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Hematological Parameter of Sicklers in Steady State at the Sickle Cell Center, Alkuwity Teaching Hospital, Elobeid City, North Kordofan Estate, West of Sudan: A Cross-Sectional Study

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Abstract

Background: Sickle Cell Anemia (SCA) is an inherited disease with high prevalence. It is a major cause of morbidity and mortality among African nations where there is no readily available effective treatment. Sickle Cell Disease (SCD) is a genetic blood disorder affecting red blood cells, with high morbidity and mortality rates.

Objective: The objective of this study was to provide baseline hematological values in sickle cell Anemia patients in steady state and to establish hematological reference values characteristics for Sudanese patients with sickle cell anemia treated at the Sickle Cell Center, Alkuwity Teaching Hospital, Elobeid City, North Kordofan Estate, West of Sudan, sickle cell disease.

Materials and Methods: A cross sectional study was carried out in the Sickle Cell Center, Alkuwity Teaching Hospital, Elobeid City, Sudan between March 2022 and December 2022. Two ml of venous blood sample was collected in Dipotassium Ethylene Diamine Tetra-acetic Acid (K2 EDTA) blood vacutainer for complete blood count from SCA patients in steady state and analyzed by automated hematology cell counter. All statistical analysis was done after recording the complete data of hematological parameters.

Results: This study had lower average values of total hemoglobin, Red Blood Cell (RBC) count and hematocrit. Higher values of Red Cell Distribution Width (RDW); and normal values of RBC indices. Higher values of Platelet count, Total Leukocyte Count, Neutrophils count, Neutrophils (%), Lymphocytes count, and Lymphocytes (%).

Conclusion: The average value of hematological parameters of SCA Sudanese patients in steady state shows moderate normocytic normochromic anemia, lower values of red cell parameters, but higher values of white blood cell and platelets counts. A larger scale work is recommended in this region for a baseline hematological profile for guiding the clinicians in management of these patients.

Keywords: Sickle cell anemia; Genetic blood disorder; Sickle Cell Disease (SCD); Hematological reference values; Red Blood Cell (RBC) count; Red Cell Distribution Width (RDW); Total Leukocyte Count; White blood cell count; Morbidity and mortality

Introduction

Sickle Cell Disease (SCD) is a genetic blood disorder of red blood cells, with high morbidity and mortality rates [1]. SCD is a genetic abnormality involving the haemoglobin. Patients with Sickle Cell Disease (SCD) have varying amounts of abnormal Haemoglobin called the sickle cell or "S" haemoglobin in their erythrocytes [2,3]. The haemoglobin S (HbS), is a structural variant of the normal Hemoglobin (HbA), be the result of a genetic mutation in the β-globin gene where thymidine replaced adenine is due to the substitution of adenine with thymine in the glutamic DNA codon (GAG→GTG), thereby encoding valine instead of glutamine in position 6 of the β-globin chain [4-7]. Sickle Cell Anemia (SCA) is the homozygous state for the Hemoglobin S (HbSS) gene, and it is the commonest and severest type of sickle cell disease. It runs a chronic steady state interrupted by episodes of acute complications. Sickle Cell Anemia (SCA) has been a prevalent disorder in Africa for many years ago. However, it was not officially described in America until November 1910 by Dr. James Herrick, a Chicago physician. Haemoglobin S causes polymerization of haemoglobin and red cell sickling on exposure to low oxygen tension (hypoxia) and unsickling on oxygenation. The repeated cycles of sickling and unsickling damages the red cell membrane reducing red cell life span as a result of membrane damage producing hemolytic Anemia. The clinical manifestations of SCD vary from mild almost asymptomatic periods of painless steady state that is periodically break off by severe painful Vaso-Occlusive Crisis (VOC) processes resulting in multisystem failure, tissue necrosis, ischemic organ damage, infections, that is associated with high mortality rates [8]. Clinical manifestations of SCA usually appear after three months of age, when the concentration of Fetal Hemoglobin (HbF) decreases. SCD leads to both quantitative and qualitative changes in RBCs. Even though sickle cell disease is mainly a disease of the red blood cell, the White Blood Cells (WBCs) and platelets are also affected by the mutation [9].

