Priapism in Sickle Cell Disease in Qatif Central Hospital

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Priapism is a common complication of sickle cell disease (SCD). Its true frequency in Saudi patients is not known. We interviewed 174 male patients or parents of young SCD patients attending sickle cell clinics at our hospital, in an attempt to estimate the frequency of this complication. Of these patients 105 were children and 69 were adults. History of at least one episode of priapism was obtained from 32 patients (18.4%). This is a much higher frequency than previously reported from the Eastern Province of Saudi Arabia. The previously reported low frequency may have been due to cultural factors or to overlooking the diagnosis at presentation.

Priapism is a common complication of sickle cell disease (SCD). A prevalence of up to 42% has been reported,1 with age of onset ranging between 2 and 55 years.2 Previous reports from the Eastern Province of Saudi Arabia suggested that this complication is rare.3,4 More recently, cases in both paediatric and adult age groups have been reported.5,6 Some patients presented with other problems, and priapism was discovered incidentally.6

These observations led us to believe that the frequency of priapism may have been underestimated in our SCD patients. Therefore, this study was conducted to estimate the frequency of this complication.

Methods

Data were collected from adult male patients or parents of children with SCD attending the sickle cell clinics at Qatif Central Hospital, during the period from 15 January 1991 to 15 January 1992. An informal private interview was conducted with each candidate. The information obtained was felt to be accurate within the limitations of cultural inhibitions. The diagnosis of priapism was subjective in all patients except for the seven patients who were admitted and the diagnosis was confirmed.

The diagnosis of SCD was based on criteria described elsewhere.7 Haematologic data were based on mean steady state values of haemoglobin (Hb), mean corpuscular volume (MCV), and reticulocytes. The proportions of HbF were estimated from cellulose acetate haemoglobin electrophoretic strips at alkaline pH (Helena Laboratories).

Priapism was defined as a painful persistent penile erection unaccompanied by sexual desire.8 Priapism was categorized as major episodes (prolonged, more than 4 hours) and minor episodes (isolated or repetitive, reversible episodes with spontaneous detumescence within 4 hours).9

The paired t-test was used to compare the haematological values in patients with and without priapism.

Results

A total of 105 paediatric and 69 adult patients were interviewed. The age at the time of the interview ranged between 1 and 40 years. A history of at least one attack of priapism was obtained from 32 patients (18.4%);
Table 1
The frequency and age of onset of priapism

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>No. of patients interviewed</th>
<th>Patients with priapism (32 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-6</td>
<td>38</td>
<td>No.</td>
</tr>
<tr>
<td>7-12</td>
<td>67</td>
<td>9</td>
</tr>
<tr>
<td>13-18</td>
<td>28</td>
<td>11</td>
</tr>
<tr>
<td>19-24</td>
<td>23</td>
<td>5</td>
</tr>
<tr>
<td>25-30</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>31-40</td>
<td>9</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
</tr>
</tbody>
</table>

Table 2
Duration of priapism

<table>
<thead>
<tr>
<th>Duration of priapism (hours)</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1</td>
<td>11</td>
</tr>
<tr>
<td>1-3</td>
<td>3</td>
</tr>
<tr>
<td>4-12</td>
<td>0</td>
</tr>
<tr>
<td>13-24</td>
<td>3</td>
</tr>
<tr>
<td>&gt; 24</td>
<td>8</td>
</tr>
<tr>
<td>Unknown</td>
<td>7</td>
</tr>
</tbody>
</table>

12 patients were adults (17.4%) and 20 were children (19.1%). The median age at onset was 11 years (range, 1-30 years). The frequency and age of onset are shown in Table 1. Duration of priapism is shown in Table 2. One patient suffered from a second major attack and five patients suffered from recurrent minor episodes.

Priapism was precipitated by sexual excitement in six patients (18.8%). In three patients (9.4%), priapism occurred in association with abdominal crises. In two others (6.3%), it was associated with bone crises and in one patient (3.1%) it was associated with an acute splenic sequestration crisis. One patient had an attack of priapism two months after splenectomy. Fifteen patients (46.9%) had some relief of priapism after urination.

Seven patients (21.9%) were hospitalized. All of these, except for one, responded to conservative management. Impotence was reported by the patient who required surgery.

Comparison of haematologic parameters in patients with and without priapism is shown in Table 3. Comparison of HbF at different age groups is shown in Table 4 and no statistical significance could be found between the priapism present and priapism absent groups.

Discussion

The frequency of priapism in Qatif Central Hospital sickle cell clinic population was 18.4% in this study. This frequency is much higher than that previously reported, indicating that priapism might have been underestimated. This previously reported low frequency may be explained by the following. Firstly, our patients or their parents may dislike disclosing symptoms related to the genitalia. Secondly, patients may present with other problems such as lower abdominal pain or major splenic sequestration crisis. Thirdly, the genitalia may not be examined during the initial evaluation of the patients, although this might not disclose prior episodes.

The median age of onset of 11 years conforms with other reports that priapism is a predominantly paediatric manifestation. A total of 34% of episodes of priapism were major, and 44% were minor. Recurrence occurred mainly among patients with minor episodes. Precipitating and associated factors were found in only 13 of the 32 patients (40.6%). Sexual excitement was the main preceding factor, while urination was the main relieving factor.

Most patients who required hospitalization, presented with other problems: four patients had abdominal crises and one patient had splenic sequestration crisis. Therefore unless the genitalia are examined, priapism may escape detection, especially in children who may point to the lower abdomen rather than the genitalia, as they do in testicular torsion.

No statistically significant differences were found between patients with and without priapism in any of the haematologic parameters tested. This is in agreement with other reports from Eastern Saudi Arabia and contrasts the findings of Emond et al. who found a lower HbF in their
priapism group.\textsuperscript{1} A higher Hb concentration in our patients\textsuperscript{3} may offset the beneficial effect of higher HbF by increasing blood viscosity.

Finally, we should address the limitations of this study. Cultural inhibition may prevent the detection of such a complication by physicians, leading to the underestimation of the problem. The problem of recall, especially with the passage of time is another complicating factor. This is supported by the fact that no episode was recalled beyond 2 years. The effect of misinterpretation of the question about priapism is unknown. The limitations of this study are unlikely to be solved completely by a prospective study, since patients may not present immediately to physicians to confirm the diagnosis.

Priapism appears to be more common than previously reported in the SCD patients in the Eastern Province of Saudi Arabia. The genitalia of SCD patients should be examined, especially when they present with abdominal crises in order to detect and properly manage priapism.

References


