SCT Among Relatives of Sickle Cell Patients in Western Sudan

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Key words sickle cell aneia, hemoglobinopathies

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Abstract

This is an analytical hospital based study carried out in relatives of patients suffering from sickle cell disease (SCD) who referred to Elobied Hospital. The aim of this study was to estimate the frequency of sickle cell trait (HbAS) among those relatives and to assess their CBC. One hundred persons of seventeen different tribes were included (48% males and 52% females) with an age ranged between 4 to 70 years. From each person, 2.5 mL of venous blood was collected into EDTA container. Blood film, complete blood count (CBC) and sickling test were performed immediately. Then haemolysate was made and stored at (– 20 ◌̊C) for electrophoresis test. The results of hemoglobin electrophoresis show that, 54% of target samples were heterozygous carrier (HbAS) while 42% were normal (HbAA) and 4% were diagnosed as sickle cell disease (HbSS). The highest distribution of sickle cell trait was among Bederia tribe 9 (23.1%) followed by Fulani and Selehab 6 (15.4% for each tribe). The results of CBC show no significant difference in the values of haemoglobin (Hb) , haematocrite (HCT%), mean cell volume (MCV), mean cell hemoglobin (MCH), mean cell haemoglobin concentration (MCHC), red cell count (RBCs), white cell count (WBCs) and platelets count (Plt) between sickle cell trait (HbAS) and normal individuals (HbAA). This work concluded that members of the studied tribes are potentially capable for spreading of SCD in western Sudan.

Background

Sickle cell trait (SCT) usually is not regarded as a disease state because it has complications that are either uncommon or mild. Nevertheless, under unusual circumstances serious morbidity or mortality can result from complications [1]–[3]. Sickle cell trait (SCT) occurs at about 8% in American Blacks and from 20- 50% among some African tribes. In sickle cell trait which is the heterozygous state for HbS gene, HbS comprises 38 – 45 percent of the total haemoglobin, and the rest being HbA, HbA_2 and HbF. The cells do not contain sufficient HbS to undergo sickling but lower oxygen tension is occurring in the body and the red cell lifespan is normal. In the stained blood film, no sickle cells can be observed and sickling can readily be demonstrated by the sickle test. If the individual has concurrent α thalassaemia, the red cells are microcytic with a mild reduction in MCV and an HbS concentration of less than 38 percent [4]-[6]. Sickle cell trait does not cause anemia and if anemia is present, it would be attributed to causes (such as iron deficiency) other than sickling statae. However, under unusual circumstances SCT can be associated with complications related to polymerization of deoxy – hemoglobin S. Pathologic processes that cause hypoxia, acidosis, dehydration, hyperosmolality, hypothermia or elevated erythrocyte 2, 3 – D PG can transform silent sickle cell trait into a syndrome resembling sickle cell disease with vaso- occlusion due to rigid erythrocytes [7]. Individuals who carry the sickle cell anemia gene are found to be more resistant to malaria which has been considered as an adaptive advantage to homozygous individuals. However, sickle cell trait is extremely common in West Africa, where malaria is also a big factor, contributing to the health problems [8]. The frequency of sickle cell trait has not been studied satisfactorily in Sudan especially in the western and the eastern parts where the gene frequency of sickle cell disease is quite prevalent. The problem becomes

augmented due to population unawareness, consanguineous marriage which employed widely in that area, lack health counseling and undertaken serious research. Accordingly, this study was conducted to detect this group in Elobied City to verify the prevalence of SCT in an attempt to decrease the spreading of the sickle cell disease.

Material and Methods

Study Population:

Sicklers relatives in Elobied City were taken randomly and interviewed by using special questionnaire in a descriptive observational and cross sectional study.

The questionnaire included demographic data (sex, age, tribe, history of disease and relationship to sickler).

2.4. Sampling and Sample Size:

In this study sampling method was a non probability sample calculated to achieve 100 specimens of blood. The sample size was determined according to available resources and facilities.

2.7. Sample:

Each member of the sickler's relatives who referred to Elobied Hospital was informed about the outcomes, objectives of the study and 2-5 mL of venous blood was collected in EDTA container. The samples were investigated for complete blood count using MYTHIC18 automated hematological analyzer, sickling test and Hb electrophoresis following laboratory routine procedures described earler 48-50.

