DIAGNOSTIC PROBLEMS OF THROMBOTIC TRROMBOCYTOPENIC PURPURA IN A PATIENT WITHOUTH THE CHARACTERISTIC CLINICAL PRESENTATION- Case report.

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Thrombotic Thrombocytopenic Purpura (TTP) is a disease characterized by a congenital or acquired deficiency of the protein von Willebrand factor cleavage called ADAMTS13, which is manifested by the "pentad": microangiopathic hemolytic anemia, thrombocytopenia, neurological manifestations, renal failure and fever.

Below we present a case that raised issues of differential diagnosis of TTP before measuring ADAMTS13 activity and dosing the anti-ADAMTS13 antibodies.

Our patient, female, aged 33 years old, without significant pathological personal history came in the hematological service accusing asthenia, fatigue, bruising to the legs and chest in the absence of trauma. On admission with easily influenced general state, skin and scleral jaundice. Paraclinically normochromic normocytic anemia (Hb = 7.5g / dL), marked reticulocytosis (281.9 ‰) with schistocytes on the the blood picture, severe thrombocytopenia (12,000 / mm^3). In the biochemical evidence LDH= 3047 U / L and significant mixed hyperbilirubinemia with predominantly indirect bilirubin.

In the absence of clinical features specific to TTP (neurological signs, fever and renal failure),a differential diagnosis with paroxysmal nocturnal hemoglobinuria (test HAM negative), a neoplastic cause(markers negative + nonsugestive imaging), with collagen diseases (immunological profile nonspecific), valvular heart disease (echocardiography in normal relations) with infectious causes (negative markers), disseminated intravascular coagulation and Evans syndrome (negative immunological tests) was made. ADAMTS13 activity assay revealed a downturn of this protein, the presence of anti-ADAMTS13 antibodies and ADAMTS13 antigen decline confirmed the diagnosis of TTP. Further therapy by plasmapheresis was initiated with the normalization of the hematological profile.

PTT clinical expression may be less suggestive, with the consequence of delaying specific treatment for a disease whose mortality rate is about 90% without treatment.