RESEARCH ARTICLE

Hematological Profile of Sickle Cell Anemic Subjects from Gadchiroli District, Maharashtra

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ABSTRACT

After Thallassemia sickle cell anemia is the major hemoglobinopathy in India. It is common autosomal recessive disease due to single nucleotide substitution (GTG-GAG). Observational study was conducted to determine the hematological values of sickle cell subjects and patients from April 2010 to April 2012. 27 carrier subjects (heterozygous Hb-AS) and 24 sufferers (homozygous -HB-SS) were studied at steady state. Our study shown Reduction in level of RBC, MCV, Hemocrit, MCH, low level of platelet found in the patients with Hb-SS pattern as compared to the Hb-AS subjects. MCHC found normal with increased level of RDW% presenting the symptoms of crisis, the mean hemoglobin found significantly lower in SS patient as compared to Hb-AS subjects this difference is because of presence of HB- A in sickle cell traits which causes less number of RBCs to undergo sickling and further hemolysis, while in Hb-SS subjects promotes sickling and increased rate of RBC destruction leading to profound anaemia. Total WBC count and differential leukocyte count showed normal to elevated in both Hb-AS subjects, as well as Hb-SS subjects.

Key words: Hemoglobin, Sickle cell anemia, Complete blood count, Gadchiroli District.

INTRODUCTION

Sickle cell anaemia is known to the medical world since the discovery of this entity (Herrick, 1910) a Chicago cardiologist. He first provided the formal description of sickle cell anemia, he reports that the blood smear of a dental student at the Chicago College of Dental Surgery contains pear shaped and elongated forms. Herrick (1910) report led not only to the recognition of hundreds of abnormalities of hemoglobin synthesis but, also to a series of remarkable scientific advances involving protein Chemistry, Cell Biology, Physiology, and Genetics. The discovery of hemoglobin- S (Hb-S) Pauling *et al.* (1949) was the first demonstrated the production of an abnormal protein could be the cause of a genetic

disorder**.** In India it is the second most dominated haemoglobinopathy after Thallassemia, it is most common in central and southern part of India. Genetically it is an autosomal recessive disorder characterized by the substitution of valine for glutamic acid at position 6 in the β-Globin chain (Hb-S). This results in a solubility problem in deoxygenated state and upon deoxygenation the affected RBCs changes from biconcave discoidal cell to crescent of sickle shaped cell (Evans and Mohandas, 1987)**.**

This major haemoglobinopathy occurs in both homozygous and heterozygous state, red cell contain both normal adult hemoglobin (Hb-A) and the variant, because they rarely have phenotypic expression of clinical significance, heterozygous is said to have the trait for that abnormality, e, g. sickle cell trait. In the homozygous state, Hb-A is totally lacking, and clinical manifestation is of variable severity; individuals so have the anemia called sickle cell anemia. Gadchiroli is a newly carved district, of Maharashtra with a major population of Gond and Madia tribes being most backward and Naxlite hit, the district lag behind in healthcare facility from rest of the region. Though various aspects of SCA are studied aspects with reference to subjects from Gadchiroli district of Maharashtra, are unclear.

MATERIAL AND METHODS

The study conducted in Gadchiroli district, of Maharashtra with a major population of Gond and Madia tribes. The district is having 45 PHC's distributed in Gadchiroli-3, Korchi-2, Kurkheda-3, Wadsa- 3, Armori-4, Dhanora-5, Chamorshi-6, Etapalli-3, Mulchera-3, Bhamragarh-3, Aheri-5, and Sironcha-5. Fifty one SCA subjects visiting different PHCs of district during April 2010 to April 2012 were studied. 27 carrier subjects (heterozygous - HbAS) and 24 sufferer subjects (homozygous - HbSS) proceeded for the complete blood count.

About 3-5 ml of blood sample from both heterozygous and homozygous subjects was made in two vials for the haematological evaluation with the help of concerned staff member with due permission. Blood collected by vein puncture, CBC is done using the Blood Cell Counter following parameters studied. Red Blood Cell Count (RBC), Mean Corpuscular Volume (MCV), Red Cell Distribution width (RDW %), Hemocrit (PCV), Total Platelet Count (PLT), Mean Platelet Volume (MPV), White Blood Corpuscles (WBCS), Hemoglobin (HGB), Mean Corpuscular Hemoglobin (MCH), Mean Corpuscular Hemoglobin Concentration (MCHC), Lymphocyte Percentage. (LYM %), Total Granulocyte Percentage (GRA%).

