A study of Blood indices in Sickle Cell Disorder

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ABSTRACT:

BACKGROUND: Sickle cell disorder is second most common hemoglobinopathy after Thalassemia. Patients present with wide spectrum of disorders because of a single-point mutation in which thymine substitute for adenine, thereby encoding valine-instead of glutamine in sixth position of beta chain. Haemoglobin S causes polymerization of haemoglobin and red cell sickles on exposure to low oxygen tension and not sickle on oxygenation. Hemolysis occurs due to quantitative and qualitative changes in red blood cell. Hemolysis could be intravascular as well as extravascular which affect the Indices of Red blood cells resulting in low Hb, low RBC count and variations in RBC indices. **AIMS and OBJECTIVE:** The study was conducted to examine the hematological profile in patients of sickle cell disorder. To study Mentzer Index and Srivastva Index in patients with sickle cell disorderand to assess whether there is prevalence of iron deficiency anaemia with sickle cell disorders. **METHODOLOGY:** This was a prospective observational study in which patients of sickle cell disorder confirmed by HPLC (High performance liquid chromatography) were taken and those with history of blood transfusion in past 3 months or with HIV infection was excluded. Total 537 patients were included in this study. **RESULT:** The mean Hb of the study population was 9.87±2.14%.The mean MCV was 73±9fl. The mean MCH was 31.68±12.87%.The mean PCV was 32±8.15%.The mean MCHC was 31.68±12.8%.The mean Srivastava Index was 5.81±2.83. The mean Mentzer Index was 18.47±8.81.

Keywords: Sickle cell disease, blood indices, Anaemia

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INTRODUCTION: The inherited disorders of blood incorporate hemoglobinopathies which are one of the significant health challenges in India [1].Sickle cell disease is the second most prevalent hemoglobinopathy, second to Thalassemia in India [2].The discoveries of the Indus valley civilization site show the pervasiveness of hereditary anaemia (sickle cell disease or β thalassemia) in the Indian subcontinent from around 2000-5000 BC [3].General infection of Sickle cell disease in India is 1-44%[4,5,6]. The average presence of haemoglobin S (HbS) is 4.3 % in India [4]. Quantitative and qualitative changes in RBCs have been observed. Haemolysis ensuing to the damaged red cell membrane could be intravascular or extravascular. The former event occurs as a result of the lysis of complement sensitive red cells [5] and haemoglobin lost during sickling-induced damage to the membrane of the RBCs[6,7]The latter event on the other hand mentioned, happens by phagocytosis of red cells that have gone through sickling [8,9] and physical entrapment of rheologically compromised red blood cells[10]. Increased vulnerability to mechanically induced cell breakdown has been reported in-vitro and in sickle cell patients undergoing strenuous physical activity [7]. Degree of haemolysis is related in an inverse manner with the concentration of hemoglobin and packed cell volume in patients of sickle cell anaemia. [11]. Various elements influence haemolysis in sickle cell anaemia, amongst which the amount of the irreversible sickle cell is of most prominent significance [11].The degree of hemoglobin polymer development, determined from the mean corpuscular hemoglobin concentration and the relative proportion of hemoglobin fractions additionally relates to a great extent with the severity of haemolysis [12,13].The uniformity of RBC population can be known from RDW. Evaluation of these values is an integral part of determining the cause of anaemia. Most of the reported cases of sickle cell disorder have documented normal or increased MCV. Few of the reports especially in India have suggested decreased in MCV There is paucity of data on blood indices of SCD

from India. Thus this study was undertaken to study to determine the profile of blood indices in patients of sickle cell disorder and to study Mentzer & Srivastava index amongst the Sickle cell patients visiting: a rural tertiary care setup in Western part of India.

METHOD: It was a prospective observational study which was carried out between February 2014 to July 2016 after approval of the Institutional Ethics Committee. All the patients who were screened by sickling solubility test & confirmed by HPLC, having sickle cell disease(homozygous) or trait were included while patients with History of blood transfusion in past 3 months HIV infected patients were excluded from the study. A total of 537 patients (417 were Sickle cell trait patients and 120 were Sickle Cell Disease patients) were enrolled in the study after taking their written informed consent. After obtaining and documenting complete and proper history, under all aseptic precautions, 2 ml of blood was drawn from antecubital vein by clean venepuncture from each patient with a sterile plastic syringe and collected in an EDTA (anticoagulant) tube for determination of investigations like Sickling test, CBC (Complete Blood Cell count), Hemoglobin electrophoresis, Reticulocyte count. Blood indices were done with Bio Rad D-10 hemoglobin testing system. Hemoglobin analysis was done with High performance liquid chromatography (HPLC).

