A True Bloody Emergency: An Unusual Case of Thrombotic Thrombocytopenic Purpura

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Background

Thrombotic thrombocytopenic purpura (TTP) is a primary thrombotic microangiopathy that is classically characterized by thrombocytopenia and microangiopathic hemolytic anemia (MAHA). Although rare with an annual incidence of 3.7 cases per one million adults, it is considered a true hematological emergency due to its fatality rate of almost 100% if appropriate treatment is not initiated immediately. This makes it vitally important to identify and treat patients with TTP, a task that becomes unusually challenging in the absence of the disorder's other characteristically diagnostic clinical features such as mucosal bleeding, fever, or presence of schistocytes.

Case Presentation

A 30-year-old gentleman with a past medical history of Autoimmune Hemolytic Anemia is admitted for severe thrombocytopenia found on routine laboratory testing. The patient endorsed generalized weakness, fatigue, headache and dark-colored urine onset three days ago. Three months ago, the patient was admitted for exacerbation of warm AHA and started on Prednisone 100 mg post-discharge. The patient reports he had been tapering off the medication and stopped completely three days ago. The patient's is unremarkable except for tachycardia and mild distress, His laboratory findings include platelet count at 16k/μL, hemoglobin 11.1 gm/dL, hematocrit 33.7%, MCV 92.2 fL, lactic acid 3.17 mmol/L and creatinine 1.3 mg/dL. Initial management plan included methylprednisolone 125 mg IV, peripheral blood smear and CBC to monitor platelet count with transfusion precautions if levels drop below 10k/μL. A few hours into admission, the patient developed confusion, hyperbilirubinemia, worsening thrombocytopenia (13k/μL) and 1.5 mg/dL to 1.7 mg/dL increase in creatinine. Although the lack of schistocytes on peripheral blood smear dismissed the possibility of MAHA, the acutely worsening condition of the patient raised concerns for acute TTP and orders for fresh frozen plasma transfusions and plasmapheresis were empirically initiated. The patient's TTP PLASMIC score was 6 and an ADAMTS13 level was ordered. The patient was also started on daily doses of rituximab 375 mg/m^2 and methylprednisolone 1000 mg. Nephrology was consulted for concerns of acute kidney injury given up-trending creatinine levels and urine studies showing microscopic hematuria with 3+ blood in urine. CT abdomen/pelvis and renal doppler ultrasound were performed which resulted in findings indicating inflammatory nephritis. Over the course of the hospital stay, the patient developed small petechiae on his abdomen and bilateral upper extremities and reported episodes of dizziness and nausea. Subsequent daily rounds of TPE, transfusions and steroids, however, resulted in the resolution of both neurological and hematological symptoms. The patient's creatinine level improved to 0.9 mg/dL, lactic acidosis resolved with lactic acid at 1.24 mmol/L, and bilirubin levels returned to normal limits as well. ADAMTS13 level was noted to be 0.03% which confirmed thrombotic thrombocytopenic purpura as the precipitating pathology in this patient's condition. With a significant improvement of the patient's thrombocytopenia (platelet counts 350k/μL), patient was deemed fit for discharge under strict recommendations to continue steroid treatment and regular follow-up appointments with his hematologist/oncologist.

Conclusion

Acute episodes of TTP are extremely life-threatening situations and immediate recognition and intervention is vital for patient mortality, especially in such non-classical presentations.