Hematological profile of sickle cell disorder in tertiary care hospital

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Abstract:
Introduction*: Sickle hemoglobin is the most common pathological hemoglobin mutation worldwide in which upon deoxygenation RBCs convert into sickle form. Heterozygotes(AS) are usually asymptomatic and Homozygotes(SS)suffer from sickle cell anemia.* ***Aim****: To study hematological profiles of sickle cell disorders from Sir.T.Hospital, Bhavnagar.* ***Materials and Methods****: Prospective cross-sectional study was done in the Department of pathology, Government medical college, Bhavnagar from January 2013 to May 2015. Blood collected in EDTA vaccute and hematological indices measured by Abott cell dyne 3700 along with peripheral smear examination and confirmed by sickle solubility test and Hb electrophoresis.* ***Results****: Among 30 cases of sickle cell disease and 15 cases of sickle cell trait, male most commonly affected with mean age group 10 year. Hematological profile shows low Hb, MCH, MCHC and Hematocrit values with high MCV. Peripheral smear demonstrate anisopoikilocytosis(98%),sickle cell(68%) and target cell.(48%). Because sickle cell disorder is most common hemoglobinopathy such type of study is required which will support for correct diagnosis and epidemiological analysis.*
Key words*: sickle cell disease, hematological profiles , peripheral smear.*

I. Introduction

Hb S(Sickle hemoglobin) is abnormal hemoglobin variant in which adenine in sixth codon of beta gene is substituted by thiamine.So, valine replace glutamic acid in sixth position of beta chain. On deoxygenation of red blood cells, Hb S form tactoid aggregates and distorts RBCs into sickle shape. Upon oxygenation, these RBCs regain their normal shape. On repeated sickling and desickling, RBCs convert into permanent sickle shape. Because of sickle red blood cell anemia, crisis and organ damage can occur.[1]

Hb S is most common mutation worldwide. Heterozygotes(AS) are usually asymptomatic and Homozygotes(SS)suffer from sickle cell anemia. Heterozygous carrier patients usually resistant against malaria.[2] 50% of world population affected by sickle cell disease resides in India. The average frequency of sickle cell disease gene ranges between 22-44%.High prevalence is in tribal community of Gujrat, India.[3]

Hematological profiles of sickle cell disease is extremely variable. There is very little data available on hematological profiles of sickle cell disease from India. Few studies were undertaken for hemat profile of sickle cell disease from surat,south gujrat,valsad.[4]

This study was conducted to study hematological profiles and burden of sickle hemoglobin among patient visiting at Sir.T.Hospital,Bhavnagar. This will helpful in better diagnosis of sickle cell disease and for epidemiological study.

II. Materials And Methods

Prospective cross-sectional study was done in the Department of pathology, Government medical college, Bhavnagar from January 2013 to may 2015. Data regarding hemoglobin, MCV(mean corpuscular volume),MCHC(mean corpuscular hemoglobin concentration),MCH(mean corpuscular hemoglobin), RBCs count, Hematocrit, peripheral blood smear examination ,sickling test and hemoglobin electrophoresis were noted. Blood collected in ethylene di-aminetetraacetic acid(EDTA) vaccutte and hematological indices were measured using ABOTT CELL DYN 3700 and peripheral smear stained with leishman stain. Sickling test done using Sickle solubility test(SICK VIEW) method. To differentiate between disease and trait electrophoresis done using cellulose acetate strip at alkaline Ph.

III. Results

In our study total 45 cases were diagnosed as sickle cell disorder. Among them males were more commonly affected then female with male:female ratio 2:1. Mean age group was 10 year. 30 cases were of sickle cell disease showing Hb S band and remaining 15 cases showing Hb A and Hb S band that were diagnosed as sickle cell trait(By cellulose acetate electrophoresis). All cases were positive for SICK VIEW.

Figure No. 1
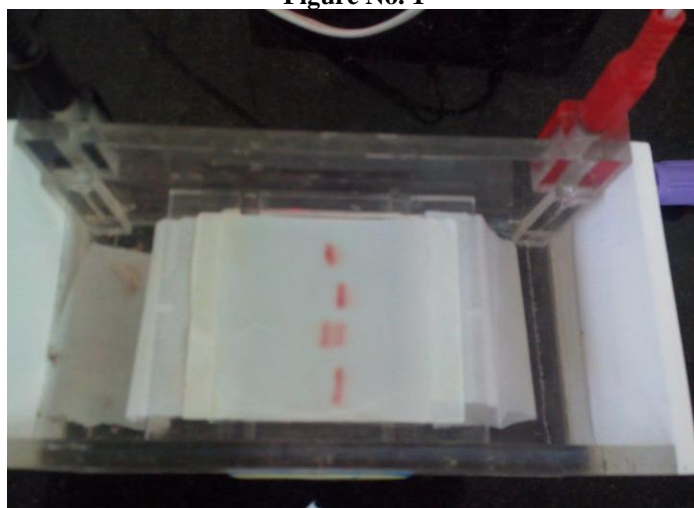
**Electrophoresis showing sickle cell trait**

