CASE REPORT

Molarization of premolar with dentin dysplasia Type Ia - A rare unilateral entity

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Abstract

Human dentition exhibits variations in anatomic features. When the size and morphology of teeth are analyzed, a range exists within which a particular tooth should be probably placed, but some teeth show variance from this range. Mandibular premolars (MnP) are among such teeth. This paper presents a rare case of unilateral megadontia with dentin dysplasia, showing the presence of molariform appearance of second MnP which appeared radiographically as rootless tooth with deformative pulpal morphology. Clinically, the patient suffered from temporomandibular joint disorder which may be because of the occlusal instability caused by the lingually placed molarized premolar and so appropriate preventive therapy was recommended.

Introduction

Tooth anomaly affects both permanent and deciduous dentition and anterior and posterior teeth including maxillary and mandibular arch. The condition involves size, shape, structure, and number of the tooth. Mandibular premolars (MnP) are highly variable in shape. A variation called human MnP tooth shape deviation (TSD) is characterized by squeezed faciolingual (FL) dimension and enlarged mesiodistal (MD) dimension reversing typical premolar proportions. MnP-TSD is 8 times more common in the second premolar than the first premolar. Males are more commonly affected than females (1.4:1). Bilateral involvement is more common (59%) than unilateral involvement. This condition causes a hindrance in eruption, disrupts the dentition, and becomes a challenge to orthodontic treatment and achievement of ideal occlusal intercuspation. Once erupted, the anatomy of such teeth predisposes them to caries and other clinical abnormalities.[1-5]

When dental size and anatomy present characteristics that deviate from the accepted range of normality, they are termed anomalies and it is important to distinguish between what can be called variation and what can be described as anomaly.[6] The anatomy of mandibular second premolar is particularly unpredictable and shows an elevated variability in morphology. Two types of extreme deviations in MnP shape or size can be ascertained from the literature: (1) Megadontism/macrodontia or molarization-the gross enlargement of both the MD and FL dimensions of the MnP and (2) tooth shape deviations (TSD) of MnP (MnP-TSD) - the product of a concomitant increase in the MD dimension and reduction in the FL dimension.[1]

Dentin dysplasia (DD) is a rare disturbance of dentin formation characterized by normal enamel, atypical dentin, and abnormal pulpal morphology. It is an inherited autosomal dominant trait and classified into two types. DD Type I is characterized by the presence of primary and permanent teeth with normal appearance of the crown but no or only rudimentary root development, incomplete or total obliteration of the pulp chamber, and periapical radiolucent areas or cysts. DD Type II is characterized by primary teeth with complete pulpal obliteration and brown or amber bluish coloration similar to that seen in hereditary opalescent dentin.[7]

This paper describes an unusual case of unilateral morphological variation, i.e., molarization of the second MnP with Type I DD.

Case Report

A 21-year-old male patient reported to the Department of Oral Medicine and Radiology, Mansarovar Dental College and Research Centre, Bhopal, with a chief complaint of pain in left temporomandibular joint (TMJ) area for 2 months.

Extraoral clinical examination revealed clicking and pain in the TMJ and intraoral clinical examination revealed the presence of all the teeth found to be normal except for the molariform appearance of permanent second premolar on the right side which was not in the occlusal plane [Figures 1 and 2]. This tooth had four vestibular cusps and deformed lingual cusps with a faciolingual and MD diameter of 12.56 mm and 12.39 mm, respectively. No other hard tissue abnormality was detected with respect to any other tooth in the oral cavity. However, the soft tissue around the permanent second premolar showed signs of gingivitis because of the poor oral hygiene. There was no reported history of such teeth in the family. The resulting morphological appearance of the tooth was similar to the mandibular molar which according to the literature suggest it of being molarized premolar.[8]

Radiographic examination (intraoral and periapical radiograph) and orthopantogram [Figure 3] showed large crown of the second premolar with increased cuspal height and without roots. There was deformative pulpal morphology with respect to the molarized premolar, whereas the premolar on the left side and all other teeth show normal tooth morphology (Reviewer 2). On the basis of clinical and radiological appearance, it was diagnosed as unilateral molarization of mandibular second premolar with DD Type I (a).

The treatment plan included was oral prophylaxis followed by the International Monetary Fund for 21 days and occlusal splint for about 1 ½ months for the internal derangement of TMJ diagnosed on dynamic transcranial view radiography. The patient was also advised to go for extraction of the molarized premolar for establishing occlusal stability but was reluctant for the removal of tooth.

