Case Report

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Website: www.ijhonline.org DOI: 10.4103/ijh.ijh_9_17 The challenge of microangiopathic hemolytic anemia

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Abstract:

Microangiopathic hemolytic anemia (MAHA) is a Coomb's-negative hemolytic anemia characterized by red cell fragmentation (schistocytes). Thrombotic microangiopathy anemia, including thrombotic thrombocytopenia and hemolytic-uremic syndrome, malignant hypertension, preeclampsia are among the most common causes. We present a case of MAHA presenting with thrombocytopenia initially diagnosed as MAHA secondary to thrombotic thrombocytopenic purpura and received five sessions plasmapheresis without improvement but with worsening of anemia and thrombocytopenia. On further inquiry, glucose-6-phosphate dehydrogenase deficiency was identified, and the patient showed dramatic recovery after the trial of B12 and folate.

Keywords:

Fragmentation syndrome, glucose-6-phosphate dehydrogenase deficiency, megaloblastic anemia

Introduction

Microangiopathic hemolytic anemia (MAHA) is a Coomb's-negative hemolytic anemia characterized by red cell fragmentation (schistocytes). Thrombotic microangiopathy anemia, including thrombotic thrombocytopenia and hemolytic-uremic syndrome, malignant hypertension, preeclampsia are among the most common causes.

Case Report

A 35-year-old male previously healthy was referred to the hematology department of Merjan Teaching Hospital with a 1-week history of progressive pallor, headache, fever and red color urine. He denied any history of recent exposure to medications. On examination, the patient was pale, jaundiced; petechia was noted on the extremities and trunk. There are no congested tonsils, lymphadenopathy, or hepatosplenomegaly.

Complete blood count (CBC): Hb 5 g/dL, white blood cell (WBC) 4.4×10^9 /L, platelet

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 12×10^{9} /L, and reticulocyte 30%. Blood film showed polychromasia, oval, and crenated cells, and many fragmented cells are seen [Figure 1] with schistocyte count 4%. The following investigations were done including Coomb's test was negative, glucose-6-phosphate dehydrogenase (G6PD) assay was deficient, lactate dehydrogenase (LDH) was 2250 U/L, haptoglobin level-low normal, while antinuclear antibody/anti-DNA and prothrombin time/partial thromboplastin time (PT/PTT) were normal. Bone marrow examination showed erythroid hyperplasia. Liver and renal function tests are normal apart from mild elevation of unconjugated bilirubin. Because of these findings, a presumptive diagnosis of thrombotic thrombocytopenic purpura was raised, and the patient was started on therapeutic plasma exchange (plasmapheresis) for five sessions without objective response in CBC/blood film regarding platelet count, schistocyte count, or LDH with hemoglobin continue to drop with rising reticulocyte count. Further tests for vasculitis including C3, C4 and pANCA and cANCA were negative. Plasmapheresis was stopped, and the patient received a trial of intravenous

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Figure 1: Blood film showed fragmented red cells (schistocytes)

 B_{12} and oral folate. After 5 days of treatment, WBC was 4.4×10^9 /L, platelet count was 35×10^9 /L, and the hemoglobin raised to 8 g/dL with disappearance of jaundice. After five days of supportive treatment ,his complete blood picture showed WBC 5×109 /L, platelet 365×109 /L, and Hb 13 g/dL.

The case showed that megaloblastic anemia may present with severe red cell fragmentation that resolved completely after treatment, and this presentation with microangiopathic hemolytic anemia (MAHA) is actually folic acid megaloblastic anemia secondary to severe hemolytic attack of G6PD deficiency.^[1-3]

In addition to folic acid deficiency, disorders of intracellular Vitamin B12 (cobalamin) metabolism can cause thrombotic MAHA with the true incidence of which is unknown. These syndromes appear to be exclusively hereditary due to mutations in the methylmalonic aciduria and homocystinuria type C (MMACHC) gene; however, patients may present with a thrombotic microangiopathy anemia as an infant or adult. Elevated homocysteine and low methionine levels are seen in plasma, and urine may show methylmalonic aciduria. A dramatic case report described a previously healthy 18-year-old control who presented with MAHA, thrombocytopenia, and renal failure who had negative testing for thrombotic thrombocytopenia (TTP) and ST-hemolytic-uremic syndrome and was subsequently found to have cobalamin C deficiency due to an MMACHC mutation. Treatment with high-dose Vitamin B12, betaine, and folinic acid resulted in a dramatic recovery. His brother had died from complications of a similar syndrome at the same age.^[4,5]

Acquired severe Vitamin B12 deficiency can cause thrombocytopenia and ineffective erythropoiesis, which may be accompanied by hemolysis and red blood cell morphology resembling MAHA. Homocysteine and methionine level are recommended by some authors in the workup of MAHA and TTP cases.^[3-5]

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Conflicts of interest

There are no conflicts of interest.

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