

INSIGHTS OF SICKLE CELL ANEMIA; CROSS SECTIONAL STUDY FROM AKOLA DISTRICT MAHARASHTRA

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Abstract

The blood contains several different types of cells. Each of these cell types is quite distinct in appearance, and each has a specific biologic function. Sickle cell disorder is a group of diseases caused by a point mutation at sixth position in beta Globin chain, Valine substituting Glutamic acid due to which in deoxygenated state, shape of erythrocytes change to sickle shape and also the fragility of cell membrane increases. Sickle cell anemia is a genetic disorder affecting haemoglobin the oxygen-transporting protein in red blood cells. Present study aims to Document Haematological profile of sickle cell anaemic subjects and Variation in pattern of haematological profile in HB-As and In HBSS subjects from Akola district of Maharashtra (India). In these findings we studied, 24 carriers Hb-AS and 7 sufferer Hb-SS subjects for complete blood count. Red Blood Cell count, (Hb-AS- 2.5559 ± 2.1422 , Hb-SS- 2.4186 ± 1.5875 , $p < 0.0001$) MCV (Hb-AS – in Hb-AS was 59.805 ± 19.813 , and in Hb-SS was 71.643 ± 34.067 , $p < 0.0001$) and HCT (47.191 ± 64.301 in AS, 22.1071 ± 12.6643 in SS patients), WBC count in present study 9.1436 ± 5.3290 in AS and in SS 10.2686 ± 4.4474 which differed statistically showing decreased level of haematological indices.

Key words: Sickle cell anemia, Hb-variants, Haematological indices

Introduction

The blood contains several different types of cells. Each of these cell types is quite distinct in appearance, and each has a specific biologic function. Erythrocytes are a nucleate, biconcave discoid cells filled with haemoglobin, the major protein that binds oxygen. Red blood corpuscles (RBCs) are the non-nucleated formed elements in the blood. Red blood cells are also known as erythrocytes (Erythro = Red). Red colour of their blood cell is due to the presence of the colouring pigment called haemoglobin. RBCs play a vital role in transport of respiratory gases. RBCs are larger in number compared to two other cells, namely white blood corpuscles and platelets. Normal value of RBCs ranges between 4 and 5.5 million/cu mm of blood. In adult males it is 5million/ cu mm and in adult females, it is 4.5 million. cu mm. (Winstrobes Clinical Haematology, 2003). Red blood cells are non-nucleated. Only mammal which has nucleated RBC is camel. Because of the absence of nucleus in human RBC, the DNA is also absent. Other organelles such as mitochondria and Golgi apparatus also are absent in RBC. Because of absence of mitochondria, the energy is produced from glycolytic processes. Normally, the RBCs are (disk shaped) dumbbell shaped. Central portion is thinner and periphery is thicker. Haemoglobin (Hb) molecules are a set of proteins formed by pairing of two alpha

(α) and two beta (β) globin polypeptide chains into a tetramer unit (Embryo *et al.*, 1994 and Schechter, 2008 and Sargent, 1997). This unit, the $\alpha_2\beta_2$ molecule, forms the major adult haemoglobin responsible for the transport of oxygen from the lungs to tissues. The α -globin gene is located on chromosome 16 and encodes the adult α -globin and the ζ -globin chain, which is the embryonic form of α -globin (Schechter, 2008). As a result of the replacement of negatively charged hydrophilic glutamic acid with the non-polar hydrophobic valine at position six on the 146 amino acid β -chain (Ingram, 1956 and Shelton and Schroeder 1960). Sick cell anemia is transmitted from one generate onto another, due to acquisition of two abnormal genes one from each parent. Sick cell anemia evolves when a heterozygote (AS) (carrier) marries either a fellow carrier or a homozygote (SS) (sufferer). Many clinical manifestation are produced due to sickle cell anemia.

Objective

Present study deals with Documentation of Haematological profile of sickle cell anaemic subjects from Akola district, Maharashtra.

Materials and Methods

Cross sectional study of diagnosed patients of sickle cell disorder was conducted in Department of Pathology of Government Medical College and district Hospital, Akola from Consecutive records one year of patients of IPD, from 01/01/2023 to 31/03/2023. Confidentiality of patients was maintained. Blood sample was collected in EDTA test tube and haematological indices HB%, MCH, MCHC, MCV, WBC, RBC, PLT, MPV RDW (RDW-CV, RDW-SD), were measured by C.B. counter machine (auto analyser) and confirmed by Sickling method and electrophoresis test. Sickling test was done by Solubility test and electrophoresis test done by using cellulose acetate strip at alkaline Ph to differentiate and confirmation of sickle cell disease and sickle cell trait. Nalbandian (1971) and Norbert Tietz (1999). The data was analyzed statistically following the method adapted by Arora and Malhan (1996).

Observation and Results

Blood components are produced by the process of haematopoiesis where production and maturation takes place; it consists of erythropoiesis, thrombopoiesis and leucopoiesis, under normal conditions, production of blood cells carried out by bone marrow a totipotent stem cell precursor for erythrocyte, lymphocytes and granulocytes. Solubility test positive subjects were considered for the electrophoresis and Hb-AS and Hb-SS patterns were determined. in present study AS and SS subjects were selected for Complete Blood Count (CBC).

