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CASE HISTORY REPORT

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Supernumerary teeth in a patient with turner syndrome: An unusual finding

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Abstract

Aims: Turner syndrome (TS) is a genetic disorder associated with abnormalities of the X-chromosome, occurring in about 1 in 2000 to 1 in 3000 live-born girls. We present a case of a 14-year-old girl with TS, who was referred to our outpatient clinic in 2016 because of an ectopic eruption.

Methods and results: Dental clinical examination and radiographic investigation revealed eight supernumerary teeth, short roots, enamel hypoplasia, increased overjet, rotation and displacement of teeth, moderate gingivitis and morphological alteration of the upper right central incisor. Dental treatment included extraction of erupted supernumerary teeth, composite resin restoration, supragingival scaling and oral hygiene for plaque control.

Conclusions: The unpublished finding of supernumerary teeth in our patient has led us to suggest the investigation of this dental developmental anomaly in other patients with Turner syndrome.

KEYWORDS

dental and craniofacial anomalies, dental management, supernumerary teeth, turner syndrome

1 | INTRODUCTION

Turner syndrome (TS) is one of the most common sex chromosome abnormalities in females, occurring in approximately 1 in 2000 to 1 in 3000 live female births.[1] This genetic disorder was first identified in 1938 by Henry Hubert Turner, who described seven female patients with sexual infantilism, webbed neck, cubitus valgus and short stature.[2]

Although the exact cause of Turner syndrome is unknown, the absence of all (45X monosomy) or part (deletions of the short and long arms) of a normal second sex chromosome, aberrations of X-chromosome (eg, isochromosomes) and various mosaicisms appear to occur as a result of a random error during formation of either ovules or sperm cells.[3,4] Additional genes can be involved in the pathogenesis, such as haploinsufficiency of SHOX genes, which explains the reduction in final height, changes in bone morphology, sensorineural deafness, and other features.[5]

The main general clinical characteristics suggesting the diagnosis of TS include short stature, webbed neck, sexual infantilism, cubitus valgus, gonadal dysgenesis, short metacarpals, shield-like chest, breast hypertelorism, cardiac and renal malformations, intestinal telangiectasia, and multiple pigmented nevi.[6,7] Craniofacial stigmata include epicanthic folds, short nose with elevated base and tip, triangular face, micrognathia with small receding chin, low-set prominent ears, low hairline at the nape of the neck, and compromised craniofacial growth.[6–8]

The final diagnosis of TS is confirmed with standard karyotyping. When TS is suspected in pregnancy, the diagnosis can be confirmed by chorionic villus sampling or amniocentesis.[4] The treatment of TS is determined according

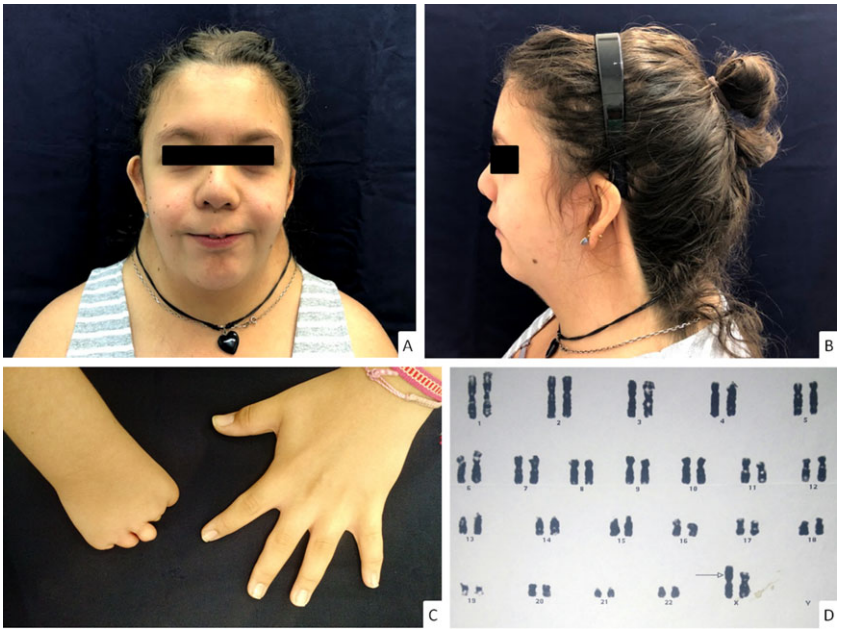

FIGURE 1 (A) Front view: triangular face, webbed neck, short nose with elevated base and tip. (B) Lateral view: webbed neck, prominent ears and low implantation of hair. (C) Atrophy in the right upper limb. (D) Karyotype 45,Xi (Xq) for Turner syndrome

to the patient's age. Cardiac or renal malformations should be treated during the newborn period. Growth hormone (GH) is often used in childhood and estrogen therapy for induction of puberty, whereas sex hormone therapy (estrogen and progesterone) is commonly used after puberty, at least until the age of menopause.[9]

The oral features in individuals with TS, as described in the literature, are high-arched palate, malocclusions, early dental development and premature eruption of permanent teeth, alterations in tooth shape and size, enamel hypoplasia, abnormalities in intercuspal distance, and abnormal root morphology.[3,7,9] Here, we describe supernumerary teeth that have not been previously reported in patients with Turner syndrome.

