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Research Article

Distribution of Sickle Cell Disease in Different Communities of Patient Visiting Out Patient Department

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Abstract

Background: The highest prevalence of Hb S is in tropical Africa and among blacks in the countries that participated in the slave trade. Results of studies of DNA polymorphisms linked to the ßs gene suggest that it arose from three independent mutations in tropical Africa. The sickle cell anemia and sickle cell trait are observed to occur in relatively high frequencies among the endogamous population of India. Here in this study we have screened the sickle cell patients visiting OPD at our institution to find the cast - wise prevalence of sickle cell disease. **Materials and Method**: Subjects included are diagnosed cases of sickle cell disease taken from OPD. Detailed history of every individual was obtained. Permission was taken from institutional ethical committee. **Results:** Most of the cases in our study are found to be from mahar community with decreasing frequencies in kunbi, otkar, halbi, koshti. Very few cases are also fund from teli muslim maheshwari, brhamin community. **Conclusion:** It is well known that sickle cell gene is widely prevalent in tribal populations. Due to recent activities of globalization, migration of population, inter-caste marrieges and mixing of gene pool it has been seen that the disease which was once thought to be limited to the tribal communities is now also spreading in other communities of the society.

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Keywords: Sickle Cell, Tribal population, globalization, Mahar, Mutation

Introduction

The sickle cell anemia and sickle cell trait are observed to occur in relatively high frequencies among the endogamous population of India. The highest of 22.2% has been found in Lohars of Orissa followed by Mahars of Madhya Pradesh (20%), Kinkars of Assam (19.3%), and Pardhans of Andhra Pradesh (18.3%). The average sickle cell gene frequency is found to be highest 9.1% in Orissa, 8.3% in Assam, 7.4% in Madhya Pradesh, 7.2% in Uttar Pradesh, 7.1% in Tamil Nadu.(1) Prevalence of sickle cell disorder is very high amongst tribal population groups of Bhil and Pawara from Nandurbar district and amongst the Madia, Pardhan and Otkar population from Gadchiroli district i.e. 20%. Here in this study we have screened the sickle cell patients visiting OPD at our institution to find the cast - wise prevalence of sickle cell disease.

Materials and Methods

Subjects included are diagnosed cases of sickle cell disease taken from OPD. Detailed history of every individual was obtained. Permission was taken from institutional ethical committee. The consent was taken from the individuals that were included in the study. A total of 1575 patients were included in the study over a period of time from Dec' 2007 to April '2009. Demographical details included in the history are shown in table No. 1.

Results

Date obtained in the study is tabulated and presented.

Table.1 Demographical details included in history

Name-	Age-
Occupation-	Sex-
Marital status-	Reg. no. -
Address -	Socioeconomic status-
Religion-	Caste-
Family History: History of - Inter-caste/Consanguial Marriages	General Disease History
General Examination	Systemic Examination

Table. 2 Showing sex-wise distribution of subjects in the study

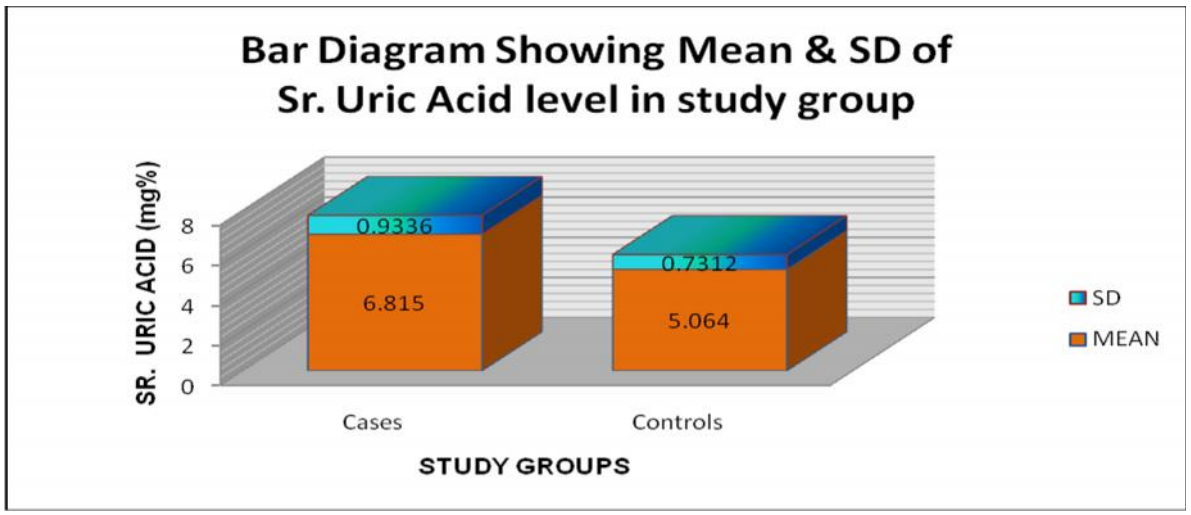
Sr. No.	Sex	Cases
1.	Male	817 (51.9 %)
2.	Female	758 (48.1%)

