

# Sickle Cell Disease and Superstitions: An Interesting Study Report

Swarnali D.P

Shri Shankaracharya group of Institutions, Faculty of Pharmaceutical Sciences, Bhilai, Chhattisgarh, India.

## ABSTRACT

Submitted: 04-07-2013

Accepted: 03-10-2013

This study focuses on four interesting cases of sickle cell anaemia in rural areas of Chhattisgarh (CG) state. CG is a young state of growing central India with highest resources of minerals, iron, herbs and rice cereals. These patients were identified during a sickle cell disease management programme conducted by Faculty of Pharmaceutical sciences, Shri Shankaracharya Group of Institutions, Bhilai, CG. The study reveals all the patients were from poor backward communities. Symptoms related to malnutrition and infections were mostly intermingled with anaemia but all of them were least interested to follow any treatment. They were following some 'jaributi' from years which had worsened their conditions. Their details history was taken from them and counselled them so that they understand the scope of the problem and participate in the management of the disease.

**Keywords:** Blood disorder, Chhattisgarh, India, Sickle cell anaemia, patient counselling.

## INTRODUCTION

Sickle cell anaemia is of hereditary in nature and is a result of an abnormal type of red blood cells. People with this disorder have atypical haemoglobin molecules called 'hemoglobin S', which can change the shape of normal red blood cells into a sickle, or crescent, shape.<sup>1</sup> When red blood cells sickle, they break down prematurely, which can lead to anaemia. These irregularly shaped cells get stuck in the blood vessels and are unable to transport oxygen effectively, causing pain and damage to the organs. One interesting fact about the disease is unlike normal RBC's sickle-shaped cells live only 10 to 20 days. Sickle Cell trait (Hb AS) is a healthy carrier state which does not give rise to a significant clinical presentation HaemoglobinS - Sickle Cell.<sup>2</sup>

In India several states are reporting Sickle cell disease from years.<sup>3</sup> The prevalence of sickle haemoglobin from various parts of Madhya Pradesh and Chhattisgarh varied from 15 to 30 percent.<sup>5</sup> Chhattisgarh is a young state of central India. The majority of the people in this state belong to a non-educational background.<sup>6</sup> Their existence is predominantly with agriculture and often lives in remote areas. They are not blessed with the shower of education. The population which are illiterate believe that this is a curse of god. Poor economic condition is also a rival of them. Previously various other researchers also have reported several data on this disease in different areas of this state.<sup>4,7,8</sup>

## CASE STUDY

In the course of studies of cases of chronic sickle cell anaemia here we reported details of four cases. They belonged to other backward caste (OBC) and schedule caste (SC) communities. Their clinical and haematological data's are presented in table I and II respectively. Following details were obtained from these patients:

**Case 1:** Mr. C.L. Sahu (caste OBC), student, suffering from this disease for more than 10 years. He belongs to a family of four members in which his mother and sister is a chronic sufferer of this disease. His father is not a patient neither a sickle trait. Mr. Sahu reported about his chronic joint pain, severe weakness and low haemoglobin count. Often he gets various other infections. He often suffers from fever and cold. The fever is malaria positive. Occasionally he undergoes with blood transfusion. He is under the treatment of some local herbal medicines.

**Case 2:** Mrs. P. Sona (caste SC), house wife of 34 years age, suffering from this disease for more than 10 years. She has four girl children but by God's grace no one is a patient of Sickle cell disease (no information about sickle trait). Her husband is not a patient neither a sickle trait. Mrs Sona is suffering from weakness, vertigo, swelling in legs and hands, low haemoglobin count. She often suffers from malaria and other infections. She used to take folic acid tablet and "jharfuk" when the condition is worse.

**Case 3:** Ms. I. Sona (caste SC) of 18 years old is a victim of this disease. She is suffering from whole body swelling, extreme low haemoglobin level and severe viral infections. Relapsing fever is a frequent trouble she faces. She was a school going student but the inferior health condition forced her to leave school. Her mother is also a patient of this disease

### Address for Correspondence:

Swarnali DasPaul, Assistant Professor, SSIPS, Junwani, Bhilai, C.G., India.

E-mail: swarnali4u@rediffmail.com

but father is safe. She often got blood transfusion and other emergency treatment. Now she is following ayurvedic treatment but no significant changes in her condition is reported.

**Case 4:** Mrs. B. Sahu (caste OBC), house wife, reported her sickness for more than 10 years. She has severe anaemia along with joint pain and weakness. She never followed any treatment except “jaributi”. She believes this disease is a curse.