2 | CASE REPORT

A 14-year-old Caucasian female was seen at the Special Dental Care Center complaining of ectopic tooth eruption. She was the single child born to healthy but consanguineous parents. Familial history reveals four close relatives with upper limb atrophy. She was diagnosed with Turner syndrome after her birth, with her karyotype determined as 45,X,i (Xq) (Figure 1D).

She reported primary amenorrhea, but no cardiovascular or renal anomalies, and exhibited short stature, cubitus valgus and multipigmented nevi. We have also observed skin folds in the neck (webbed neck), low implantation of hair (Figures 1A and B) and atrophy of right upper limb (Figure 1C).

Intraoral examination revealed supernumerary tooth erupted laterally to the upper right central incisor, hypoplastic enamel affecting several teeth, increased overjet, rotation and displacement of teeth, moderate gingivitis and morphological alteration of the upper right and left central incisors (Figures 2A and B). Radiographic evaluation showed four supernumerary teeth in the premaxilla and four supernumerary premolar teeth in the mandible. Furthermore, lower incisors and lower right second molar exhibited short roots (Figure 2C).

We have extracted one erupted supernumerary tooth and periodical clinical monitoring of other supernumerary teeth was planned. A direct resin composite restoration of maxillary central incisors was carried out to obtain a proper anatomical morphology. We have also performed periodontal scaling and orientation of oral hygiene for biofilm control. Additionally, the patient was referred for evaluation and subsequent orthodontic treatment. The treatment plan included maxillary expansion and dental alignment. But the beginning of the orthodontic treatment would depend on the success of the periodontal treatment to which the patient was submitted, as well as the improvement of oral hygiene and biofilm control. The patient was scheduled to receive a 6-month follow-up evaluation periodically. In the first return, it was observed that one of the supernumerary teeth was erupting toward the hard palate (Figure 2D) and therefore it was extracted. This patient has been under regular monitoring for three years.

3 | DISCUSSION

To our knowledge, this is the first report of multiple supernumerary teeth in a patient with Turner syndrome (TS). Many authors have already reported other tooth abnormalities in individuals with TS before, such as tooth size, alterations in the shape and size of the crown, abnormal root morphology,

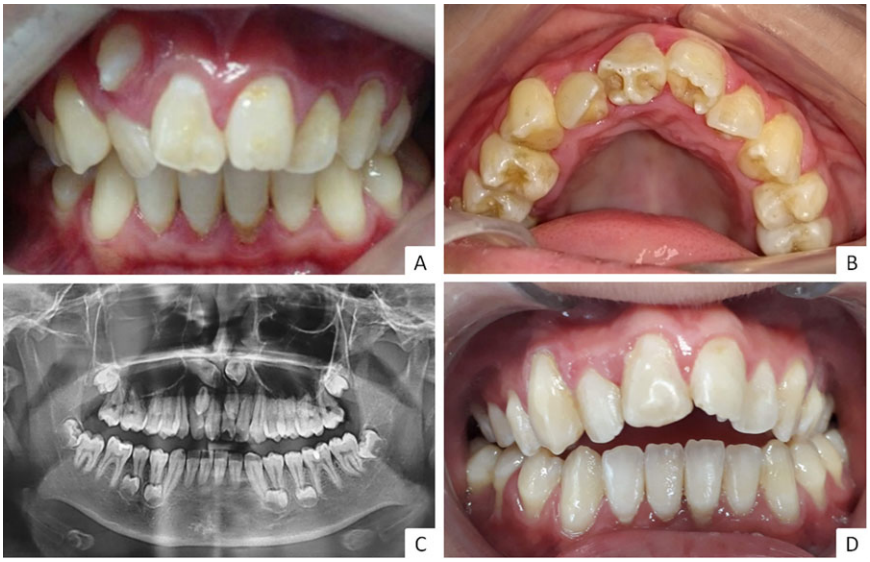

FIGURE 2 (A) Erupted supernumerary tooth laterally to the upper right central incisor and hypoplastic enamel affecting several teeth. (B) Morphological alteration of upper right and left central incisors and teeth rotations and displacements (note: at that time, the supernumerary canine had already been extracted). (C) Radiographic evaluation showing several supernumerary teeth. (D) Last follow-up, after 12 months, showing worsening of maxillary atresia and open bite

enamel hypoplasia, root resorption and early eruption of permanent teeth,[5,8–11] but none reported supernumerary teeth.

