Original Research Article

Prevalence of sickle cell disorder in patients visited to tertiary care hospital

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Received: 22-11-2020 / Revised: 06-01-2021 / Accepted: 04-02-2021

Abstract

Background: Sickle cell disease (SCD) is prevalent in many countries of the world. In India, it is more common in central and southern parts of the country. Because of this fact, this study was initiated to study the prevalence of sickle cell disease.**Methods:** Diagnosis of SCD was done by sickling slide tests, solubility test and haemoglobinopathies were determined by cellulose acetate membrane tests.Results: 2,386 patients were screened by sickling slide test, and 109 SCD cases (4.55%) were detected. The same cases were also positively identified by solubility test. Electrophoretograms showed 4 (3.66%) of the study subjects were having SS type of Hb pattern while 105 (96.33%) were AS of Hb pattern.**Conclusion**: Sickle Cell Disease is prevalent in this area which is around 4.55%. There should be evaluation of relatives for the purpose of early diagnosis.

Keywords: Sickle cell disease, sickling, hemoglobin, HbS.

Introduction

Sickle cell disease (SCD) is prevalent in many countries of the world, including India. In India, it is more common in central and southern parts of the country. Sickle cell anemia (SCA) in Indians which is linked to the Arab-Indian haplotype has a mild clinical presentation which sometimes goes unnoticed throughout life. This has been attributed to the high fetal hemoglobin and associated alfa-thalassemia commonly seen among these patients.[1]Most individuals with SCD are healthy at birth and become symptomatic later in childhood after fetal haemoglobin levels decrease. Sickle cell anaemia (SS): An inherited disorder of the red blood cells in which the haemoglobin is different from the normal haemoglobin. This unusual haemoglobin results in the production of unusually shaped cells and is referred to as "HbSS." Sickle cell disease (SCD): An inherited disorder of the red blood cells in which one gene is for sickle haemoglobin(S) and the other gene is for unusual haemoglobin such as S, C, and Thalassemia.

Sickle cell trait: A person carrying the defective gene HbS, but it has some normal haemoglobin HbA. Haemolysis causes actual anaemia of the illness, abnormal shape of red cells inside the spleen leads to their destruction. The bone marrow attempt to recover by forming new red cells, which does not compensate the rate of destruction. Healthy red blood cells typically live 90 - 120 days, but sickle cells survive 10-20 days. [2,3] This study was planned to know the epidemiology of sickle cell anemia in our region.

Materials and methods

All the chemicals were of an analytical grade and supplied by s. d. fine chemical limited,Mumbai,India.To confirm Sickle Cell Disease, a sickling slide test(using 1gm/dL sodiummetabisulphite)was performed. [4] This study involved prior informed consent from the participants and was found to be within ethical standards as the Helsinki Declaration was followed. Blood was to be collected in an EDTA-K2 vial. For solubility test procedure, we mixed 1.0 ml of solution into10 uL of packed RBCs in a glass tube. We observed to look for the development of transparency or opacity of the red-colored solution after five minutes. The test was read as positive, if the turbidity impaired the visibility of dark, bold lines on a white paper held against bright source of light at one inch distance. Negative test was indicated by visible lines. Sickling positive tests were run on haemoglobin electrophoresis using cellulose acetate membrane method.[4] The control haemoglobin samples of HbA and HbS were kept in every batch. Haemoglobin was estimated by 3 parts Automated Haematology Analyzer. The study cases of Sickle cell anaemia having haemoglobin less than 12 gm/dl were graded for anaemia- as mild (10-10.9 gm %), moderate (7-9.9 gm %) and severe (<7 gm%) anaemia based on their initial haemoglobin level.The study was conducted in a span of six months (May-October, 2017) at DUPMC hospital, Jalgaon. We screened 2,386 suspected patients referred by consultants from different departments in our laboratory.

Statistical Analysis: Statistical method used for analysis of data. Statistical softwares used for the analysis of the data were SPSS version 20.

Results

Table 1 shows that, prevalence of disease in the study subjects was found to be 4.55%. Out of total 2386 study subjects 2277 (95.43%) were found to be negative while 109 (4.55%) were found to be positive on both Solubility test as well as Sickling slide test for the screening of Sickle Cell Disorders. Table 2 shows, the distribution in males was found to be 60 out of 109 (55.04%) while it was (44.95%) in female patients. Distribution of 109 study subjects (who have shown positive screening test) according to pattern of Hemoglobin electrophoresis. It was found that 4 (3.66%) of the study subjects were having SS type of Hb pattern i.e. they were having the sickle cell disease and 3 of them were males and 1 is female patients.

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Remaining 105(96.33%) study subjects were having AS type of the Hb pattern i.e. they were having Sickle Cell Trait. Out of these 105 study subjects, 57 were males and the remaining 48 were females.Table 3 shows, the severity of anemia amongst the study population. 2.75% patients were severely anemic, 43.11% patients were moderately anemic while 54.12% were mildly anemic.Table 4 gives the idea of hematological parameters while table 5 shows the clinical presentation of study subjects.