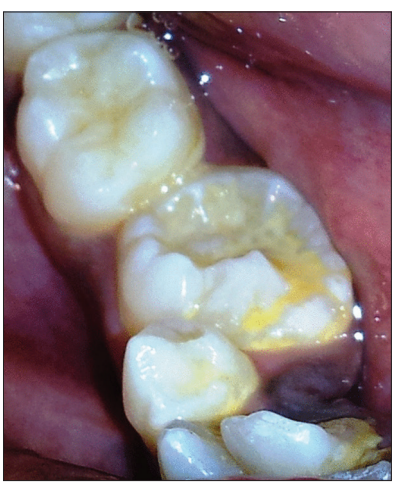

Figure 1: Clinical picture showing molarization of premolar

Discussion

Two types of extreme deviations in MnP shape or size can be ascertained from the literature: (1) Megadontism/macrodontia or molarization-the gross enlargement of both the MD and FL dimensions of the MnP and (2) tooth-shape deviations (TSD) of MnP (MnP-TSD) - the product of a concomitant increase in the MD dimension and reduction in the FL dimension.

As per the mentioned classification in the literature,[1] our case was of unilateral molarization of premolar which showed greater FL dimension than MD and hence cannot be categorized in the mentioned classification. This variability can be attributed to the strong genetic influence in the development of MnP and their position in dental arch. The morphology of mammalian teeth is diverse and is determined genetically. The characteristic

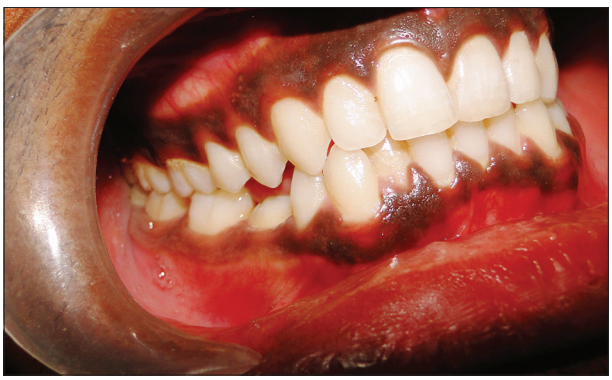

Figure 2: Clinical picture showing molarization of premolar

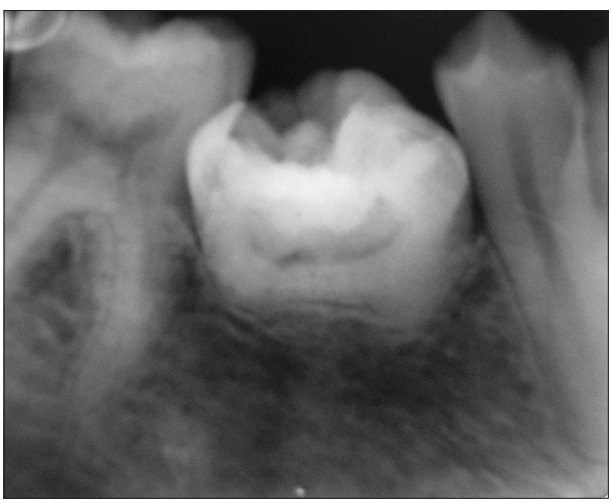

Figure 3: Intraoral periapical radiograph showing dentin dysplasia Type I

crown morphology in mammalian teeth is produced by the folding of inner enamel epithelial-mesenchymal interface and growth of dental papillae which leads to the formation of "primary enamel knot."

The formation of primary enamel knot may commence at the cap stage with a cluster of non-dividing epithelial cells, but "secondary enamel knot" structures are observed at early bell stage of tooth development at the sites of future secondary cusps under the influence of primary knot.[9-13]

As discussed by Thomas and Sharpe in 1997, the mechanism of patterning of dentition is based on the spatial and restricted expression of homeobox genes. As the tooth develops, a signaling center, known as the enamel knot, is set up at the center of the tooth germ, which controls the shape (morphogenesis) of the tooth and the number of cusps which are due to form. The genes involved in these patterning processes are starting to be elucidated in the enamel knot which initially expresses BMP-2, BMP-7, and SHH signaling molecules and later expresses BMP-4 and FGF-4 transforming factors, which was evident in an experiment done over the knockout mice. These signaling molecules affect both epithelial and mesenchymal cells and also their reciprocal interactions.[11-14]

