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Demographic and Geographic Distribution of Sickle Cell Disease in Gondia District of Central India; A Hospital Based Study.

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ABSTRACT

Background: Sickle cell disease is prevalent in central India. Gondia district is located in the Vidarbh region of Maharashtra of central India. Objective of this study is to find the geographic (block level and primary health center level) and demographic (age, sex, caste, sub-caste) distribution of sickle cell carriers and cases which presented to the hospitals for last 10 years.

Methods : All the symptomatic patients visiting five hospitals of Gondia were tested by solubility test. Those found positive in solubility test were tested by hemoglobin electrophoresis. Results were categorized into sickle cell carriers/traits (AS pattern) and sickle cell cases (SS pattern).

RESULTS:

Total 1318567 patients were tested by solubility test and 18152 were found positive. These underwent hemoglobin electrophoresis and revealed 11209 carriers/traits and 1088 cases. More females were carriers and more males were cases. Most of the patients presented were between 11-20 years of age. Unmarried cases were significantly higher than unmarried carriers. With around 20% of disease burden, Gondia block have highest number of patients. Schedule casts have highest number of patients followed by other backward classes and schedule tribes.

Conclusion: Maximum number of sickle cell patients was from Gondia block followed by Arjuni Morgaon and Sadak Arjuni block. Sickle cell disease is most prevalent in Mahar sub-caste of SC, Gond, Halbi sub-caste of ST and Kunbi, Mali, Teli, Kalar sub-caste of OBC. Public health authorities need to focus more in these blocks and sub-castes to promote awareness and improve quality of life.

Keywords: sickle cell, gondia, caste.

Introduction

Sickle cell disease has an autosomal recessive pattern of inheritance [1]. The gene defect is a single nucleotide mutation of the ß-globin gene, which results in glutamic acid being substituted by valine at position 6. Under low oxygen concentration, HbS polymerizes and forms fibrous precipitates because the deoxy form of hemoglobin exposes a hydrophobic patch on the protein [2]. In people heterozygous for HbS (carriers/ traits), the polymerization problems are minor because the normal allele is able to produce half of the hemoglobin. In people homozygous for HbS (cases), the presence of long-chain polymers of HbS distorts the shape of the red blood cell and makes it fragile and susceptible to break within capillaries. Carriers have symptoms only if they are deprived of oxygen (for example, while climbing a mountain) or while severely dehydrated. The allele responsible for sickle cell anemia is present on the short arm of chromosome 11 [3].

Hemolysis is the actual cause of anemia. Healthy red blood cells function for 90–120 days, but sickled red blood cells last only for 10–20 days [4]. Clinical signs usually begin in early childhood and the severity of symptoms varies from person to person. Sickle cell disease may lead to various acute and chronic complications such as anemia, vaso-occlusive pain crises, risk for pneumococcal infections, acute chest syndrome, stroke and organ failure with high morbidity and mortality. Infection, dehydration, oxidative stress and acidosis promotes sickling [5].

Sickle cell anemia is prevalent among people in malaria endemic areas such as Mediterranean, Africa, India, and the Middle East due to adaptive advantage of heterozygote [6]. In India, the disease is prevalent among tribal populations in Maharashtra, Madhya Pradesh, Chhattisgarh, south Gujarat and western Odisha with a smaller focus in the southern region in Andhra Pradesh, northern Tamil Nadu, Karnataka and Kerala. It is also prevalent in some of the non-tribal populations, scheduled castes and other backward classes mainly in central India [7,8]. It is estimated that India has 50% of the world's sickle cell patients. While sickle cell is prevalent in many ethnic groups, the highest prevalence appears to be in scheduled castes, scheduled tribes, and other backward classes [9]. In India, the prevalence of sickle cell carriers in different tribes varies from 0-35 percent [10].

The sickle cell gene is widespread in all the eastern districts (vidarbha region) of Maharashtra, Satpura range and in some parts of Marathawada. In Maharashtra, a study reported prevalence of disease from 1.9% to 33.5% in different

communities [11]. In central India, the sickle cell anemia incidence was found highest in the scheduled caste group(1:50) [12]. Another study was the first to report the disease in vidarbha region of Maharashtra showing prevalence from 9.4% to 22.2% in non-tribal population [13]. Other study reported prevalence of 5.5% from few villages of Wardha [14]. In a study performed in Vidarbh region, 35,636 people were screened, in which 5466 were identified with sickle cell trait and 1010 were found to have sickle cell disease. This study also showed sickle cell trait prevalence of 13.0% in the SC, 12.0% in the ST and 3.4% in the OBC population [15]. Gondia district is in the Vidarbh region of Maharashtra. As per 2011 census, total population of Gondia district is 1322507 and SC, ST population is 355484, 309822 respectively. In this study, we tried to figure out the geographic and demographic prevalence of sickle cell trait and sickle cell disease in Gondia district.