The hematological profile consisted of: Hemoglobin (Hb), Hematocrit (HCT), RBC, Platelet count, WBC count, WBC differential count, MCH (Hb/RBC count), MCV (HCT/RBC count), MCHC (Hb/ Hematocrit), and RDW (RBC volume) in CV. The following RBC Discriminate indices were calculated: Mentzer Index: MCV/RBC count, Srivastava Index: MCH/RBC count,

STATISTICAL ANALYSIS

Data wasanalyzed using SPSS version 16.0 (Statistical Package for Social Sciences, Inc., Chicago, Ill). The descriptive data were given as means ± standard deviation (SD)

RESULTS: There were 537 subjects who participated in the study. The mean Hb of the study population was9.87±2.14gm%. The mean Hb of the patients with SCD (sickle cell disease) was8.36±2.16 while mean Hb of patients with SCT (sickle cell trait) was 10.30±1.90. The mean MCV of the study population was 73±9 fl . The mean MCV of the patients with SCD was75.04±10.69 while mean MCV of patients with SCT was72.44±9.07. The mean MCH of the study population was 23±3.66pg. The mean MCH of the patients with SCD was 24.37±3.78 while mean MCH of patients with SCT was22.55±3.53. The mean PCV of the study population was 32±8.15%. The mean PCV of the patients with SCD was 26.77±9.07while mean PCV of patients with SCT was 33.37±7.79. The mean MCHC of the study population was 31.68±12.8%. The mean MCHC of the patients with SCD was31.78±2.05 while mean MCHC of patients with SCT was4.29±24.30. (Table 1)

Indices	SCD+SCT	SCD	SCT	Normal values
Mean Hb	9.87±2.14gm%	8.36±2.16	10.30±1.90	13-17gm%
Mean RBC	4.4±1Mil/µl	3.52±1.00	4.61±0.90	4.4-5.9Mil/µl
Mean PCV	32±8.15%	26.77±9.07	33.37±7.79	Males:40-53% Females:36-47%
Mean MCV	73±9fl	75.04±10.69	72.44±9.07	82-92fl

Mean MCH	23±3.66pg	24.37±3.78	22.55±3.53	27-31pg
Mean MCHC	31.68±12.87%	31.78±2.05	31.65±14.56	32-36%
Mean Platelet count	4.1±5.2lac/cumm	3.452.05	4.29±24.30	1.5-4.5lac/cumm

Table 1: Mean Hb, RBC indices and Platelet counts of the study population along with the normal reference range

The mean WBC count of the study population was11304±10328The mean WBC count of the SCD patients was11759±7960 while that of SCT patients it was11172±10921. (Table2)

Indices	SCD+SCT	SCD	SCT	Normal values
Mean WBC count	11,304±10,328	11759±7960	11172±10921	4000-11000
Polymorph	67±14%	60.77±17.13	69.00±12.81	60-70%
Lymphocyte	24±12%	30.20±14.50	22.28±10.89	20-35%
Eosinophil	4±3%	4.11±3.56	3.73±2.41	1-4%
Monocyte	4±1%	4.02±1.40	4.03±1.37	2-6%

Table 2: Mean WBC counts of the study population.

From the obtained hematological values, the Mentzer index, Srivastava index and RDW CV was calculated. The mean Mentzer index of study population was 18.47±8.81. It was for SCD

patients and for SCT patients. The mean Srivastava index of study population was 5.81±2.83. It was for SCD patients and for SCT patients. The mean RDW CV of study population was 19±14.7%. It was for SCD patients and for SCT patients. (Table 3)

Indices	SCD+SCT	SCD	SCT	Normal values
Mentzer	18.47±8.81	24.46±13.56	16.39±5.87	13
Srivastava	5.81±2.83	7.94±4.28	5.14±1.95	3.8
RDW-CV	19±14.7	20.22±3.82	18.61±16.56	13-15%

Table 3 Mentzer index, Srivastava index and RDW CV of the study population.

Index	IDA	Beta Thalessemia	Our study
RBC count	<5	>5	4.4±1Mil/µl S/O IDA
RDW-CV	>14	<14	19±14.7. S/O IDA
Mentzer Index	>13	<13	18.47±8.81 S/O IDA
Srivastava Index	>3.8	<3.8	5.81±2.83 S/O IDA

Table 4 Various indices and comparison between IDA and Beta thalassemia and correlation with current study

DISCUSSION:

In the present study, mean Haemoglobin, MCV and MCH was lower which was suggestive of microcytic hypochromic anaemia. This indicated that sickle cell disease could be associated with Iron deficiency anaemia. Rao et al. had obtained similar results in a study conducted in 2009-10 in a tertiary care hospital in south Gujarat (14). Similar findings were found in a couple of other studies in different parts of our country (15, 16). We derived the Mentzer index and Srivastava index and found that they were higher which again was hinting towards iron deficiency anaemia. Also, the mean RDW was higher suggestive of nutritional iron deficiency associated with sickling disorder (Table 4) The Mentzer index provided the highest reliabilities for differentiating β-TT from IDA. Sensitivity (98.7%), specificity (82.3%)[17]The Srivastava and Bevington Index had a sensitivity of (IDA:79, β-TT:74%) and specificity (IDA:74% β-TT 79%)[18]

CONCLUSION: In our study population MCV and MCHC was lower. RDW- CV was higher Mentzer and Srivastava index was higher. The above findings are suggestive that patients of Sickle cell disorder might have associated Iron deficiency anaemia. Similar large scale studies are required nationwide especially in areas with a strong predominance of Sickle cell disorders so as to better establish a strong correlation between sickle cell disorders and Iron deficiency anaemia and ultimately to enhance our knowledge and understanding about this blood disorder.

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