IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 2 Ver. 6 February. (2018), PP 82-85
www.iosrjournals.org

Study of hematological profile in sickle cell patient.

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Abstract
Background: *Sickle cell disease is one of the most common autosomal recessive diseases in the world caused by a single nucleotide substitution (GTG - GAG) and is located at the sixth codon of the human-globin gene.*
Aims & Objectives: *This study was designed to study hematological profile in sickle cell patient and compare hematological profile in sickle cell traits with sickle cell disease patient*
Materials and Methods: *This study is an observational study done in 70 sickle cell patients in steady state, at Acharya VinobaBhave Rural Hospital, Sawangi Meghe, Wardha, MaharashtraIndia. The complete blood counts (CBC) were analyzed using the Automated Coulter Counter & Sickling pattern were identified on Hbelectrophoresis, at a pH of 8.6oncellulose acetatepaper.*
Results: *Haemoglobin concentration and the MCV was significantly low (≤0.05) in sickle cell diseased patients as compared to sickle cell traits. TLC count was not significantly higher while platelets & red cell distribution width (RDW) were significantly higher (≤0.05) in sickle cell disease (SCD) patients, as compared to traits.*
Conclusion: *Our study has explained haematological parameters & their role in sickle cell trait and sickle cell diseased which could be used in designing of the better management of sickle cell patients.*
Key Words: *Sickle Cell Anaemia; Complete Blood Counts (CBC); Blood Indices.*

Date of Submission: 01-02-2018 Date of acceptance: 17-02-2018

I. Introduction

The inherited disorders of blood include hemoglobinopathies which are one of the major public health problems in India.[1] Sickle cell disease is the second most common hemoglobinopathy next to Thalassemia in India.[2] The findings of the Indus valley civilization site indicate the prevalence of hereditary anemia (sickle cell disease or β thalassemia) in the Indian subcontinent from about 2000-5000 BC.[3] General incidence of Sickle cell disease in India is 1-44%.[4,5,6] The average frequency of hemoglobin S (HbS) is 4.3 % in India.[4] Sickle cell trait occurs in approximately 300 million people worldwide, with the highest prevalence of approximately 30% to 40% in sub-Saharan Africa.[7] Sickle cell disease refers to a group of genetic disorders, .characterized by presence of sickle hemoglobin (HbS), anemia, and acute and chronic tissue injury secondary to blockage of blood flow by abnormally shaped red cells**.** Herrick first described a case of sickle cell disease in 1910.[8]

The clinical manifestations arise from the tendency of the hemoglobin (HbS or sickle hemoglobin) to polymerize and deform red blood cells into the characteristic sickle shape. This property is due to a single nucleotide change in the β -globin gene leading to substitution of valine for glutamic acid at position 6 of the β-globin chain (β6glu→val or βs).[9]

The homozygous state (HbSS or sickle cell anaemia) is the most common form of sickle cell disease, but interaction of HbS with thalassaemia and certain variant hemoglobins also leads to sickling. The term sickle cell disease (SCD) is used to denote all entities associated with sickling of hemoglobin within red cells.[10]The normal hemoglobins are hemoglobin A (α2β2), hemoglobin A2 (α2δ2), and hemoglobin F (fetal hemoglobinα2γ2). Sickle hemoglobin is designated S because erythrocytes with hemoglobin S can transform into a sickle shape. [11] Hemoglobin S tends to polymerize into long rigid structures, which distort the cell into the characteristic sickle shape. Anything that causes deoxygenation of hemoglobin predisposes to sickling, including hypoxia, acidosis, and increased temperature.

The polymerization of hemoglobin S is reversible, and cells that have sickled may return to normal shape with reoxygenation. However, the repeated cycles of sickling and unsickling damage the cell, and, eventually, the erythrocytes becomes irreversibly sickled. The rigid elongated sickle cells obstruct small blood vessels, resulting in tissue infarction. Sickled erythrocytes are also “sticky” and adhere to endothelial cells, predisposing to thrombosis. Common sites of infarction include the spleen, bone and bone marrow, the medulla of the kidney, mesenteric vessels, and pulmonary vessels .[12] Sickle-cell disease may lead to various acute and chronic complications, several of which have a high mortality rate.[13] Many patients with SCA are inreasonably good health most of the time and achieving a steady state level of fitness. This state of relative well-being is

periodically interrupted by crisis of which the vaso- occlusive crisis (VOC) is the most common and hallmark of patients with SCD.

Based on this background, this study was aimedtoTo study hematological profile in sickle cell patient and compare hematological profile in sickle cell traits with sickle cell diseased patient

II. Material and methods

70 patients, age group of 16-50 years, withdiagnosedSCD, attending the outpatient clinic at the AcharyaVinobaBhave Rural Hospital, Sawangi Meghe,Wardha,Maharashtra, were taken as subjects. All subjectsstudiedwerediagnosedasSCAbasedonpositivesicklingtests,and haemoglobin electrophoresis at a pH of 8.6oncellulose acetatepaper.

