Haematological parameters in Sudanese children with sickle cell disease

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Abstract

Background In Africa, SCD is the most common inherited hematological disease with high mortality rate at age one to five years. This disease was discovered early in Sudan, the peak occurrence of SCD is among the population from the Western Sudan. The objective of this study was to determine the haematological parameters changes in Sudanese patients with SCD.

Methods This study was conducted in Omdurman between August 2011 - Aril 2012, in hundred patients with homozygous SCD of age between 6 month –17 years. Questionnaires were used to collect demographic and clinical data. About 5 ml of venous anticoagulated blood were collected for complete blood count.

Results A total of hundred cases were enrolled. More than 97% belong to tribes from Western Sudan. The Hb, RBCs count and Hct level were low in all patients. The normal MCH and MCHC was observed except in age group below two years. The TWBCs were significantly increased in all cases.

Conclusions This study provides haematological reference ranges for homozygous Sudanese patients with SCD. As well is known, the results of this study showed lower

values of Hb, RBCs count and Hct among patients. The remarkable result in this study was the significant increase in leucocytes count.

key words: SCD.

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Introduction

Sickle cell disease (SCD) is caused by an autosomal recessive inheritance of an abnormal beta globin gene (sickle cell gene) leads to substitution of thymine by adenine in glutamic acid which in turn results in the substitution of valine to glutamic acid at the sixth position on the beta globin chain. Hemoglobin S (HbS), is the defective Hb that produced as a result of this defect, is a tetramer (alpha2/beta2) that is poorly soluble and polymerizes when deoxygenated (1). The most common characteristic features in the pathophysiology of SCD is the chronic hemolytic anemia and the vaso-occlusion (2).

SCD is believed to be the most frequent inherited blood disorder on the globe affecting an estimated 100 million people world-wide and, in particular, the black races and persons of Mediterranean origin (3). In Africa, SCD is the most prevalent genetic disease with high mortality rate at age one to five years (4,5). In the United States of America (USA), sickle cell anemia has been found to be the most frequent autosomal recessive gene disorder affecting approximately 1:375 persons of African ancestry (6).

The SCD was early detected in Sudan (7). The highest prevalence of SCD in Sudanese is among the population from the Western Sudan (8-12) (Figure .1). It is believed that the sickle cell gene has brought to Sudan through immigrants from West African tribes, especially from Hosa, Folani and Bargo (13).

The objective of this study was to determine the haematological parameters changes in SCD Sudanese patients.

Figure 1. Map of Sudan. The red circles demonstrate the distribution of tribes with high prevalence of SCD.

Materials and methods

It is a cross-sectional descriptive hospital based study conducted at Albuluk pediatric teaching hospital and Omdurman pediatric hospital (from August, 2011 to April, 2012).

The study population included 100 (HbSS) SCD Sudanese patients, fifty seven male and forty three female (age 6 month to 17 years). Questionnaires were used to collect demographic and clinical data . After appropriate ethical approval and written consent form participants prior the commencement of the study. About 5 ml of venous blood in K2 ethylenediaminetetraacetic acid (EDTA) were collected, well mixed and an investigated for complete blood count (CBC) by using Sysmex KX-21 hematology analyzer (manufactured by Sysmex corporation Kobe, Japan) a three- part auto analyzer able to run 19 parameters per sample including haemoglobin concentration, packed cell volume, red blood cell concentration, mean corpuscular haemoglobin, mean cell volume, mean corpuscular haemoglobin concentration, white blood cells and platelet parameters.

Data were entered in the computer and Statistical software packages (Excel 5.0, Microsoft, Redmond, WA; and Statistical Package for the Social Sciences 20.0, SPSS, Inc., Chicago, IL) were used for data management and analysis, chi square test was used to compare percentages. P-value <0.05 was considered significant.

Results

All subjects who were recruited to the selected hospitals were diagnosed to had homozygous sickle cell disease. The mean age of the study subjects was (6.1 ± 3.3) years old; the minimum age was 6 months year old and the maximum one was 17 years old. Seventy five (57%) of the study subjects were males and forty three (43%) were females, there is no significant difference between them (P=0.119). The highest incidence of the

disease in both sexes was found among the age group (2 - 6 years) (Figure .2) , there were no statistically significant differences regarding to age (*P*=0.302)**.**