Table.3 Showing family history of sickle cell in subjects in the study.

Sr. No.	Family History	
1.	present	1543
2.	absent	32

Table.4 Showing caste- wise distribution of subjects in the study.

Sr. No.	Caste	Cases(n=1575)
1.	Mahar	977
2.	Kunbi	283
3.	Otkar	97
4.	Halbi	76
5.	Halba-Koshti	57
6.	Gond	37
7.	Madia	33
8.	Teli	07
9.	Muslim	03
10.	Powar	03
11.	Maheshwari	01
12.	Brhamin	01



Graph No.1: Bar Diagram Showing Mean & SD of Uric Acid level in study group

Table 5 Prevalence of sickle cell diseases by race or ethnic group.

Race or ethnic group	Average prevalence per 100,000 live births
White	1.72
Black	289
Hispanic, total	5.28
Hispanic, eastern states	89.8
Hispanic, western states	3.14
Asian	7.61
Native American	36.2

Table 6 Showing prevalence of Sickle Cell Hemoglobinopathy in some States of India (Hospital attending patients).

State	Prevalence of sickling (%)
Andhra Pradesh	34.6
Bihar	0.6
Gujarat	30
Karnataka	25
Kerala	29.7
Maharashtra	45.4
Madhya Pradesh	48.5
Orissa	12.4
Tamil Nadu	35.3
Uttar Pradesh	32.6
West Bengal	1.1
Adopted from Kamble et al 2000 (12)	

Table.7: Summary of the distribution of sickle cell trait among different tribal groups of Maharashtra (13)

Sr. No	Tribal Group	District	Sickle Cell Carriers (%)
1	Otkar	Gadchiroli	35
2	Pardhan	Nanded, Yeotmal	33.7
3	Pawara	Dhule, Jalgaon	25.18
4	Madia, Gond	Gadchiroli, Yeotmal	20.8
5	Bhil	Nandurbar	20.6
6	Halbi	Gadchiroli	13.93
7	Malhar Koli	Thane	13.88
8	Rajgond	Gadchiroli	10.88
9	Korku	Amravati	9.49
10	Tandvi	Jalgaon	8.33
11	Kolam	Yeotmal	8.33
12	Warli	Thane	8.04
13	Katkari	Pune, Raigad, Ratnagiri	5.90
14	Kokana	Dhule, Nasik	3.50
15	Andha	Nanded	1.97
16	Mahadeo Koli	Pune, Nasik	0.81
17	Thakur	Pune, Thane, Raigad, Ahmednagar	0.00

Discussion

The highest prevalence of Hb S is in tropical Africa and among blacks in the countries that participated in the slave trade. Results of studies of DNA polymorphisms linked to the ßs gene suggest that it arose from three independent mutations in tropical Africa. (2) The most common ßs chromosome is found in Benin and Central West Africa. A second haplotype is prevalent in Senegal and the African West coast. A third haplotype is seen in the Central African Republic (Bantu speaking Africa). The same three types of haplotype are associated with the sickle gene in black Americans and Jamaicans.(3) The Hb S gene in the Eastern Province of Saudi Arabia and in Central India is associated with different DNA structure not encountered in Africa and it represents a fourth independent occurrence of sickle cell mutation (4) Only the Benin and Senegal haplotypes are prevalent among North Africans, Greeks and Italians suggesting that the ßs mutation has spread to the Mediterranean basin from West Africa.(5) In the United States, Latin America and the Caribbean, (6,7) approximately 8% of blacks carry the gene.

