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An Unusual Presentation of Sickled Cell Anemia with Splenomegaly –A Case Report

Authors
Dr K.Shalini (PG), Dr K.Ambedkar Raj MD Professor, Dr V. Sarada Professor &HOD
Trichy Srm Medical College and Research Centre

Introduction

Sickle cell anemia is very common single gene disorder. 50% of world population affected by sickle cell anemia resides in India. The average frequency of sickle cell gene ranges between 22-44%. High prevalence has been found in various tribal community of India. Splenomegaly usually evident at six months of age but undergoes autosplenectomy by 8 years of age.

Clinical History

A 11 year old girl presented with history of high coloured urine ,yellowish discolouration of sclera, fatigability, failure to thrive since 2 years .Also complained of abdomen distension with mild pain and intermittent low grade fever since 6 months.

Examination

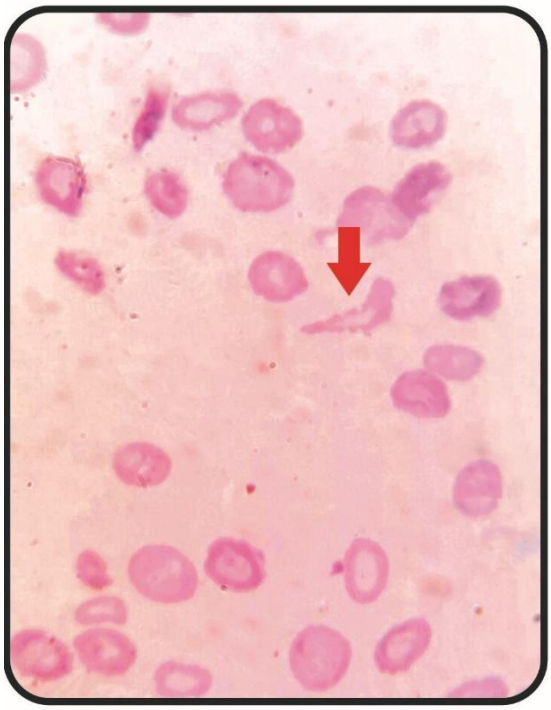
On Examination – Pallor +, Icterus +, Hemolytic faces +
CVS –S1S2+, RS –BAE +
P/A -Soft , Splenomegaly INVESTIGATION:

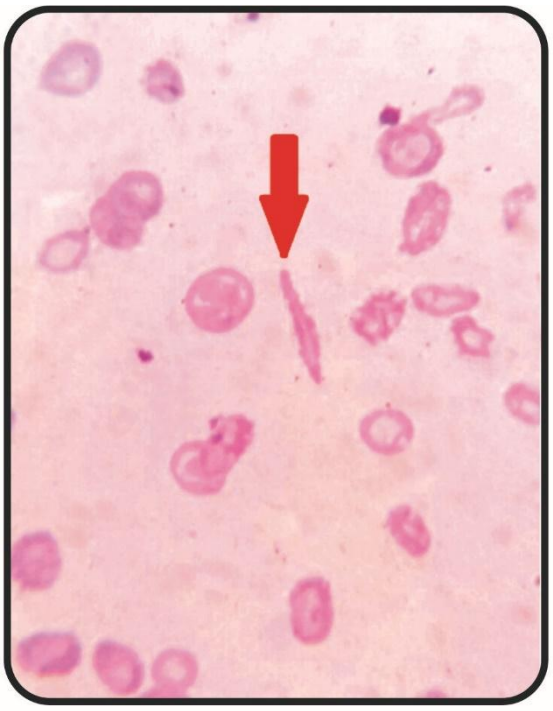
Complete Hemogram

Hb-3.5g/dl, RBC- 1.23millions/ml, PCV – 12%, MCV- 97.6 fl, MCH – 28.9pg, MCHC -29.6g/dl. WBC-5420cells/cmm, Platelet – 41000cells/microlitre

