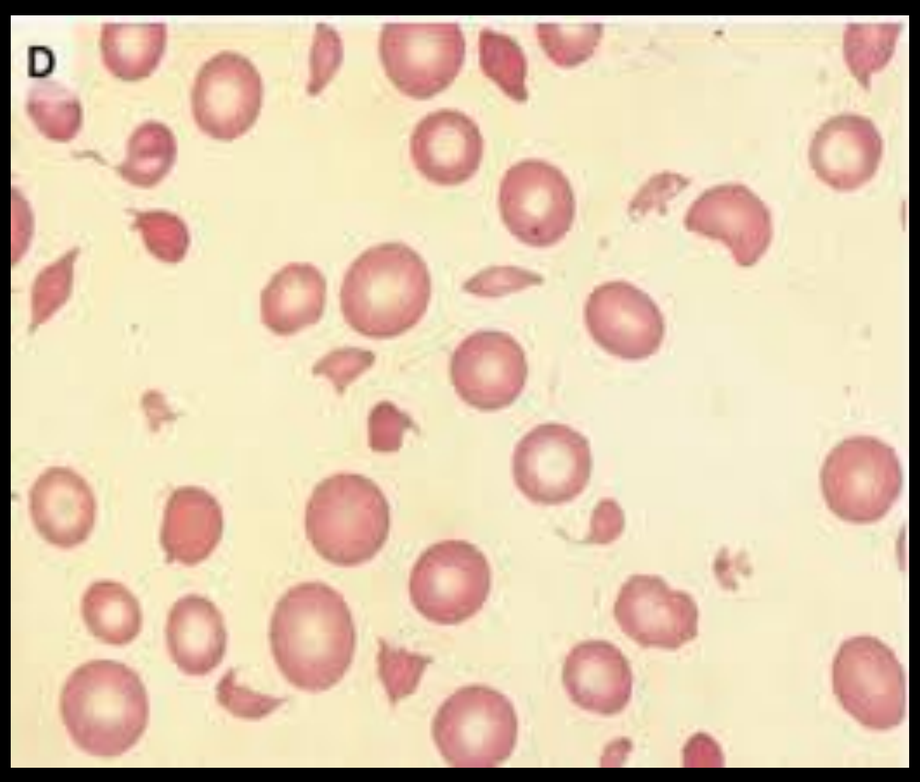
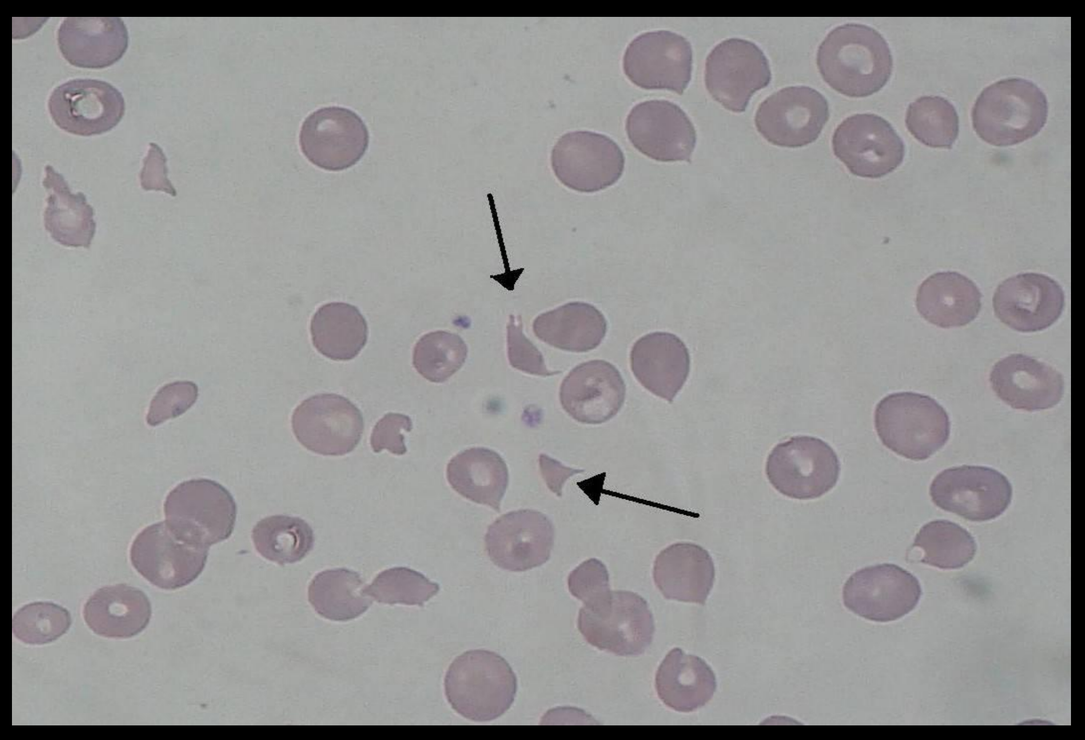
Introduction: Thrombotic Thrombocytopenic Purpura (TTP) causes fibrin deposits and platelet aggregates in capillaries and arterioles. Usually the affected people's age ranges between 10 and 40; and they're predominantly women. Often it is an idiopathic disease. The illness causes the circulation of high quantities of von Willebrand factor, which regulates the platelets aggregation and takes part to the blood coagulation. That depends on metalloprotease ADAMTS 13 shortage, which normally cuts the factor in small pieces in sane subjects. The uncut factor makes a circle around which the pathologic aggregates are built up. The consequences are platelet reduction (thrombocytopenia) and formation of thrombi and platelet aggregates, which occlude the microcirculation vessels. The following ischemia involves many organs, first of all the nervous system. The red blood cells, passing in the occluded vessels, undergo mechanical destruction with haemolytic anaemia. Fragmented red blood cells (schistocytes) appear in the circulatory system. The haemolysis and the ischemic zones necrosis release intracellular enzymes; which are revealed by the increasing of circulating LDH.

Clinical Case: M.B., 54 years old, female. In the history: Sjogren's Syndrome and coeliac disease. She was transferred after having been hospitalized for pulmonary thickening and fever, with confusion and aphasia symptoms. She had gone under a PM installation a few days earlier. The blood chemistry exams pointed out anaemia and marked thrombocytopenia. The following urgent brain TAC was negative and the CSF exam resulted normal. Thinking about a encephalitis with autoimmune or viral origin, A therapy was started based on Acylovir (750 mg x3/die) and Methylprednisolone (60 mg/die). The woman went through moments of lucidity and others of confusion and aphasia, an EEG protracted exam was therefor tried to point out critical paroxysms, but the exam was only slowed down. BOM excluded autoimmune thrombocytopenia, and the peripheral blood smear showed schistocytes, giving credit to the TTP hypothesis. Moved to Hematology Departmet she went under 9 plasmaphereses. The ADMTS 13 activity level was 3% (n.v. ranged 65-130); the anti-ADMTS antibodies were 48 U/ml (n.v. under 17). After the sixth plasmapheresis, the LDH became normal and the platelets were 150.000. The patience is now fine.

Conclusions: neurological symptoms, anaemia, thrombocytopenia, increasing of LDHs all lead to a TTP diagnosis. It is a potentially lethal haematological emergency, if not promptly dealt with. An association between this condition and heart interventions is described.





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