Assessment of hematological values of Sudanese sicklers is of great value. Any population is different from the other and there are many factors which can influence normal physiological values for a given set of population making it necessary to have reference values of the local population. These data will provide a basis to compare alarms caused by intercurrent complications like vaso-oclusive crisis and will

help health care providers in planning the proper intervention. In addition, the results of the present study were expected to fill the gap in the literature about the hematological values of Sudanese sicklers as a record as literature is scarce in local epidemiological studies in Sudan and will be used as "population norms". This is especially important if we consider the relatively high prevalence rates of sickle cell anemia in Sudan.

Materials and Methods

The present cross sectional study was carried out in the sickle cell clinic at Alkuwity Teaching Hospital, Sickle Cell Center, Elobeid City, North Kordofan Estate, West of Sudan between March to December 2022. The Center serves the population residing in the neighboring districts of West Sudan.

Inclusion criteria

- Confirmed patients of sickle cell anemia (SCA HbSS) diagnosed by alkaline haemoglobin electrophoresis at pH (8.6).
- Sickle cell patient in the steady state established by a steady hematocrit and haemoglobin values over a given period of 2-3 clinic visits at 4-6 weeks intervals and a state of well-being without any symptoms or signs suggestive of crisis established by a careful history and complete physical examination.
- Informed consent was taken from each participant.

Exclusion criteria

- Patients with history of complications in past two years.
- Patients with chronic conditions like renal failure, which can affect the hematological findings. Any patient with disorders that may affect the hematological values such as renal disease and pregnancy
- Patients who had received blood transfusion within past three months.
- Cases from whom informed consent could not be obtained.
- The heterozygous and double heterozygous patients haemoglobin phenotype SC patients.

Sample collection

Two ml of venous blood sample was taken under sterile conditions into Ethylene Diamine Tetra-Acetic Acid (EDTA) coated containing vacutainer tubes for Complete Blood Count (CBC) and analyzed by automated hematology cell counter (Sysmex KN-21 N,(manufactured by Sysmex corporation Kobe, Japan). Parameters including Hemoglobin (Hb), Red Blood Cell Count (RBC ct), Hematocrit (Hct), Red Blood Cell (RBC) Indices (Mean Cell Volume (MCV), Mean Cell Hemoglobin (MCH), Mean Cell Hemoglobin Concentration (MCHC)), Red Cell Distribution Width (RDW), Platelet Count (Plt Ct), Total Leukocyte Count (TLC), Neutrophil Count, Neutrophil (%), Lymphocyte Count, and Lymphocyte (%) obtained were noted. Result of analysis was displayed after about 30 secs. A printout copy of result is released on the thermal printing paper.

Statistical analysis

The data were analyzed by the Statistical Package of Social Science (SPSS Inc., Chicago, Illinois, USA) software version 24.0 in windows 10. Continuous variables are presented as means and standard deviation (M) ± Standard Deviations (SD) or median and interquartile range if the variable showed non-parametric distribution. Proportions of the studied groups were expressed in percentages (%) and absolute number (n) of frequencies. The confidence limit was kept at 95%, hence values less than 0.05 on the Chi-square distribution table is statistically significant (a P-value ≤ 0.05) was considered to be statistically significant. While greater than 0.05 (a P-value>0.05) on the Chi-square distribution table is not significant.

Results

A total 130 SCA patients in steady state attending the sickle cell clinic at Alkuwity Teaching Hospital, Sickle Cell Center, Elobeid City, North Kordofan Estate, and West of Sudan between Marchs to December 2022 were included. There were 58 males and 72 females (Table 1) (Figure 1) with age ranging from 2-29 years with a mean of 8 ± 5.804 years. 25.9% of sicklers were Falata, 15.8% were Misaria, 8.3% were Bidariaa, 3.5% were Gawamaa, 3.9% were Barno, 2.6% were Bargo, 35.5% were Hausa 2.6% were Silihab, 2.6% were Shanabla. Figure 2. Table 1 shows the general characteristics of the study group.

