Prevalence of Glucose-6-Phosphate Dehydrogenase Deficiency among Sickle Cell Patients of Chhattisgarh Region

Mohd Jafar Memon¹, Renuka Gahine², Nighat Hussain³, Yashita Gupta⁴

¹Assistant Professor, Department of Pathology, Pt. JNM Medical College, Raipur, Chhattisgarh, India, ²Professor and Head, Department of Pathology, Government Medical College, Rajnandgaon, Chhattisgarh, India, ³Associate Professor, Department of Pathology & Lab Medicine, AIIMS, Raipur, Chhattisgarh, India, ⁴Senior Resident, Department of Pathology & Lab Medicine, AIIMS, Raipur, Chhattisgarh, India

Abstract

Background: About 50% of world population of sickle cell disease (SCD) is found in India, predominant among the tribal population of central India. Sickle cell (SC) hemoglobinopathy and glucose-6-phosphate dehydrogenase (G6PD) enzyme deficiency are important genetic and public health problems in central-eastern part of India. This study aims to determine the prevalence of G6PD deficiency in SCD patients.

Materials and Methods: The study was carried out prospectively over a period of 1 year from September 2010 to August 2011 in the Department of Pathology at our institute. The material for the present study consisted of 100 cases including 76 patients with SCD and 24 of control group. Solubility test for sickling followed by hemoglobin (Hb) electrophoresis and qualitative dye decolorization test for G6PD deficiency were performed on all the samples.

Results: A total of 100 patients were studied, 76 were of SCD including 38 patients each of SC trait (AS) and SC anemia (SS) and remaining 24 patients were of the control group. The majority of AS patients were females (69.4%) while the SS and control patients had almost equal number of males and females. The prevalence of G6PD deficiency was observed to be 5%, predominantly in males i.e. 4 (9.1%) with only one female (1.79%). The prevalence of G6PD deficiency in SC patients (5.26%) was almost similar to that of control group (4.17%).

Conclusion: G6PD deficiency should be looked for in all subjects with SC anemia. Individual with HbSS is uniquely unfit to tolerate increased hemolysis and when the two problems coexist, particular care should be exercised in the administration of drugs known to initiate hemolysis in patients with G6PD deficiency.

Key words: Central India, Drugs, Hemoglobin electrophoresis, Hemolysis, Qualitative, Solubility test

INTRODUCTION

Glucose-6-phosphate dehydrogenase (G6PD) and sickle cell disease (SCD) are two inherited red blood cell (RBC) disorders which could be encountered in the same person due to the following features common to both these disorders:

1. More commonly seen in certain geographical areas and ethnic groups
2. In stable conditions, both these disorders do not alter the hemoglobin (Hb) levels, RBC count, and indices
3. Patients usually remain asymptomatic in both.

Hence, detailed clinical history and screening tests are necessary to detect these RBC disorders.¹

The interaction between SCD and G6PD has been studied in different populations. Many studies have suggested the higher incidence of G6PD deficiency in SCD patients as compared to general population. However, other studies do not confirm this association.²

Corresponding Author: Dr. Yashita Gupta, F-12, Block-II, Pt. J.N.M. Medical College, Jail Road, Raipur, Chhattisgarh, India.
Phone: +91-7389066467. E-mail: yashigupta@gmail.com
The prevalence of sickle Hb varies from 15% to 30%, affecting nearly 3 million people of Chhattisgarh. Hence, we performed this study to know the association and prevalence of G6PD in SCD patients so that to judiciously administer drugs during the general treatment or episodes of crisis.

MATERIALS AND METHODS

The present study was carried out in the Department of Pathology at our institute. The study was spread over a period of 1 year from September 2010 to August 2011.

The material for the present study consisted of 100 cases including 76 patients with SCD and 24 of control group.

With all standard aseptic precautions, blood was collected from anti-cubital vein using 21 or 22-gauge needle from each individual, after obtaining informed and written consent. Blood was delivered into ethylene diamine tetraacetic acid pilot, and solubility test for sickling was performed on all the samples, followed by Hb electrophoresis. In the entire cases, qualitative dye decolorization test for G6PD deficiency had been performed adjusting for the Hb content of the patient.

RESULTS

Of the total 100 patients studied, 76 were of SCD including 38 patients each of AS and SS. The 24 control patients selected had anemia but found to be negative for solubility test and Hb electrophoresis, of which 11 cases were of malaria, 8 cases of neonatal jaundice, 2 of β-thalassemia, and 3 cases of miscellaneous group. Of the 100 patients, 5 were found to be G6PD deficient showing the prevalence rate of 5%. In the individual group studied the prevalence was found to be 2 (5.3%), 2 (5.3%), and 1 (4.2%) in control, AS and SS patients, respectively (Table 1).

