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Case Study

MEGALOBLASTIC CRISIS IN SICKLE CELL DISEASE–A RARE PHENOMENA WITH AVAILABLE REVIEW OF LITERATURE

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ABSTRACT

Sickle cell disease (SCD) is an inherited disease where some red blood cells are shaped like sickles or crescent moons. This condition affects the blood and various organs of the body. These sickle cells also become rigid and sticky, which can slow or block blood flow, resulting in episodes of sickness that produce pain and other crisis. There are different types of crisis. SCD with a megaloblastic crisis is rare. The frequency of the carrier state determines the prevalence of sickle cell anemia. Here we describe a 20 y old female patient with sickle cell disease who experienced a megaloblastic crisis.

Keywords: Crisis, Megaloblastic crisis, Sickle in disease, Vitamin B12, Folic acid, Deficiency

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INTRODUCTION

A subset of genetic red blood cell diseases known as sickle cell disease (SCD) affects around 1 in 500 African American children and 1 in 36,000 Hispanic American youngsters [1]. The "acute painful crisis," which frequently necessitates hospitalization, is the primary clinical characteristic of sickle cell disease [2] "Sickle cell crisis" refers to a group of acute disorders that include dactylitis, acute chest syndrome, splenic sequestration crisis, aplastic crisis, hepatic crisis, hemolytic crisis, and vaso-occlusive crisis (acute painful crisis).

Avascular necrosis, priapism, meningitis, pneumonia, sepsis, osteomyelitis, stroke, and venous thromboembolism are among more acute consequences [3]. Though megaloblastic crisis is a known phenomenon in thalassemia and hereditary spherocytosis [4-7]. SCD with megaloblastic crisis is rare. The nutritional deficit of folate and vitamin B12 could be the cause. We describe a 20 y old female patient with sickle cell disease who experienced a megaloblastic crisis.

CASE REPORT

A 20 years female patient presented to our hospital with complaints of loose motion, lower abdominal pain, vomiting and intermittent fever since 4 days. Pallor and bilateral pedal edema were noted throughout the examination. On examination, splenomegaly and mild glossitis was present. Rest other systemic examination was unremarkable. All vitals were normal. Routine investigation revealed Hemoglobin of 6.8 gm% with microcytic hypochromic blood picture and features of hemolysis. Total leukocyte count showed neutrophilic leukocytosis and thrombocytopenia was present. The total amount of bilirubin in the serum was 3.06 mg%, of which 2.8 mg% was unconjugated and 1.0 mg % was conjugated. Other blood investigations were within normal limit. In view of the fever, blood and urine cultures were sent, which came negative.

Further on evaluation of bicytopenia, serum folic acid, serum cobalamin and hemoglobin electrophoresis was sent. Serum folate level was low 3.2 nmol/l (normal range 4.5–45.3 nmol/l), while serum cobalamin level was 126 ng/l (normal range 180–914 ng/l) level. In view of hemolysis, Hemoglobin electrophoresis was performed, which showed a sickle cell disease pattern. With above investigations, it was assumed that patient had bicytopenia due to underlying combined B12 and folic acid deficiency. Bone marrow aspiration and biopsy was done, which showed erythroid hyperplasia with megaloblastoid changes. Patient was treated with antibiotics, analgesics, vitamin B12 and folic acid. Additionally, the patient had two blood transfusions.

Repeat peripheral smear after 8 days showed normocytic red blood cells, suggesting response to vitamin B12 and folate therapy. Repeat hemoglobin after 5 days was 9.2 gm%. The patient was released with instructions to continue taking zinc supplements, injections of vitamin B12 1000 µg once a month, and 5 mg of folic acid once a day.

DISCUSSION

Two main problems are raised by the development of megaloblastic blood image in sickle cell anemia. The first is whether the megaloblastic anemia in this instance, is caused by a deficit of isolated folic acid, vitamin B12, or both. Whether megaloblastosis and SCD are related etiologically or if this is just a fortuitous observation is the second question.

The second question is a rare assumption because hemolytic anemias are unlikely to 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 a combined vitamin B12 deficiency and folate deficiency. 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 vegetarian.

Another school of thought contends that during periods of enhanced erythropoiesis, folic acid and other hematopoietic components are needed in greater amounts. Whenever a sickle cell crisis is associated with hemolysis, the demand for increased erythropoiesis consumes more folate, leading to a relative folate deficiency and megaloblastosis [8].

CONCLUSION

Sickel cell disease with megaloblastic crisis is rare. Estimation of serum vitamin B12 and folate levels will be helpful in diagnosing the underlying etiology and prompt treatment.

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AUTHORS CONTRIBUTIONS

All authors have contributed equally

CONFLICT OF INTERESTS

Declared none

REFERENCES

1. Lim SH, Fast L, Morris A. Sickle cell vaso occlusive crisis: its a gut feeling. J Transl Med. 2016 Dec 1;14(1):334. doi: 10.1186/s12967-016-1092-5, PMID 27906010.
2. Jeremiah ZA. Abnormal haemoglobin variants ABO and Rh blood groups among student of African descent in Port Harcourt Nigeria. Afr Health Sci. 2006 Sep;6(3):177-81. doi: 10.5555/afhs.2006.6.3.177, PMID 17140342.
3. Hiran S. Multiorgan dysfunction syndrome in sickle cell disease. J Assoc Physicians India. 2005 Jan;53:19-22. PMID 15857006.
4. Reisner EH JR. The nature and significance of megaloblastic blood formation. Blood. 1958 Apr;13(4):313-38. doi: 10.1182/blood.V13.4.313.313, PMID 13522835.
5. Grunland S. Megaloblastic hemolytic anemia. Act Med Scand. 1950;138 Suppl 239:101.
6. Leibetseder F. Die erythropoese beim hamolytischen icterus. Sang. 1950;21:155.
7. Schlagetter K. The importance of bone marrow examinations in the diagnosis of anemia in obstetrics. Arch Gynakol. 1955;185(6):641-74. doi: 10.1007/BF00977016, PMID 13303155.
8. Westerman MP, Bacus JW. Red blood cell morphology in sickle cell anemia as determined by image processing analysis: the relationship to painful crises. Am J Clin Pathol. 1983 Jun;79(6):667-72. doi: 10.1093/ajcp/79.6.667, PMID 6846257.