MATERIAL AND METHODS

This is retrospective descriptional study based on records of five hospitals namely KTS District general hospital, Bai Gangabai womens hospital, Rural hospital of Rajegaon, Deori and Amgaon for last 10 years i.e. from april 2009 to december 2019. As this region has high prevalence of sickle cell disease, an extensive screening of all the symptomatic patients visiting these five hospitals was done for sickle cell trait and disease under sickle cell control programme run by government of Maharashtra. All the patients presenting with anemia, intermittent jaundice, joint pains, painful vaso-occlusive crisis and splenomegaly were tested by solubility test. A mixture of RBC containing HbS in a reducing solution (such as sodium dithionite) gives a turbid appearance, whereas normal Hb gives a clear solution. Hb Electrophoresis of solubility test positive patients was done. Results were categorized into Normal (AA) Trait (AS) and disease (SS). Based on available records results were grouped into geographic prevalence i.e. Blocks, Primary health center (PHC) and demographic prevalence i.e. Age, Sex, Caste, marital status.

RESULTS

Table 1: Number of Tests and Results

Total Solubility test performed	1318567
Positive solubility tests	18152
Total Hb Electrophoresis performed	18152
Normal (HbAA)	5855 (32.25%)
Carrier (HbAS)	11209 (61.75%)
Cases (HbSS)	1088 (5.99%)

Table 2: Gender distribution

Carrier	Male	4723	42.14%
	Female	6486	57.86%
	Total	**11209**	
Cases	Male	561	51.56%
	Female	527	48.44%
	Total	**1088**	

Table 3: Age distribution

Age	**Carrier**		**Cases**	
0-10 yrs.	2114	18.85%	298	27.38%
11-20 yrs.	4230	37.73%	381	35.01%
21-30 yrs.	2685	23.95%	267	24.54%
31-40 yrs.	1407	12.55%	101	9.28%
41-50 yrs	511	4.55%	26	2.38%
51-60 yrs.	198	1.76%	12	1.1%
>60 yrs.	64	0.57%	3	0.27%
	11209		**1088**	

Table 4: Marital status

	Married	**Unmarried**	**Total**
Carrier	4742 (42.3%)	6467 (57.69%)	**11209**
Cases	290 (26.65%)	798 (73.34%)	**1088**

Table 5: Geographic distribution according to Blocks and PHC

Blocks	**PHC**	**Controls**			**Total (%)**
Amgaon	Bangaon	296		16	135 (12.4%)
	Kalimati	341		55	
	Tigaon	218		28	
	Thana	181		36	
Gondia	Bhanpur	312		31	243 (22.33%)
	Ekodi	223		17	
	Rawanwadi	417		76	
	Kamtha	310		37	
	Dawaniwada	201		18	

	Dasgaon	254		22	
	Morwahi	226		21	
	Kati	235		21	
Salekasa	Kawarabhandh	213	913 (8.14%)	21	107 (9.83%)
	Satgaon	303		36	
	Darekasa	288		39	
	Bijepar	109		11	
Arjuni Morgaon	Channa Bhakti	447	1997 (17.81%)	40	173 (15.9%)
	Dhabepawani	353		35	
	Korambhitola	397		28	
	Mahagaon	304		24	
	Gothangaon	249		29	
	Keshori	247		17	
Sadak / Arjuni	Pandhari	799	1921 (17.13%)	43	156 (14.33%)
	Dawwa	278		22	
	Shenda	444		38	
	Soundad	400		53	
Deori	Mulla	399	1261 (11.24%)	68	116 (10.66%)
	Kakodi	324		17	
	Futana	378		19	
	Ghonadi	160		12	
Goregaon	Soni	265	1346 (12%)	11	85 (7.81%)
	Chopa	253		15	
	Kawalewada	159		16	
	Kurhadi	375		34	
	Tilli Mohagaon	294		9	
Tiroda	Indora	129	557 (4.96%)	13	73 (6.7%)
	Mundikota	116		21	
	Wadegaon	187		28	
	Sukadi	125		11	