Most of the patients of AS were in age range of 11-20 years and 21-30 years, whereas most patients of SS were in between 0 and 10 years of age. Of the control patients, majority were in between 0 and 10 years. Both the patients of AS with G6PD deficiency were in age range of 11-20 years while both the SS patients in 0-10 years and the only control patient was above 40 years of age (Table 2).

Of the AS patients, females were more than males while among the SS patients equal number of males and females were present. Both the SS patients and the one control patient with G6PD deficiency were males (100%) while of the AS patients with G6PD deficiency one each (50%) was male and female (Table 3).

DISCUSSION

In the present series of 100 patients studied, the prevalence of G6PD deficiency was observed to be 5% (Table 1), predominantly in males i.e., 4 (9.1%) with only one female (0.79%) (Table 3).

Different studies conducted in the past indicate that the incidence of G6PD deficiency in India ranges from <1% to 27.94% (Table 4). The prevalence rate observed in the present study were similar to those of Swaroop et al., Kalra et al., Da Costa et al., Meera Khan, Choubisa et al., Reddy et al., and Pant et al. Discrepancies found with other may have resulted from different techniques used. The G6PD enzyme deficiency in the present study was detected using dichlorophenol indophenol dye as described by Bernstein.
In the present study, the prevalence of G6PD deficiency in sickle cell (SC) patients (5.26%) was almost similar to that of control group (4.17%). Similar findings of equal prevalence of G6PD deficiency among SC and control group was observed in studies by Heller et al.,43 Naylor and associates36 in Chicago, observed 14.3% G6PD deficiency in 56 SS males, as compared to 15% of 54 AA Afro-American males; Bouanga et al.,2 also found equal prevalence in HbSS patients (22.2%) and in HbAA (22.5%) (Table 5).

However, an increase in the prevalence of G6PD deficiency among SC patients as compared to control patients was observed in the studies by Lewis et al.,37 in Ghana; Beutler et al.,39 Bienzle.40 Piomelli et al.38 (33.3% among SC patients and 10.8% among controls); Diop et al.46 (21.6%
in SC patients and 12.3% in normal subjects); Andoka et al.\textsuperscript{41} found 11% patients to be G6PD deficient, of which 10 were of SC anemia and 2 of SC trait. Kar et al.\textsuperscript{38} screened 60 cases of malaria and observed that sickle Hb was found in 7 (11.5%) patients and G6PD deficiency in 3 (5%) cases. One patient with falciparum malaria had both SC trait and G6PD deficiency. Balgir\textsuperscript{34} observed 12 cases showing compound heterozygosity for SC hemoglobinopathy and G6PD deficiency. The author demonstrated an inverse relationship of SC allele with G6PD deficiency and beta thalassemia in a cross-section of malaria endemic (Plasmodium falciparum) tribal communities in Orissa. Balgir\textsuperscript{32} found 52.2% patients with SCD to be G6PD deficient in hemizygous/heterozygous/homozygous condition in Dhelli Kharia tribal community of Orissa (Table 6).

Variations in the results can also be due to racial, ethnic, and geographic distribution. Variable prevalence of erythrocyte G6PD enzyme deficiency can also be explained by the fact that Indian population is composed of many heterogeneous religion and caste groups.

A difference in the frequency of G6PD deficiency among SCD patients can occur by post-zygotic selection, indeed

### Table 5: Prevalence of G6PD deficiency in patients with and without sickle cell disease in various studies outside India