In present study and 24 Hb-AS and 7 Hb-SS patients were subjected for complete blood count. Following results were obtained for Blood count as given in table 1 and 2.

Table no. 1: Showing Hematological Profile of Hb-AS patients (Carrier, N = 24)

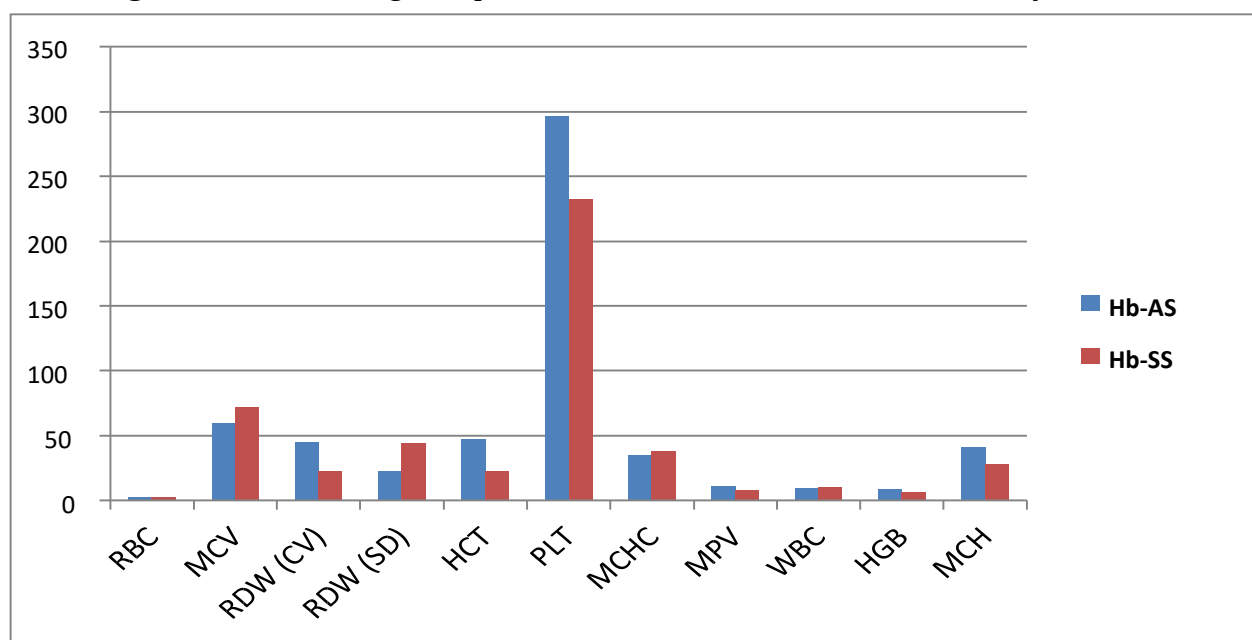
Sr. No.	Parameters	Mean	S.D.	SEM
1	RBC	2.5559	2.1422	0.4567
2	MCV	59.805	19.813	4.224
3	RDW (CV)	44.955	28.857	6.152
4	RDW (SD)	22.505	7.593	1.619

5	HCT	47.191	64.301	13.709
6	PLT	296.82	173.07	36.90
7	MCHC	34.627	17.888	3.814
8	MPV	11.123	1.986	0.423
9	WBC	9.1436	5.3290	1.1362
10	HGB	8.7323	3.7372	0.7968
11	MCH	40.700	16.393	3.495

Table no. 2: Showing Hematological Profile of Hb-SS patients (Diseased, N = 7)

Sr. No.	Parameters	Mean	S.D.	SEM
1	RBC	2.4186	1.5875	0.6000
2	MCV	71.643	34.067	12.876
3	RDW (CV)	22.957	7.491	2.831
4	RDW(SD)	44.157	22.209	8.394
5	HCT	22.1071	12.6643	4.7867
6	PLT	232.71	180.60	68.26
7	MCHC	37.843	21.860	8.262
8	MPV	7.7629	3.9071	1.4768
9	WBC	10.2686	4.4474	1.6809
10	HGB	6.5729	3.9802	1.5044
11	MCH	27.929	4.701	1.777

Figure no. 1: Showing Complete blood count in Hb-AS and Hb-SS Subjects.