Table 6: Caste distribution

Carrier	SC	4556	40.64%
	ST	2757	24.59%
	OBC	3552	31.68%
	Others	344	3.06%
Cases	**Total**	**11209**	
	SC	553	50.82%
	ST	179	16.45%
	OBC	283	26.01%
	Others	73	6.7%
	Total	**1088**	

Table 7: Subcaste distribution

Caste	Subcaste	Carrier		Cases	
	Mahar	4352	95.52%	537	97.1%
	Chambhar	126	2.76%	11	1.98%
	Holiyaa	78	1.71%	5	0.9%
	Total	**4556**		**553**	
ST	Gond	1530	55.49%	131	73.18%
	Halbi	859	31.15%	35	19.55%
	Gowari	346	12.54%	13	7.26%
	Nagarchi	8	0.29%	0	0%
	Bhill	7	0.25%	0	0%
	Binjhwar	7	0.25%	0	0%
OBC	**Total**	**2757**		**179**	
	Kunbi	1238	34.85%	111	39.22%
	Mali	561	15.79%	34	12.01%
	Teli	547	15.39%	55	19.43%
	Kalar	489	13.76%	37	13.07%
	Dhiwar	243	6.84%	19	6.71%
	Powar	131	3.68%	8	2.82%
	Lohar	95	2.67%	6	2.12%
	Lodhi	75	2.11%	4	1.41%

	Sonar	68	1.91%	5	1.76%
	Kohali	57	1.6%	2	0.7%
	Muslim	34	0.95%	2	0.7%
	Koshti	3	0.08%	0	0%
	Kumbhar	5	0.14%	0	0%
	Ahir	3	0.08%	0	0%
	Thakur	3	0.08%	0	0%
	Total	**3552**		**283**	

DISCUSSION

Total 1318567 patients were tested for solubility test in during last 10 years and 18152 patients were tested positive. All these underwent Hb electrophoresis in which 5.99% diagnosed as sickle cell cases, 61.75% as sickle cell trait or carrier and 32.25% as normal (Table:1). Female (57.86%) dominated as carrier while male (51.56%) dominated as cases (Table:2). Most of the cases and carriers were under 30 years of age with majority in between 11-20 years. Significantly more cases were found below 10 years as compared to carriers (Table:3). This shows early appearance of clinical symptoms in homozygotes as compared to the traits. More unmarried cases (73.34%) were reported as compared to unmarried carriers (57.69%) and it was statistically significant (Table:4). This is due to higher number of homozygotes below 10 years of age as compared to carriers. Problem in marriage may also contribute to this due to more severe symptoms.

Gondia district has 8 Blocks which further have 39 primary health centers. We observed that maximum number of patients was from Gondia block followed by Arjuni Morgaon and Sadak Arjuni block. Amgaon, Deori and Goregaon have approximately equal distribution followed by Salekasa and Tiroda (Table:5).

Sickle cell disease was found to be most prevalent in schedule casts with 40.64% of carrier and 50.82% of cases (Table:6). Among schedule casts Mahar sub-caste predominates with more than 95% of cases and carriers (Table:7). Schedule tribe population contributes to 24.59% of carriers and 16.45% of cases. Gond sub-caste contribute more than half of ST carriers and more than 2/3rd of ST cases followed by sub-caste Halbi and Gowari. OBC population contributes 31.68% of carriers and 26.01% of cases with major contribution from sub-caste Kunbi (more than 1/3rd of OBC cases and carriers) followed by sub-caste Mali, Teli, Kalar, Dhiwar, Powar, Lohar, Lodhi, Sonar etc.

CONCLUSION

Maximum number of Sickle cell patients was from Gondia block followed by Arjuni Morgaon and Sadak Arjuni block. Schedule cast (Mahar sub-caste) predominate the total number of sickle cell traits and cases followed by OBC (Kunbi sub-caste) and ST (Gond sub-caste). As far as sub-castes are concerned, Mahar is a way ahead in number of cases and carriers followed by Gond, Kunbi, Halbi, Mali, Teli, Kalar, Gowari, Dhiwar etc. Public health authorities need to focus more in these blocks and sub-castes to promote awareness and improve quality of life.

LIMITATION

This is retrospective descriptional study based on hospital records of last 10 years and it does not give clear image of community prevalence because people come to the hospital when they are symptomatic. Real prevalence of the disease would be more as many people will be asymptomatic also. More community based extensive screening programmes are needed to get the real picture.

DECLARATIONS

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