SHORT COMMUNICATION

Sickle Cell Anemia in Garasia Tribals of Rajasthan

SANJAY MANDOT, VINAY LAXMI KHURANA AND JITYENDRA KUMAR SONESH

From Department of Pediatrics, J Watumull Global Hospital and Research Centre, Mount Abu, Rajasthan, India.

Correspondence to:
Dr Sanjay Mandot,
Consultant Pediatrician,
J Watumull Global Hospital and Research Centre,
Mount Abu 307501, Rajasthan, India.
E-mail : drmandot@yahoo.com
Manuscript received: January 7, 2007; Initial review completed: February 20, 2008; Revision accepted: April 26, 2008.

Our objective was to document the prevalence of sickle cell anemia among scheduled tribe (Garasia) of Sirohi district in Rajasthan state and study the clinical and hematological profile of the patients with sickle cell disease (Hb SS). In this prospective cross-sectional study, 1676 Garasia tribals attending the hospital or the mobile clinic were screened for sickle cell anemia by sickling test followed by confirmation with hemoglobin (Hb) electrophoresis. Prevalence of sickle cell anemia was found to be 9.2% (155/1676) of which 0.8% (14/1676) were homozygous (disease, Hb SS) whereas 8.4% were heterozygous (carrier, Hb AS). Common presentations of sickle cell disease were anemia, pain, recurrent infection and splenomegaly.

Keywords: *India, Prevalence, Sickle cell disease, Sickle cell trait, Sirohi.*

Nearly 20 million people suffer from sickle cell anemia in India. The sickle cell gene in India was first described among tribal groups in South India(1) but is now recognized to be widespread, especially in Central India, where the prevalence in different castes and communities varies between 9.4-22.2%(2).

Our hospital caters to tribal population (Garasia tribe) of Southern Rajasthan (Sirohi district). According to 2001 census, population of Sirohi district was 0.8 million of which 0.2 million were scheduled tribes. We conducted this study to assess the prevalence and pattern of sickle cell disease in this community.

METHODS

The study was conducted as a cross sectional survey in a hospital setting. All tribal patients who attended the OPD and mobile clinic of J Watumull Global Hospital and Research Centre, Mount Abu between December 2006 to September 2007 were screened for sickle cell anemia by sickling test (with freshly prepared sodium metabisulphite). Details were also recorded including age, sex, clinical and laboratory parameters, and morbidity. A total of 1676 subjects were studied. These included 1168 cases from pediatric age group (≤15 years) and 508 cases from the adult age group (>15 years). Of the total 1168 (69.6%) from pediatric age group, 428 (25.5%) cases were from 0-5 yr age group, 550 (32.8%) cases were from 5-10 yr age group and 190 (11.3%) cases were from 10-15 yr age group. Male:female ratio in this group was 1.3:1. Of the total 508 (30.3%) cases from adult age group, 210 (12.5%) cases were in 15-20 yr age group, 174 (10.3%) in 20-25 yr age group and 124 (7.3%) in >25 yr age group. Male:female ratio in this group was 1.1:1. The blood sample was collected by finger prick. Sickling positive patients were tested by haemoglobin electrophoresis (on cellulose acetate membrane) to confirm the diagnosis and classify them as Hb SS (sickle cell disease) and Hb AS (carrier). Privacy and confidentiality of patients was ensured to protect them from stigmatization or discrimination.

WHAT THIS STUDY ADDS?

- The prevalence of sickle cell anemia was 9.2 % in tribal (Garasia) community of Sirohi district, Rajasthan in this hospital based study.

RESULTS

Out of 1676 patients, 155 were found to be sickling positive (9.2%). Of these 0.83% (14) had homozygous state (Hb SS) whereas 8.4% (144) had heterozygous state (carrier, Hb AS) diagnosed by hemoglobin electrophoresis. Sickle cell anemia was more common in males, the male: female ratio being 3.6:1 in sickle cell disease patients and 1.38:1 in carriers (Hb AS).

Main complaints included recurrent fever 11 (78%), generalized weakness 11 (78%), musculoskeletal pain 9 (64%), abdominal pain 5 (35%) and chest pain 1 (7%). Pallor was the commonest clinical sign observed in 13 (92%) patients followed by splenomegaly in 10 (71%) patients. Massive splenomegaly (>9 cm) was seen in 3 patients. Jaundice was observed in 5 (35%) patients and gall stones in 1 patient. 2 patients presented with acute infection (pneumonia). Severe anemia (<7 g/dL) was observed in 7 (50%) patients. 12 sickle cell disease patients required admission in hospital because of painful crisis, respiratory infections and severe anemia. Specific features like sequestration crisis, hyperhemolytic crisis, aplastic crisis, priapism, epistaxis etc. were not observed in any of the patients.

DISCUSSION

Sickle cell disease in this tribal area poses difficulty in diagnosis and management, as the sign and symptoms of this disease overlap with other common diseases. Recurrent attacks of musculoskeletal pain, anemia, frequent respiratory infections, jaundice and splenomegaly are the typical features which should arouse suspicion of sickle cell disease.

The prevalence of sickle cell anemia in this study (9.2%) is similar to that reported from Orissa and Maharashtra i.e. between 5.5 to 16.5% (2-8). We acknowledge that a community based study is ideal to know the true prevalence of the disease but because of ethical/social issues involved, it is difficult to conduct a community based study involving invasive procedure (blood collection), especially in pediatric age group. However, this hospital based survery is important for sensitization and can serve as a baseline for generating more data.

ACKNOWLEDGMENTS

Dr Partap Midha, Medical Superintendent and trustee, J Watumull Global Hospital and Research Centre, Mount Abu for providing material support, guidance and encouragement.

Contributors: SM: Concept, design, analysis and writing work. VLK: Concept, design and analysis. JKS: Field work, collection of blood samples, laboratory work, acquisition of data and follow-up.

Funding: J Watumull Global Hospital and Research Centre and Indian Council of Medical Research.

Competing interests: None stated.

REFERENCES

1. Lehmann H, Cutbush M. Sickle cell trait in Southern India. BMJ 1952; 1: 404-405.
2. Shukla RM, Solanki BR. Sickle cell trait in Central India. Lancet 1985; 1: 297-298.
3. Kar BC. Sickle cell disease in Orissa. J Assoc Phys India 1991; 39: 954-960.
4. Kamble M, Chaturvedi P. Epidemiology of sickle cell disease in rural hospital of Central India. Indian Pediatr 2000; 37: 391-396.
5. Sahu T, Sahani NC, Das S, Sahu SK. Sickle cell anemia in tribal children of Gajapati district in South Orissa. Indian Comm Med 2003; 28:180-183.
6. Kar BC, Devi S. Clinical profile of sickle cell disease in Orissa. Indian J Pediatr 1997; 64: 73-77.
7. Kar BE, Satapathy RK, Kulozik AE, Kulozik M, Sergeant BE. Sickle cell disease in Orissa state, India. Lancet 1986; 2: 1198-1201.
8. Balgir RS, Mishra RK. Clinical and hemoglobinopathies in two tribal communities of Sundergarh district in Orissa, India. Int J Hum Genet 2003; 3: 209-216.