A case report of atypical haemolytic-uremic syndrome (aHUS) in a patient with type IIB von Willebrand disease (VWD): management and differential diagnosis of thrombotic microangiopathies (TMA) disorders in pregnancy

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Objective. TMA disorders in pregnancy and postpartum (thrombotic thrombocytopenic purpura, aHUS and haemolysis-elevated liver enzymes-low platelets, syndrome -HELLP-) are characterized by partly overlapping clinical and laboratory features: microangiopathic haemolytic anaemia, thrombocytopenia and organ injury. While differential diagnosis is not always obvious, these conditions require different management.

Materials and Methods. A 48-years-old woman affected by type IIB VWD showed reduced platelet count, mild anaemia and haemolysis, initial liver and renal damage at 33+6 weeks of her medically assisted dichorionic-diamniotic twin pregnancy complicated by foetal growth restriction of both foetuses. Caesarean section was performed for maternal indication. On first postoperative day patient showed new onset hypertension, severe haemolysis (LDH > 1,000 U/L; LDH/AST > 10), severe thrombocytopenia (12,000/μl), anaemia (8.4 g/dl) and high creatinine levels (1.74 mg/dL) suggesting a TMA disorder.

Results. Negative direct Coombs test, low C3, C4 and haptoglobin levels, schistocytes on peripheral blood smear, ADAMTS13 activity > 10% led to the final diagnosis of aHUS, treated with Eculizumab, a complement blockade, with progressive resolution of the clinical features.

Conclusions. TMA are rare but severe disorders of pregnancy and postpartum. In this case VWD hampered the diagnosis, as it itself causes thrombocytopenia. In such critical cases a systematic and multidisciplinary team approach (nephrology, haematology, obstetrics, anaesthesiologist) is recommended to avoid delays in differential diagnosis and in treatment that may be life-threatening.