

Atypical presentation of sickle cell disease

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Article Info

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Abstract

A 20 year old female presented with a history of frequent syncopal attack since her childhood. Each episode persisted 5-10 min without having any aggravating factor or prodrome. She had persistent generalized bodyache aggravating during the winter. She had jaundice and episodic abdominal pain. She received 1 unit of blood transfusion 4 months back and improved symptomatically. Patient was mildly anemic, moderately icteric and had mild splenomegaly. Over these long periods of her illness she was thoroughly evaluated several times. Her biochemical and neurological evaluation revealed no abnormalities. But she was treated with anticonvulsant for long time empirically without significant improvement. Her CBC showed microcytic hypochromic anemia. She was negative for Wilson's disease. Reticulocyte count was high. Coomb's test was negative. Osmotic fragility test was positive. Hemoglobin electrophoresis revealed Hb-S 60%. Sickling test was found positive. Finally it was diagnosed as a case of HbS/ β +

Introduction

Sickle cell disease is highly prevalent in Sub-Saharan and equatorial Africa with lesser prevalent in India and Middle East.¹ It is an inherited chronic hemolytic anemia due to formation of abnormal hemoglobin (HbS). It is due to substitution of valine for glutamic acid at position 6 of the beta globin chain.²

Manifestations of sickle cell disease are mostly due to vaso-occlusive events rather than anemia itself. It is present with diverse clinical manifestation ranging from mild anemia along with jaundice to cerebrovascular manifestation. In the earlier age may present with acute confusional state but not much manifestation found in adult. In adult, it is present with stroke both ischemic and hemorrhagic. Nocturnal hypoxemia may cause seizure or transient ischemic attack.³ But syncopal attack in sickle cell anemia is not well documented. Recently we have experienced a patient of sickle cell anemia presented with frequent syncopal attack since her childhood.

Case Report

A 20 year old lady hailing from Thakurgaon attended with the history of repeated syncopal attack since her childhood. She also had generalized bodyache aggravated during the winter. Along with jaundice, she had occasional

abdominal pain and respiratory distress. For syncope she was evaluated thoroughly biochemically and radiologically but no definitive diagnosis could be reached. However, she was treated with anticonvulsant empirically but didn't improve symptomatically. Meanwhile, she developed anemia and jaundice and evaluated for Wilson's disease and hemoglobinopathies by the estimation of copper, CBC and hemoglobin electrophoresis without obtaining any conclusion. But improved symptomatically by single unit of blood transfusion first ever 4 months back. The patient was re-evaluated and found HbS (60.8%), HbA (2.4%), HbF (35.8%) in hemoglobin electrophoresis. Sickling test was positive. Finally it was confirmed as a case of HbS/ β +

Discussion

Sickle cell disease is not a frequently diagnosed disease in Bangladesh. The disease is caused by formation of HbS. When de-oxygenated HbS molecule stack into polymers causing microvascular occlusion and tissue damage occurs.⁴ Clinical manifestation due to vaso-occlusion and hemolysis. Patient presents with anemia, jaundice, painful episode, frequent infection due to loss of splenic function, neurological complication, pulmonary and hepatobiliary complication are more marked.² Among CNS manifestations more marked manifestation are transient ischemic attack, stroke (ischemic and



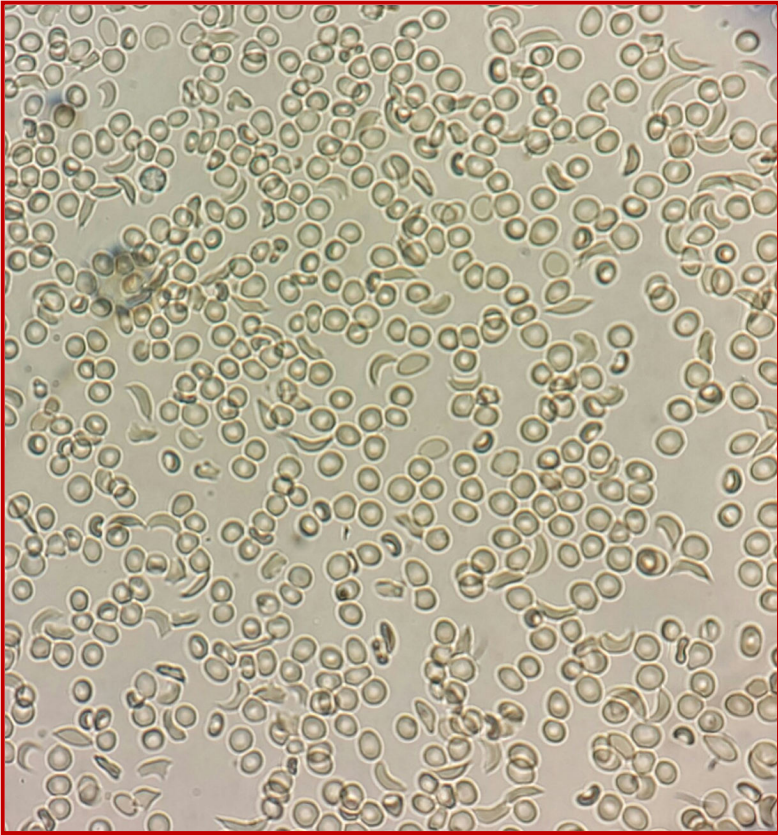


Figure 1: Photograph shows numerous sickled red cells in the sickling test of the patient

hemorrhagic). But syncopal attack is not so common. The above mentioned patient has got anemia, jaundice which are feature suggestive of

hemolysis. She also has generalized bodyache, abdominal pain, chest discomfort indicating vaso-occlusive pathology. But she has history of repeated syncopal attacks since her childhood manifested by sudden loss of consciousness and frequent fall. She was treated empirically by different medication without having any improvement. But after giving blood transfusion she is free from these sorts of symptoms. Interestingly although the patient has got multiple symptom she still has splenomegaly rather than splenic atrophy.

Conclusion

Any atypical neurological manifestation may need thorough hematological evaluation.

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