RESULTS AND DISCUSSION

A complete blood count (CBC) gives important information about the kinds and numbers of cells in the blood, especially red blood cells , white blood cells , and platelets. A CBC helps in provide baseline haematological values in sickle cell disease patients that can be used in monitoring the status and management of sickle cell anemia patients. The count suggested that Sickle cell disease is a genetic abnormality primarily, involving the hemoglobin and red cell. The white blood cells and platelets are also affected by the mutation.

Table 1: Showing Hematological Profile of Hb-AS patients (Carrier, N = 27)

Sr. No.	Parameters	Mean	S.D.	SEM
1	**RBC**	3.6337	± 1.3203	0.2541
2	**MCV**	76.589	± 10.497	2.02
3	**RDW %**	17.341	± 2.276	0.438
4	**HCT**	26.444	± 7.958	1.532
5	**PLT**	217.93	± 101.46	19.53
6	**MCHC**	34.969	± 1.294	0.249
7	**MPV**	7.619	± 0.914	0.176
8	**WBC**	7.089	± 2.769	0.533
9	**HGB**	10.33	± 2.511	0.483
10	**MCH**	25.785	± 5.776	1.12
11	**LYM%**	49.026	± 15.505	2.984
12	**GRA%**	40.726	± 16.419	3.16

Table 2: Showing Hematological Profile of Hb-SS patients (Diseased, N =24)

Sr.No.	Parameters	Mean	S.D.	SEM
1	**RBC**	1.954	± 0.4602	0.0939
2	**MCV**	72.658	±11.467	2.341
3	**RDW %**	20.079	± 4.350	0.888
4	**HCT**	15.329	± 3.469	0.708
5	**PLT**	149	± 56.17	11.47
6	**MCHC**	37.063	± 3.825	0.781
7	**MPV**	7.604	± 1.094	0.223
8	**WBC**	4.587	± 2.573	0.525
9	**HGB**	5.454	± 1.016	0.207
10	**MCH**	24.921	± 3.212	0.656
11	**LYM%**	48.183	± 13.038	2.661
12	**GRA%**	41.621	± 11.935	2.436

In these findings we studied, 27 carriers Hb-AS and 24 sufferer Hb-SS subjects for complete blood count. Red Blood Cell count, (Hb-AS – 3.637± 1.3203, Hb-SS – 1.954 ± 0.4602, $p < 0.0001$) MCV (Hb-AS – 76.589 ± 10.497, Hb-SS – 72.658 ± 14.558 , $p < 0.0001$) and Hemocrit (Hb-AS – 26.444 ± 7.958, Hb-SS- 15.329 ± 3.469 % $p < 0.0001$), differed statistically showing decreased level of hematological indices Davies *et al.,* (1983) reported the same, but Powars *et al.,* (1980) reported no correlation between above parameters. Our findings showed decreased level of RBC, MCV, and Hemocrit (PCV), Kar *et al.,* (1986) observed lower mean corpuscular volume. Balgir (2000) found low MCV (60-113 ft) Roy *et al.,* (1996), Rao *et al.* (2012) also observed low level of above parameters and supports to our studies Akuzawa *et al.* (1989) also reported the low level of above parameters indicating hemolytic anaemia and pathological erythrocytes. In our studies observed MCH was low in Hb-SS patients as compared to Hb-AS (Hb-AS-25.785 ± 5.776, SS- 24.921 ± 3.212) with MCHC (Hb-AS-34.959 ± 1.294, Hb-SS-37.063 ± 3.825) and increased red Cell Distribution Width (AS- 7.341 ± 2.276, SS-20.079 ± 4.350%) Low MCH and MCHC reported by Kar *et al.,* (1986) and Balgir, (2000) (low MCH in India it ranges from 0.25-0.42g/dl) Rao *et al.,* (2012) reported low level of MCH and MCHC increase in percentage of red cell distribution width reported by Schweiger, (1981), Webster and Castro, (1986) and Sayed and Tawfik (1994).