Table No. 1 Hematological profiles in sickle cell disorder

Serial no.	Parameters	Mean value
1	Hemoglobin value	8.6 gm/dl
2	RBCs count	2.8 million/cumm
3	Hematocrit	25%
4	MCV	101 fl
5	MCH	24 pg
6	MCHC	26 gm/dl

Table No. 2 Peripheral smear examination in sickle cell disorders

Serial no.	Parameters	Total no. of case showing particular changes
1	Anisocytosis	44(97.77%)
2	Poikilocytosis	45(100%)
3	Target cells	22(48.88%)
4	Sickle cells	31(68.88%)
5	Polychromasia	27(60%)

Diagram No. 1
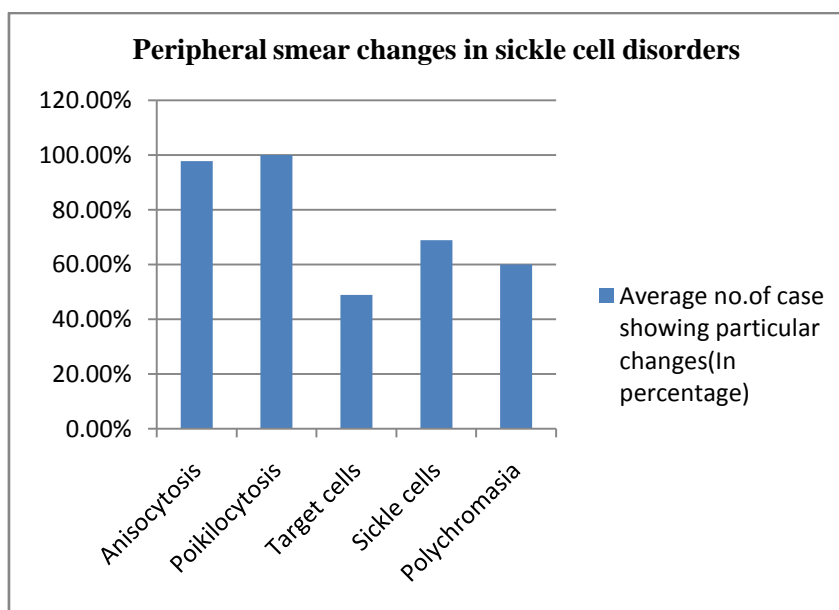
Bar diagram illustrate peripheral smear changes in sickle cell disorder

Figure No. 3
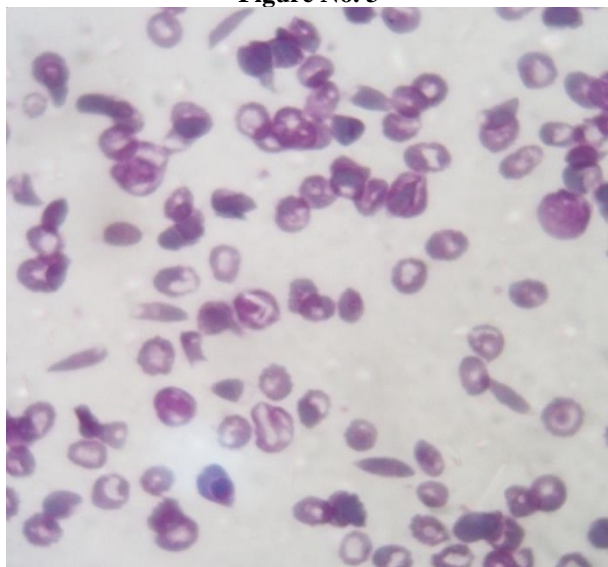
Peripheral smear showing sickled RBCs,Normoblast and target cells.

IV.   Discussion

In our study males were more as compared to female, which may be due to male child get more attention as compared to female. Male : female ratio 2:1.Similar result seen in some other study also.[5,6]Mean age group was 10 year which correlates with other study by Shrikhade AV et al.[7] in which maximum number of male patient seen up to age 14 year.

Mean Hb value 8.6 gm/dl that is nearby to other study`s average Hb value which is 7.11 gm/dl.[7] Total no. of case showing sickle RBCs were 31 while sick view test was positive in all cases. Five cases studied by shukla RN et al. observed presence of sickle cells and positive for sickling test in all 5 cases.[8] Total RBCs count, MCH, MCHC are low in our study which is comparable to other study.[9,10]MCV is higher than normal, reason may be because of increased need of erythropoiesis due to chronic hemolysis leading to macrocytosis.[11,12]All sick view positive cases were confirmed as sickle cell disorder by cellulose acetate agar electrophoresis at alkaline ph. In our study 30 cases(66.66%) were diagnosed as sickle disease and 15 cases(33.33%) were sickle cell trait by electrophoresis. Study done by Kamble M et all observed 61.6% cases of Hb SS and 38.4% cases of Hb AS.[5]

V.   Conclusion

This study shows that MCH, MCHC, RBCs count, hematocrit, hemoglobin value low in sickle cell disease patients. These patient`s peripheral smear shows anisopoikilocytosis, target cells, sickle RBCs. Further study can be done using HPLC(High performance liquid chromatography) for quantification of Hb S, if facilities available for HPLC.

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