All procedure were conducted with due consent of the subjects and institutional ethics committee approval. 5 ml of venous blood was collected by cleanvenipuncture from each patient via the ante-cubitalvein,using a plastic syringe with minimum stasis,intocommercially prepared concentrations ofsequestreneEthyleneDi-amineTetraaceticAcid(EDTA)bottles.

The complete blood counts (CBC) were analyzed using the automated COULTER (Beckman Coulter). The CBC included haemoglobin (Hb) haematocrit (HCV), total white blood cell count (TWBC), platelet count, mean cell volume (MCV), mean cell haemoglobin (MCH), mean cell haemoglobin concentration (MCHC) and red cell distribution width (RDW).

Sickling pattern wereindentified on Hb electrophoresis , at a pH of 8.6oncellulose acetatepaper.

Data was analyzed using SPSS version 16.0. The results were expressed as mean ± standard deviation

III. Results

In this study total 70 patients of sickle cell were taken in which Male to female ratio is 1:2.33. Out of 70 patients 50 (72%) was belongs to sickle cell trait(AS group) , and 20(28%) sickle cell diseased (SS group). In both group maximum no of patients belong to age group 21-30year.

Table 1:- Gender wise distribution of patients.

Gender	No of patients	Percentage (%)
Male	21	30%
Female	49	70%
Total	70	100%

Table 2: Age wise distribution of patients according to diagnosis

AGE	AS pattern	SS pattern
11-20 year	18%	26%
21-30 year	68%	60%
31-40 year	14%	14%
Total	100%	100%

Table3: Hematological parameters in sickle cell patients.

	AS pattern (n= 50)	SS pattern (n= 20)	p-value
Hb(gm%)	12.2± 0.86	8.94 ± 0.65	≤0.05
TLC (cells / µl)	9810.12± 1456	11312.6± 1010	≥0.05
Platelet (lac/ µl)	1.92± 0.50	3.12± 0.48	≤0.05
MCV (fl)	70.24± 9.56	64.92± 4.12	≤0.05
MCH (pg)	27.16± 4.70	20.82 ± 3.90	≥0.05
MCHC (g/dl)	33.05± 1.86	34.65± 3.80	≥0.05
RDW (%)	12.52± 2.77	19.94± 2.56	≤0.05

There was a statistically significant difference in the haemoglobin concentration ($P<0.05$), mean cell volume ($P<0.01$), red cell distribution width ($P<0.05$) and platelet count between two groups. However TLC, MCH & MCHC were not significantly different.

IV. Discussion

In present study, the haemoglobin level was significantly low in patients of SCD. Haemoglobin concentration remained significantly low in SCA patients.[6,7,10,14]The SCD patients suffer from continuous hemolysis of red cells, with a short survival rate of the erythrocytes between 12-14 days. Hence, the haemoglobin values are usually low[6]

The mean total white blood cell count (WBC) in this study were similar to other earlier studies.**[2,6,7]** Studies reported rise in TLC in patients of SCD, as compared to control groups. But in our study, it was not statistically significant (>0.05). WBC counts are correlated with the severity of crisis and risk of premature death. Insignificant rise in leucocyte count shows the steady state of the patients of our study.[11]

The mean platelet count for steady state value is similar to other studies. The platelet count in patients of SCD was significantly higher than sickle cell trait ($P \leq 0.05$) – which may be due to loss of splenic platelet pool function in adult sickle cell patients consequent upon autosplenectomy. This result is in agreement with previous studies.[6,7,10]Platelet aggregate formation activity is normal in SCD patients in steady state, but abnormally high in each SCD patient with vaso-occlusive crisis. It is a matter of further research that whether increased platelet aggregate formation during acute thrombotic event is primarily responsible for the initiation of the event, or is a secondary phenomenon in response to tissue injury.[12]

In this study, the mean corpuscular volume (MCV) value was significantly lower in cases of SCD, as compared to sickle cell traits groups – which is similar to previous studies conducted by Omoti C E, Ahmed S G &Tripette J. In contrast, there was no significant difference in the MCH & MCHC values in present study.

The RDW, which is a measure of erythrocyte anisocytosis, was significantly higher in as compared to sickle cell traits ($P<0.05$). This is in agreement with previous studies, which showed that SCD is associated with marked anisocytosis. This may be because of more rapid erythropoiesis, in which, cells at different stages of maturation, with different sizes, and are present at the same time.

However, this study is limited by the fact that we have taken patients of steady state and not of crisis state. The picture might have been slightly different if patients of steady state and sickle cell crisis both have been taken. The age group which is considered was also very broad. Other biochemical parameters which can assess oxidative and anti-oxidative properties could have also been included. Further studies including all confounding factors are required to assess the effect of these hematological findings with patients of sickle cell anemia and its effective management

V. Conclusion

Recent therapeutic approaches to SCD focus on attempt to reduce intracellular HbS polymerization by altering the haemoglobin species. So monitoring all the haematological indices has a great diagnostic & prognostic value in patients of sickle cell anaemia either in steady or crisis state. The health status and life expectancy of these patients can be improved considerably by monitoring these parameters.

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