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Original Article

Sickle Cell Anemia In Sheedi Population of Lyari: Hemoglobinopathy Seen in a Neglected Population

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Abstract:

Background: Hemoglobinopathies are global health burden. Some geographical areas are more severely hit by this problem than others, this is partly related to the genetics and partly to the custom of inter-marriages seen in some populations. Counseling could play an important determinant in combating the increased incidence of hemoglobinopathies by reducing family inter-marriages.

Material & Methods: This descriptive cross-sectional study was conducted in Lyari General Hospital Karachi for six months. Two hundred(200) subjects of Sheedi/Makrani population
were inducted and Complete blood |Count (CBC) with Red blood cell indices and absolute values, peripheral smear and electrophoresis was performed.

Results: Most of the study participants were females with female to male ratio of 1:0.59. Out of 200 study participants, 9% (n=18) were found to be Sickle carriers, while 5% (n=10) were found to have Sickle cell disease with total frequency was 14% in the study population.

Conclusion: Sickle cell anemia was found to be affecting 14% of study population belonging to the Sheedi/Makrani caste residing inLyari. High percentage of interfamily marriages relates as one of the possible reasons of high frequency of Hemoglobinopathies in that cast.

Key Words: Sheedi/Makrani, Sickle cell Anemia, Sickle cell trait, Hemogram, Anemia, Hemolytic anemia

Introduction

The inherited single gene disorders in which there is either functional or structural deformity in hemoglobin molecule are termed as Hemoglobinopathies. These include a variety of diseases such as sickle cell anemia and thalassaemia.[1]Accordingto World Health Organization (WHO) report around 5% of world population carry abnormal hemoglobin gene as a result more than 12,000 pregnancies produce children with hemoglobinopathies yearly and around 42 million carriers are born each year.

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Sickle cell anemia is one the most common hemoglobin disorder after thalassemia and approximately >300 000 people are effected per year globally.[3]It is estimated that around 312000 homozygous sickle cell births occur in a year. In developed countries where better health care system is available, life threatening complications of the disease are avoided and quality and span of life is improved.[4]On the other hand approximately more than 90% of patients with sickle cell anemia are seen in low resource income areas and with no or poor medical facilities, therefore they succumb to the complication of disease before their 5th birthday.[5]WHO statistics display that approximately 5% of the children fatality results from sickle cell disease.[6]

SCA is mostly compounded to Asian and African countries; the major factor for this increasing incidence in these areas could lie in the fact that custom of inter-

family marriages is very high in these populations.[7-8] Lyari is one of the backward areas of Karachi with marked illiteracy, lack of basic facilities and very limited health facilities. Currently there is dearth of data regarding burden of hemoglobinopathies in Pakistan and especially in Sheedi cast which originated from African population and is at major risk for hemoglobinopathies[9]

Screening of general and specifically high risk population is the need of the day to reduce the disease burden by counseling for inter-familial marriages in carriers.

Methodology

The present study was carried out on 200 subjects including both patients and attendants of Sheedi cast who were visiting or admitted to the Lyari General Hospital. Children less than Six (6) months of age and patients who received blood transfusions within two weeks were excluded. This was a prospective cross sectional study which was carried out for one year with the approval of ethical review board of Dow University of Health Sciences.

Calculatedsample size was 185, which was calculated for 95% confidence interval in population size of 607,992 at 22% prevalence of Sickle cell anemia with 5% error[11]. But we included 200 subjects.Individual consent and interview were taken through questionnaire first, then 2 ml intravenous blood was collected in EDTAanticoagulaed tube after maintaining all aseptic measures.

The samples were transferred to Dow Ishrat-ul-ebad Khan Institute of Blood diseases (DDRRL) maintaining the cold chain, for peripheral smear with complete blood counts and electrophoresis.

Peripheral smears were prepared and stained in laboratory, Complete blood count was obtained using automated hematology cell counter, Cell TAC-α and 18 parameter haemogram indices were obtained and electrophoresis was performed on all the samples using Genio-S (fully automated system) by INTERLAB (G-26).