Hematological parameters	Mean (± SD)
Red blood cell count (x 106/l)	2.77 ± .63
Hemoglobin (g/dl)	8.41 ± 1.60
Hematocrit (%)	24.11 ± 4.55
Mean cell volume (fl)	86.78 ± 10.05
Mean cell haemoglobin (pg)	29.80 ± 2.99
Mean cell haemoglobin concentration (g/dl)	32.87 ± 0.77
Red cell distribution width-cv (%)	23.44 ± 4. 70
Red cell distribution width-sd	68.32 ± 13. 13
White blood cell count (x 103/l)	13.89 ± 5.48
Neutrophil (x 103/l)	6.96 ± 4.11
Neutrophils%	46.48 ± 13.02
Lymphocytes (x 103/l)	6.90 ± 4.57
Lymphocytes%	47.56 ± 12.64
Platelets (x 103/l)	453.71 ± 189.52

Table 1: Mean (± SD) of hematological parameter values of 130 patients with sickle cell disease (HbSS).

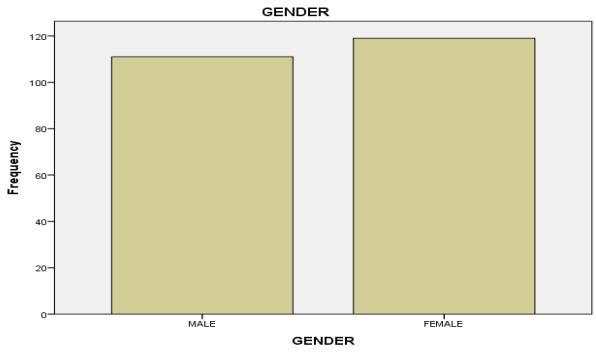

Figure 1: Gender of the participants.

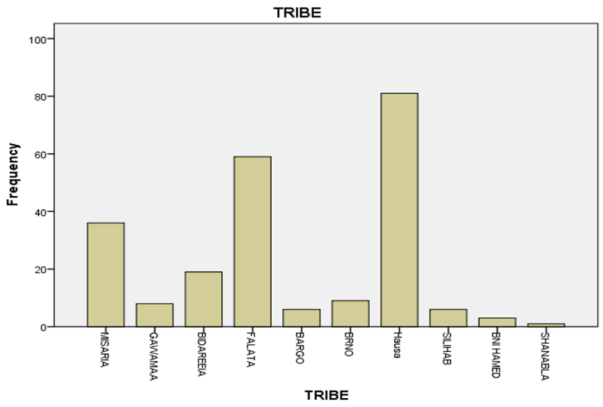

Figure 2: Tribe of the participants.

Hematological indices in study population

The hematological values obtained in steady state is shown in Table 1.

Red blood cells indices in study population

The overall mean RBCs count was 2.77 ± .63 (x 106/l). Most of the patients 126 of 130 patients (97%) had Red Blood Cells (RBCs) Count less than 3.9 (x 106/l). Mean haemoglobin concentration was 8.41 ± 1.60 g/dl. Most of the patients 119 of 130 patients (92%) had haemoglobin concentration less than 11(g/dl) (55 males, 64 females). positive correlation was found between total hemoglobin and total RBCs count (Table 2).

Chi-square tests			
	Value	**df**	**Asymptotic significance (2-sided)**
Pearson chi-square	5359.869[a]	5096	0.005
Likelihood ratio	893.258	5096	1
Linear-by-linear association	78.308	1	0
N of valid cases	130		

Table 2: Correlation between total hemoglobin and total RBCs count in sickle cell anemia (HbSS). **Note:** a. 5247 cells (100.0%) have expected count less than 5. The minimum expected count is .01.

Haematocrit (%) was 24.11% ± 4.55%. Majority of the patient's 127 of 130 patients (98%) had haematocrit % less than 34% (57 males, 70 females). The Mean Red Cell Distribution Width-CV (%) was 23.44 ± 4. 70 and the Mean Red Cell Distribution Width-SD was 68.32 ± 13.13 (Table 1). RBCs count is low in our study and significant

RBC indices

Mean cell volume 86.78 ± 10.05 fl, mean cell haemoglobin 29.80 ± 2.99 pg, and mean cell haemoglobin concentration 32.87 ± 0.77 g/dl.

