Taurodontism and Klinefelter’s syndrome

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SUMMARY The incidence of taurodontism in 31 patients with XXY Klinefelter’s syndrome was studied. Taurodont molars were observed in 6 of the 31 cases (19.4%), a significantly higher rate than among the controls. Though taurodontism is not an obligatory finding in Klinefelter’s syndrome, it is believed to be one of the anomalies frequently observed in connection with this condition.

Taurodontism is an anomaly of the shape of the teeth. It is found in multirooted teeth and is characterised radiologically by short roots, elongated body, and enlarged pulp chamber. The word ‘taurodontism’ was first used by Keith (1913) to describe the teeth of prehistoric man, the Neanderthals, and the Heidelberg. Anthropologically, it has been viewed with interest in connection with the evolution of man (Shaw, 1928; Hamner et al., 1964; Barker, 1976).

It is believed that taurodontism was first discussed in relation to Klinefelter’s syndrome when Law et al. (1969) reported that the 2 conditions often appear concurrently. Since then, cases of Klinefelter’s syndrome accompanied by taurodontism have been reported by several authors. These cases have included not only the standard XXY type, but also the XXXY/XXXX, XXXY, XXXXY, and XXXXY/XXXX variants (Witkop, 1971; Keeler, 1973; Stewart, 1974; Feichtinger and Rossiwall, 1977).

Taurodontism, however, is not an obligatory finding in Klinefelter’s syndrome and its incidence is, as yet, unclear. In this report, 31 patients with XXY Klinefelter’s syndrome were studied to ascertain the incidence of taurodontism in this condition.

Materials and methods

Thirty-one patients with Klinefelter’s syndrome, treated at the Infertility Clinic of the Department of Urology, Kyoto University Hospital, were studied. Their ages ranged from 21 to 42. Chromosome studies were performed on cells from peripheral blood cultures, according to the method of Moorhead et al. (1960). In each case, 30 cells were examined and a minimum of 16 cells with the modal chromosome number were karyotyped. The karyotype of all the cases was 47,XXY.

The controls consisted of 48 males and 63 females, without phenotypic anomalies, chosen from patients treated at the Oral Surgery Clinic of this hospital. The ages of the controls, both males and females, ranged from 25 to 42.

Panorex radiographs were used to measure the length of the ‘body’, as defined by Shaw (1928), as well as that of the crown of the permanent mandibular molars. A molar with a crown/body ratio of more than 0.5 was designated as a taurodont tooth. Maxillary molars were not used for this measurement because of the difficulty of obtaining clear radiographs. In all cases, the measurements were performed by one examiner (AF) who did not know whether the patient belonged to the Klinefelter’s syndrome group or to the control group.

Results

Taurodontism was observed in 6 of the 31 Klinefelter’s syndrome patients (19.4%), while it was seen in 2.1% of the control males and 4.8% of the control females, indicating a significantly higher ratio in the Klinefelter’s syndrome group (Table 1). Of the 31 Klinefelter’s syndrome patients with taurodontism, 6 were affected bilaterally (Fig.) and 4 unilaterally. Of the 9 taurodont teeth found in the 6 cases, 7 were

<table>
<thead>
<tr>
<th>Klinefelter’s syndrome</th>
<th>No. of affected cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>n = 31</td>
<td></td>
</tr>
<tr>
<td>Control males</td>
<td>1 (2-1)</td>
</tr>
<tr>
<td>n = 48</td>
<td></td>
</tr>
<tr>
<td>Control females</td>
<td>3 (4-8)</td>
</tr>
<tr>
<td>n = 63</td>
<td></td>
</tr>
</tbody>
</table>

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Table 2  Classification of 6 cases of Klinefelter’s syndrome with taurodontism

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Karyotype</th>
<th>Affected teeth</th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) ST</td>
<td>30</td>
<td>XXY</td>
<td>8 8</td>
</tr>
<tr>
<td>(2) TU</td>
<td>30</td>
<td>XXY</td>
<td>7 7</td>
</tr>
<tr>
<td>(3) KK</td>
<td>21</td>
<td>XXY</td>
<td>7</td>
</tr>
<tr>
<td>(4) SK</td>
<td>32</td>
<td>XXY</td>
<td>7</td>
</tr>
<tr>
<td>(5) TM</td>
<td>23</td>
<td>XXY</td>
<td>7</td>
</tr>
<tr>
<td>(6) NV</td>
<td>39</td>
<td>XXY</td>
<td>7</td>
</tr>
</tbody>
</table>

mandibular second molars, while the remaining 2 were mandibular third molars (Table 2). The average crown/body ratio of the taurodont teeth of the Klinefelter’s group (0.72 ± 0.04) and the controls (0.62 ± 0.03) were computed to compare the degree of taurodontism in the 2 groups. No significant difference was observed.

Pectus excavatum was noted in one Klinefelter’s syndrome patient with taurodontism (Case 4, SK), while the remaining 30 patients examined did not have any clinically or radiographically visible structural defect of the thoracic cage, the spine, or the hand.

Discussion

It has been said that racial factors are important in the incidence of taurodontism, and that this incidence is low in modern Caucasians and high in Capoids, Australoids, and Mongoloids (Witkop, 1971).

The only study on the incidence of taurodontism among the Japanese was conducted on deciduous teeth. It was reported that the incidence is 0.5% to 1-6% and that it is higher among females than males (Daito and Hieda, 1971; Hitomi et al., 1971). However, there has been no population study on permanent teeth, and no indisputable data on the incidence of taurodontism among the Japanese is available.

In 1971, on the basis of reports by Law et al. (1969), Hieda and Daito undertook a chromosomal analysis of a 4-year-old whose mandibular first deciduous molar was a taurodont tooth, and reported that in this case the karyotype was 47,XXY. Thus, the relation between Klinefelter’s syndrome and taurodontism was noted even in Japan.

Keeler (1973) reported that in 6 cases of X chromosome aneuploidy (3 XXY, 1 XXX, 1 XXXY and 1 XXXXY/XXXY), taurodont molars were observed in each. Stewart (1974) reported on 1 XXY and 1 XXXXY case, both with taurodontism, and speculated, on the basis of this and past studies, that the greater the number of X chromosomes, the greater the incidence and degree of taurodontism would be. Feichtinger and Rossiwall (1977) studied 8 individuals with X chromosome aneuploidy and found taurodontism in only 1 XXY/XXXXY case. They also reported that, among 6 XXY cases and 1 XXX case, taurodontism was not observed. Based on these results, they concluded that there is some relation between additional X chromosomes and the appearance of taurodontism, but that taurodontism is not an obligatory finding in standard XXY Klinefelter’s syndrome.

In the 31 XXY cases we studied, the incidence of taurodontism was approximately 20%, slightly less than that of gynaecomastia in patients with Klinefelter’s syndrome. Thus, we believe this is one of the anomalies frequently associated with this condition.

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References


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