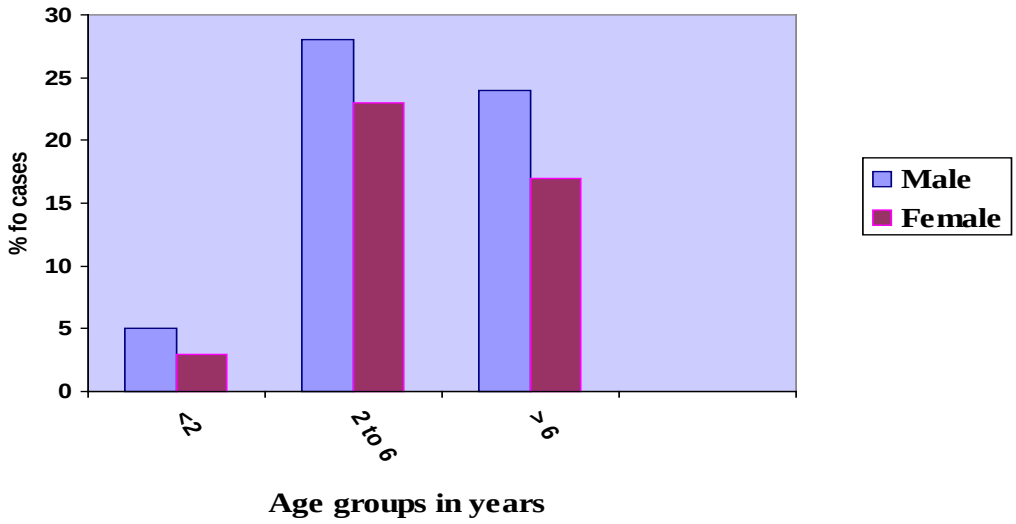

Figure 2. Distribution of study subject according to age and sex.

The study subjects constitute of 13 different tribal stocks, more than 97% are tribes from Western Sudan. 70% of the subjects belong to only four tribal stocks namely; the Miserria tribal stock (28%), Marareet stock (22%), Rizegat stock (12%), Bargo stock (8%) Other tribes represents low percentage include: Tama, Habbania, Jawamaa, Zagawa, Masalma, Manasra, Slihab, Bne Halba and Kwahla (Figure 3).

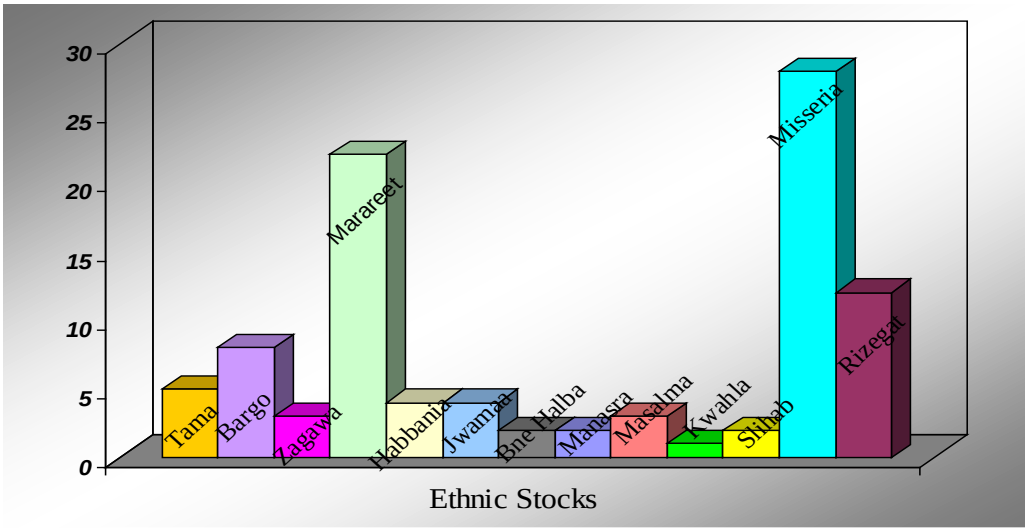

Figure 3. Tribal distribution of study subjects.

The most frequent clinical presentation of patients was anemia (44%) and they had been indicated for blood transfusion just to increase the Hb level, followed by painful crises (19 %). About (18%) of the cases had joint pain in addition to anemia, (15%) had hemolytic crises with or without joint pain, and the other minor causes include: infection, sequestration, CVA and acute heart failure (figure 4).

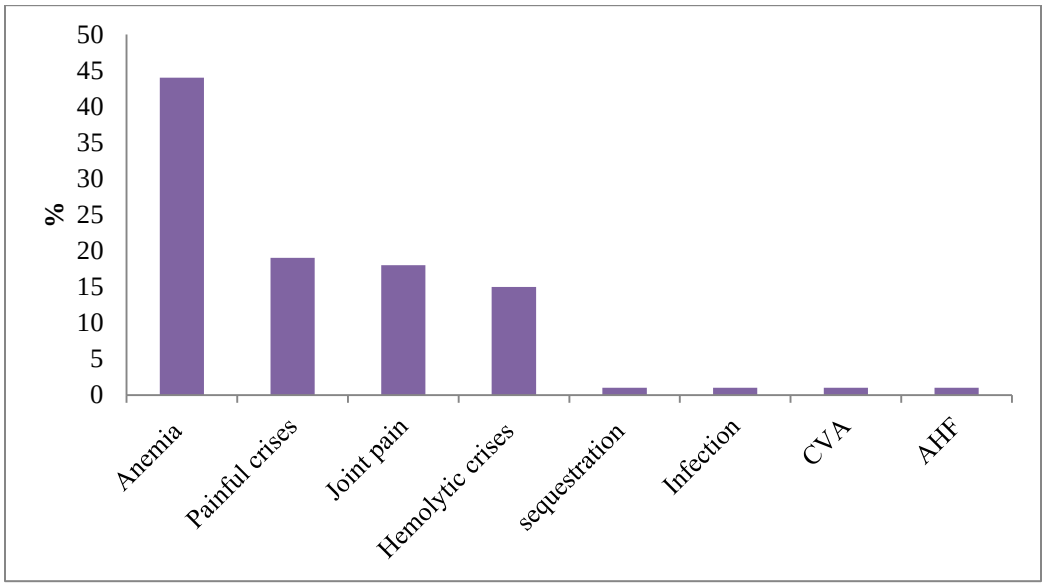

Figure 4. Clinical presentations of patients.