Fewer of the patients 9 of 130 (7%) had mean cell volume less than 75.0 fl (4 males, 5 females). Fewer of the patients 5 of 130 (4%) had mean cell haemoglobin less than 24.0 pg (2 males, 3 females). Fewer of the patients 4 of 130 (3%) had mean cell haemoglobin concentration less than 31.0 (g/dl) (2 males, 2 females) (Table 3). The average value of hematological parameters brings to mind moderate normocytic normochromic anemia in SCA patients in steady state.

Hematological parameters	**Reference range**
Red blood cell count (x 106/l)	1.57-5.48
Hemoglobin (g/dl)	5.0-17.0
Hematocrit (%)	15.3-43.0
Mean cell volume (fl)	39.1-109.6
Mean cell haemoglobin (pg)	21.1-35.3
Mean cell haemoglobin concentration (g/dl)	29.5-34.6
Red cell distribution width-cv (%)	13.2-36.6
Red cell distribution width-sd	36.5-95.0
White blood cell count (x 103/l)	2.4-32.6
Neutrophil (x 103/l)	0.8-28.1
Neutrophils%	16.1-72.3
Lymphocytes (x 103/l)	1.4-37.2
Lymphocytes%	19.4-79.0
Platelets (x 103/l)	66.0-1331

Table 3: Reference Range of hematological parameter of 130 patients with sickle cell disease (HbSS).

White blood cells in study population (Sickle cell disease)

The overall mean white blood cells count was 13.89 ± 5.48 (x 103/l) the mean neutrophils count was 6.96 ± 4.11 (x 103/l), the mean neutrophils percent (%) was 46.48 ± 13.02, the mean lymphocytes count was 6.90 ± 4.57 (x 103/l) the mean lymphocytes percent (%) was 47.56 ± 12.64 (Table 1). Most of the patients 91 of 130 patients (70%) had white blood cell count more than 11,000/l (41 males, 50 females).

Platelets in study population

Platelet counts was found to be of 453.71 ± 189.52 (x 103/l) (Table 1). Half of the patients 65 of 130 (50%) had platelet count greater than 450,000/l (31 males, 34 females).

Discussion

This study was designed to provide baseline hematological values in sickle cell Anemia patients at the Sickle Cell Center, Alkuwity Teaching Hospital, Elobeid City, North Kordofan Estate, and West of Sudan. According to the results of this study, most sicklers were female (55.4%), predominantly aged between 2 and 29 years of age with the mean age was 8 ± 5.804 years. Similar findings were reported in study conducted in Brazil in which the majority of cases were

women. Also in study conducted in Benin City, Nigeria in which there were more females 130 (52.8%) than males 116 (47.2%) in this study.

CBC can be a more useful diagnostic tool [10]. The use of reference values must not generalize as one result which can be normal for one group of population, might not be normal for the other living in a totally different area. Evidence was derived from the observation that normal Caucasians have higher Hb levels than black individuals matched for age and sex. The reference value is the value gained by measuring individual parameters according to the reference healthy population group [11].

Comparison with the hematological reference range of Sudanese healthy adults

In a comparison with the hematological reference range of Sudanese healthy adults, the red cell indices in our results were generally lower [12,13]. These results were anticipated taking into account the degree of chronic haemolysis. The rate of chronic haemolysis associated with sickle cell Anemia patients could give a reason for these lower values. Whereas, the white blood cells (leukocytes) and platelet (thrombocytes) counts were higher. These results were predictable considering the higher risk of infections and chronic pain in sickle cell patients. The majority of SCD patients had reduced values of hemoglobin and hematocrit. The Hemoglobin (Hb) level of normal subjects is greatly variable and is determined by both genetic and environmental factors. Besides age and sex, acquired factors such as diet, smoking, body weight, hypoxia and infections influence the Hb levels. The normal Sudanese Hemoglobin (Hb) concentration values in healthy adult Sudanese is; Hemoglobin (Hb) level ranged from 13.4-16.4 g/dl with a mean value of 14.9 g/dl in males, and 10.7-13.7 g/dl with a mean value of 12.2 g/dl in females respectively. This in comparison with our results, Hemoglobin (Hb) level ranged from 5.0-17.0 g/dl with a mean value of the mean haemoglobin concentration was found to be (8.41 ± 1.60) (g/dl).