In our study, out of 1575 case nearly 51.9 % were male and the rest 48.1 % were females. The higher number of males can be explained by the sex ratio in our region, and the fact that males seek medical attention more than females. Similar findings were noted in Peterson et al (1975) (8). A. M. Hussain et al (1978) (9) Sally Davies et al (1983) (10).

In India, in 1952 Dunlop and Muzumdar reported eight cases, three of which had sickle cell anemia and five cases of sickle cell trait (11) The cases were also found in tea laborers of Assam. The sickle cell anemia and sickle cell trait are observed to occur in relatively high frequencies among the endogamous population of India. The highest of 22.2% has been found in Lohars of Orissa followed by Mahars of Madhya Pradesh (20%), Kinkars of Assam (19.3%), and Pardhans of Andhra Pradesh (18.3%). The average sickle cell gene frequency is found to be highest 9.1% in Orissa, 8.3% in Assam, 7.4% in Madhya Pradesh, 7.2% in Uttar Pradesh, 7.1% in Tamil Nadu.(1)

In our study, we found that most of the cases are from mahar (62%) community with decreaseing order of frequency in kunbi (15%), otkar (6%), halbi (5%), halba - koshit (3.6%), gond (2.3%), madia (2.1%) communities of this region.

R N Shukla and A S Parande in 1956, during their routine work in Government Medical College, Nagpur found a case of sickle cell anemia and 3 cases of sickle cell trait. Case of sickle cell anemia was a 13 year old male from Mahar community of Nagpur. (14)

R N Shukla and B R Solanki in 1958 examined 1010 adult labourers of Model Mills, Nagpur and reported incidence of sickle cell trait as 22.2% in Mahars, 9.4% in Kunbis and 11.3% in Telis. (15) Das et al in 1962 reported the incidence in Mahars to be 18.1 %.Lele et al 1962 found the prevalence of HbS in Mahars is 5.14 %. (16)

Abhyankar D et al (2000) reported the prevalence in general population of Central India as 12 %. (17) Population survey indicates the prevalence of sickle cell disorder in the overall population of Maharashtra is less than 0.1% while it is very high amongst the tribal population from Nandurbar and Gadchiroli districts of the states. The same tribal population groups residing in the neighboring states of Gujarat, Madhya Pradesh and Andhra Pradesh have a similar prevalence. (18)

Prevalence of sickle cell disorder is very high amongst tribal population groups of Bhil and Pawara from Nandurbar district and amongst the Madia, Pardhan and Otkar population from Gadchiroli district i.e. 20%.
The highest recorded prevalence is among the Otkar group i.e. 35%. The sickle cell gene is practically absent in the Mahadeo Koli, Thakur and other tribal groups from Western Maharashtra. The overall prevalence among tribal population is about 10% for the carrier state and 0.5% for sufferers.

In our study even communities like teli, muslim, powar also showed the presence of sickle cell cases. We have also found one case each from brhamin and maheshwari community is also detected. This was probably associated due to mixing of gene pool as a result of inter caste marrieges. Similarly few cases from communities like brahmin and muslim has been detected by Gangakhedkar et al (1989) (19). Also communities like marathas and koshti have also shown the prsence of sickle cell cases in a study according to Ambekar et al (2001) (20).

Out of 1575 cases screened in our study 1543 cases were having positive family history for either sickle cell disease or sickle cell trait. Rest of the 32 cases were negative for family history of sickle cell anemia as well as for sickle cell trait. Transmission of sickle cell disease is autosomal recessive pattern. The cases in our study with negative family history may be a result of germline mutation which remains as a alternate mode of causation of disease that fact needs to be verified.

Conclusion

It is well known that sickle cell gene is widely prevalent in tribal populations. Although large number of population groups have been screened to find the prevalence of the gene still many are yet to be screened. They should be screened with the help of a competent hematology laboratory. Once the prevalence is established, awareness and understanding about the disease should be generated amongst them. Due to recent activities of globalization, migration of population, inter-caste marrieges and mixing of gene pool it has been seen that the disease which was once thought to be limited to the tribal communities is now also spreading in other communities of the society. The simplest way to prevent the birth of affected children is the avoidance of marriage between ‘trait’ persons.

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