Early 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 the 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.

**Keywords:** India, Prevalence, Sickle cell disease, Sickle cell trait, Sirohi.
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 survey is important for sensitization and can serve as a baseline for generating more data.

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