<table>
<thead>
<tr>
<th>Name of author</th>
<th>Year</th>
<th>Place</th>
<th>Total number patients of SCD</th>
<th>Total number of G6PD deficient patients with SCD</th>
<th>Total number of G6PD deficient patients without SCD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Naylor et al.\textsuperscript{36}</td>
<td>1960</td>
<td>Chicago</td>
<td>56</td>
<td>8</td>
<td>14.3</td>
</tr>
<tr>
<td>Lewis et al.\textsuperscript{37}</td>
<td>1966</td>
<td>Ghana</td>
<td>95</td>
<td>41</td>
<td>43.1</td>
</tr>
<tr>
<td>Pirmelli et al.\textsuperscript{38}</td>
<td>1972</td>
<td>New York</td>
<td>15</td>
<td>5</td>
<td>33.3</td>
</tr>
<tr>
<td>Beutler et al.\textsuperscript{39}</td>
<td>1974</td>
<td>Los Angeles</td>
<td>21</td>
<td>4</td>
<td>19.1</td>
</tr>
<tr>
<td>Bienzle et al.\textsuperscript{40}</td>
<td>1975</td>
<td>Nigeria</td>
<td>100</td>
<td>16</td>
<td>16</td>
</tr>
<tr>
<td>Nhonoli et al.\textsuperscript{41}</td>
<td>1978</td>
<td>Male-53</td>
<td>12</td>
<td></td>
<td>14</td>
</tr>
<tr>
<td>Gibbs et al.\textsuperscript{42}</td>
<td>1980</td>
<td>Jamaica</td>
<td>5072</td>
<td>566</td>
<td>11.16</td>
</tr>
<tr>
<td>Heller et al.\textsuperscript{43}</td>
<td>1979</td>
<td>Illinois</td>
<td>19</td>
<td>28.3</td>
<td></td>
</tr>
<tr>
<td>Anderson et al.\textsuperscript{44}</td>
<td>1988</td>
<td>Congo</td>
<td>14</td>
<td>31.8</td>
<td>128</td>
</tr>
<tr>
<td>Bouanga et al.\textsuperscript{2}</td>
<td>1998</td>
<td>AS-44</td>
<td>188</td>
<td>42</td>
<td>22.2</td>
</tr>
<tr>
<td>Mohammad et al.\textsuperscript{45}</td>
<td>1998</td>
<td>Bahrain</td>
<td>125</td>
<td>59</td>
<td>47</td>
</tr>
<tr>
<td>Diop et al.\textsuperscript{46}</td>
<td>2005</td>
<td>France</td>
<td>319</td>
<td>69</td>
<td>21.6</td>
</tr>
<tr>
<td>Nouraie et al.\textsuperscript{47}</td>
<td>2010</td>
<td>USA</td>
<td>261</td>
<td>8.7</td>
<td></td>
</tr>
</tbody>
</table>

G6PD: Glucose-6-phosphate dehydrogenase, SCD: Sickle cell disease

### Table 6: Prevalence of G6PD deficiency in patients with sickle cell disease along with controls in various studies in India

<table>
<thead>
<tr>
<th>Name of author</th>
<th>Year</th>
<th>Place</th>
<th>Total number patients of SCD</th>
<th>Total number of G6PD deficient patients with SCD</th>
<th>Total number of patients without SCD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Praharaj et al.\textsuperscript{15}</td>
<td>1977</td>
<td>Orissa</td>
<td>12</td>
<td>200</td>
<td>22</td>
</tr>
<tr>
<td>Kar et al.\textsuperscript{48}</td>
<td>1990</td>
<td>Western Orissa</td>
<td>1</td>
<td>60</td>
<td>3</td>
</tr>
<tr>
<td>Balgir\textsuperscript{34}</td>
<td>2006</td>
<td>Orissa</td>
<td>12</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Balgir\textsuperscript{49}</td>
<td>2008</td>
<td>Bhubaneswar, Orissa</td>
<td>29</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Balgir\textsuperscript{50}</td>
<td>2010</td>
<td>Sundarghar, Orissa</td>
<td>23</td>
<td>12</td>
<td>52.2</td>
</tr>
<tr>
<td>Present study</td>
<td>2012</td>
<td>Raipur, Chhattisgarh</td>
<td>76</td>
<td>4</td>
<td>5.26</td>
</tr>
</tbody>
</table>

SCD: Sickle cell disease, G6PD: Glucose-6-phosphate dehydrogenase
by postnatal selection, since the phenotypic expression of the homozygous S condition is not significant until at least a few months after birth.

CONCLUSION

The present study has been conducted in the tertiary care hospital, which may or may not represent a true cross-section of the normal population. Yet in a preliminary study of this nature, the observation of 5% could be taken to consider that the problem of G6PD deficiency exists in this region and should be of concern as the enzyme deficiency remains obscure, there being no overt clinical manifestation. It would be important to keep in mind that an intrinsic anomaly of enzyme would make these deficient subjects vulnerable to some of the common drugs, which in therapeutic doses are harmless for persons with normal enzyme; hence, the detection of this enzyme deficiency is important for protecting such individuals.

The number of patients screened in the present study is not sufficiently large to make the observation of the enzyme G6PD deficiency highly significant, and it would be proper to screen a larger and representative sample of this region to know the exact nature and magnitude of the problem.

ACKNOWLEDGMENT

I am highly obliged to my institute and all faculty members of the department especially Dr. Pratima Kujur for guiding me in this study.

REFERENCES


Memon, et al.: Prevalence of Glucose-6-Phosphate Dehydrogenase Deficiency among Sickle Cell Patients

Memon, et al.: Prevalence of Glucose-6-Phosphate Dehydrogenase Deficiency among Sickle Cell Patients


Source of Support: Nil, Conflict of Interest: None declared.