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Research Article

FREQUENCY OF SICKLE CELL DISEASE AMONG ALHASSANIA TRIBE IN THE WHITE NILE STATE, SUDAN

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ABSTRACT

Sickle cell anemia (SCA) is a hereditary blood disorder (autosomal recessive), characterized by red blood cells that assume an abnormal, rigid, sickle shape. Sickling decreases the cells' flexibility and results in a risk of various life-threatening complications.Two hundred and fifty subjects were randomly selected and enrolled in this cross-sectional study, from general population of Alhassania tribe who are live in the White Nile state in Sudan, Two ml of venous blood was collected in EDTA anticoagulant from each subject for haematological analysis and haemoglobin electrophoresis.4.8% of the study population has an abnormal haemoglobin variants, the haemoglobin electrophoresis showed HbSS with 2%, HbAS with 2.8% and HbAA with 95.2% frequency, no other abnormal Hb variant was detected . All patients with Hb SS are severely anaemic with maximum Hb level of 7.4 g/dl; and 80% of them have leukocytosis with TWBC count higher than $11.0X10^9$/L. In conclusion, our study reported low frequency of Hb S among Alhassania tribe in the White Nile state in comparison with previous reports in Sudan, in particular those from western Sudan.

Key words: Hb S, Alhassania tribe, Sudan.

INTRODUCTION:

Sickle cell anemia (SCA) is a hereditary blood disorder, characterized by red blood cells that assume an abnormal, rigid, sickle shape (1). Sickling decreases cells flexibility and results in a risk of various life-threatening complications(2). This sickling occurs because of a mutationin the beta goblin gene which results in an abnormal haemogloinwith a substitution of valine for glutamic acid at position six in the betaglobin chain (3). The homozygosity of sickle cell genes (HbSS) results in SCA, while the heterozygosity results in other sickle cell diseases (SCD), which include sickle cell trait with one sickle cell gene and a normal haemoglobin gene (HbAS), and a double heterozygosity of a sickle cell gene with other abnormal haemoglobin variants gene (e.gHbSC)(4).In sickle cell disease, low oxygen tension promotes red blood cell sickling, and repeat episodes of sickling damage the cell membrane and resulting in chronic haemolysis (5). The rigid blood cells are unable to deform as they pass through narrow capillaries, leading to vessel occlusion and ischemia. Although the bone marrow attempts to compensate by creating new red cells, it does not match the rate of RBC destruction. (6,7)

Sickle cell anemia is one of the most common genetic disorder found worldwide, mainly among people of African origin.(8)The sickle mutant gene has the highest frequency of occurrence in Central Africa. SCA is particularly common among people whose ancestors come from Sub-Saharan Africa, South America, Cuba, Central America, Saudi Arabia, India, and Mediterranean countries such as Turkey, Greece, and Italy (5).

In Sudan, sickle cell anaemia is one of the major types of anaemia**.** The high prevalence was reported from Western and Southern regions, particularly Hosa, Folani, Bargo and Baggara tribes group that includes Hawazma and Meseria.

Alhassaniatribe, one of the biggest branch of Alkawhala tribe who descended from Arab Peninsula**,**is a semi closed tribe, they marry among themselves and the consanguineous marriage is more common throughout the tribe.This study aimed to determine the prevalence of sickle cell disease (AS or SS), and the other abnormal haemoglobin variants, among Alhassania tribe in Sudan (10).

MATERIALS AND METHODS:

Two hundred and fifty subjects were randomly selected and enrolled in this cross-sectional study, from general population of Alhassania tribe who are live in the White Nile statein Sudan, Two ml of venous blood was collected in EDTA anticoagulant from each subject for haematological analysis (hemoglobin, PCV,RBC count, TWBC count, platlets count and Reticulocyte count) and haemoglobine lectrophoresis. Laboratory investigations

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were performed at the department of haematology, faculty of medical laboratory sciences, Alneelain University, Sudan.CBC was performed immediately by automated cell counter (Sysmex Kx21N). Different haemoglobin types were determined by Cellulose Acetate membrane electrophoresis at alkaline pH. Statistical analysis was performed using statistical package for social science (SPSS) software. Descriptive data analysis was used to determine the frequency of Hb variants and to calculate the mean of the haematological values.

RESULTS:

The study included 250 subjects; there median age was 38 year, with minimum age of 1 and maximum of 75 years. All subjects were tested for blood countand Hb electrophoresis. Results of the blood count were as follows: Mean Hb level 12.2±2.3g/dl,mean PCV 36.3±7%, mean TWBC count 6.7±2.8X10^9/L and mean platelets count 277±109.7X10^9/L. 4.8% of the study population has an abnormal haemoglobin variants, the haemoglobin electrophoresis showedHbSS with 2%, HbAS with2.8% and HbAA with 95.2%frequency, no other abnormal Hb variant was detected . All patients with Hb SS are severely anaemic with maximum Hb level of 7.4 g/dl; and 80% of them have leukocytosis with TWBC count higher than 11.0X10^9/L. Results of the blood count of patients with Hb SS were as follows: Mean Hb level 6.8±0.56g/dl, mean PCV 19.4±7%, mean TWBC count 13.8±3.2X10^9/L and mean platelets count 432.6±138.1X10^9/L. While for those with Hb AS were as follows: Mean Hb level 12.4±1.6g/dl, mean PCV 36.3±4.5%, mean TWBC count 5.3±0.9X10^9/L and mean platelets count 293±97.3X10^9/L.

DISCUSSION:

Sickle cell disease is a major public health with a great impact on both individuals' and societies. Sudan includes variable ethnic groups that range from Arabs to African and Afro-Arabs tribes. These ethnic groups include groups with Negroid genetic characteristics with an established history in the area such as Nuba and Nilotes. Other groups include Arab, Hausa and Copt who migrated to the area in different times in history, as well as the Arab-negroid admixture tribes (9,10). In this study we determined the frequency of Hb S (SS and AS) among Alhassania tribe in the White Nile state in Sudan.250 individuals of Alhassania tribe were investigated for full blood count and haemoglobin electrophoresis. 4.8% of the study population has an abnormal Hb, with 2.8% Hb AS and 2% of HB SS, no other abnormal haemoglobin variant was detected. Previous reports revealed the wide range of SCD among different areas of Sudan, ranging from 0% among Shagia and Manasir tribes in northern region (11) to 30.4% among the Messeryia tribe (a branch of the Baggara tribes) in Western Sudan (12). Alhassania considered as the biggest branch of Alkawhala tribe who descended from Arab Peninsula, they moved from the Red sea region into Sudan. The large family size, high rate of consanguinity in conjunction with tribe/clan endogamy, makesAlhassania tribe unique from the point of view of genetic analysis. Our study reportedlow frequency of Hb S among Alhassania tribe in the White Nile state in comparison with previous reports inSudan, in particular those from western Sudan.

Allpatients with HbSS were severely anaemic with maximum haemoglobin value of 7.4g/dl. The mean Hb and PCV values are in agreement with previous study done in Sudan (*13,*14).Most of patients (80%) have an elevated TWBC, leucocytosis was also noticed in previous study done in Sudan (*13,*15). This result was expected considering the degree of chronic haemolysis, vulnerability to overwhelming infections and chronic pain in sickle cell patients.

CONCLUSION

We studied the frequency of abnormal haemoglobin variants among Alhassaniain the White Nile state in Sudan. Our study reported a low frequency of Hb S, with 2.8% Hb AS and 2% of Hb SS, among the study group in comparison with previous reports in Sudan, in particular those from western Sudan. Our study also reported a haematological feature of HbSS patients similar to many previous reports in Sudan.

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