Thrombotic Thrombocytopenic Purpura-like Illness in a Patient With Anti-Glomerular Basement Membrane Antibody Disease

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Anti-glomerular basement membrane (anti-GBM) disease or Goodpasture's disease is characterized by the presence of anti-GBM antibodies causing glomerulonephritis often in conjunction with diffuse alveolar hemorrhage. A small number of cases in the literature report anti-GBM disease presenting with thrombotic thrombocytopenic purpura (TTP)-like illness. We report an unusual case of anti-GBM disease with thrombocytopenia and microangiopathic hemolytic anemia (MAHA) in the setting of normal ADAMTS13 activity exacerbated by therapeutic plasma exchange (TPE). A 50-year-old African American female presented to our institution for TPE and dialysis after recent diagnosis of biopsy confirmed Goodpasture's disease. At admission, she reported no hemoptysis; platelet count was 131,000/µL and quickly decreased to 89,000/µL. After 2 TPE procedures, using 5% albumin as replacement fluid, thrombocytopenia worsened and a microangiopathic process emerged, indicating possible TTP. The 3rd TPE was performed using thawed plasma as replacement fluid. The patient’s plasma was noted to be dark reddish-brown indicating hemolysis. Despite consecutive TPE and platelet transfusions, platelet count decreased to 21,700/µL. Her condition also declined with development of diffuse alveolar hemorrhage requiring intubation and multiorgan failure; TPE was held and continuous renal replacement therapy was initiated. The patient received 3 units of platelets and her count stabilized (~80,000/µL). TPE resumed 2 days later and after 3 procedures, her platelet count rapidly decreased to 39,000/µL; TPE was subsequently discontinued. At discharge the platelet count was 79,000/µL. There is no clear etiology for thrombocytopenia or MAHA in this patient nor do we understand why TPE appeared to exacerbate these processes. This case of Goodpasture's disease with TTP-like illness is similar to other cases described in the literature, and illustrates the need for multidisciplinary management of these patients. We recommend early ADAMTS-13 testing and caution when activity is normal as TPE may be contraindicated and unnecessarily expose patients to blood products.

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