Statistical analysis was performed using SPSS16.Age, gender, different haemogram indices; peripheral smear morphology and electrophoresis were our study variables.

Results

Most of the study participants were females with female to male ratio of 1:0.59.

Out of 200 study participants 9% (n=18) were found to be Sickle carriers, while 5% (n=10) were found to be having Sickle cell disease on Hemoglobin electrophoresis with total frequency of 14% in our study population.

Most common age group was 11-20 and 1-10 years for sickle cell disease and trait respectively.

More than half (59%) of study participants provided positive history of inter-family marriages among their parents, 15 out of 18 (83.3%) for carriers and 10 out of 11 (90.9%) for Sickle cell disease (Fig 1).

The Sickle trait showed occasional Sickle cells on peripheral smear morphology; while in diseased cases there were target cells, histocytes and significant number of sickle shaped cells (Fig 2).

The mean hemoglobin for sickle cell trait and disease was 8.5 g/dL and 7.9g/dL respectively, while other indices were red cell count 3.99 x 10^{12}/Land 3.22x 10^{12}/L, hematocrit 29.2% and 25.06%, mean corpuscular volume 71.1 fL and 82.7 fL, mean cell hemoglobin 21.1 pg and 25.7 pg, mean cell hemoglobin concentration 29.15g/dLand 30.3 g/dL. The Hemogram indices of sickle trait and diseased subjects are shown in Table1.

Hemoglobin electrophoresis results and representative graphs are shown in Figure 3 and Figure 4.

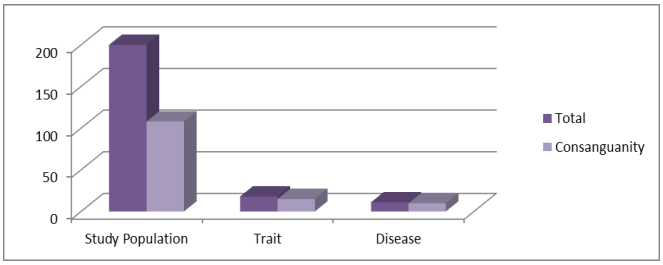

Figure 1: **Graph showing high incidence of Family marraiges (Consanguanity).**

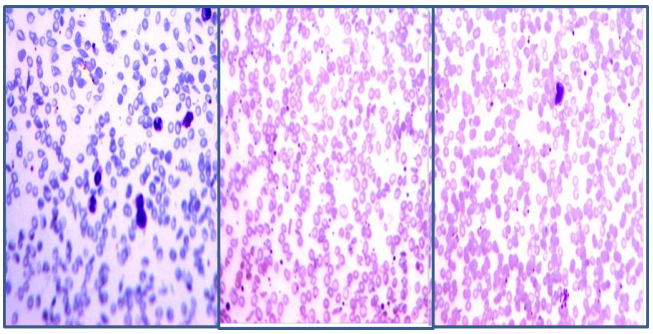

Figure 2: Peripheral smear morphology of (a) Sickle cell disease (b) Sickle Trait and (c) Normal.

Table 1: Hemogram parameters of Sickle trait and Disease patients

HAEMOGRAM INDICES ± SD	STUDY POPULATION (n=200)	SICKLE CELL TRAIT (n=18)	SICKLE CELL DISEASE(n=10)
Haemoglobin (g/dL)	10.2 (2.6-16.8)	8.50 (3.3-14.0)	7.9 (6.3-9.2)
Red cell count (10^{12}/L)	4.37 (1.6-6.4)	3.99 (1.66-5.9)	3.22 (2.2-5.9)
Haematocrit (%)	32.5 (30.0-49)	29.2 (12.6-49)	25.06 (20-32)
Mean corpuscular volume (fL)	75.69 (47-101)	71.1 (52.6-100)	82.7 (54-101)
Mean corpuscular haemoglobin (pg)	23.48 (10-57)	21.1 (11.5-33.3)	25.7 (16-31.7)
Mean corpuscular haemoglobin concentration (g/dL)	30.78 (21-84)	29.15 (21.9-33.3)	30.3 (27-33.7)