Comparison with the hematological reference range of Sickle cell disease patients in other countries.

Red blood cells in sickle cell disease

The mean haemoglobin concentration in our study was found to be (8.41 ± 1.60 g/dl), this in agreement with previous studies in sicklers, for example a retrospective study conducted at the blood center of the Hemominas Foundation in Divinópolis, Brazil from August 2012 to August 2014. The study includes 50 patients with sickle cell anemia. The hemoglobin value was 8.55 ± 1.33 g/dl. Also, in concordance with our result is a retrospective cross-sectional cohort study conducted in Maiduguri, Borno State, northeast Nigeria, 2006-2007 and in Kano, northwest Nigeria, 2009-2010. Includes 352 adult patients with SCD. The sicklers were divided into blood group O (n=187) and non-O blood groups (A+B+AB) (n=165). The haemoglobin concentration for blood group O sicklers was 8.30 g/dl ± 1.2 and for .non-O blood group was 8.10 g/dl ± 1.4. In a retrospective study carried out in populations of Gujarat and adjacent states Madhya Pradesh and Rajasthan, India. From June 2013 to May 2016. Out of total 173 patients homozygous sickle cell disease 108 (62.4%) were males and 65 (37.6%) were females. Mean Hb concentration was 8.21 ± 2.07 g/dl. In a cross-sectional study carried out in Central India over the period of one and half years. A total 65 SCA patients in steady state attending Out Patient Department (OPD) during the study period were included. There were 31 males and 34 females with age ranging from 11 months to 43 years. Average hemoglobin is 8.47 ± g/dl.

In comparison with other studies conducted to determine the mean haemoglobin concentration, our result (8.41 ± 1.60) (g/dl) is found to be slightly more than that in many previous studies. For example, in study conducted at the Sickle Cell Center in Benin City, Nigeria, includes two hundred patients with a diagnosis of SCA in their steady state, the Haemoglobin concentration (g/dL) was 7.54 ± 2.26. In a case–control study conducted amongst homozygous sickle cell patients attending the sickle cell clinics in Lagos, Nigeria, comprised a total of 103 patients the overall mean haemoglobin concentration was 7.93 ± 1.47 g/dl. In the study, conducted amongst homozygous Sickle cell disease (HbSS) Sudanese patients including 52 patients, enrolled from Abnaof Paediatric Hospital, Khartoum, Sudan, between February and May 2014, the overall mean haemoglobin concentration was 7.59 ± 10.3 g/dl [14]. Also, in study conducted in India composed of total 81 'HbSS' cases registered from specialty clinic of RHDMC from Central India and other neighboring states 44 males and 37 females. Age ranges from 6 months to 64 years. In a period of Jan 2003-Dec 2005. Average hemoglobin in males is 7.11 ± 2.13 g/dl and in females 6.75 ± 1.85 g/dl [15]. On the other hand, in a large cohort study of adults 780 homozygous for HbS gene conducted in Saudi Arabia to determine the mean haemoglobin concentration, included patients from the Eastern Province of Saudi Arabia, average hemoglobin was 9.0 (7.9, 10.3) (g/dl).

The haemolysis which is a consequent of the red cell membrane damaged could be intravascular or extravascular. The intravascular haemolysis results from the lysis of complement-sensitive red cells and haemoglobin lost during sickling-induced membrane damaged. The extravascular haemolysis, occurs by phagocytosis of RBCs that have undergone sickling. The degree of haemolysis in sickle cell anemia patient is inversely proportional to haemoglobin (Hb) concentration and Packed Cell Volume (PCV).

The haematocrit value in our study was 24.11% ± 4.55%. In Agreement with our results was a study conducted in Nigeria by Akinbami et al., the overall mean hematocrit in their study was 24.44% ± 4.68%. However, our result was slightly lower than many studies for example, a retrospective study in Brazil includes 50 homozygous Hb SS patients. The overall mean hematocrit was 25.7% ± 4.4%.