The mode of evolutionary modification in molarization of premolar is because of the overlapping expression of these signaling molecules and transcription domains in enamel knot (as seen in seals and ungulates).[15] The literature suggests that all six Dlx genes exhibit complex expression patterns in the developing teeth during the formation of transient structures within the enamel organ known as the enamel knot, enamel cord, and enamel navel in the cap stage, suggesting its role in tooth morphogenesis. The expression of Dlx2 in the underlying mesenchyme at the bud stage and Dlx2/Dlx3/Dlx5 Dlx7 at the cap stage raises the possibility of a role of these genes in enamel knot induction or maintenance. Moreover, Thomas and Sharpe stated that the patterning of dentition is also under the control of Dlx-*1* and Dlx-*2* genes which are specifically involved in patterning of molar tooth development, so any aberrancy in expression of this group of genes is also responsible for cuspal and morphological defects.[11-15]

DD, a rare anomaly of unknown etiology, is classified into two types, radicular DD as Type I and coronal DD as Type II. In Type I, both the deciduous and permanent dentitions are affected. The crowns of the teeth appear clinically normal in morphology, but defects in dentin formation and pulp obliteration are present. Radiographic examination is important for the identification of DD Type I. The abnormal root morphology in DD Type I is postulated secondary to the abnormal differentiation and/or function of the ectomesenchymally derived odontoblasts. There are four subtypes for this abnormality. In Type 1a, there is no pulp chamber and root formation; Type 1b has a single small horizontally oriented and crescent-shaped pulp; Type 1c has two horizontal or vertical and crescent-shaped pulpal remnants surrounding a central island of dentine, and in Type 1d, there is a visible pulp chamber and canal with near-normal root length, and large pulp stones. According to this classification, our case was of DD Ia subtype.[16]

During the transition from cap to bell stage, the cranial neural crest derived dental papillae mesenchymal cells come in contact with the basement membrane lining the developing tooth pulp chamber which undergo cytodifferentiation under a process known as dentinogenesis.

Dlx3 is expressed in these mesenchymal teeth forming cells that give rise to dentin and the pulp. It is a potent regulator for proliferation and odontoblastic differentiation of human dental pulpal cells. Hence, any aberrancy in the expression of Dlx *3* gene can be responsible for the defective dentin and pulpal morphology found in DD. Choi *et al.* demonstrated that the mutant Dlx *3* gene disrupts the cytodifferentiation of odontoblasts leading to its apoptosis, resulting in aberrant dentinal tubule formation and dentin matrix production. Both these changes lead to decreased dentin and taurodontism. Moreover, immunohistochemical expression patterns of dentin matrix proteins, especially dentin sialophosphoprotein (DSPP), showed aberrant expression in transgenic (mice expressing MT-DLX 3) driven by a mouse 2.3 col1A1. Some reports have shown that three different net-1 bp frameshift mutations early in DSPPs repeat domain caused DD, whereas six more 3' frameshift mutations were associated with DGI. Here, it is appreciated that DD is because of the frameshift mutation in DSPP gene which previously was found to be associated with DGI phenotype only.. This new frameshift mutation shows that overlapping DSPP mutations can give rise to either DGI or DD phenotypes. Thus, abnormal expression of DSPP in mutant Dlx 3 transgenic mice can be one of the important factors responsible for the disrupted odontoblastic differentiation in DD.[17-19]

At the late bell stage, transcription factors and signaling molecules related to Dlx3 expression are shifted to the inner enamel epithelium which forms the Hertwig's root sheath, necessary to establish root morphology. Failure of Hertwig's root sheath to invaginate at the appropriate time leads to failure of root formation in DD Type Ia which might be due to any defect (mutation) in Dlx *3* gene. However, the pathogenesis of this rare radicular dysplasia and its molecular bases remains uncertain and needs to be evaluated at a genetic level.[10]

Conclusion

The clinical problem with molarization of premolars is the failure of acquisition of occlusal plane which may lead to instability and subsequent complications, among which the problem of chronic internal derangement of TMJ is the most common. The same problem was encountered in the present case. This can be because of the traumatic bite in the presence of lingually placed molarized premolar disturbing the occlusal plane. Management of patients with dentinal dysplasia has also presented dentists with many problems. Extraction has been suggested as a treatment alternative for such teeth.

Hence, it is important in diagnosis that the size, shape, and occlusal harmony of the teeth should be established so as to avoid any unforeseen complications.

Clinical Significance

From a clinical perspective, the early recognition of these disorders is crucial to prevent the onset of various complications. Cystic formation and lesions from impaction with neighboring teeth are the most common complications. Associated risks related to orthodontic treatment include the delayed or impossible closure of the space, interference with the root torque, and reabsorption of the roots of adjacent teeth. They are highly disposed to caries, because of their malposition and intricate occlusal morphology.

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