Result

Study population:

A total number of 100 participants (48% were males 52% were females) represented 12 western Sudanese tribes with ages ranged from 4 to 70 years (figures 1) were tested in this study.

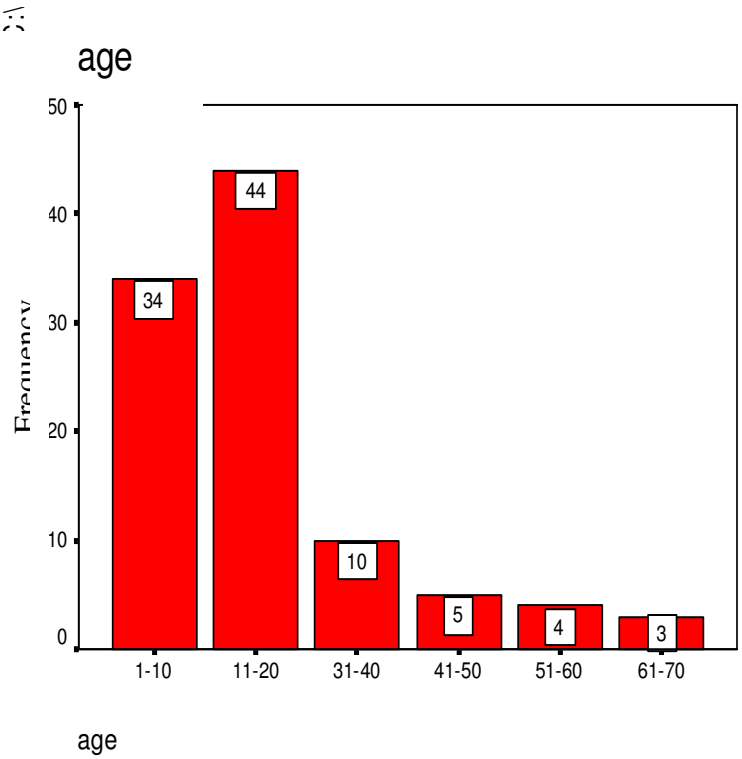

Fig. 1. Age group distribution

Frequency of SCT among studied population:

Sickling test was positive in 58 samples and negative in 42 and haemoglobin electrophoresis result revealed that 54 samples were heterozygotes or sickle cell trait (HbAS), 42 sample were (HbAA) and 4 samples were (HbSS) (figure 2). The highest distribution of SCT were among Bederia tribe which got 9 individials or 24.75% of SCT followed by Selehab and Fulani with equal frequencies 7 each or 12.96% of SCT (table 1).

TABLE I

Frequencies and percentages of SCT among study population

Tribes	Frequency of SCT	Percentage of SCT
Bederia	13	24.75
Selehab	7	12.96
Folani	7	12.96
Kenana	4	7.47
Benhelba	4	7.47
Gawamaa	4	7.47
Ashraf	3	5.55
Darhamed	3	5.55
Hosa	3	5.55
Shewehat	2	3.70
Barno	2	3.70
Rezegate	2	3.70

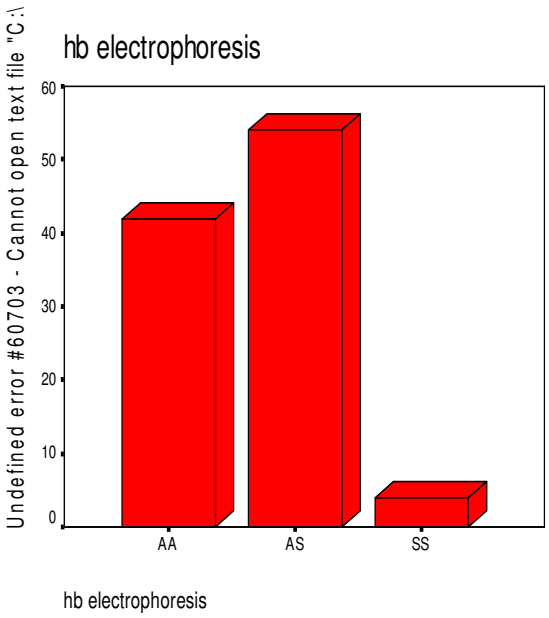

Fig. 2. Hb electrophoresis result

RBC morphology:

The blood film for normal subjects (HbAA) was normocytic normochromc and that for sickle cell trait (HbAS) was microcytic hypochromic. The blood film appearance for sickle cell disease (HbSS) was normocytic normochromic (figure 3) with sickle cell, target cell, and normoblast.

