International Research Journal of Medicine and Medical Sciences
Vol. 5(4), pp. 64-68, November 2017
ISSN: 2354-211X
Full Length Research Paper

Ethnicity of sickle cell anaemia - Study at Tertiary Care Hospital of Yavatmal

Kalpana B. Rathod* and Jayshree Tijare

Department of Pathology, Government Medical College, India.

Accepted 9 November, 2017

ABSTRACT

Sickle cell anaemia is a hereditary haemolytic anaemia, posing important health problem in the community. In India, this disease has been shown to be prevalent among the three socio-economically disadvantaged ethnic groups, the scheduled tribes, scheduled castes and other backward communities. India has largest tribal population and vidarbha region of Maharashtra state, which includes Yavatmal, contributes significant number of sickle cell cases. The prevalence of sickle cell carriers varies from 0 to 35 percent in different tribes. This study was carried out to find out extent of sickle cell disorders in tribal community in and around Yavatmal. The community wise distribution of sickle cell disorder cases showed maximum cases belonged to Mahar community (Scheduled caste). The tribal communities included were Pardhan, Gond, Banjara, Kolam Gowari and Andh. Amongst the tribes Pardhan showed maximum cases 10%, followed by Gond 7.9%, and Banjara 6.4%. There were also double heterozygous Sickle - β thalassemia in this community.

Keywords: Sickle cell anaemia, tribes, ethnicity, Yavatmal.

*Corresponding author. E-mail: rkalpanaa11@rediffmail.com.

INTRODUCTION

Sickle cell anemia is a hereditary haemolytic anaemia, with autosomal recessive inheritance. Long before the sickle cell anemia was known in Western hemisphere sickling disorders were known in Africa by onomatopoeic names, denoting the recurrent, unrelenting, and painful nature of crisis. Africa has highest prevalence of sickle cell anemia (Hankins and Wang, 2014). This dreaded disorder results from mutation that substitutes thymine for adnine in the sixth codon of the β gene (GAG to GTG). As a result of this mutation, glutamine is replaced by valine in the sixth position of β chain. This minor change in structure is responsible for change in molecular stability and solubility. The tendency of deoxygenated sickle haemoglobin to undergo polymerisation underlies the innumerable expressions of the sickling syndromes.

The sickle gene is widespread amongst tribes in India. In India, this disease has been shown to be prevalent among the other socio-economically disadvantaged ethnic groups like scheduled castes and other backward communities in addition to scheduled tribes (Bhatia and Rao, 1987; Rao, 1988; Kaur et al., 1997; Kate and Lingojwar, 2002; Patra et al., 2011; Urade, 2012; Kaur et al., 2013; Colah et al., 2014). According to the census of India 2009, the tribal population of India is 8.4 percent of the total population which is about 67.2 million people (Census of India, 2009). India has largest tribal population and vidharbha region of Maharashtra state, which includes Yavatmal, contributes significant number of sickle cell cases. Yavatmal district is located in east central part of Maharashtra. It has thirteen Tehsils. The important tribes in Yavatmal district include Kunbi (22%), Andh, Gond and Pardhan (10%), Kolam, banjara (6%) and mali (4%) (Colah et al., 2014). The prevalence of sickle cell gene in the different communities ranges from 9.4 to 22.2% (Shukla et al., 1958). According to studies carried out by Kate (2001), the overall prevalence amongst scheduled caste, scheduled tribes and other backward communities is 10% (Kate and Lingojwar, 2002). Zade et al. (2011) have studied and reported the high prevalence of Sickle cell disease in tribals of Amaravati District.

The inheritance of sickle cell anaemia is controlled by a single gene that transmits from one generation to another. Sickle cell anaemia manifests either as carrier /

heterozygous form, sickle cell disease / homozygous or double heterozygous form. The primary pathophysiology is based on the polymerization of deoxyHbS with formation of long fibers within the RBCs causing a distorted sickle shape which eventually leads to increased hemolysis and vaso-occlusion of sickle red cells (Odièvre et al., 2011).

MATERIALS AND METHODS

Study design: Observational study
Study settings: Tertiary care hospital
Selection of study subjects: Solubility positive cases referred from primary health centres around Yavatmal.
Inclusion criteria: Solubility positive cases and their family members.
All individuals aged 1 to 60 years referred for screening of sickle cell anaemia.
Exclusion criteria: Children < 1 year

Study procedure

Government of Maharashtra has a structured program for control of sickle cell anaemia. Public health department is carrying out screening of population at peripheral (rural) level. Solubility is used as screening test. All positive cases are referred to district hospitals and medical colleges.