Supernumerary teeth may occur as an isolated event or in association with syndromes such as cleidocranial dysplasia, Gardner's syndrome, Down's syndrome, Noonan's syndrome and Fabry-Anderson disease.[12]

Supernumerary teeth might cause clinical consequences such as displacement of other teeth, failure to erupt, overcrowding, root resorption, cyst formation, and ectopic eruption. Early diagnosis and appropriate intervention can reduce supernumerary teeth related complications, and both clinical and imagiological exams, such as radiography and computerized tomography, are essential for diagnosing supernumerary teeth The usual treatment is to extract the supernumerary tooth, but there is no consensus on the best time to remove it. Early removal of unerupted supernumerary teeth seems to be advantageous up to approximately age 6-7 years after which further complications are expected. Another option is to keep the supernumerary tooth under observation if there is no complication or interference with function or aesthetics.[13,14]

In addition, the presence of asymptomatic supernumerary teeth near areas of risk (ie mandibular canal, nasal fossa, maxillary sinus, basilar mandible), where the surgical risk does not exceed the benefits of extraction, justifies the nonperformance of elective procedures and reinforces the need for follow-up.[15,16]

In this case we have extracted the ectopic supernumerary teeth because its presence was aesthetically unacceptable and it was traumatizing the upper labial mucosa, leading to ulceration from time to time. The remaining 7 supernumerary teeth were kept under follow-up, since they were asymptomatic and there were no pathological conditions associated with them.

Another oral feature of this syndrome is the small mesiodistal diameter of permanent teeth, particularly the crown width of the lower first permanent molar. The reduced size of the tooth crown probably is caused by the thin thickness of the enamel.[17] In the present case, the patient presented shape alterations in the permanent upper right and left central incisors, besides poor oral hygiene. Morphological changes in the dental crown have been previously described and are characterized by differences in mesiodistal and vestibulolingual width when compared with healthy individuals.[18,19] Enamel hypoplasia has been described in TS patients, which was also found in the present case, such as talon cusp, enamel opacities and enamel hypoplasia.[3,5,17]

To improve the shape of the central incisors and seal the existing pits in these teeth, we proposed two types of treatment for the patient. The first one, more invasive, more time consuming and more expensive, included the endodontic treatment and the zirconia/porcelain crown. The second option, more conservative, proposing the direct restoration in composite resin. The patient chose the second treatment plan, and there was no pulp involvement during the execution of operative dentistry.

Enamel defects have been correlated with the karyotype 45,X and patients with structural aberrations of X-chromosome exhibit changes in the excretion of amelogenin quantitatively and qualitatively.[3] Lopez et al[9] reported lower values of calcium and phosphate blood levels in most of the patients with TS, and this finding might probably influence the hypoplastic enamel of TS teeth. In the present case, the patient had karyotype 45,Xi(Xq) and enamel hypoplasia in upper anterior teeth, thus affecting the esthetics. The treatment for these defects is very well established and good esthetic results can be obtained with resin composite restoration, like in the present case report.

Individuals with TS are characterized by reduced maxillary growth, mid-face hypoplasia, anterior open bite, high-arched palate, narrow upper arch and micrognathic mandible.[11] In

addition, they have shorter posterior length and increased cranial base angle along with bi-maxillary retrognathism. This indicates that deficiency of the X-chromosome genes has a direct influence on cranial base and jaw bones, causing irregular growth. The retrognathic position of the jaw in TS patients might be associated with direct influence of the deficiency of X-chromosome genes, possibly SHOX gene deficiency.[8]

Rate and timing of growth, use of GH therapy and development of craniofacial structures are factors involved in the stomatognathic system development (SHOX gene). Craniofacial and dental morphology in TS patients influences significantly the response to orthodontic treatment. Sometimes, growth modification techniques associated or not with orthognathic surgery are required.[11,20]

In the present case report, we have observed increased overjet, rotation and displacement of teeth as well as presence of one erupted supernumerary teeth and 7 unerupted supernumerary tooth. The patient was referred for evaluation and subsequent orthodontic treatment. The treatment plan included maxillary expansion and dental alignment. But the beginning of the orthodontic treatment was related to the success of the periodontal treatment to which the patient was submitted, as well as the improvement of oral hygiene and biofilm control.

Although changes in tooth morphology and structure have been well described in the current literature, this seems to be the first case report on a patient with Turner syndrome having multiple supernumerary teeth. It is important that the dentist knows this possibility in order to better plan the dental treatment of the patient.

4 | FINAL CONSIDERATIONS

Dental treatment of patients with Turner syndrome is necessary for oral health esthetically and functionally. Dentists should be aware about the characteristics of Turner syndrome and modify the patient's treatment plan accordingly. The unpublished finding of supernumerary teeth in our patient suggests that further studies are needed to investigate this dental developmental anomaly in other patients with Turner syndrome.

CONFLICT OF INTEREST

There is no conflict of interest.

ETHICS STATEMENT

Data from the patient included in this case report were treated anonymously and a statement of informed consent was signed by him allowing the use of their medical and dental records.

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