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 11.0X10⁹/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 mutation in the beta globin gene which results in an abnormal haemoglobin with 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.g HbSC)(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.

Alhassaniat tribe, 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 state in Sudan. Two ml of venous blood was collected in EDTA anticoagulant from each subject for haematological analysis (hemoglobin, PCV, RBC count, TWBC count, platelets count and Reticulocyte count) and haemoglobin electrophoresis. Laboratory investigations
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 count and Hb electrophoresis. Results of the blood count were as follows: Mean Hb level 12.2±2.3 g/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 the haematological feature of HbSS patients similar to many previous reports in Sudan. Our study reported a 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.

All patients 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 Alhassania tribe in 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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REFERENCES: