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Original Research Article

Study of prevalence of sickle cell disorders in and around Yavatmal district

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Abstract

Sickle cell disease (SCD) is the most common inherited disorder of haemoglobin worldwide, predominantly affecting the tribal peoples of central India. The present study was carried out to find out prevalence of sickle cell disordersin and around Yavatmal district. A total of 67,930 individuals were screened under the sickle cell disease control program held on Jan 2004 to Dec 2017 by SVNGMC Yavatmal. Screenings of samples for the presence of the sickle gene was done by solubility test and confirmation was done by cellulose acetate membrane electrophoresis and HPLC method. Among the total samples were screened, 3120 (4.59%) were found heterozygous (Hb AS) and 1725 (2.53%) were homozygous (Hb SS) for SCD, 450 (0.66%) cases were found with other Hb. The prevalence of SCD was more common in the age group of 0 to 20 years with female predominance. The caste wise distribution of solubility test confirmed samples were SC-51.95%, ST-20.21%, NT -16.2%, OBC-8.8%, SBC-1.84% and Open 1.0%. In conclusion, a sickle cell disorder was most prevalent in tribals of Yavatmal district withhighly predominant in backward classes especially in schedule caste (SC).

Keywords: Sickle cell disease, Haemoglobin, Prevalence, Electrophoresis, HPLC, Heterozygous.

1. Introduction

Red blood cells of adult healthy human individual consist of mixture of three unique respiratory proteins known as hemoglobins. The alteration of sequence of amino acids in any of the four globin chains is termed as abnormal hemoglobin. A well-known example of abnormal hemoglobin is sickle cell hemoglobin (Hb S) in which 6th amino acid (i.e. glutamic acid) is replaced by valine. (Hb S = α2β2 6-Glut-Val) [1]. Sickle cell anemia (SCA) is a recessive genetic disorder that affects the proper functioning of red blood cells. The abnormal hemoglobin is less soluble than normal hemoglobin HbA and therefore tends to crystallize out, resulting in deformation of cell which instead of being round become sickles shaped. The sickle shape red blood cells are oxygen deficient [2], confirmed the intimate relationship between the sickling of red blood cell and a reduced supply of oxygen.

Prior to 1952, no information was available about existence of Sickle Cell Hemoglobin in India. In 1952 it was recorded for the first time simultaneously amongst tribal population groups of Nilgiri Hills and laborers in the tea gardens of Assam [3,4]. Now it is firmly established that this gene harbor amongst different caste groups but very high prevalence amongst Scheduled Caste (SC), Scheduled Tribe (ST) and Other Backward Communities (OBC) [5-13].Taking into our huge population size, more than 50 % of the world's sickle cell anaemia cases are in India. It is estimated that most of the cases are in the Central and South India. During last 50 years, because of simple, reliable and inexpensive laboratory methods are available [14], the large number of population genetic surveys conducted by different scientific groups and data on geographical distribution, clinical manifestation along with its variations, available from the state of Maharashtra [5-13].

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 [15].

Hence the preset study was carried out in the Yavatmal district of Vidarbh region, located in Satpuraranges in North. About sixteen talukas comes under the Yavatmal district. This study was done on 67,930 volunteered subjects carrying the sickle cell gene.

2. Materials and Methods

In the present study, screening of large-scale population was done for the sickle cell gene in different areas of Yavatmal District during the period of Jan 2004 to December 2017. A total of 67,930 blood samples from individuals belonging to different tribal caste were collected by organizing screening camps in co-ordination with the officials from Primary Health Centers. Few drops of blood were collected by bold finger prick for performing the solubility test [16] for preliminary diagnosis of SCD. The positive samples were subjected for cellulose acetate membrane electrophoresis (4903 subjects) at pH 8.8 to confirm the diagnosis and classify Hb SS and Hb AS pattern. Samples were collected into tubes containing dipotassium EDTA (vacutainers). The samples were run on cell counter to obtain hemoglobin value and red cell indices. The same sample was used for HPLC.

A total of 6039 samples were analyzed on HPLC. After collection, the samples were stored at 2-80c and tested within a week of collection. The samples were run on an instrument manufactured by BIO RAD laboratories. The instrument is known as BI0-RAD variant (Beta Thalassemia short program) utilizes the principle of high performance liquid chromatography. An HbA2F calibrator and two levels of controls (BIO-RAD) were analyzed at the beginning of each run. The total area acceptable was between one to three million.

