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Multiple dens invaginatus, mulberry molar and conical teeth. Case report and genetic considerations

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Abstract

Dens in dente, also known as dens invaginatus and dilated compound odontoma, is a malformation that can occur on primary, permanent, or supernumerary teeth that is characterized by a deep invagination of the surface of a crown or root covered with enamel. This abnormality in tooth morphology generally affect the maxillary lateral incisors but several cases of multiple dens invaginatus have been reported in the literature. A 15 year-old female patient is reported here presenting five dens invaginatus: four in the permanent mandibular incisors and one in the permanent, maxillary left central incisor, additionally the following dental findings were observed: a permanent mandibular left mulberry molar, molarization of some premolars, several microdontic conoid teeth, retention of five primary teeth, absence of several permanent teeth germs, a macrodontic molar with abnormal roots and several periapical radiolucencies associated to the dens invaginatus. There was no family history of similar dental findings to those observed in the patient. There are several genes that participate in the development of teeth, of those, the following five genes could be implicated as responsible or co-participators for some of the dental anomalies present in this patient: MSX1 (Muscle segment homeobox 1), DLX1 & DLX2 (Distal-less homeobox 1 & 2 genes), PAX9 (Paired box gene) and PITX2 (Pituitary homeobox transcription factor 2).

Key words: *Dens invaginatus*, *dens in dente*, *conical teeth*, *mulberry molar*.

Introduction

Dens in dente, also known as dens invaginatus, dilated compound odontoma or telescopic tooth, is a malformation that can occur on primary, permanent, or supernumerary teeth (1,2). Dens in dente is characterized by a deep invagination of the surface of a crown or root covered with enamel (2) and has an incidence of 0.04 to 10% in the general population (3). According to the extent of the invagination, dens in dente has been classified as: type I where the invagination is confined to the crown; in type II the invagination extends from under the cemento-enamel junction ending in a blind sac that can or cannot communicate with the pulp and type III with an invagination that extends through the root perforating the root apex or its vicinity without communicating with the pulp (1,4-6).

Mulberry molars are irregular teeth generally affecting the first molars and are characterized by a grossly deformed crown imitating, as the name implies, the surface of a mulberry. These molars are frequently seen in patients with congenital syphilis.

Microdontia, macrodontia and conoid teeth represent variations in crown morphology that in some families are inherited traits.

Case Report

A 15-year-old female was seen at the Pediatric Dentistry Clinic at the University of Baja California Tijuana complaining of pain and slight swelling in the area of the permanent maxillary left central incisor area of several days duration.

The extraoral exam revealed an increase in size of the upper lip caused by what it seemed to be a periapical abscess in the permanent maxillary left central incisor. The intraoral exam revealed the following: the maxillary right first molar was microdontic and its crown was in a mesodistal, vestibulolingual and cervical-occlusal orientation; the primary maxillary right second molar, primary maxillary right canine and primary maxillary right lateral incisor were retained and the maxillary right first premolar and the permanent maxillary right central incisor were conoid in shape. The permanent maxillary left central incisor was fused to the permanent maxillary left lateral incisor forming a dens in dente. The primary maxillary left canine and the primary maxillary left second molar were retained; the maxillary left first premolar and the permanent maxillary left first molar were conoid in shape (Figure 1).

The permanent mandibular right first permanent molar was microdontic, the mandibular right second premolar had a grade 2 group I caries, the mandibular right first premolar and the permanent mandibular right canine were conoid, and the permanent mandibular right lateral and central incisors presented a dens in dente type III as well as the permanent mandibular left central and lateral



Fig 1. Note the various dental anomalies in shape as described in the text.



Fig 2. The abnormal shape of the mandibular teeth presenting dens-in-dente can be seen in this photo, as well as the left mulberry molar.



