TAURODONTISM AND KLINEFELTER SYNDROME: CASE REPORT AND REVIEW OF THE LITERATURE

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CASE REPORT

Taurodontism and Klinefelter syndrome: case report and review of the literature

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ABSTRACT

This paper presents a seven-year old patient with Klinefelter Syndrome with dental treatment needs. The patient had mild mental retardation, learning and speaking difficulties, taurodontic teeth, missing premolars and incisor teeth. This paper emphasizes the importance of regular dental visits with panoramic radiography for early recognition of the systemic disorder and to improve quality of patient's life.

INTRODUCTION

Klinefelter's syndrome (KS) is a disorder first described by Dr. Klinefelter et al. in 1942 that is characterized by an extra X chromosome in human cells.[1-3] The patient with KS have two X chromosomes with Y.[2,4-8] Approximately one in 1000 boys is born with an additional X chromosome and the most common type of this syndrome is 47,XXY, which represents 82% of these cases and the remaining cases show a variety of aneuploidies like 48,XXXY; 48,XXYY; 49XXXXY and mosaic patterns like 46,XY/47,XXY, 47,XXY/48XXXY.[9-11] KS is probably the most frequent cause of male hypogonadism. The presence of two X chromosomes, one carrying a small translocated fragment of Y chromosome, makes men with sex reversal syndrome very similar to men with KS. Both syndrome groups are characterized with eunicoid habitus, but men with sex reversal syndrome are shorter than men with KS.[5,6,12] The diagnosis of KS is rare because the men with KS do not differ from healthy men till puberty.[2,6,13] The characteristic clinical findings of this syndrome may vary but most of the patients have infertility, tall, slender body habitus with long legs and shorter torso, a coarse face, skeletal anomalies, hypogonadism, osteoporosis, mental retardation, aspermatogenesis, variable gynecomastia, small testis and penis. In addition to these findings, language

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delays, learning problems, behavioral difficulties and cardiovascular problems may occur.[4,6,7,11,14,15]

Oral findings of KS reported in many cases include cleft lip and palate, taurodontism, larger pulp chamber and crowns, maxillary and mandibular prognathism, congenital absence of permanent teeth, bifid uvula, shovel incisors and enamel defects.[2,9,16,17]

The most common dental finding of KS is taurodontism. Taurodontism is observed with several syndromes and anomalies including amelogenesis imperfecta, ectodermal disturbance, Down syndrome, Klinefelter syndrome, trichodento-osseous syndrome, Mohr syndrome, Wolf-Hirschhorn syndrome and Lowe syndrome.[2,8,18,20] Tauorodontism was first proposed by Sir Arthur Keith[18] in 1913. The teeth were classified as normal, hypotaurodont, mesotaurodont or hypertaurodont by Shaw in 1928[2,19]. Hypotaurodontism is the least pronounced form, in which the pulp chamber is enlarged. Mesotaurodontism is the moderate form in which the roots are divided only at the middle third. Hypertaurodontism is the most severe form in which bifurcation or trifurcation occurs near the root apices.[20] Taurodontism leads to constriction of the cemento-enamel junction, which results in vertically elongated pulp chambers, apical displacement of the pulpal floor, and bi or trifurcation of the root. This trait can be seen both in primary and permanent teeth, in single or several molars of the same quadrant and can be seen unilateral or bilateral.[12,20]

The aim of this case report was to describe a case with Klinefelter's syndrome and to discuss the oro-dental findings with current literature.

CASE REPORT

A seven-year old boy was referred to the Department of Pediatric Dentistry with a history of pain in his teeth. The patient had diminutive speech, sluggish movement. The mother of the child reported that he was premature (34 weeks) with 1680gr birth weight and had the chromosome anomaly KS, mild mental retardation, learning and speaking difficulties. He was having special education for his complaints. Mother was 25, the father was 31 year old and they have consanguinity. They were healthy, made intermarriage and had one another boy who was healthy. There was no other person in the family that had chromosome anomally. A visual examination of the dentition revealed multiple caries, rednees and swelling of the vestibulum of the left mandibulary second primary molar (75) and right first primary molar (54). The maxillary arch was larger than mandibular arch and the right mandibular lateral incisor was missing (82) (Figure 1) and attrition of maxillary and mandibulary anterior teeth was noted. In the panoramic radiograph examination (Figure 2), congenital absence of four permanent teeth; maxillary second premolar (15), mandibular left second premolar (35), mandibular right lateral incisor (42) and mandibular right second premolar was noticeable (45). Maxillary second (55) and mandibular first primary molars (74, 84) were hypertaurodont; maxillary left first primary molar (54) and mandibular right second primary molar (85) were mesotaurodont. The roots of the permanent molars are not yet formed