Table 1: Agewise distribution

Sr No	Age(years)	Total patients Screened	Total positive		Percentage(%)
			Solubility	Sickling Slide Test	
1	0-5	89	4	4	4.49
2	6-15	254	12	12	4.72
3	15-25	312	13	13	4.16
4	26-35	571	27	27	4.72
5	36-45	726	33	33	4.54
6	46-55	391	18	18	4.60
7	>56	43	2	2	4.65
Total		2386	109	109	4.55

Prevalence of disease in the study subjects was found to be 4.55%.

Table 2: Hemoglobin Electrophoresis results.

Sr No	Electrophoretic pattern	Male	Female	Total
1	HbSS	3	1	4
2	HbAS	57	48	105
Total		60(55.04%)	49(44.95%)	109

4 (3.66%) of the study subjects were having SS type while 105(96.33%) study subjects were having AS type of the Hb pattern.

Table 3: Distribution of study subject according to severity of anemia

Hemoglobin Level	Total	%
< 7 gm%	3	2.75
7-9.9 gm%	47	43.11
10-10.9 gm%	59	54.12
Total	109	100

Table 4: Haematological parameters in Sickle Cell patients

Parameters	Range	Mean
Hb	4.3-12.6	9.03
RBC	1.47-5.98	4.04
HCT	14-38.2	32.03
MCV	63.2-98.4	72.71
MCH	19.2-31.6	22.72
MCHC	27.2-34.6	31.2

Mean Hb concentration was 9.03 gm/dl, mean HCT 32.03 %, mean MCHC- 31.2 g/dl, mean MCV- 72.71fl.

Table 5: Clinical picture of study subjects

Clinical Features	No of patients	Percentage %
Pallor	107	98
Fever	32	29.35
Abdominal Pain	23	21.11
Chest pain &/Or joint pain	42	38.53
Vascular occlusive crisis	1	0.91
Hemolytic crisis	1	0.91

Pallor was 98 %, Fever - 29.35%, abdominal pain (21.11%), chest and joint pain (38.53 %).

Discussion

2-34% prevalence of sickle cell gene has been found in India.The highest frequency of sickle cell gene in India is reported in Orissa followed by Assam, Madhya Pradesh, Utter Pradesh, Tamilnadu and Gujarat.[5] Patel AB et al reported older age of presentation, absence of severe anemia, male preponderance in their study. [6]The prevalence of the disorder was found to be 4.55%. Kamble reported prevalence in Wardha to be 5.7% in a clinic based study. [7] The differences may be attributed to the differences in study designs. In the present study, the prevalence of sickle cell disorders was maximum in the age group 36-45 yrs. Leikin et al in his cooperative study also had patients of sickle cell disorders which were distributed more in higher age groups.[8] Ankushe and Kamble also noted the similar findings.[7,9] The prevalence of the disorder was 55% in males and 44% in females. As regards to sex distribution of the disorder, Wintrobe stated that sickle cell trait is more common in females.[10] But we did not find any such correlation. Although the proportion of Sickle Cell disorder in other caste was found to be on higher side in this study, less number of

study participants belonging to the other castes can't be ignored. Deshmukh PR et al.(2006) has also reported presence of Sickle cell disorders in many castes and tribes of Central India.[11]Pallor was recorded in 98 % and fever in 29.35%, abdominal pain (21.11%), chest and joint pain (38.53 %) & was found in sickle cell patients. Vascular occlusive crisis 0.91 % was the commonest crisis encountered followed by hemolytic crisis 0.91%. The commonest precipitating factor for painful crisis is cold, education and proper clothing which may minimize its effect.[3] 3 of 109 patients had clinically detectable splenomegaly and 0.91 % had hepatomegaly. The peak prevalence for both occurred in the 5-10 year age group which is similar to Milena Magalhaes Aleluia et al .[12] In our study the mean haemoglobin concentration was 9.03 gm/dl, mean HCT%, mean MCHC- 31.2 g/dl, mean MCV- 72.71fl, which is similar to the study of Rinku et al.[3]High prevalence is observed in the rural area and diagnostic and treatment facilities are also not properly available. Modern interventions like Bone Marrow Transplantation(BMT),Gene Therapy(GT),Preimplantation Genetic Diagnosis(PGD) and Prenatal diagnosis is beyond their capacity (the population with sickle cell anaemia inheritance). It indicates limited availability of education and counseling available to susceptible populations,[13] which was also observed in present study.

Conclusion

The prevalence of the Sickle cell disease in this region was found to be 4.55%. SCD is prevalent in this area. Evaluation of relatives for the purpose of early diagnosis and to avoid complications. Proper and adequate counseling should be given to couple before marriage and haemoglobin genetic counseling and education should be included in the society. Lack of knowledge and awareness enhances superstitions about the disease. It is necessary to establish community control programme involving people, doctors and social workers.

Acknowledgment: Authors are grateful to Dr Ulhas Patil Medical College and Hospital, Jalgaon for this work. The authors are grateful to the technical staff for their cooperation and help rendered. This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

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Conflict of Interest: Nil
Source of support:Nil