This study includes all the referred cases of haemoglobinopathies to tertiary care hospital at Yavatmal, during the January 2009 to December 2009. This tertiary care Hospital located in Yavatmal, which is a city and municipal council in Indian state of Maharashtra. It is administratively headquarter of Yavatmal district. During the above period, total number of 1543 cases referred to tertiary care Hospital, were screened for haemoglobin S.

Solubility test was repeated. 10 µl blood samples were drawn from finger prick for each test and mixed thoroughly with phosphate buffer solution. Result for haemoglobin S variant was noted down after 5 minutes. The sample with turbidity was considered positive for sickle cell disorder (Wild and Bain, 2006). Sample from known case of sickle cell trait was used as positive control.

All the samples were subjected to haemoglobin electrophoresis using cellulose acetate membrane in alkaline TEB buffer at pH 8.9 for electrophoresis pattern (Wild and Bain, 2006). Control of known sickle cell trait (AS) patient was used for electrophoresis. HPLC was used in suspected cases of double heterozygous states like HbS – β Thalassemia.

Ethics committee approval

The present study was approved by the Institutional Ethics Committee.

RESULTS

Total number of 1543 cases were referred to this centre. Females (1023) outnumbered males (520) (Table 1). Amongst 1543 cases majority cases were having sickle cell trait (AS) 71.8% (Table 2). Homozygous sickle cases were 8.8%. Double heterozygous cases of sickle with βThalassemia were also seen and were confirmed by HPLC. Double heterozygous cases were below 30 years (Table 3). Amongst the solubility positive cases referred from PHC 18.3% of c were negative for any hemoglobinopathy (AA pattern) (Table 4).

The community wise distribution of sickle cell disorder cases showed maximum cases belong to Mahar community (35.1%) (Table 5). There were also double heterozygous cases in this community. The tribal communities which were referred include Pardhan, Gond, Banjara, Kolam Gowari and Andh. Amongst the tribes, Pardhan showed maximum cases 10%, followed by Gond 7.9%, and Banjara 6.4% (Table 5).

DISCUSSION

The first description of sickle hemoglobin in India was by Lehman and Cutbush in 1952 in the tribal populations in the Nilgiri hills in south India (Lehman and Cutbush, 1952). In the same year, Dunlop and Mazumdar also reported the presence of sickle hemoglobin in the tea garden workers of Upper Assam who were migrant laborers from tribal groups in Bihar and Odisha (Dunlop and Mazumber, 1952). Since then, many population groups have been screened and the sickle cell gene has been shown to be prevalent among three socio-economically disadvantaged ethnic groups, the scheduled tribes, scheduled castes and other backward classes in India with a prevalence range of 0 to 35 percent (Bhatia and Rao, 1987; Rao, 1988; Kaur et al., 1997; Kate and Lingojwar, 2002; Patra et al., 2011; Urade, 2012; Kaur et al., 2013; Colah et al., 2014).

The presence of the deleterious gene HbS in some groups and the complete absence in some groups indicates that the independent mutation might have taken place during early life of human being. It is evident from the literature that the several ethnic groups with varied genetic elements have been assimilated into the main-stream, resulting in population diversity with the passage of time (Russel and Lal, 1916).

India have nearly 20 million people with sickle cell disease (Ghai, 2000). In India, the sickle cell gene has been reported in various studies as 73% in tribal people, 17% among lower castes, 9 % among middle castes and 1% among higher castes (Rao, 1998). In Maharashtra, the sickle cell gene is widespread in all eastern districts also known as the Vidarbha region, in the Satpura ranges in the north and in some parts of Marathwada. The tribals with high prevalence of sickle cell gene are 20 to 35%. The tribes include Bhils, Madias, Pawaras, Pardhans and Otkars. It has been estimated that Gadchiroli, Chandrapur, Nagpur, Bhandara, Yavatmal and Nandurbar districts would have more than 5000 cases of sickle cell anemia (Colah et al., 2015).

The present study has shown a very high frequency among the Mahar (35.1%) followed by Pardhan (10%), Gond (7.9%), Banjara (6.4%), kunbi (5.4%), Kolam

Table 1. Age wise distribution of study subjects.

