A Review on Pervasiveness of Sickle Cell Anemia in India with special reference to Madhya Pradesh

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ABSTRACT:

Madhya Pradesh is said to be the heart of India. It is occupied largely with rural area where agriculture is the main source of income. In these places tribal areas are also included where the sickle cell disease is prevalent. Initially these people were almost cut off from the other areas, but now the government is bringing them in to the main stream and efforts are made to solve their problems and have better life for them. One of the most common scary disease and found to be is Sickle Cell Disease (SCD) in people of these tribes. A ceaseless trial is going on for the treatment and spreading awareness among them. Survey reports are showing its prevalence but 100% cure and prevention is yet unavailable or out of reach of tribes.

Key word: *Sickle Cell Disease, prevalent. tribes, awareness*.

INTRODUCTION

Sickle cell disease (SCD) is currently the most prevalent severe monogenic condition in the world, with a high prevalence in sub-Saharan Africa, areas of the Mediterranean, India, and the Middle East. From South-Eastern Gujarat to South-Western Odisha, Central India has the highest frequency of the s allele (up to 10%). India has been rated as the nation with the second-highest total anticipated SCD births, with 42,016 babies predicted to have been born with sickle cell anaemia in 2010 (Piel et al, 2013).

In India, more than 20 million SCD sufferers reside, whereas the largest prevalence of the illness in South Asia. SCD prevalence in India has been calculated using a variety of methods, with a primary focus on those populations where the S allele is very prevalent. These methods include village-level prevalence studies and state level-wide screening schemes (Kato et al, 2018).

At the point of care, the Hb solubility test is typically used for screening, however it cannot differentiate between sickle cell trait (HbAS) and SCD, necessitating additional testing. Pilot studies of new-born screening programmes for SCD are currently being conducted in the States of Gujarat, Maharashtra, and Chhattisgarh, and they have yielded estimates of HbAS prevalence ranging from two to forty percent (Hockham et al., 2018).

Clinical testing on SCD reveals significant variability. Genetic modifiers, such as those that impact HbF level and co-inheritance of -thalassaemia, can help to explain some of this heterogeneity. The Arab-India haplotype is widespread along the eastern coast of Saudi Arabia and East Africa to India and is linked to high HbF levels (Kato et al, 2018). It typically has a milder phenotype compared to the four haplotypes linked with Africa (Benin, Bantu, Cameroon, and Senegal haplotypes), and it is more prevalent in Indian tribal tribes (Jain and Mohanty 2018). Early reports of SCD in India concentrated on these tribal communities since SCD there was first believed to be milder than in other nations (Hockham et al., 2018; Jain and Mohanty 2018).

Later findings, however, imply that the severity of the disease can be likened to that in African nations. In a study conducted in Nagpur, 833 kids, including newly born SCD patients, were examined for common SCD-related problems. When compared to the Cooperative Study of SCD, which was conducted in the United States between 1978 and 1988 and prospectively examined the natural history of SCD (Brousse et al 2021; Jain et al 2016), it is clear that symptoms like severe anaemia, acute pain, splenic sequestration, stroke, acute chest syndrome, and meningitis were more common (Jain and Mohanty, 2018). Lehman and Cutbush (1952) in the tribal communities of the Nilgiri Hills in south India are given credit for being the first to describe sickle haemoglobin in India. The same year, Dunlop and Mazumder reported the occurrence of sickle haemoglobin in the migrant labourers from

tribal groups in Bihar and Odisha who worked in the Upper Assam tea gardens (Dunlop and Mazumber,1952). Since then, other demographic groups have undergone screening, and it has been discovered that three socioeconomically disadvantaged ethnic groups—the scheduled tribes, scheduled castes, and other backward classes in India—are more likely to carry the sickle cell gene (Bhatia and Rao,1987; Rao 1988, Kaur 1997; Kate, Lingojwar 2002, Patra, 2011; Urade, 2012; Kaur et al 2013; Colah et al 2014.

Anaemia is a symptom of sickle cell disease (SCD), an illness that alters the structure of haemoglobin. It prevents a person's natural growth, produces excruciating pain, severe chest syndrome, high temperature, and the patient exhibits recurrent swelling in various body regions. Due to a genetic mutation or change that is passed from an infected parent to a kid, it is a hereditary disease. It is primarily reported among the central and southern Indian tribal people (Yadav et al. 2016) Additionally, in Madhya Pradesh, sickle cell disease affects 27 of the 45 districts of the state, and the prevalence of sickle haemoglobin (HbS) varies from 10% to 33%.

