Microcytic Hypochromic Anaemia in Sickle Cell Disease – Think Again!

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Abstract

We report a combination of an alpha thalassaemia homozygous state in a patient with sickle cell anemia presenting with a hypochromic microcytic anemia.

Introduction

Sickle cell anemia is widely prevalent in India. Usually, patients of sickle cell anemia do not have a hypochromic microcytic anemia unless it is associated with other causes. These causes vary from iron deficiency anemia to a combination with a haemoglobinopathy, namely thalassaemia. Such a combination is presented here which is clinically associated with a less aggressive disease.

Case Report

A 26 year male of Banjara community was admitted to our ward for evaluation of anemia with complaints of easy fatigability, generalized weakness and pain in right upper abdomen. Physical examination revealed a thin, pale, icteric and febrile patient in pain. He had hepatomegally but no palpable gall bladder. Patient was hemodynamically stable and was treated with hydration, antibiotics, oxygen and analgesics.

Investigations revealed hemoglobin 9.9 gm/dl (N:12-17), TLC 9.2x10⁹/l (N:4-11) with differential of neutrophils 57%, lymphocytes 20%, eosinophil 10%, monocyte 11%, basophil 2% and platelet count 383x10⁹/l (N:150-400). Peripheral smear revealed hypochromic microcytic picture with sickle cells and few target cells. There was no other red cell (RBC), white blood cell (WBC) or platelets abnormality on smear. RBC indices were, RBC- 5.95 millions/cu.mm, PCV- 40.6%, MCV- 68.2 fl, MCH-22.1 pg, MCHC-33.7%, HbF-<0.1%, HbA- 71%, HbA2-3.1%; Brother: Hb-13.7 gm/dl, RBC-5.95 millions/cu.mm, PCV- 40.6%, MCV- 68.2 fl, MCH-23 pg, MCHC-33.7%, HbS%- 27.5%, Hbf<0.1%, HbA- 69.4%, HbA2-3.1%.

Patient’s father had unfortunately expired a few years ago. Patient’s mother and brother are sickle cell trait. Their RBC indices were,

- Mother: Hb-11.1 gm/dl, RBC- 5.15 millions/cu.mm, PCV-35.5%, MCV- 68.9 fl, MCH-21.6 pg, MCHC-31.3%, HbS- 25.9%, Hbf<0.1%, HbA- 71%, HbA2-3.1%;
- Brother: Hb-13.7 gm/dl, RBC-5.95 millions/cu.mm, PCV- 40.6%, MCV- 68.2 fl, MCH-23 pg, MCHC-33.7%, HbS%- 27.5%, Hbf<0.1%, HbA- 69.4%, HbA2-3.1%.

Discussion

Sickle cell anemia (HbS>50%) or sickle cell trait (HbS<50%) do not exhibit hypochromic microcytic picture. Above, we have presented a case of hypochromic microcytic anemia with sickle cell disease in a young male. Iron deficiency, the commonest cause of a hypochromic microcytic anemia in our country, was ruled out. His peripheral smear did not show features of basophilic stippling or dimorphic picture suggestive of lead toxicity or sideroblastic anemia, which could be two other causes of a hypochromic microcytic anemia. His red cell indices were classically thalassaemic but his conventional hemoglobin electrophoresis did not show beta thalassaemia trait. With this in mind, we chose to assess his alpha thalassaemic state which was confirmed by investigating his family. His brother had a raised HbF. This finding was important to understand his pathophysiology where it was noted that the patient had very few sickle cell crises (only three episodes of vaso-occlusive crisis in his lifetime with long intervals between episodes) which is unusual for a sickle cell disease patient. While there is heterogeneity in the presentation of sickle cell patients it appears that the alpha thalassaemia appeared to protect him from recurrent crises. The latter is found
commonly in Southeast Asia, particularly Thailand where 20% population is affected. It also occurs in Middle East, Greece, Italy, Africa and pacific islands and American blacks (25-30%).

Sickle cell anemia is mostly common in tribal areas of India.

Review of literature revealed that patients with coexisting α-thalassaemia showed a significant increase in erythrocyte deformability and change in the erythrocyte density profile, possibly due to inhibition of polymerization-related increases in cell density to explain the beneficial effect of α-thalassaemia on hematological parameters and clinical events in homozygous sickle cell disease.

While it was noted that osteonecrosis and sickle retinopathy occurs more often in sickle cell patients with co-existent alpha thalassaemia, the incidence of stroke is lower in the same subjects. However, a subsequent larger study demonstrated that co-existent alpha thalassaemia and sickle cell anemia is associated with diminished mortality risk in patients older than 20 years of age indicating milder crises episodes.

**Conclusion**

We conclude that every hypochromic microcytic anemia with or without sickle cell anemia isn’t always an iron deficiency. If the history confirms that for homozygous sickle cell anemia, the vaso-occlusive crises are not as frequent and there is a hypochromic microcytic anemia, then a differential diagnosis of haemoglobinopathy especially homozygous alpha thalassaemia two gene deletions should be considered after the common causes discussed above have been ruled out.

**References**


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