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A rare case of multiple impacted teeth in a Klinefelter patient

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ABSTRACT

Klinefelter syndrome includes a group of chromosomal disorders with at least one additional X chromosome in male karyotype (46,XY). Up to now, different dental manifestations such as taurodontism, congenital absence of permanent teeth, shovel incisors, occlusal anomalies and increased permanent tooth size have been reported. A case of Klinefelter syndrome with very rare dental features is presented.

Keywords: Klinefelter syndrome, Multiple impacted teeth, Gardner syndrome, Cleidocranial syndrome.

Introduction

Klinefelter syndrome includes a group of chromosomal disorders with at least one additional X chromosome in male karyotype (46,XY). The most common and classic type of karyotype in Klinefelter syndrome is 47,XXY in which there is one extra X chromosome. Other less common karyotypes include 48,XXYY, 46,XY/ 47,XXY, 48XXXY, and 49,XXXXY. Klinefelter syndrome is the most common sex chromosome disorder [1].

Clinical presentations of Klinefelter syndrome may be widely different according to the age; leading to a late or missed diagnosis [1]. The Classic Klinefelter syndrome is characterized by gynecomastia at late puberty, hypogonadism (small testes, azoespermia/oligospermia, and androgen deficiency), infertility due to hyalinization and fibrosis of the seminiferous tubules, elevated urinary gonadotropines, sparse body hair and increased stature [1].

Psychosocial problems, developmental and learning disabilities including delayed speech and language acquisition are other manifestation of Klinefelter syndrome. In addition, some maxillofacial and dental features have been described in these patients, which are of interest of dentists. Smaller calvarial size, smaller cranial base angle and larger gonial angle, maxillary and mandibular prognathism, bifid uvula or cleft palate and enamel defect are maxillofacial complications of Klinefelter syndrome [2]. Up to now different dental manifestations such as taurodontism, congenital absence of permanent teeth, shovel incisors, occlusal anomalies and increased permanent tooth size have been reported.

A case of Klinefelter syndrome with very rare dental features is presented.

Case Presentation

A 24 year-old man with the chief complaint of the undesirable dental status and missing of some teeth referred to our clinic. On initial clinical examination, medical history and laboratory tests showed no systemic disease.

In oral examination, there were some abnormal deciduous and permanent teeth with spacing and unfavorable angle. Oral soft tissue seemed normal, but to some extent fibrosis was seen (Figure 1).

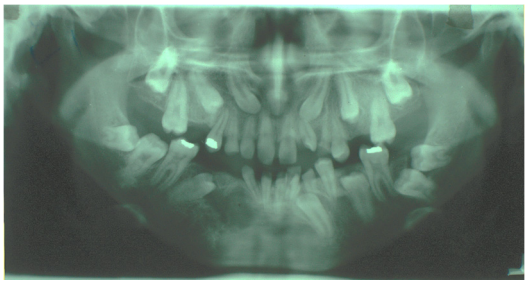

Figure 1. In Panoramic X-ray multiple impacted teeth with radiolucent lesions around some of them were seen.

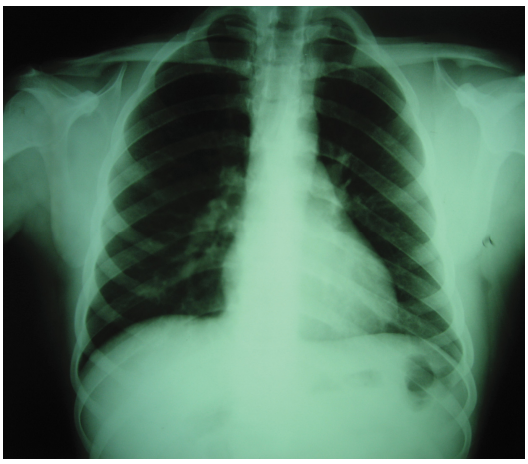

Figure 2. In patient's chest X-Ray both clavicles were intact.