Age (years)	Male (%)	Female (%)	Total (%)
13 months - 5	61 (3.9)	66 (4.2)	127 (8.2)
6 - 10	100 (6.4)	112 (7.2)	212 (13.7)
11 - 20	178 (11.5)	386 (25)	564 (36.5)
21 - 30	86 (5.5)	308 (19.9)	394 (25.5)
31 - 40	61 (3.9)	94 (6.0)	155 (10)
41 - 50	26 (1.6)	47 (3.0)	73 (4.7)
≥ 50	8 (1.4)	10 (0.6)	18 (1.1)

Figures in parentheses indicate percentage.

Table 2. Hb electrophoresis pattern in study subjects.

Hb electrophoresis pattern	Total (%)
AA	282 (18.3)
AS	1108 (71.8)
SS	136 (8.8)
β-Thalassemia minor	2 (0.1)
SS with *β*-Thalassemia	15 (1)

Figures in parentheses indicate percentage.

Table 3. Age wise distribution of Hb Electrophoresis pattern.

Age (years)	AA	AS	SS	*β* -Thalassemia minor %	SS with *β*-thalassemia %
13 months – 5	18 (1.1)	98 (6.3)	8 (0.5)	-	3 (0.1)
6 – 10	31 (2)	152 (9.8)	26 (1.6)	-	3 (0.1)
11-20	103 (6.6)	396 (25)	58 (3.7)	1 (0.06)	6 (0.3)
21-30	85 (5.5)	279 (18)	27 (1.7)	-	3 (0.1)
31-40	22 (1.4)	121 (7.8)	12 (0.7)	-	-
41-50	19 (1.2)	48 (3.1)	5 (0.3)	1 (0.06)	-
≥ 51	4 (0.2)	14 (0.9)	-	-	-

Figures in parentheses indicate percentage.

Table 4. Gender wise distribution of Hb electrophoresis pattern among study subjects.

Hb electrophoresis pattern	Male (%)	Female (%)
AA	78 (5)	204 (13.2)
AS	381(24)	727 (47)
SS	54 (3.4)	82 (5.3)
β-thalassaemia minor	--	2 (0.1)
SS with *β*-thalassaemia	7 (0.4)	8 (0.5)

Figures in parentheses indicate percentage.

(4.7%), Gowari (2.5%), Andh (2.3%), Teli (1.1%) and Mali (0.8%). The present findings are in agreement with the findings where they had reported a very high frequency of HbS among the Mahar and Kunbi reported by Shukla et al. (1958).

Also these findings are consistent with the findings of other investigators. The study carried out by Bhobate et al. (1983) in Chandrapur district of Maharashtra in their study found sickle cell trait, 9.42%, that is, maximum in Mahar caste and 5.06% in Teli (21). Kar (1983) also

Table 5. Caste wise distribution off haemoglobinopathies of study subjects.

Caste	% of sickle cell disorder (AS & SS)	% *β*- Thalassaemia
Mahar	542 (35.1)	9 (0.5)
Pardhan	161 (10)	1(0.06)
Gond	121(7.9)	1 (0.06)
Banjara	98 (6.4)	1 (0.06)
Kunbi	89 (5.7)	-
Kolam	74 (4.7)	-
Gowari	39 (2.5)	2 (0.12)
Andh	34 (2.3)	1 (0.06)
Teli	18 (1.1)	-
Mali	13 (0.8)	1(0.06)

Figures in parentheses indicate percentage.

reported high prevalence of sickle cell trait in Mahar (19.8%) (Kar, 1983). Another study carried out by Shukla et al. (1958) among 1010 subjects in Nagpur found the prevalence of sickle cell trait about 22.2% in Mahar and 11.3% in Teli caste.

In the present study, frequency of β-Thalassaemia in Mahar is 0.5 percent followed by Gowari is 0.12 percent. Sporadic occurrence of *β*-thalassaemia has also been found among other Hindu caste population in Vidarbha region. However, a very high frequency (8 to 17%) of the β thalassaemia gene has been reported among the Sindhi population of Nagpur (Jain et al., 2003; Jawahirani et al., 2007).

Tribal population of India have their own culture, custom and even the language used is different. The languages that are spoken in Yavatmal district are Marathi, Gormati/Banjara, Gond, Hindi, Telagu and Kolam. This demands social leader taking up the responsibility of communication in their own language and counseling. As importance of counseling and education about genetic disease cannot be overlooked the information material also need to be translated in local language in order to achieve effective communication. Considering this aspect more of such studies should be planned so as to assess the burden of hemoglobinopathy and planning of effective measures for eradication.

REFERENCES

Bhatia HM, **Rao** VR, **1987** Bombay: Institute of Immunohaematology (ICMR). Genetic atlas of Indian Tribes.