Also, a retrospective cross-sectional cohort study in Borno State, northeast Nigeria, 2006-2007 and in Aminu Kano Teaching Hospital, Kano, northwest Nigeria, 2009-2010. The mean hematocrit for blood group O sicklers was 26% ± 0.03% and for non-O blood group was 25% ± 0.04%. In a retrospective study carried out in Gujarat and adjacent states Madhya Pradesh and Rajasthan, India during June 2013 to May 2016. Included 173 patients homozygous sickle cell disease. Mean Hematocrit (HCT) % was 25.87% ± 8.67%. In a cross-sectional study carried out in Central India over the period of one and half years. Average HCT (%) is 26.55% ± 3.74%. On the other hand, our result was slightly higher than study, conducted amongst homozygous Sickle cell disease (HbSS) Sudanese patients by Awoda et al., the overall mean hematocrit was 22.9% ± 3.0%. The majority of SCD patients had reduced values of hemoglobin and hematocrit.

The Mean Cell Volume (MCV), Mean Cell Haemoglobin (MCH) and Mean Cell Haemoglobin Concentration (MCHC) values in our study, were as MCV was 86.78 ± 10.05 fl, MCH was 29.80 ± 2.99 pg, and MCHC was 32.87 ± 0.77 g/dl. In agreement with our results is the study, conducted by Awoda et al., amongst 52 homozygous Sickle cell disease (HbSS) Sudanese patients, the overall MCV was 86.5 ± 7.1 fl, MCH was 28.6 ± 3.0 pg and the MCHC was 33.1 ± 1.4 (g/l).

However, our result was slightly higher than that in many studies for example, in a retrospective study carried by Shah et al., out in the rural and tribal populations of Gujarat and adjacent states Madhya Pradesh and Rajasthan, India. Mean MCV was 71.22 ± 11.96 fl, MCH 22.48 ± 3.70 pg, MCHC was 32.97 ± 2.63 gm/dl. Also, in a prospective study in Benin City, Nigeria. The MCV, MCH and MCHC in steady state were 79.38 fl ± 22.41, 28.31 pg ± 3.58 and 32.56 g/dl ± 2.27. Again, in a large cohort study in Saudi Arabia included 780 homozygous for HbS gene. Mean MCV was 80.3 (72.4, 86.9) fl. Also, in a cross-sectional study in Central India with a total 65 SCA patients in steady state were included. Mean MCV was 81.73 ± 13.55 fl, MCH 26.43 ± 5.22 pg. MCHC was 32.65 ± 5.99 gm/dl. In their case-control study, Akinbami et al., found that the overall MCV 81.52 ± 7.89 fl, and MCH 26.50 ± 3.20 pg. On the other side, our result was slightly lower than that in two studies, the first in Nigeria was a retrospective cross-sectional cohort study Ahmed et al., found that, the overall MCV of blood group O sicklers was 87 (fl) ± 50, for non-O blood groups was 85(fl) ± 60, overall MCH of blood group O was 30(pg) ± 1.2, for non-O blood groups was 31 ± 1.3, the overall mean MCHC of blood group O was 32.7(g/dl) ± 1.1, for non-O blood groups was 33 (g/dl) ± 1.2. The second study was conducted in India by Dani and Agrawal the average MCV in males sicklers was 85 ± 13.1 fl and that in females was 89.8 ± 13.4 fl. Average MCHC in males sicklers was 31% ± 64%, while that in females was 31.02% ± 1.76%.

The overall mean RBCs count in our study was 2.77 ± .63(x 106/l). In agreement with our result was the study, conducted amongst homozygous Sickle cell disease (HbSS) Sudanese patients by Awoda et al., the overall mean RBCs count was 2.70(x 106/l) ± 0.50. Also, in agreement with our finding was a result of study conducted in India composed of total 81 sicklers 44 males and 37 females. Mean RBC count was 2.58 ± 0.75 (106/l). However, our result was slightly lower than two studies in India, for example the first was a retrospective study carried out in Gujarat and adjacent states Madhya Pradesh and Rajasthan, India. Out of total 173 patients homozygous sickle cell disease 108 (62.4%) were males and 65 (37.6%) were females. Mean Average RBC count was 3.82 ± (3.42) (106/l). The second was a cross-sectional study carried out in Central India included a total of 65 SCA patients in steady state. Mean RBC count was 3.29 ± 0.60(106/l). SCD leads to both quantitative and qualitative changes in RBCs. Anemia is a main cause of both morbidity and mortality in SCD, and many patients die in hospital emergency rooms and wards even before blood transfusions can be started.