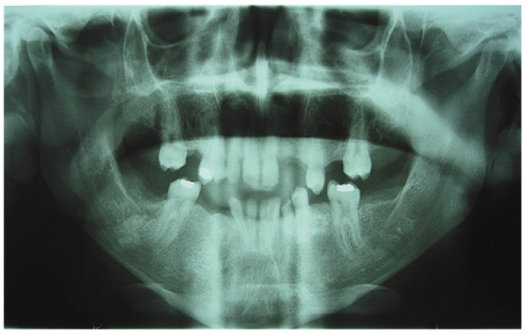

Figure 3. All of impacted teeth (and related pathologic lesions) were surgically extracted.

Panoramic X-ray revealed that teeth 13, 15, 17, 23, 25, 27, 33, 35, 37, 38, 45, 47, 48 were impacted and teeth 18, 22, 28, 43, 44 were missing (Figure 2). There were no supernumerary teeth.

Patient was treatment planned for forced eruption of impacted teeth using orthodontic forces. However, after consultation with orthodontists, it seemed impossible because his teeth did not have the ability to erupt and there was no desirable anchorage in his mouth. Therefore due to presence of pathological lesions (e.g. around right second premolar) it was decided to extract the impacted teeth and then prepare the alveolar ridge for denture or implant placement.

Considering patient's feature and radiographic findings, further investigation into presence of syndrome was carried at the time of extraction of impacted teeth.

In the first step, cleidocranial dysplasia and Gardner syndrome were suspected. In clinical and radiographic examination, both clavicles were intact. Patient's fontanels were closed. Thus, cleidocranial dysplasia was ruled out.

Furthermore, in order to evaluate the presence of Gardner syndrome (an autosomal dominant familial adenomatous polyposis with premalignant lesions), colonoscopy was performed and it was negative for rectosigmoid and colonic polyps or mass. Considering the result of colonoscopy and consultation with the internists Gardner syndrome was ruled out.

Later, patient was referred for genetic examinations and determining genotype and karyotype. The patient's karyotype was 47,XXY.

In clinical examination, it was revealed that patient has small testicles with azoespermia and scanty head hair, large gonial angle, mandibular prognathism and enamel defects in the erupted teeth. These symptoms also confirm the diagnosis.

For further medical treatment and consultation, the patient was referred to a geneticist and an urologist (because of azoespermia and probably infertility).

Unfortunately, after surgical extraction of impacted teeth (Figure 3), the patient failed to visit for further treatment.

Discussion

Tooth impaction is a situation that mechanical blocking causes eruption failure and the tooth remains unerupted in the normal time of eruption. Impacted teeth usually involve permanent dentition. This condition is caused by systemic or local etiologic factors or may be related to syndromes and metabolic disorders. Multiple impacted teeth have been reported as a feature of cleidocranial dysosotosis, Gardner syndrome, Yunis-varon syndrome, Gorlin-sedano syndrome and Gorlin syndrome [3-7]. This condition has also been reported in mucopolysaccharidoses [8].

Lapeer and Fransman reported hypodontia, impacted permanent teeth, spinal defects, and cardiomegaly in a previously diagnosed case of Yunis-Varon syndrome [5].

Another condition which can be considered in the differential diagnosis in multiple impacted teeth is Nevoid basal cell carcinoma syndrome (NBCCS) or Gorlin syndrome. Oral manifestations of Gorlin syndrome are impacted teeth, dental agenesis, malocclusion and cleft palate and lip [7].

Klinefelter syndrome is an under diagnosed chromosomal disorder with important challenges for health and medical management. By now various dental features in these patients have been reported but to our knowledge this is the first report of multiple impacted teeth in the Klinefelter syndrome.

Previous studies on the sex chromosome abnormalities show that the X chromosome primarily influences enamel thickness, whereas the Y chromosome promotes both enamel deposition and dentine growth [9]. However, the role of sex chromosomes in the formation of impacted teeth is unknown and has not been studied yet.

Hata et al reported dentofacial manifestations of XXXXY syndrome [10]. The most common dental manifestation which was reported by Hata et al was Taurodontism. Other dental manifestations were missing teeth and delayed development of the permanent teeth. However, they did not report impacted teeth in this syndrome, as seen in other reports.

Therefore in each case with abnormal dental feature, further work-up should be performed to rule out any metabolic disorders or syndromes.

Conflict of Interest: *There are no conflicts of interest or financial interests to be disclosed.*

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