AN UNUSUAL CASE OF THROMBOTIC THROMBOCYTIC PURPURA AND SEVERE VITAMIN B12 DEFICIENCY PRESENTING AS WEAKNESS AND ALTERED MENTAL STATUS

Sanu Paul, MD[1] (sanusarapaul@gmail.com); Ponnu M Paul, MBBS[2]; Abbi Johns, MD[3] , Mathew Mathew, MD[1]

1. Department of Internal Medicine, Suburban Community Hospital, East Norriton, PA
2. Department of Medicine, MOSC Medical College, Kerala, India
3. Department of Medicine, Temple University Hospital, Jeanes campus, PA

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Background

Thrombotic thrombocytopenic purpura (TTP) is a rare hematological emergency that is invariably fatal if left untreated. Severe vitamin B12 deficiency can mimic TTP because of microangiopathic changes. There have been case reports of patients with severe B12 deficiency misdiagnosed as TTP and received plasmapheresis.

Case Report

A 35-year-old female with no significant past medical history presented to the emergency room with complains of left upper extremity weakness, headache and intermittent confusion. On arrival, vitals were Temp 99.5 F, Heart rate 112, blood pressure 119/82, respiratory rate 22, saturating 98% on room air. She was awake, alert and oriented x 3 and neurological examination was normal.
CT head and CT angiogram of head and neck was negative. Labs were significant for severe thrombocytopenia and anemia. WBC count 5.6, hemoglobin 7, platelet count 18,000, reticulocyte count 10.8%. Peripheral smear showed schistocytes. HIV was negative.

Patient denied any history of anemia/thrombocytopenia. She denies taking new medications/ herbal supplements. She consumes meat. Hematology was consulted. LDH , Coombs test, vitamin B12, hepatitis panel, ADAMTS 13 were ordered. Coombs test and hepatitis panel came back as negative. LDH 549

On the next day, she had a T max of 100.7, was more confused and only oriented to place. Platelets trended down to 14. Haptoglobin was less than 10 mg/dL-all suggestive of hemolysis. TTP was strongly suspected due to fever, confusion, Hemolytic anemia, thrombocytopenia.

She was started on plasmapheresis and IV steroids. Next day platelets improved to 39,000 and LDH levels improved. Patient had on and off episodes of confusion and altered mentation during her stay. Another 2 plasma exchanges were also done. Her mentation and weakness improved. Platelets improved to 120,000. Vitamin B12 levels came as low 117. Intrinsic factor antibodies were positive suggesting pernicious anemia.

Initial ADAMTS 13 was 9% which improved to 18% after the plasmapheresis. Rituximab was started for treatment of TTP. She also was started on IM vitamin B12 injections. She was discharged on day 5 of hospitalization and was asked to follow up with gastroenterologist and hematologist as outpatient

Conclusion

Severe B12 deficiency and TTP together can pose a diagnostic and therapeutic dilemma to clinicians, but when in doubt always treat TTP as it is life threatening with high mortality.

Discussion

Thrombotic thrombocytopenic purpura (TTP) is a rare hematological emergency characterized by pentad of microangiopathic hemolytic anemia, thrombocytopenia, neurological symptoms, renal injury, and fever. TTP without treatment has a mortality of about 90%[1]. .Prompt intervention with plasma exchange minimizes mortality and is the cornerstone of therapy.

Vitamin B12 deficiency is a relatively benign diagnosis that can mimic microangiopathic hemolytic anemia, characterized by the presence of anemia, thrombocytopenia, indirect hyperbilirubinemia, markers of hemolysis, and schistocytes. The mechanism of thrombotic microangiopathy in vitamin B12 deficiency is multifactorial, but most closely tied to hyper-homocysteinemia[2]. Vitamin B12 deficiency induced pseudo- TTP is a rare condition that resembles the clinical features of TTP.

Rapid recognition of the underlying etiology of microangiopathic hemolytic anemia is necessary as treatment approaches differ greatly. Treatment for TTP should be started with plasmapheresis in cases of doubt even if ADAMS13 levels are pending. But other rare causes of hemolytic anemia like Vitamin B12 deficiency should also be kept in mind by physicians.

References

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