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There's No Ecstasy in Thrombotic Thrombocytopenic Purpura

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INTRODUCTION: Thrombotic thrombocytopenic purpura (TTP) is a rare and life-threatening clinical syndrome traditionally linked to a pentad of hemolytic anemia, thrombocytopenia, fever, renal impairment, and neurologic dysfunction. All five symptoms are observed in only 40% of patients. Despite its many reported causes, acquired TTP continues to develop with newer drug exposures.

CASE PRESENTATION: 25 year-old man without prior medical history was admitted to the hospital for abdominal pain, vomiting, and diarrhea symptoms thought to be consistent with pancreatitis. Two days prior, he ingested four ecstasy tablets with alcohol. Physical examination was significant for fever, tachycardia, bilateral scleral icterus, and no skin rashes. Systems evaluation revealed acute kidney injury, indirect hyperbilirubinemia, elevated lactate dehydrogenase, low haptoglobin with normal PT and INR, marked anemia with numerous schistocytes and thrombocytopenia, negative Coombs, and elevated D-dimer. Urine drug screen was positive for tetrahydrocannabinol and amphetamines. Infectious stool studies were negative. MRI brain revealed bilateral basal ganglia and posterior midbrain hyperintensity compatible with toxic metabolic poisoning. The patient's constellation of signs and symptoms progressed to worsening encephalopathy prompting intubation, empiric plasma exchange (PLEX) and methylprednisolone (1mg/kg) for suspected TTP. Within days, encephalopathy and laboratory abnormalities reversed with daily plasma exchange and high-dose corticosteroids. However, thrombocytopenia and hemolysis relapsed after discontinuation of therapy. He remains in the hospital currently after receiving fifteen sessions of PLEX in conjunction with rituximab. Final serum analysis (pre-PLEX) revealed severe ADAMTS13 deficiency (activity 3%, inhibitor level 1.1), consistent with TTP.

DISCUSSION: TTP can be congenital, idiopathic, or acquired secondary to infections, drug-induced, autoimmune diseases, malignancies, pregnancy, or bone marrow transplantation, with an annual incidence of 4 cases per million people. Systemic clumping of platelets is due to the lack of ADAMTS13, a protease that cleaves von Willebrand factor (vWF). Ecstasy is the "street" name for 3,4-methylenedioxymethamphetamine (MDMA). To our knowledge, our case is the fifth to describe a link between MDMA and acquired TTP. High clinical suspicion of TTP is enough to warrant urgent PLEX and administration of high dose steroids. Common side effects after ingestion include tachycardia, hypertension, and hyperthermia. Recent literature hypothesizes that MDMA-induced TTP is the result of the molecular similarity, an aryl-ring with a propane side chain, of MDMA and the thienopyridines known to cause TTP. Ticlopidine-linked TTP cases had a detectable autoantibody against ADAMTS13, while clopidogrel-linked TTP led to a deficiency in ADAMTS13 activity without detectable antibodies. Our patient's inhibitor level was greater than reference cut off, consistent with autoantibody mediated rather than binding inhibition. Our case is also the first reported administration of rituximab for relapse TTP after MDMA use. ADAMTS13 activity of 3% not only classified our patient with severe TTP but identified him as high risk for relapse TTP. Recent studies reported that persistently undetectable ADAMTS13 activity (<5%) early in the course of remission (<7 days) was associated with an increased risk for relapse over 18 months. If there is suboptimal response after PLEX and steroids, such as in our patient, rituximab inhibits production of autoantibodies against ADAMTS13.

CONCLUSIONS: In our patient's case, TTP was likely due to development of autoantibody after MDMA use. MDMA is a synthetic compound with a chemical structure similar to thienopyridine, a known precipitator of TTP. PLEX and steroids is standard therapy but does not halt autoantibody production. If relapse MDMA-associated TTP occurs, the addition of rituximab may be effective in decreasing the sessions of PLEX and duration of steroids.

Reference #1: de Fallois J et al. Fulminant thrombotic thrombocytopenic purpura (TTP): association with amphetamine consumption? Ann Hematol 2015; 94: 337-338.

Reference #2: Kayar Y et al. Thrombotic thrombocytopenic purpura and focal segmental glomerulosclerosis associated with the use of ecstasy. Indian J Crit Care Med 2015; 19: 230-232.

Reference #3: Schirren C et al. Thrombotic Thrombocytopenic Purpura After Ecstasy-Induced Acute Liver Failure. Annals of Internal Medicine 1999; 130; 163.

DISCLOSURE: The following authors have nothing to disclose: Victoria Tang, Nitya Kumar, Brian Hsi, Kamran Boka

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