The Reliability of ADAMTS13 Activity for Diagnosis of Thrombotic Thrombocytopenic Purpura

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Thrombotic thrombocytopenic purpura (TTP) is a life-threatening disorder classically characterized by the pentad symptoms of microangiopathic hemolytic anemia, thrombocytopenia, fever, neurological impairment, and renal dysfunction. TTP is initially diagnosed based on clinical and laboratory findings. Then the diagnosis is confirmed by deficiency of ADAMTS13 (a disintegrin and metalloproteinase with thrombospondin motif-13) activity. An elderly female was unresponsive on scene, had a witnessed new onset tonic-clonic seizures requiring intubation for airway protection, and refractory hypotension requiring pressor support. She was also found to have anuric acute kidney injury (AKI) and thrombocytopenia. Septic work up was negative. Cerebrospinal fluid studies were benign. The electroencephalogram and MRI Brain results were negative. Hospital course was worsened by increasing creatinine and hemolytic anemia with severely elevated LDH of 1300 and nondetectable haptoglobin. Peripheral smear was significant for multiple schistocytes and thrombocytopenia. The elderly female was clinically diagnosed with idiopathic TTP and received one session of plasmapheresis. Her clinical status immediately improve: resolving neurological status, anuria AKI, thrombocytopenia, anemia, and shock. Interestingly her ADAMTS13 activity level was within normal limits. As a result, further plasmapheresis sessions were discontinued, and supportive care was resumed. The significance of ADAMST13 deficiency for diagnosis of TTP is a controversial issue. TTP usually presents with severe ADAMTS13 deficiency; however, this abnormality is only found in about 60% of patients with TTP. Additionally, there has been increasing evidence questioning the reliability of ADAMTS13 assays. Studies had shown that the sensitivity and specificity might be lower than expected for ADAMTS13 assays. More research is needed to explore prevalence of ADAMTS13 deficiency within TTP and to evaluate the efficacy of the ADAMTS13 assays.