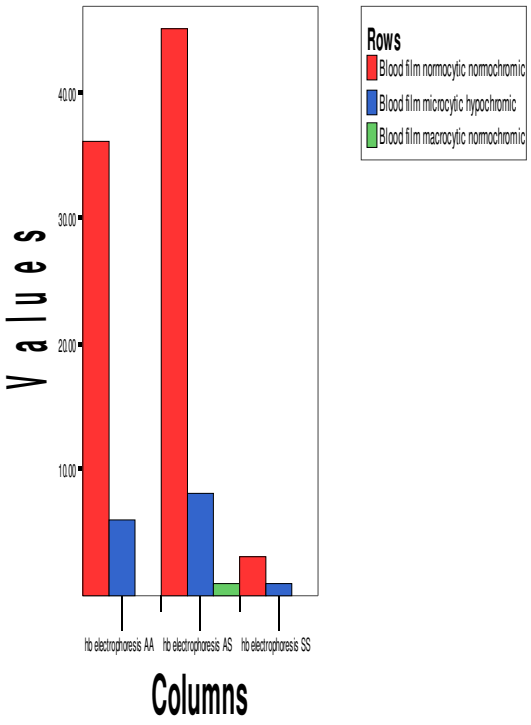

Fig. 3. Morphology of RBC among study population

CBC results:

The results of CBC components (Hb, HCT, RBC count, WBC count, MCV, MCH, MCHC and platelets count) in sickle cell traits show no significant differences when compared to results of healthy control (table 2).

TABLE II

CBC components of trait (HbAS) and control (HbAA)

Parameters	Subjects	Means	SD	P. value
Hb g/dl	SCT	12.06	1.339	0.228
	Control	11.70	1.56	
HCT%	SCT	38.12	3.60	0.181
	Control	36.95	4.79	
RBC count	SCT	4.64	0.38	0.546
Million/cmm	Control	4.58	0.58	
MCV in fl	SCT	82.23	6.83	0.596
	Control	81.52	6.16	
MCH in pg	SCT	26.08	2.70	0.569
	Control	25.79	1.97	
MCHC	SCT	31.61	1.35	0.774
gm/dl	Control	31.68	0.99	
WBC count/	SCT	7.730	2.36	0.832
cmm	Control	7.600	3.26	
Platelets	SCT	267.72	94.55	0.569
count/cmm	Control	258.41	104.06	

Discussion

Sickle cell anaemia occurs in the homozygous state (HbSS) in which the haemoglobin is nearly all haemoglobin S and the remainder is HbF. There is no (HbA) in homozygous individuals, while in hetrozygotes sickle cell trait half or less of circulating haemoglobin is haemoglobin S, the rest being normal haemoglobin (HbA) [1] [2]. Sickle cell disease (SCD) is an important cause of morbidity and mortality among indigenous and immigrant ethnic groups in western Sudan [8]. The assessment of