Bhobate SK, Kabinwar N, Sonule SS, **1983**. Incidence of sickle cell disease in Chandrapur area. Indian J Med Sci, 37(11): 201-203.

Census of India, **2009**. Office of Registar General and Census Commissioner, Ministry of Home Affairs, Govt of India. Available from: http://www.censusindia.gov.in. Accessed on March 27, 2015.

Colah R, Mukherjee M, Ghosh K, **2014**. Sickle cell disease in India. Curr Opin Hematol, 21: 215–223.

Colah R, Mukherjee MB, Snehal M, Ghosh K, **2015**. Sickle cell disease in tribal populations in India. Indian J Med Res, 141: 509-515.

Dunlop KJ, **Mazumber** UK, **1952**. The occurrence of sickle cell anemia among a group of tea garden labourers in Upper Assam. Indian Med Gaz, 87: 387–391.

Ghai OP, **2000**. Essential Paediatrics. New Delhi, Interprint, 5th Edn., 100.

Hankins JS, **Wang** WC, **2014**. Sickle Cell Anemia and Other Sickling syndromes. In Wintrobe Clinical Hematology, Ed John P Greer, Alan F List, George M Rodgers et al, thirteenth edition. Lippincott Williams and Wilkins. pp: 823-862.

Jain M, Das K., Padmanabham PBSV, Dhar P, Rao VR, **2003**. A1A2BO, Rh (D) blood groups and haemoglobinopathies among neo-Buddhist (Mahar) of Nagpur City. Man in India, 84(1-2): 77-83.

Jawahirani A, Mamtani M, Das K, Rughwani V, Kulkarni H, **2007**. Prevalence of *β*-Thalassaemia in subcastes of Indian Sindhi - Results from a two phase survey. Public Health, 121(3): 193-198.

Kar BC, **1983**. Sickle cell anaemia in tribes of Orissa. JAPI, 31(5): 321-325.

Kate SL, **Lingojwar** DP, **2002**. Epidemiology of sickle cell disorder in the state of Maharashtra. Indian J Hum Genet, 3: 161–167.

Kaur M, Dangi CBS, Singh M, Singh H, Kapoor H, **2013**. Burden of sickle cell disease among tribes of India: A burning problem. Int Res J Pharm App Sci, 3: 60–80.

Kaur M, Das GP, Verma IC, **1997**. Sickle cell trait and disease among tribal communities in Orissa, Madhya Pradesh and Kerala. Indian J Med Res, 55: 104–109.

Lehman H, **Cutbush** M, **1952**. Sickle cell trait in southern India. Brit Med J, 1: 404–405.

Odièvre MH, Verger E, Silva-Pinto AC, Elion J, **2011**. Pathophysiological insights in sickle cell disease. Indian J Med Res, 134: 532–537.

Patra PK, Chauhan VS, Khodiar PK, Dalla AR, Serjeant GR, **2011**. Screening for the sickle cell gene in Chhattisgarh state, India: an approach to a major public health problem. J Commun Genet, 2: 147–151.

Rao VR, **1988**. Genetics and epidemiology of sickle cell anemia in India. Indian J Med Sci, 42: 218–222.

Rao VR, **1998**. Variation of HbS frequency in Indian population role of P.Falciparum and other factors. Indian J Hum Genet, 4(1): 23-31.

Russel RV, **Lal** H, **1916**. Tribes and Castes of the Central Provinces of India. Vol. 4, McMillon Company, London.

Shukla RM, Solanki BR, Parande AS, **1958**. Sickle cell disease in India. Blood J, 13: 552-558.

Urade BP, **2012**. Incidence of sickle cell anemia and thalassemia in Central India. Open J Blood Dis, 2: 71–80.

Wild B, **Bain** BJ, **2006**. Investigations of abnormal hemoglobins and thalassaemia. In Dacie & Lewis – Practical Haematology, Ed. Barbara J Bain, Imelda Bates, Sheena Blackmore et al. 10th edition. Churchill Livingstone Edinburgh. pp: 278-292

Zade VS, Chede S, Thakare VG, Warghat NW, **2011**. The prevalence of sickle cell disease phenotypes and sickle cell gene frequency in some tribals of Melghat forest region of Amravati, Maharashtra

(India). Biosci Biotech Res Comm, 4(1): 70-73.

Citation: Rathod KB, Tijare J, 2017. Ethnicity of sickle cell anaemia - Study at Tertiary Care Hospital of Yavatmal. Int Res J Med Med Sci, 5(4): 64-68.