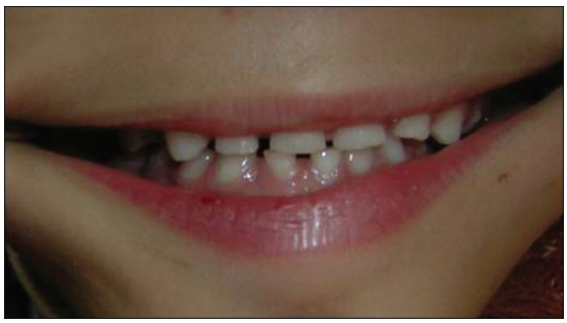

Figure 1. Large maxiller arch, attrition of the both mandibular and maxillary teeth, absence of the primary lateral can be seen.

but four permanent molars had a large pulp chamber. Because of the because of the difficulties in behavior guidance of the patient, the treatment was performed under general anesthasia. The tooth 54, 75 extracted, 64, 65 were restored with compomer and the teeth 55, 36, 46 were restored with amalgam (Figure 3).

DISCUSSION

The relationship between an extra chromosome and taurodontism has been widely described. Taurodontism is an isolated anomaly and is detectable before puberty. It can be associated with several syndromes and anomalies like KS and trisomy21 or Down syndrome.[18] The endodontic treatment of taurodontic teeth may be complicated. The shaping and obturation of the root may be difficult because of the canal orifiences.[2,18] Identification of the patients with multiple taurodontic teeth could lead to early recognisation of systemic disorder and improve quality of life. If the treatment of these teeth is delayed, the teeth could be exracted. Many of the primary teeth, particularly second primary molars, have absence of their successors as in this case and the extraction of these teeth could lead to orthodontic malformation. And also the extraction of the teeth is difficult because of the dilated apical third.[18]

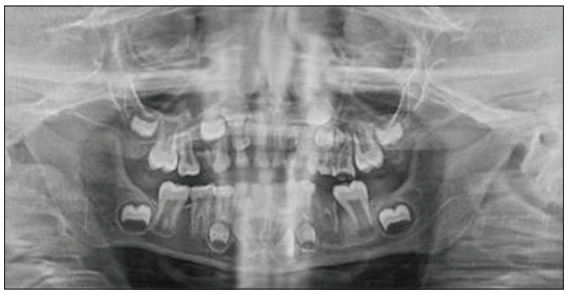

Figure 2. Panoramic view of the patient before dental treatment.

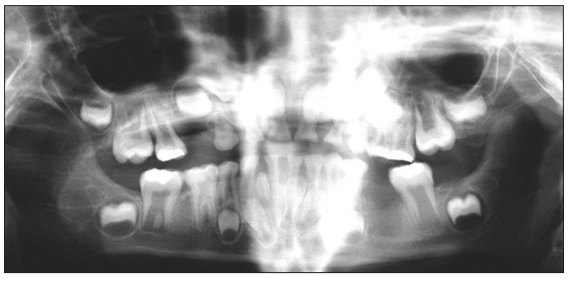

Figure 3. Panoramic view of the patient after dental treatment.

Children with KS have some dental anomalies like hypodontia. In the present case, four permanent teeth were absent. As described by some authors[13,18] that the patients with KS have mental retardation as in this patient. Most of the authors described prognatism about the jaws but only a large maxiller arch was seen in this patient.[2,5,8,10,13,14]

The patients with KS are undiagnosed because the clinical findings of the KS are often varied. It was found that nearly three quarters of estimated cases in the general population were undiagnosed.[2,9] The patients with meso- or hypertaurodontic teeth who do not have a syndrome known to be associated with taurodontic teeth should be consulted for chromosome analysis because there is a high association of taurodontic teeth with X-chromosome aneuploidy syndromes and the children may benefit from early diagnosis of KS.[2,7,8]

CONCLUSION

Although taurodontism is a dental rarity, the patient should be examined carefully if there are other clinical findings related with this syndrome and the dentist should refer the patient for chromosome analysis to provide predictable longterm oral health care and improve the life quality.

Why this paper is important for pediatric dentist?

The patients with KS are undiagnosed because the clinical findings of the KS are often varied.

Taurodontic teeth could be a clinical sign of this syndrome and the pediatric dentist should refer for chromosome analysis.

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