3. Observations and Results

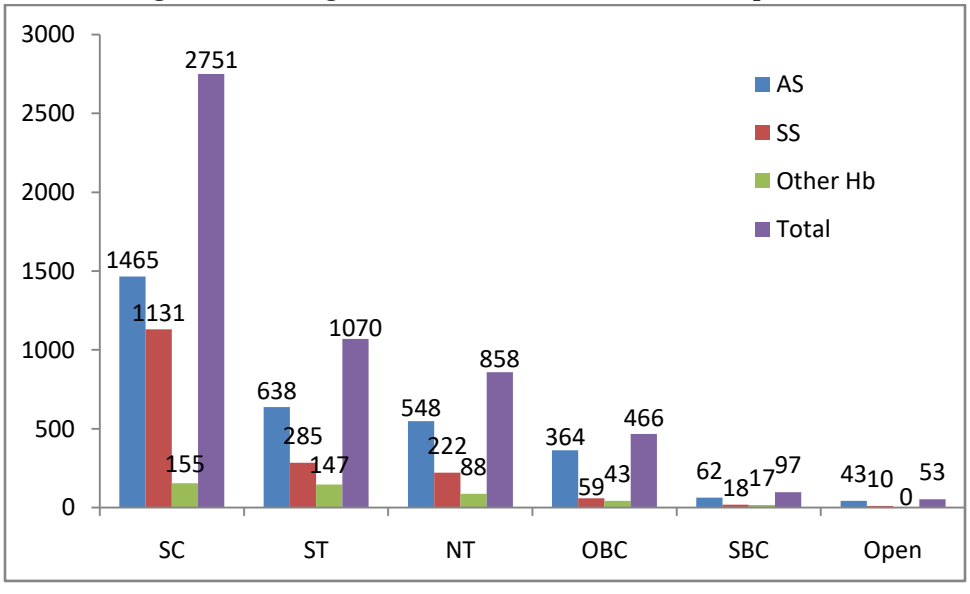
During the study period, a total of 67,930 subjects were screened for determining the prevalence of sickle cell disorders in and around Yavatmal district. A total of 5,295 (7.79%) individuals were found to be positive for sickle cell disorder, of which 3,120 (4.59%) with heterozygous genotype AS (Carrier) and 1,725(2.53%) with homozygous genotype SS (Disease) as well as 450 (0.66%) cases were found with other Hb. The sex wise distribution of sickle cell patients was shown in table 1. Out of total positive cases majority of subjects i.e. 2,909(54.93%) were females and 2,386 (45.06%) were males.

Table 1: Gender wise distribution of sickle cell patients

Sickle Cell Genotype	Sex Distribution		Total
	Male	Female	
AS	1,281 (41.05%)	1,839 (58.94%)	3,120 (4.59%)
SS	636 (36.86%)	1089 (63.13%)	1,725 (2.53%)
Other Hb	221 (49.11%)	229 (50.88%)	450 (0.66%)

The prevalence of SCD was more common in the age group of 0 to 20 years (56.46%) followed by 21 to 40 years age group (38.59%) and then 41 and above year age group (4.95%). The caste wise distribution of solubility test confirmed samples were SC-51.95%, ST-20.21%, NT - 16.2%, OBC-8.8%, SBC-1.84% and Open 1.0%, (Figure 1).

Figure 1: Showing caste wise distribution of Sickle cell patients



4. Discussion

In the present study, solubility test was used as a screening test detecting 5,295 subjects with the sickle cell trait. The confirmation of SCD was done by cellulose acetate membrane electrophoresis and HPLC method. Capillary Electrophoresis results distinguished the individuals into AS, SS and other Hb individuals. In each age group and in each caste group the number of AS individuals was quite higher than the SS individuals; this result was compared with study done by Deore *et al* [17]. The prevalence of sickle cell trait was found to be 7.79 % in our study; this prevalence was similar to Doshi *et al* [18] study, in which they found prevalence of SCD was 7.86%.

As regards to sex distribution of the disorder, Wintrobe states that sickle cell trait is more common in females. Similarly, in the current study number of affected females was much more than males. Also in previous studies [19,20**]** affected female was more than males but Deshmukh *et al* did not find any such correlation [21]. The prevalence of SCD was more common in youngest age group (0 months-20 years) had 29.04% of SS and 26.28% of AS individuals. However, the next two age groups, 21-40 years had 20.09% of SS and 20.81% of AS individual as well as 40 and above age group had 3.0% of SS and 2.9% of AS. These two age groups showed a less frequency of SCD than the 0 months-20 year's age group. In the study conducted in a population of eastern part (Vidarbha) of Maharashtra also showed similar percentage i.e. high prevalence between the age 0 and 30 years and its severity declined with increasing age [17].

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 [22]. 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%. It has been estimated that Gadchiroli, Chandrapur, Nagpur, Bhandara, Yavatmal and Nandurbar districts would have more than 5000 cases of sickle cell anemia [23].The present study has shown a very high frequency among the SC category especially Mahar (51.95%) followed by ST-20.21%, NT -16.2%, OBC-8.8%, SBC-1.84% and Open 1.0%. These findings were in agreement with the findings of other studies [15,19] where they had reported a very high frequency of HbS among the Mahar.

High prevalence of SCD is observed in the rural area from Eastern part of Maharashtra and hence population is at high risk in this area. In this rural area general practitioners have very little knowledge about this disease. Moreover, diagnostic and treatment facilities are not 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) [1]. Lack of knowledge and awareness enhances superstitions about the disease. It is necessary to establish community control programme involving people, doctors, social workers, and sympathizers. This programme will undertake diagnosis, treatment, management and counseling. Government of Maharashtra is aware of these facts but unable to undertake major projects because of financial constraint. Similarly there is need to have Central Institute to study epidemiology and clinical course aspects in detail. It needs support from Central agencies.

5. Conclusion

The present study is clearly shows that the backward communities of Yavatmal district are seriously affected by sickle cell disease. Out of six different castes, the scheduled castes was found to be most affected group in the community and next to it were ST, NT, OBC, SBC and Open in a descending order.

Tribal populations 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.

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