Total platelet count and mean platelet volume (Total platelet count in Hb-AS- 217.93 ± 0.914, Hb-SS- 149 ± 56.17 and Mean platelet volume Hb-AS – 7.619 ± 0.914 and Hb-SS- 7.604 ±1.094) shown little difference in the observation total platelet count and mean platelet count and mean platelet count found normal in Hb-AS subjects whereas in diseased (Hb-SS) patients it was little decreased in mean of total platelet count and mean platelet count Ibanga, (2006) found significant rise in platelet count in steady state patients 224+/- 46.3X10(9) L when compared with others of 196.6 +/- 39.3X10(9) L ($p < 0.05$) and platelet fall during Vasoocclusive crises to 140.6+/- 36.3 X (10)/L ($p<0.05$), Haut (1973) studied platelets survival recovery, the result indicated that during the stable period platelets function is normal and that platelets survival is normal or greater than normal. Variation in platelet activity and function also studied by Henstell *et al.,* (1965), Noroha *et al.,* (2007), Elderdery *et al.,* (2011).

The mean difference in hemoglobin level of sickle cell anaemia subjects found in trait (Hb-AS-10.33 ± 2.511 and Hb-SS- 5.454 **±** 1.016) is significantly higher than Hb-SS subjects of ($p<0.0001$) this difference is because of the presence of one normal Hb-A in sickle cell trait subjects which causes less number of RBCs to undergo sickling and further hemolysis in Hb-AS subjects while in diseased subjects both Hb-S promotes sickling and increased rate of RBC destruction leading to profound anaemia. The mean hemoglobin in Hb-AS female was significantly lower than male Hb-AS subjects may be because of average Hemoglobin levels are normally 1.5 gm% lower in females as compared to males. This may be also because of low socioeconomic status, in previous studied similar observation were noted by Serjeant *et al.,* (1968) they found 18.33%) subjects with Hb of ≥10 gm%, (46.67%) patients in the range of 8-9.9

gm% and (31.67%) subjects in the range of 6-7.9 gm% while (3.3%) subjects with Hb below 6 gm% of the total 60 Hb-SS subjects studied Kar *et al.,* (1986) found mean Hb of 8.73 ± 1.69 gm% (range 3.9 – 13.5 gm%) amongst 131 Hb-SS subjects studied. Kar and Devi (1997) reported 6-10 g/dl mean with which they thrived well Davies *et al.,* (1983) found 7.1 – 9.2 g/dl and Rao *et al.,* (2012) reported low hemoglobin in 33 subjects. From observation it is evident that normal WBCs in Hb-AS and Hb-SS patients (Hb-AS – 7.089 ± 0.533 and 4.587 ± 0.525), with difference in Lymphocyte percent and Granulocyte percentage (Lym%, in Hb-AS-49.26 ± 15.505 %, Hb-SS - 48.183 **±** 13.038% and Gra%, in Hb-AS- 40.726 ± 16.419 % in Hb-SS- 41.612 ± 11.935%). The rate of chronic haemolysis associated with sickle cell anaemia subjects could account for all these disturbed values. In view of Sherwood *et al.* (1987) there is a blunted response to erythropoietin secretion in sickle cell anaemia, hence the rate of increase is not proportional to the degree of anaemia. Morris *et al.*, (1991) pointed out that it may be due to right shifted haemoglobin dissociation curve seen in sickle cell disease. Similarly lower values were obtained by Diggs (1965) in his hematological studies in sickle cell disease found that people in stable condition do not have any significant hematological deviation. The studied patients also did not show much deviation from normal in stable condition. This study provides hematologic reference ranges for sickle cell disease patients compared with normal controls in the district of Gadchiroli. It is our hope clinics involved in supervision of sickle cell anaemia patients would become more up to date and make use of the result in this study in their practice ranges for sickle cell disease patients compared with normal controls here. The study would make expert involved in inspecting sickle cell anaemia subjects to become more knowledgeable and help them make use of these findings in their skill to manage this genetic menace in the region.

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