PREVALENCE IN MADHYA PRADESH AND OTHER REGIONS OF INDIA

Around 75% of the current instances of sickle cell anaemia are found in four tribal districts in the Madhya Pradesh state, specifically Alirajpur, Anuppur, Chhindwara, and Dindori, according to the annual report for 2020–2021 (https://tribal.nic.in/downloads/Statistics/AnnualReport/AREnglish2021.pdf).

Children and expectant mothers have perished in these areas as a result of incorrect diagnoses, a lack of access to medical care, or miscarriages that took place there. Due to their predominance in forested areas, vector-borne diseases, particularly malaria, are very common in the tribal community. Due to parasitic diseases, both liver cells and red blood cells (RBC) are impacted by malaria. To avoid malaria, RBCs must transform from their original round shape to a sickle shape, but this shape change causes RBCs to age prematurely, before the age of 90–120. The frequency of sickle cell disease varies from 1 to 40% across various tribal groups' carriers (Bhatia and Rao 1987). The biggest number is thought to be in Madhya Pradesh, where there are 9, 61,492 sickle heterozygotes and 67,861 sickle homozygotes. Additionally, the sickle cell belt affects 27 districts of Madhya Pradesh, and the frequency of HbS ranges from 10 to 33% (Rao,1988). Additionally, it has been calculated that 13,432 pregnant women in this State (M.P.) may be at risk of giving birth to a child with sickle cell disease, with 3358 homozygote sickle births anticipated per year (Gupta, 2006). The Gonds and Bhils are the two biggest tribal communities in central India. The annual report on health and family for 2020–21 out of the 6649 sickle cell disease cases that have been reported in Madhya Pradesh as a whole, 4955 cases—including Alirajpur, Anuppur, Chhindwara, and Dindori—have been found in these districts. Sickle cell illness is not a single symptom-only condition but rather a collection of genetic blood abnormalities that reduce the oxygen-carrying capacity of haemoglobin. Red blood cells with a disc form are distorted in sickle cell disease, preventing blood from flowing through blood arteries.

People from Scheduled castes like Jharia, Mehara, and Dehariya as well as Scheduled tribes like Pradhan, Panika, and Bhilala are those who are most afflicted by sickness, according to a Times of India report (https://timesofindia.indiatimes.com/city/indore/75-of-sickle-cell-anaemia-cases-from-4-tribal-districts-of-madhya-pradesh/articleshow/87671049.cms). According to the ICMR report, Jhabua, Alirajpur, Dhar, Barwani, Khandwa, Khargone, Burhanpur, Betul, Chhindwara, Jabalpur, Mandla, Dindori, Katni, Umaria, Shahdol, and Anuppur are among the districts with a high index of sickle cell anaemia.

Under the direction of the governor of M.P., the state government had already started a State Haemoglobinopathy Mission in November of last year. Currently running in the districts of Alirajpur and Jhabua is a prototype initiative. Since that time, routine screenings of pregnant women and all children under the age of 18 have been conducted. The project was also introduced in November, although testing didn't start until February. In Alirajpur and Jhabua, about half of the 8.26 lakh people who are targeted for the pilot project's first phase are examined for sickle cell anaemia. Recently, camps for sickle cell anaemia screening were also held in the Khandwa district. Phase two of the plan, which involves screening in additional districts, will also take place. Phase two is being planned in the 14 impacted tribal regions by the responsible authority. According to data, the MP alone reports over 6500 instances of sickle cell anaemia each year (https://timesofindia.indiatimes.com/city/indore/6-tribes-in-15-dists-most-hit-with-sickle-cell-anaemia-in-state-icmr-study/articleshow/92465665.cms).

According to the National Health Mission-MP's figures, only 18725 (1.91%) of the population in Alirajpur and Jhabua collectively have sickle cell characteristics. In those two tribal districts, the numbers of people with sickle cell disease are even lower. Records suggest that just 1997 people (0.20%) who were screened in these areas (Alirajpur and Jhabua) were diagnosed with sickle cell disease.

All the eastern districts of Maharashtra, generally known as the Vidarbha region, the Satpura mountain ranges to the north, and a small portion of Marathawada are affected by the sickle gene spread. Between 0% and 35% of members of certain tribes are sickle cell carriers. The Bhils, Madias, Pawaras, Pardhans, and Otkars are among the tribal tribes having a high frequency of HbS (20-35%). According to estimates, Gadchiroli, Chandrapur, Nagpur, Bhandara, Yoetmal, and Nandurbar districts have more than 5000 cases of sickle cell anaemia Kate and Lingojwar 2002).