Fig 3:Hb Electrophoresis
(T= Sickle trait, D= Sickle disease, -ve C= Negative control, +ve C= Positive control)

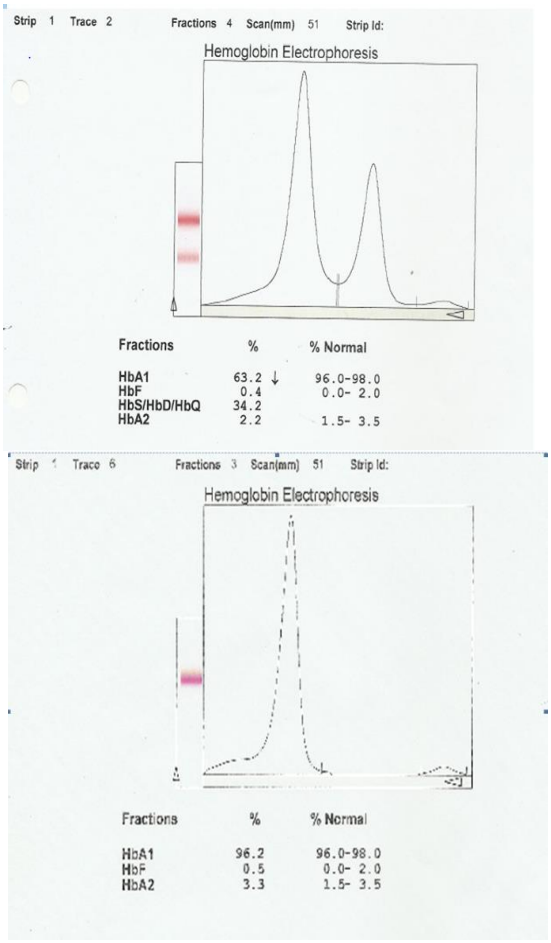

Fig 4: Graph of sickle cell trait and disease patients on electrophoresis

Discussion

Sickle cell anemia is a neglected disease in Pakistan although there is a dearth of data regarding its prevalence, but it is present in Pakistan quite significantly. Despite less than 1% overall prevalence reported for sickle cell disorders in Pakistan, the prevalence pattern exhibits heterogenous distribution with concentration in certain regions of the country, particularly Balochistan where prevalence as high as 10% has been reported.[9]In Karachi, it is centered to lower socio-economic class, including Sheedi/Makrani population. These people find it difficult to maintain their day to day need so spending money on health-care is a luxury. Due to this fact true picture of sickle cell anemia prevalence in our population has not yet been determined.[9]

The overall frequency of sickle cell disease in this study is 14% which is in contrast to previous study

performed in Jinnah Postgraduate Medical Centre, Karachi (JPMC) by Kazmi A et al during a period of 2 years and detected only 8 cases of sickle cell disorder. Another study conducted by Amanat et al., on pattern of heamoglobinopathies also displayed very low percentage of sickle cell disease patients (only 0.3%). This difference may be since present study targeted only at-risk population, i.e. Sheedi/Makrani race.[10]

Ghani et al in a study claimed that 5% of Karachi and 22% of Lyari population is suffering from Sickle cell anemia but in our study, we found 14% incidence in latter. This could be due to change in sampling technique, or size of study population.[11]

Last study conducted by Hashmi.N. K. et al., showed that Pakistan has 1.92% prevalence of sickle cell anemia and 46.4% of the patients are residing in Balochistan, whereas 33.1% of the patients belong to the province of Sindh.[9]Thus, our study and other studies showed that the frequency of sickle cell disease is quite significant in our country but specifically in certain areas of Pakistan.

Consanguinity was also found to be one of the major problems of the study population and this custom of inter-family marriages should be discouraged so as to reduce the new cases of hemoglobinopathies in at-risk population.

Conclusion

Sickle cell anemia was found to be affecting 14% of our study population belonging to Sheedi/Makranicaste of Lyari. High percentage of interfamily marriages relates as one of the possible reasons of high frequency sickle cell anemia in that cast. Further studies with large sample size are needed to analyze the prevalence of this hemoglobinopathy in that population..

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