Fig 3. Note the various dens-in-dente in the mandibular arch and also the maxillary one.

incisors. The rest of the left mandibular arch was occupied by a permanent conoid canine and first premolar, molarization of the second premolar and a permanent mulberry first molar (Figure 2). A Panoramic radiograph as well as intraoral X-rays were ordered (Figure 3) and they revealed: retention of the primary maxillary right second molar, primary right canine, primary right lateral incisor and primary left canine. The permanent germs corresponding to the aforementioned primary teeth were absent. Periapical radiolucencies were observed at the level of the permanent mandibular central incisors, permanent mandibular left lateral incisor and the permanent maxillary left central incisor. Those were the teeth presenting dens in dente. A macrodontic molar with radicular anomalies, was seen at the level of the

permanent mandibular second right molar, the possibility of this lesion being an odontoma will be ascertained after its surgical removal and pathology evaluation.

The patient was the product of a seventh pregnancy. The mother did not report any chronic disease and denies taking any drug before or during pregnancy. She also denies any family history of similar dental findings to those observed in the patient. Four family members were examined intraorally and were found to be within normal limit. The patient's clinical history is positive for neonatal hypoxia. She has not undergone any surgical intervention and does not have any history of allergic reactions. Two years previously she was diagnosed with a heart murmur without specifying place or etiology. A consultation with a cardiologist was sought who ruled out the murmur but found pulmonary valve insufficiency, which does not require care and does not present a health risk.

Discussion

Kantaputra and Gorlin (7) in 1962 reported a case of a female patient who in addition of having severe congenital sensorineural deafness also presented double dens invaginatus of the molarized maxillary central incisors, premolarization of maxillary lateral incisors, multituberculism of the mandibular incisors, canines and first premolar.

The patient was evaluated by an otolaryngologist who performed an audiometry, ruling out the presence of sensorineural deafness, thus establishing a differentiation with the case reported by Kantaputra and Gorlin (7).

Casamassimo and coworkers (8) reported a 12 year-old boy, which have been seen previously by them at age 9, presenting microdontia, taurodontia and dens invaginatus with a family history of this association inherited as an X-linked recessive; this differ from our case in that there is no family history of the findings presented by the patient. There are several genes that participate in the development of teeth, of those, the following five genes could be implicated as responsible or co-participants for some of the dental anomalies present in this patient.

MSX1 (Muscle segment homeobox 1) which map to the short arm of chromosome 4 (4p16.1), is critical for the development of specific human teeth i.e. 2nd premolars and 3rd molars, it also causes Witkop syndrome also known as Tooth and Nail syndrome, which is characterized by diminished number of teeth with a conoid shape as well as nail dystrophy (9).

DLX1 & DLX2 (Distal-less homeobox 1 & 2 genes) map to the long arm of chromosome 2 (2q32), these genes are required for early tooth and craniofacial development (10,11).

PAX9 (Paired box gene) mapped to the long arm of chromosome 14 (14q12-q13) is active in the formation of molars as well as mandibular 2nd premolars and incisors. It is essential in promoting the inductive capa-

bility of the tooth mesenchyme and it has been shown to have a functional relationship with MSX1 during teeth development (12).

PITX2 (Pituitary homeobox transcription factor 2) which maps to the long arm of chromosome 4 (4q 24-25) could also be considered because it is responsible for Rieger syndrome type I in which patients present eye anomalies, hypodontia and conoid teeth.

It must be bear in mind that different mutations in the same genes will produce different clinical manifestations (13).

The treatment plan for our patient included consultations with Endodontia, Prostodontia, Periodontia, Oral Surgery and Oral Pathology. It was recommended to start with endodontic treatment of the teeth affected by dens invaginatus. The success of such treatments will predict the possibilities of prosthetic treatment. Periodic observation of the abnormal permanent mandibular right second molar is recommended to determine if there is a possibility of its eruption to properly ascertain if it is an abnormal tooth or a possible odontoma. During July 2006, successful root canal treatments of the maxillary and mandibular dens invaginatus were performed together with bone grafts to fill the defects.

When confronted with cases of apparently unique and infrequent pathology, where a variety of morphological anomalies appear in the majority of teeth, as in this patient, a comprehensive multidisciplinary approach should be the treatment of choice.

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