The whole 1,25,000-person tribal community in Kerala's Wayanad district was screened, and all affected individuals received genetic counselling with the recommendation that they avoid getting married to other HbS carriers (Verma, 2004). These tribes exhibit a relatively high incidence of HbS (18.2 to 34.1%) (Feroze and Aravindan, 2001).Gujarat, including the Dhodia, Dubla, Gamit, and Naika tribes, has a relatively high frequency of HbS (13–31%) (Bhatia and Rao 1987). More recently, the Gujarat State Branch of the Indian Red Cross Society conducted very thorough demographic surveys, screening 1,68,498 tribal people from 22 districts. The prevalence of sickle cell carriers was found to be 11.37 percent overall (Patel et al. 2012). The tribal Chaudry families Gamit, Rohit, Vasava, and Kukana have both demonstrated a high incidence of HbS (6.3 to 22.7%) in south Gujarat. Additionally, they have a 6.3 to 13.6% -thalassaemia trait (Patel et al. 2012). These two genes appear to be shared by these ethnic groups.

The screening of 15200 people from 14 primitive tribes in Maharashtra, Gujarat, Tamil Nadu, and Odisha was done as part of a comprehensive multicenter study (Mohanty et al 2015). The screening revealed that the allele frequencies for HbS ranged from 0.011 to 0.120 and those for -thalassemia ranged from 0.005 to 0.024. Iron deficiency is a typical illness sign that was observed in 26.2% of sickle heterozygotes as well as in 67.7% of sickle homozygotes in this study (Mohanty et al 2008).

The world's highest concentration of tribal peoples is found in India. They are thought to be the original occupants of the country and the first people to settle there. The tribal community makes up 8.6% of India's overall population, or around 67.8 million people, according to the Census of India 2011. The scheduled tribe population makes up about 83 percent of all tribes in some Indian states, including Madhya Pradesh, Maharashtra, Odisha, Gujarat, Rajasthan, Jharkhand, Chhattisgarh, Andhra Pradesh, West Bengal, and Karnataka. The majority of these tribal groups reside in rural areas. There are 461 scheduled tribes in India as a whole, and each one has its own distinct cultural characteristics (Singh,1992). The subcontinent as a whole had a period of enormous intermarriage, shuffling its population's genetic deck so thoroughly that it left clears traces even in the genomes of today's most remote tribes (Reich, 2009).

If these socially disadvantaged patients have access to the proper screening and diagnostic tools, they will be able to get the proper management, treatment, and counselling tools, and lead normal lives with their families. The genetic counselling card can be made available for early detection and prompt action, which will be essential in the fight against the disease. If the partners had matching cards before getting married, it will be clear how likely it is that their offspring will contract the illness. Additionally, a sickle cell couple will be able to predict whether their subsequent child would also have the illness. It will be crucial for family planning and raising awareness among the tribes. Depending on the first outcomes of the intervention, a number of important stakeholders can also participate in awareness campaigns at the state level and offer suggestions for improvements to the mission. This initiative by the Honourable Prime Minister and the Madhya Pradesh Government will be of utmost importance because tribal health in Madhya Pradesh has suffered due to SCDs in the tribes that have not yet been affected, and intervention programmes like this mission will gain a thorough understanding of the disease and the function of various interventions in disease prevention (Bijlwan, 2022). Hydroxyurea is the main SCD medication recommended to patients, but it has a number of negative side effects, including a higher risk of infection and tumour growth. According to research, this medication is unsuccessful in one-third of SCD patients. Therefore, it is essential to create new medications with minimal to no adverse effects for the effective treatment of sickle cell anaemia (SCA). The distribution of HbS in various ethnic groups from several States has been summarised by Kaur et al. in their study (Kaur et al, 2013). There are still a lot of gaps in our knowledge regarding the distribution of the HbS gene in tribal societies, despite the fact that a sizable number of tribal groups have been taken in India for HbS screening (Colah, 2015). Records indicate that the original goal for the project was to examine 8.26 lakh people from 12 blocks in two districts in six months (https://timesofindia.indiatimes.com/city/indore/2-of-9-75l-screened-tribal-people-have-sickle-cell-trait/articleshow/95463268.cms.).

CONCLUSION

In Madhya Pradesh and other regions of India, there are a few distinct, castes and geographic areas where sickle cell anaemia is more common. These are primarily tribes from the interior or the countryside. Before a decade, these people were notably uneducated and ignorant of the sickness, but now the government is implementing numerous experimental initiatives to reduce the occurrence of disease. A number of social organisations, non-governmental organisations (NGOs), commercial financing organisations, and media are collaborating with the government in this approach. To completely eradicate the disease, however, further efforts are needed for adequate screening, diagnosis, and awareness programmes.

FUTURE SCOPE

Being a genetic disorder, the effect of Sickle cell disease is mainly on tribes, who are unaware of its lethality. Although there are many studies on causes symptoms and treatment of the disease, there is still gap of awareness, treatment and studies to eradicate the disease.

Conflict of interest: None

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