incidence and diagnosis of haemoglobin S married couples or in prenatal stage is an important step towards reducing this gene distribution in future generation. The frequency of sickle cell trait in this study was 54 which means that 54% of the studied population were carrier of sickle cell gene. This frequency is comparable and within the range of reported frequency in other African continents [9]. But frequencies lower than that were reported in Bahrain, Sultanate of Oman and Turky in which SCT frequencies were 7%. 10% and 0.5% respectively [10]-[12]. One of the sharp results immerging from this study is that, sickle cell trait is occurring in a high frequency in the study area and hence putting expected new born at risk of getting SCD. In our study population as other African communities, common factors were acting to reflect high frequency of SCT. These factors include illiteracy, consanguineous marriage, closure societies and lack of medical counseling. In order to avoid risk of having SCD, serious efforts are needed to improve social culture and economical status to help reducing this vast spreading and getting rid of sickle cell disease in that region. The results of this work also showed that, the highest frequency of sickle cell trait were among Bedaria tribe (9 individuals or 23.1%) followed by Fulani and Selehab which have equal frequencies (6 individuals or 15.4% for each) while the rest of the tribes got the lower frequencies. Complete blood components investigation revealed that SCT in the study area, like other SCT individuals, were normal and not suffering or complaining from blood disorder. Blood films results from sickle cell trait (AS) and normal (AA) individuals in the present study showed normocytic normochromic and microcytic hypochromic. This indicated no morphological differences between sickle cell trait (AS) and normal (AA) individuals. These results agreed with that obtained by Frank et al 1990 and John et al 2003 who reported that, blood pictures in sickle cell trait group and normal individuals were normocytic normochromic and microcytic hypochromic [13]-[14]. However, this type of cell morphology was attributed to be due to iron deficiency anaemia, thalassemia or other types of hypochromic anaemia. Extra intermarriage between displace relatives and foreign tribes may decrease the incidence of sickle cell disease but unlikely peoples of study area have strange link with their customs which prevent them to marry members of other tribes.

Acknowledgements:

The authors acknowledged all participants who were so glad to join this work and family of Alobied hospital for providing place for prelaboratory investigations. The authors also appreciate the financial and logistic supports offered by Sudan university of sciences and technology.

References

1. Sears DA: The morbidity of sickle cell trait: a review of the literature. Am J Med 1978;64: 1021-36.
2. Sears DA. Sickle Cell trait. In: Embury SH, Hebbel RP, Mohandas N, Steinberg MH, eds. Sickle cell disease: basic principles and clinical practice. New York, Raven Press, 1994: 381-94.
3. Biedrzycki OJ, Sheaff M. Sudden death during prolonged religious fasting. Am J Forensic Med Pathol. 2010. 31(4):385-7.
4. Hoffbrand A.V, P.A. H. Moss, J.E. Pettit. (2006). *Essential Haematology* 5th edition. Victoria, Australia. Blackwell Publishing Ltd, p24-26.
5. N. C. Hughes. John. (2001) Lecture Notes Haematology, 5th edition. London, Oxford, Black Well scientific publishing.
6. Frank Firkin, C.Chesterman, D.Penington, B.Rush (1990), *De Gruchy Clinical Haematology in Medical Practice*. 5th edition.

London. Blackwell Publishing Ltd. p29-32,172-181.

7. Wood WG, Pembrey ME, Serjeant GR, Perrine RP, Weatherall Dj. Hb F synthesis in sickle cell anemia: a comparision of Saudi Arabia cases with those of African origin Br J Haematol. 1980; 54 (3) 431 – 45
8. Roth Jr, EF, Friedman M, Ueda Y, Tellez I, Trager W Nagel RL. 1978. Sickling rates of human AS red cells infected in vitro with Plasmodium falciparum malaria. Science 202: 650-652.
9. Fleming AF, Storey J, Molineaux L, Iroko EA, Attai ED. Abnormal haemoglobins in the Sudan savanna of Nigeria. I. Prevalence of haemoglobins and relationships between sickle cell trait, malaria and survival. Ann Trop Med Parasitol. 1979. 73(2):161-72.
10. Guler E, Garipardic M, Dalkiran T, Davutoglu M. Premarital screening test results for β-thalassemia and sickle cell anemia trait in east Mediterranean region of Turkey. Pediatr Hematol Oncol. 2010 Nov;27(8):608-13.
11. Rajab AG, Patton MA. Modell B. study of hemoglobinopathies in Oman Through a national register. Saudi Med J. 2000; 21 (12): 1168 -72.
12. Blair D. Gregory WB. Nutritional Vs hereditary anaemias appropriate screening procedures for Bahrain. Bahrain Med Bull. 1986; 124-7.
13. Frank Firkin, C.Chesterman, D.Penington, B.Rush (1990), *De Gruchy Clinical Haematology in Medical Practice*. 5th edition. London. Blackwell Publishing Ltd. p29-32,172-181
14. John P. Greer, John Foerster, John N. Lukens. (2003). *Wintrobe's Clinical Hematology*. 11th edition. New York. Lippincott Williams & Wilkins Publishers.P1224-1235