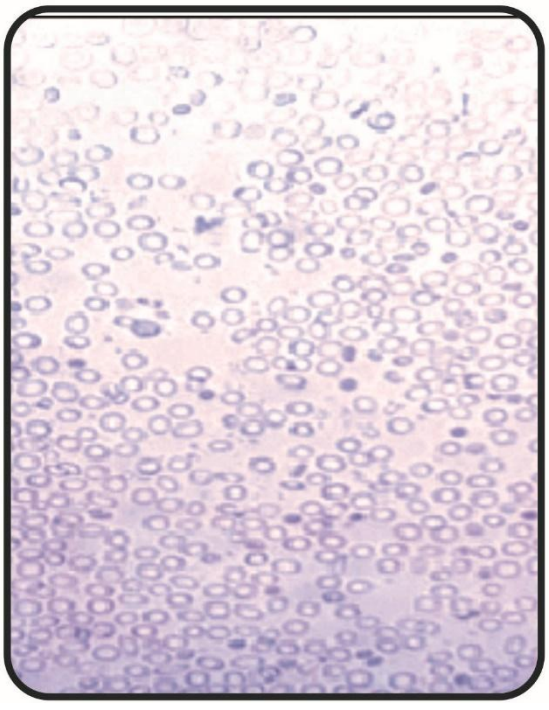
Peripheral Smear Study

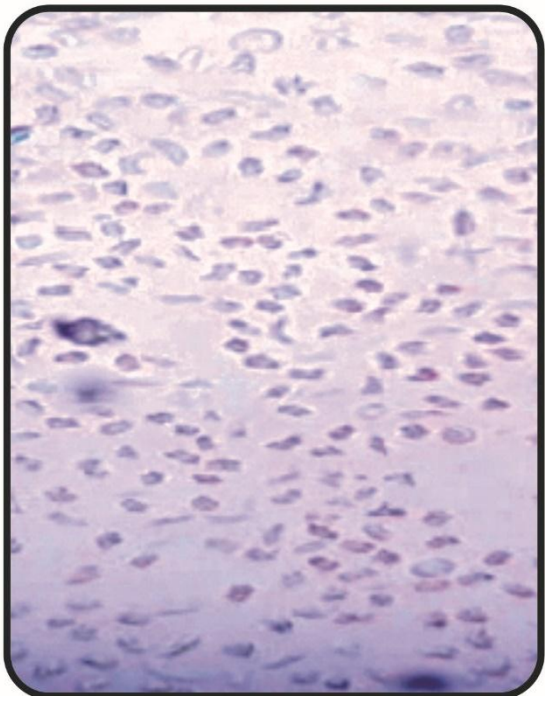
RBC – Predominantly Microcytic Hypochromic red cells with moderate anisopoikilocytosis. Sickle cells, target cells and nucleated RBCs seen. Occasional macrocytes and polychromatophils seen.
WBC- Normal in number, morphology and distribution.
Platelet- Reduced in count. Giant platelets seen.
Impression–
1. Hemolytic Anemia.
2.Thrombocytopenia.



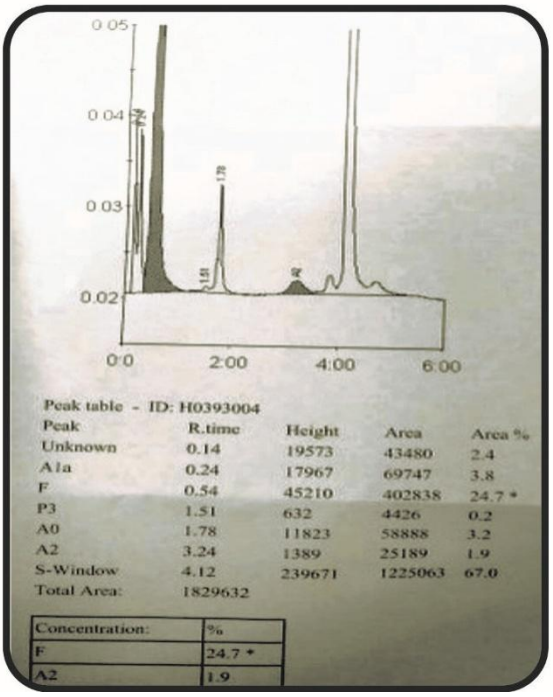


Reticulocyte Count – 5.8%
MP / MF – Negative
Sickling Test – Positive





Osmotic Fragility Test- Decreased
Hemoglobin Electrophoresis
HbA1- 4%, HbA2 -1.9% ,HbF -24.7%, HbS -67% HbD -0%



Discussion

Sickle cell anemia is the homozygous state usually manifest earlier in life. Rarely it can present in early adulthood. Though splenectomy occurs at most of the cases before 10 years of age, in patient with Sickle cell anemia (HbSS) splenomegaly may be present thereby determing the severity and frequency of vasoocclusion .The incidence of splenomegaly in Sickle cell anemia (HbSS) appears to be around 10% after 10 years of age. In our case family screening was recommended to rule out sickle cell disorders.

Conclusion

This is a case of Sickle cell anemia (HbSS) with high HbF which is an important etiology of persistence splenomegaly and late onset of symptoms.

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