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CASE REPORT

Sickle Cell Anemia with Megaloblastic Crisis: Deficiency or Demand?

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ABSTRACT

Megaloblastic erythropoiesis has been associated with hemolytic anemias. Sickle cell disease (SCD) presenting as megaloblastosis is a rare phenomenon. We present a case of sickle cell anemia presenting with painful crisis along with megaloblastic blood picture.

Keywords: Anemia, Erythropoiesis, Megaloblastosis, Sickle cell disease.

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INTRODUCTION

Sickle cell disease usually presents with vaso-occlusive, aplastic, hyperhemolytic, splenic sequestration, and acute chest syndrome as predominant crisis. Though megaloblastic crisis is a known phenomenon in thalassemia and hereditary spherocytosis,[1-4] SCD with megaloblastic crisis is rare. The possible explanation may be the nutritional deficiency of folate and vitamin B12.

CASE REPORT

A 34-year-old male, a diagnosed case of SCD since 18 years, presented to us with complaints of bone pains, myalgia, and fever of 3 days duration. On examination, pallor was present, icterus was positive, jugular venous pressure was normal, and there was no edema feet. Mild glossitis was present on tongue. Systemic examination was normal. Neurologic examination was normal. Patient was diagnosed as a case of SCD since 18 years by hemoglobin (Hb) electrophoresis, which showed SS pattern. Episodes of crisis were eight in number, which required hospitalization in the past 18 years. This was the ninth hospital admission with painful crisis. Patient was taking intermittent folic acid and zinc supplementation. He was a pure vegan.

Investigation revealed Hb 7.8 gm%, total leukocyte count 14,000 with neutrophilic leukocytosis, mean cell volume (MCV) 118 fL. Reticulocyte count was 3.2%, and repeat Hb electrophoresis revealed SS pattern. Serum lactate dehydrogenase was 850 U (normal range 140–280 U/L). Total serum bilirubin was 3.8 mg%, out of which unconjugated was 2.8 mg% and conjugated was 1.0 mg%. Serum folate level was 3.2 nmol/L (normal range 4.5–45.3 nmol/L). Serum cobalamin level was 126 ng/L (normal range 180–914 ng/L). Cultures of blood and urine were negative. Further testing uncovered very high plasma methylmalonic acid (1640 nmol/L), homocysteine (135.9 µmol/L). Serum antibodies to intrinsic factor was negative.

Patient was treated with antibiotics, analgesics, injection vitamin B12 1000 µg intramuscularly for 7 days and folic acid 5 mg postoperative once a day. Repeat peripheral smear after 5 days showed normocytic red blood cells (RBCs) suggesting response to vitamin B12 and folate therapy (Fig. 1). Hemoglobin after 1 week was 9 gm% and MCV was 98 fL. Patient was discharged on folic acid 5 mg

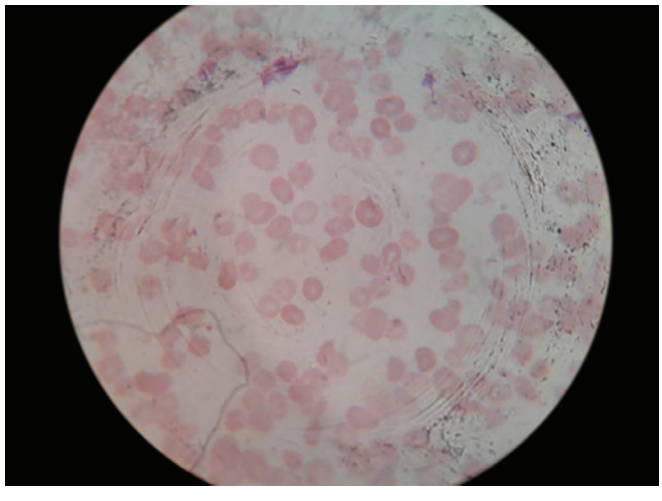

Fig. 1: Peripheral smear showing macrocytic RBCs with mild anisopoikilocytosis

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once a day to be continued, injection vitamin B12 1000 µg once a month to continue, and zinc supplementation.

DISCUSSION

The occurrence of megaloblastic blood picture in sickle cell anemia primarily raises two basic questions: the first question is whether the megaloblastic anemia in this case is due to deficiency of isolated folic acid or vitamin B12 or both. The second question is whether there is an etiological relationship between SCD and megaloblastosis or it is a coincidental finding. The second question is a rare assumption because hemolytic anemias unlikely present with macrocytosis unless the patient has autoantibodies against intrinsic factor or against islet cells. Literature shows that many of the patients have normal B12 levels, suggesting that the actual culprit is folate deficiency.[5,6] Unlike the above scenario, our patient had vitamin B12 deficiency also. We ruled out pernicious anemia with negative antibody screening.[7] The cause of cobalamin deficiency in our case was dietary deficiency, as the patient was a pure vegan.

Another school of thought argues that folic acid and other hematopoietic factors are required in increased amount at times of increased erythropoiesis. Whenever sickle cell crisis is associated with hyperhemolysis, the demand of increased erythropoiesis consumes more folate, leading to a relative folate deficiency and megaloblastosis.[8]

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