For 100 cases, the overall haemoglobin concentration (Hb) was 6.30 ± 1.35 g/dl, red blood cell count (RBCs) 2.34 ± 0.52×10^{12} cell/L , packed cell volume (PCV) 19.3 ± 4.2%, mean cell volume (MCV) 84 ± 16 fl, mean cell haemoglobin (MCH) 27.5 ± 5.4 pg, mean cell haemoglobin concentration (MCHC) 32.8 ± 2.5 g/dl, red cell distribution width (RDW) 25.7 ± 6.6, total white blood cell count (TWBCs) 21.7 ± 10.3×10^{9} cell/L, neutrophils 11.5 ± 6.3×10^{9} cell/L, lymphocytes 8.5 ± 5.2×10^{9} cell/L, mixed (monocytes, eosinophils and basophils) 1.9 ± 1.4×10^{9} cell/L, and the mean platelets count 319.2± 135.7×10^{9} cell/L (Table 1&2).

Table 1. Mean values of complete blood count parameters for all patients

Parameters	>2		2-6		<6	
	M	F	M	F	M	F
Hb (g/dL)	5.5	4.1	6.4	6.3	6.6	6.4
RBCs ($\times 10^{12}$ cell/L)	2.2	2.1	2.4	2.4	2.4	2.3
Hct (%)	16.4	13.3	19.4	19.7	19.9	19.3
MCV (FL)	78.9	65.6	82.2	84.8	87.7	84.3
MCH (Pg)	27.6	20.4	27.2	27	28.9	27.8
MCHC (g/dL)	34.1	31	33.1	32	33	33.1
WBCs ($\times 10^{9}$ cell/L)	36.5	24.9	19.6	20.6	21.9	21.2
Platelets ($\times 10^{9}$ cell/L)	287.8	363.3	300.9	316	330.9	339

Table 2. Mean values of differential leukocytes count parameters for all patients

Variables	Age< 2 years n=8	Age between 2 -6 years n=51	Age > 6 years n=41
TWBCs ($\times 10^{9}$ cell/L)	32.2	20.1	21.6
Neutrophils ($\times 10^{9}$ cell/L)	15.5	10.6	11.7
Neutrophils (%)	47.6	51.9	53.7
Lymphocyte ($\times 10^{9}$ cell/L)	14	7.8	8.1
Lymphocyte (%)	43.9	40.5	37.9
Mixed ($\times 10^{9}$ cell/L)	2.6	1.7	1.9
Mixed (%)	8.5	7.9	8.9

Discussion

SCD is the most prevalent genetic disease in African region, this study highlights the demographic and the hematological parameters of SCD Sudanese patients.

In the current study, over than (97%) of the patients belong to tribes from western Sudan, Elderdery A *et al* also noticed the high frequency of homozygous HbSS in his study

among western Sudan tribes (14). Misseria tribe, showed the highest percentage of the entire study group of patients in this study (28%), this tribe showed the prevalence of SCD to be 30%, 16% among immigrants from the province of Blue (15). The prevails of SCD among western tribes in the current study is in agreement with other studies conducted in Sudan (11,14,16,17).

The Hb, RBCs and PCV results were generally low in the patients, together with the observable high RDW reflected the degree of chronic hemolysis and anaemia. High TWBCs was observed in the current study, ($21.7 \pm 10.3 \times 10^9$ cell/L). Leucocytosis and neutrophilia also noticed in SCD Nigerian children and found to be related to the disease severity (18). In the same vein increase neutrophils activation in SCD patients was observed by Lard (19). The high value of TWBCs and neutrophilia could be attributed mainly due to infections. Other factors have been reported to cause leucocytosis in the absence of infection including pain, nausea vomiting and anxiety, leucocytosis was thought to be due to redistribution of granulocytes from the marginal to the circulating pool (20-22). In patients with age below 2 years, the Hb and Hct levels (5±1.3 g/dl and 15.3±4.5 %) respectively was significantly lower than the other age groups (P= 0.0230, 0.0332) respectively. Furthermore, there was an observable decrease in MCV and MCH values in the same age group, this findings is not concur with the results from the same age group SCD Kenyan patients (23), presumably due to the increase iron demand in growth and poor intake of iron. The RBCs morphology in other age groups are generally normocytic this reflected by the normal MCV, this observation is inconsistent with the raised MCV noticed in previous studies (24-26).

This study provides haematological reference values for homozygous Sudanese SCD patients. As well is known, the results of this study showed lower values of Hb, RBCs count and Hct among patients. On the other hand, significant increase in leucocytes count was observed.

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Disclosure of Conflicts of Interest:

The author discloses that there was no conflict of interest.

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