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Ethnic and Geographical Distribution of Sickle Cell Disease Patients Referred to Al-gadarif Teaching Hospital (East of Sudan)

Tariq. Elfatih. Elmisbah

Department of Clinical & Biotechnology Sciences, College of Applied Medical Sciences, Taif University, Kingdom of Saudi Arabia

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Abstract

Background: *This is a descriptive, cross –sectional, analytical study has been carried out in Algadrif hospital (East of Sudan) to detect the ethnic and geographical distribution of sickle cell anemia (SCD) patients referring to hospital.*
Study design*: This study was designed to determine the ethnic and geographical distribution of sickle cell anemia in patients referring to hospital East of Sudan and consent was taken from the health authority.*
Materials and methods*: Hundred blood samples were collected into EDTA blood containers (5 ml) from different SCD patients. All samples were investigated by mythic 18 for CBC and also investigated for Hb type using of Hb electrophoresis.*
Results: *The study showed that the frequency of SCD was found in Hosa tribe with frequency of 57% Followed by Falata, Burno, four, Masalet , Tama and Messeria with frequencies of 18%,9%7%,5%,3% 1% respectively .Geographic distribution showed that the highest frequencies of SCD were found in tribes originated from outside Sudan (Hosa , Falata ,Tama, Burno) with percentage of 87 % &the lowest were found in tribes originated from Kurdufan state (Messeria) with frequency of (1%). Also the study showed that no case was recorded from tribes of North , South and East of Sudan. The result showed that the percentage distribution of patients according to Hb type was 53% for AS and 47% for SS.*
Conclusion: *Most common tribes that had been affected with SCA in ALgdarif state was Hussa tribe that originated from abroad Sudan, where the lowest one was Misseria that originated from the West of Sudan.*

Keywords: *Sickle cell disease, Sudanese tribes, East of Sudan*

1. Introduction

In 1910, james B. Herrick reported “peculiar, elongated, sickle-shaped red corpuscles”in”a case of severe anemia “in a black student.emmel,in 1917, observed the transformation of the biconcave red corpuscle to the “sickle form in vitro. **Sickle-cell disease** (**SCD**) is a group of blood disorders typically inherited from a person's parents.[1] The most common type is known as **sickle-cell anaemia** (**SCA**). It results in an abnormality in the oxygen-carrying protein haemoglobin found in red blood cells. This leads to a rigid, sickle-like shape under certain circumstances.[2]

Problems in sickle cell disease typically begin around 5 to 6 months of age. A number of health problems may develop, such as attacks of pain ("sickle-cell crisis"), anemia, bacterial infections, and stroke.[2] Long term painmay develop as people get older. The average life expectancy in the developed world is 40 to 60 years.[4]

Sickle-cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent.[3] Several subtypes exist, depending on the exact mutation in each haemoglobin gene.[1] An attack can be set off by temperature changes, stress, dehydration, and high altitude.[2] A person with a single abnormal copy does not usually have symptoms and is said to have sickle-cell trait.[5] Such people are also referred to as carriers.[4] Diagnosis is by a blood test and some countries test all babies at birth for the disease. Diagnosis is also possible during pregnancy. The terms "sickle-cell crisis" or "sickling crisis" may be used to describe several independent acute conditions occurring in patients with SCD. SCD results in anemia and crises that could be of many types including the vaso-occlusive crisis, aplastic crisis, sequestration crisis, haemolytic crisis, and others. Most episodes of sickle-cell crises last between five and seven days.[14] "Although infection, dehydration, and acidosis (all of which favor sickling) can act as triggers, in most instances, no predisposing cause is identified [15].

Sickle cell disease in Sudan

The Origin of Sickle cell gene in Sudan: Based on analysis of Y-chromosome haplogroups, the sickle cell gene may

have been preferentially introduced through males of migrating West African tribes, particularly Hausa-Fulani, and Bagara in the large migrations that began in the eighteenth century and escalated during the nineteenth and early twentieth century [10]. The haplotypes associated with the S gene in Sudan are most likely to be the Cameroon, Benin, Bantu and Senegal haplotypes rather than Saudi-Asian haplotype. Among 40 clinically and electrophoretically confirmed SCA cases, the Cameroon and Benin haplotypes accounted for 25% each of the samples [11]. The most frequent haplotype among 143 chromosomes with S gene was the Cameroon (35.0%), followed by the Benin (29.4%), the Senegal (18.2%) and the Bantu (2.8%). The Indian-Arab haplotype was not observed. Three atypical haplotypes were identified in 17 patients, occurring at a combined frequency of 14.6%. One of these, found at the high frequency of 11.8%, possibly represented a new Sudan haplotype [12].

Materials and Methods

One hundred venous blood samples were collected from SCD patients of different areas of East of Sudan who were referred to Algadarif teaching hospital. The data was collected by direct interview through designed questionnaire for each patient included all tests results have been identified in addition of the special information about the patient.

EDTA blood samples were analysed CBC by Sysmex mythic 18 automated hematologyical analyzer.

Electrophoresis method

Principle

Known amounts of Hb are added to serum. Hb-haptoglobin complex is separated by electrophoresis on cellulose acetate; the presence of bound and free Hb is identified in each sample, and the amount of haptoglobin is estimated by noting where free Hb appears. At alkaline pH, Hb is a negatively charged protein when subjected to electrophoresis it migrate toward the anode (+). Structural variants that have a charge in the charge on the surface of the molecule. At alkaline pH it separate from Hb A. Hb variants that have amino acid substitution that is internally sited may not separate, and those that have an amino acid substitution that has no effect on overall charge will not separate by electrophoresis.

Results

Table 1 Distribution of SCD patients according to the age

Age	Number	Percentage
Infant	5	5%
Child	47	47%
Adult	48	48%
Total	100	100%

Table 2 Distribution of SCD patients according to the sex

Sex	Number	Percentage
Female	**64**	**64%**
Male	**36**	**36%**
Total	**100**	**100%**

Table 3 Distribution of SCD patients according to the type of Hb

Type of Hb	Number	Percentage
Sickle cell trait (AS)	53	53%
Sickle cell disease (SS)	47	47%
Total	100	100%

Table 4 Distribution of SCD patients according to the tribe

Tribe	%
Hosa	57
Falata	18
Foor	7
Masaleet	5
Barno	9
Tama	3
Messeiria	1

Table 5 Distribution of to the tribeS according to the origin

Origin	%
Abroad Sudan	87
Inside Sudan	13

Discussion

The results showed that the highest frequency of SCA was found in Hosa tribe while the lowest was found in Messeria tribe.

According to the geographical distribution it was found the highest frequency of sickle Anemia in the tribes that originated from outside Sudan while the lowest frequency was found in Sudanese tribes. Similar results were reported by Abd El raheem said that SCA common in African tribes.

The study showed that the majority of SCD patients in Algadarif city originated from West of Sudan, while no single case reported from tribes originated from North, South and East of Sudan.

The result showed that the frequency of sickle cell trait patients (AS) more than sickle cell disease patients (SS) due to intermarriage between different tribes. The traditional tribal society is still existent in Sudan. The lack of public health measures and services for the prevention of genetic disorders in general.

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