Red Cell Distribution Width-CV (%) in our study was 23.44 ± 4. 70. In agreement with our result is a prospective study of 200 patients with a diagnosis of SCA in Benin City, Nigeria. The Red Cell Distribution Width (RDW) was 23.76% ± 6.49%. However, our results were higher than that in a cross-sectional study carried out in Central India includes a total 65 SCA. The Red Cell Distribution Width (RDW) % was 16.78 ± 4.03. RDW is a measure of red blood cell size variation (anisocytosis). Elevated mean RDW values were found in the anemic patients, with the highest value seen in sickle cell anemia. The clinical value of the RDW lie in its ability for reflecting active erythropoiesis. Patients with high RDWs may be suspected of having an elevated reticulocyte count that may indicate a hemoglobinopathy, such as sickle cell trait [16].

White blood cells in sickle cell disease

The overall mean white blood cells count in our study was 13.89 ± 5.48 (x 103/l). In agreement with our result is the study conducted amongst homozygous Sickle cell disease (HbSS) Sudanese patients (n=52), the overall mean white blood cell Count (x 103/l) was 13.30 ± 3.4. Again, in same line with our result is a retrospective cross-sectional cohort study conducted in the University of Maiduguri Teaching Hospital, Maiduguri, Borno State, northeast Nigeria, 2006-2007 and in Aminu Kano Teaching Hospital, Kano, northwest Nigeria, 2009-2010 (2 years). Includes 352 adult patients with SCD in Nigeria. The overall mean white blood cell Count (x 103/l) was 13.20 ± 2.5. However, our result was higher than results obtained in many studies, for example in a prospective study of 200 patients with a diagnosis of SCA attending the consultant outpatient clinic at the University of Benin Teaching Hospital, Central Hospital and Sickle Cell Center in Benin City, Nigeria between August 2001 and July 2002. White Blood Cell Count (x 103/l) 12.72 ± 7.98. Also, in a retrospective study carried out in rural and tribal populations of Gujarat and adjacent states Madhya Pradesh and Rajasthan, India. A total 173 patient's homozygous sickle cell disease. The overall mean white blood cell Count (x 103/l) was 12.32 ± (6.00). Again, in a retrospective study conducted at the blood center of the Hemominas Foundation in Divinópolis, Brazil. The study includes 50 patients with sickle cell anemia. The mean white blood cells count was 10,502 (8361-14,776) (x 103/l). In a case–control study conducted amongst homozygous sickle cell patients attending the sickle cell clinics of Lagos State University Teaching Hospital, Nigeria. A total of 103 cases were enrolled. The overall mean white blood cell Count (x 103/l) 10.27 ± 3.94. In a cross-sectional study carried out in the Department of Pathology at a Medical Institute in Central India over the period of one and half years. A total 65 SCA patients in steady state attending OPD during the study period were included. Mean white blood cell count (x 103/l) was 7.08 ± 0.898. In a large cohort study of adults (n=780 homozygous for HbS gene) in Saudi Arabia. Mean white blood cell count (x 103/l) was 9.8 (6.1, 13.9).

The mean neutrophils count in our stud was 6.96 ± 4.11 (x 103/l). Our result was higher than result of two studies. The first study was a prospective study of 200 patients attending the consultant outpatient clinic at the University of Benin Teaching Hospital, Central Hospital and Sickle Cell Center in Benin City, Nigeria. The mean neutrophils count was (x 103/l) 5.2 ± 1.6. The second study was a retrospective study conducted at Brazil. The study includes 50 patients with sickle cell anemia. The mean neutrophils count was 5118 (4000-7005).

The mean lymphocytes count in our study was 6.90 ± 4.57 (x 103/l. In agreement with our result was a prospective study of 200 patients with a diagnosis of SCA attending the consultant outpatient clinic at the University of Benin Teaching Hospital, Central Hospital and Sickle Cell Center in Benin City, Nigeria. Lymphocytes (x 103/l) 6.5 ± 1.6.

Even though sickle cell disease is primarily a disease of the red blood cell, leucocytes, because of their sizes obstruct blood vessels more efficiently than red blood cells when attached to the endothelium. Bacterial infection associated with leukocytosis is a well-known predisposing factor to sickle cell disease crises. In sickle cell patients, moderate leukocytosis is very common in both critical and non-critical phases.

The overall mean white blood cells count amongst homozygous sickle cell disease patients doubles the value obtained in HbAA controls. The steady-state leukocytosis in HbSS limiting the usefulness of this measurement in the assessment of infection. Leukocytosis may be a result of leukocyte progenitor cell stimulation, asplenia, and granulocyte demargination [17]. Undoubtedly,

leukocytosis in sickle cell disease patients may owing to auto splenectomy resulting from recurrent splenic vessels occlusion, which make patients more susceptible to irresistible infections mainly, encapsulated organisms like Streptococcus pneumonia and Haemophilus influenzae. A high absolute neutrophil count exhibited statistically significant relationship with clinical severity of sickle cell Anemia. Neutrophilia represent a risk factor for early sickle cell disease-related death.

Platelets in sickle cell disease

The overall mean platelets count in our study was 453.71 ± 189.52. Our result was slightly higher than a retrospective study conducted at Brazil. The study includes 50 patients with sickle cell anemia. The mean platelets count was 431,541 ± 149,615 (x 103/l). Again, in a case–control study conducted amongst homozygous sickle cell patients attending the sickle cell clinics of Lagos State, Nigeria. A total of 103 cases were enrolled. The overall mean platelet count was (x 103/l) 412.71 ± 145. However, our result was higher than results of many studies; for example a prospective study of 200 patients with a diagnosis of SCA attending Sickle Cell Center in Benin City, Nigeria. The mean platelet count (x 103/l) 342.62 ± 143.03. Again in a retrospective study carried out in Gujarat and adjacent states Madhya Pradesh and Rajasthan, India. Includes 173 patients homozygous sickle cell disease. The overall mean platelet count (x 103/l) was 285.48 ± 193.20 [18]. In a cross-sectional study carried out in Central India over the period of one and half years. A total 65 SCA patients in steady state attending OPD during the study period were included. Mean Platelet Count (x 103/l) was 244.88 ± 111.55 [19]. In a large cohort study in Saudi Arabia included 780 homozygous for HbS gene. Mean platelet count (x 103/l) was 263 (158, 411).

On the other hand, our result was lower than the result of two studies, the first was a retrospective cross-sectional cohort study conducted in Maiduguri, Borno State, northeast Nigeria, 2006-2007 and in Aminu Kano Teaching Hospital, Kano, northwest Nigeria, 2009-2010. Includes 352 adult patients with SCD in Nigeria. The overall mean platelet count (x 103/l) of blood group O was 562 ± 45, for non-O blood groups was 554 ± 49. The second study conducted amongst homozygous Sickle cell disease (HbSS) Sudanese patients (n=52), the overall mean platelet count (x 103/l) was 533.6 ± 98.7.

During steady state in patients with SCD, there is increased percentages and activation of platelets and this accelerates during vaso-occlusive crisis (VOC) [20]. An association between stroke in sickle cell disease and platelet count >450,000/l has been reported.

Conclusion

The average value of hematological parameters of SCA Sudanese patients in steady state shows moderate normocytic normochromic anemia, lower values of red cell parameters, but higher values of white blood cell and platelets counts. A larger scale work is recommended in this region for a baseline hematological profile for guiding the clinicians in management of these patients. Additionally it demonstrates that white blood cells and platelet counts tend to be elevated in SCD patients, potentially reflecting the higher risk of infections and pain crises they face. These findings not only contribute to the local epidemiological knowledge but also underscore the importance of tailored medical interventions and care strategies for individuals with SCD in Sudan, where the prevalence of the disease is relatively high.

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