## DISCUSSION

The cases which we discover were belong to Teli and Dom communities. Dom is a social group also called as 'chandala' scattered across India. Its presumed root, *dom*, which is connected with drumming, is linked to damara and damaru.<sup>9</sup> The Dom community is traditionally an occupational caste. Their main occupation is making a variety of baskets and sells them. They are Sudra and known as Achhut (Untouchable). Teli is a caste traditionally occupied in the pressing of oil in India, Nepal and Pakistan. The word Teli comes from Tel, which means oil in Marathi, Hindi, and Oriya languages.<sup>10</sup> All these castes are very backward with their education and believe till date. Their community is used to practice endogamy and consanguineous marriages. This is a prime cause of spreading the disease because if one parent has sickle-cell anaemia (SS) and the other has sickle-cell trait then there is a 50% chance of a child's having sickle-cell disease and a 50% chance of a child's having sickle-cell trait. When both parents have sickle-cell trait a child has a 25% chance of sickle-cell disease.

In all these cases there was a history of antecedent anaemia, jaundice, and episodes of bone, joint and abdominal pains characteristic of crises. Their features showed facies with prominent frontal bosses and cheek bones. Fever was a distinctive feature in all these patients. Malaria is endemic in these areas but it was ruled out by repeated peripheral blood examination for the presence of parasites. In the case of sickle cell anaemia it has been postulated that malarial infection precipitates crisis.<sup>11</sup> The previous history of fever, bouts of crises and low haemoglobin count may be due to antecedent malarial infection which was difficult to rule out except at the time of investigation of the patients.

The interesting point was after having all these serious manifestations they were indifferent. They love to believe in jaributi, jharfuk and similar superstitions. They think the disease is spreading due to God's revenge. Even after getting

counselling about the disease management programme two of them (Mrs. B. Sahu and Mrs. P. Sona) were less interested about pre-marriage blood test and pre-natal diagnosis. But the young group was rational and they were encouraged by this management programme.

## CONCLUSION

It must be emphasized that reports of sickle cell anaemia in India are rare and in Chhattisgarh is few. This is because here has been a lack of awareness of the disease entity in these regions, together with the difficulty in making a rapid and certain diagnosis. Mass awareness programme and patient counselling are needed to manage the disease in micro level. The infra-structural facilities and technical knowhow for diagnosis of the disorder and its clinical management should be generated at district hospital level depending upon the disease load.

## ACKNOWLEDGMENT

The author acknowledges the help of Shri Shanakaracharya Group of Institutions, FPS, Bhilai. The author has no conflict of interest.

## REFERENCES

1. Ashley-Koch A, Yang Q, Olney RS. Sickle hemoglobin (HbS) allele and sickle cell disease: a HuGE review. *Am J Epidemiol* 2000; 151: 839-45.
2. Schnog JB, Duits AJ, Muskiet FA, Cate H, Rojer RA, Brandjes DP. Sickle cell disease; a general overview. *Neth J Med* 2004; 62: 364-74.
3. Awasthy N, Aggarwal KC, Goyal PC, Prasad MS, Saluja S, Sharma M. Sickle cell disease: Experience of a tertiary care center in a nonendemic area. *Annals of Trop Med and Pub Health* 2008; 1:1-4.
4. Bhatia HM, Rao VR. Genetic Atlas of the Indian Tribes, Published by Institute of Immunohaematology, (ICMR), Bombay, India. 1987.
5. Patel DK. Epidemiology & Clinical aspects of Sickle Cell Disease in India, Sickle Cell Research Project, V.S.S. Medical College, Burla, India.
6. Patra PK, Chauhan V S, Khodiar PK, Dalla AR, Serjeant GR. Screening for the sickle cell gene in Chhattisgarh state, India: an approach to a major public health problem. *J Community Genet* 2011; 2: 147-51.
7. Gupta RB. Sickle Cell Disease in Central India - Need for Micro Level Planning, Proceeding of National Symposium on Tribal Health, 110-5.
8. Shukla RN, Solanki BR, Parande AS. Sickle Cell Disease in India. *Blood* 1958; 13: 552-8.
9. Chetty K. Caste and Religions of Natal Immigrants 1860. Available from: <http://www.britannica.com/EBchecked/topic/168403/Dom>.
11. Whitby LEH, Britton CJC. Disorders of the Blood. 7th ed., J. & A. Churchill Ltd.; 1953. p.689.