Reduction in the number and concentration of red blood cells and content of haemoglobin is the cause of anaemias, erythrocytes and other blood cells are of extreme importance in relation to blood transfusion. As depicted in table 1 and 2 calculated Mean $\pm$ S.D in Hb-AS- 2.5559 ± 2.1422 , Hb-SS- 2.4186 ± 1.5875 . Normal level of red blood cell count is 3.50 -5.50 10¹²/l. It is the indicator of size of red blood cells, as shown in table 4 and 5 MCV in Hb-AS was 59.805 ± 19.813 , and in Hb-SS was 71.643 ± 34.067 , $p < 0.5$ MCV is 75-100 fl in normal. It occurs between 11-16% red cell distribution is measured as a coefficient of variation of red cell size distribution. As given in table Mean $\pm$ S.D of RDW % in Hb-AS was 44.955 ± 28.857 in CV and in SD it was found to be 22.505 ± 7.593 and in Hb-SS, 22.957 ± 7.491 % CV and SD was found to be 44.157 ± 22.209 . HCT was found to be 47.191 ± 64.301 in AS subjects whereas in SS diseased subjects it was found to be 22.1071 ± 12.6643 . As given in table 1 and 2 mean $\pm$ S.D of total platelet count in Hb-AS is 296.82 ± 173.07 and in Hb-SS is 232.71 ± 180.60 . In normal person it is 150 – 450. It is determined by computing arithmetic mean volume, MPV in Hb-AS – 11.123 ± 1.986 and Hb-SS - 7.7629 ± 3.9071 fl. calculated with mean $\pm$ S.D, in normal person it is 811 fl. It is the count of total WBCs cu/mm of blood. WBCs count in Hb-AS was 9.1436 ± 5.3290 and in Hb-SS was 10.2686 ± 4.4474 . Normal person is having 3.5 –10. Decrease in Haemoglobin concentration below normal range is the indication of anemia. As shown in table 1 and 2 haemoglobin in Hb- AS - 8.7323 ± 3.7372 and Hb-SS- 6.5729 ± 3.9802 g/dl, in normal person HGB is 11-16.5 g/dl.

It is the amount of haemoglobin in average red cell, as given in the table 1 and 2 in Hb-AS patients it was 40.700 ± 16.393 , and in Hb-SS patients it was 27.929 ± 4.701 pg. In normal person it is 25-35 pg. As shown in the table 4 and 5 mean corpuscular haemoglobin concentration in Hb-AS patients was 34.627 ± 17.888 and 37.843 ± 21.860 in Hb-SS patients. Mean corpuscular haemoglobin concentration is an estimate of the concentration of haemoglobin in a given number of packed red blood cells; it is 31-38 g/dl in normal person.

Discussion

In these findings we studied, 24 carriers Hb-AS and 7 sufferer Hb-SS patients for complete blood count. Red Blood Cell count, (Hb-AS- 2.5559 ± 2.1422 , Hb-SS- 2.4186 ± 1.5875 , $p < 0.0001$) MCV (Hb-AS – in Hb-AS was 59.805 ± 19.813 , and in Hb-SS was 71.643 ± 34.067 , $p < 0.0001$) and HCT (47.191 ± 64.301 in AS, 22.1071 ± 12.6643 in SS patients), WBC count in present study 9.1436 ± 5.3290 in AS and in SS 10.2686 ± 4.4474 which 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 Kar *et al.*, (1986) observed lower mean corpuscular volume. Balgir (2000) found low MCV (60-113 fl) 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 Parasitol, (1994). Total platelet count and mean platelet volume (Total platelet count in Hb-AS is 296.82 ± 173.07 and in Hb-SS is 232.71 ± 180.60) shown little difference in the observation total platelet count and mean platelet count found normal in Hb-AS patient 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 $224 \pm 46.3 \times 10^9$ L when compared with others of $196.6 \pm 39.3 \times 10^9$ L ($p < 0.05$) and platelet fall during Vasoocclusive crises to $140.6 \pm 36.3 \times 10^9$ L ($p < 0.05$). The mean difference in haemoglobin level of sickle cell anaemia patients found in trait (8.7323 ± 3.7372 and Hb-SS- 6.5729 ± 3.9802 g/dl) is significantly higher than Hb-SS patients of ($p < 0.0001$) this difference is because of the presence of one normal Hb-A in sickle cell trait patients which causes less number of RBCs to undergo sickling and further hemolysis in Hb-AS patients while in diseased patients both Hb-S promotes sickling and increased rate of RBC destruction leading to profound anaemia. This may be also because of low socioeconomic status, in previous studied similar observation were noted by Kar *et al.*, (1986) found mean Hb of 8.73 ± 1.69 gm% (range 3.9 – 13.5 gm%) amongst 131 Hb-SS patients studied. Kar and Devi (1997) reported 6-10 g/dl mean with which they thrived well. Difference in RDW was seen among as subjects and in SS patients, RDW-CV in AS 44.955 ± 28.857 in SS 22.957 ± 7.491 and in RDW-SD AS, 22.505 ± 7.593 , in SS 44.157 ± 22.209 these finding was shown similarities with the studies of Kar and Devi (1997) and Balgir also reported quite similarity in 2000. Gaikwad *et al.*, (2017) and Kohchale and Misar (2021) documented the hematological profile of sickle cell subjects which shows similarity with our result and interpretation. Complete blood count gives idea of different symptoms of sickle cell anemic subjects. There for CBC plays role preferring treatment to the sickle cell anemic subjects.

Conclusion

Present study showed the documentation of hematological profile of sickle cell disorder patients has low value of Hb%, HCT, total RBC count, MCH, MCHC with normal MCV, MPV and platelets count and high RDW%. These all parameters are suggestive of different symptoms and different suffering to the victim suffering from sickle cell disorder.

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