Dentin Dysplasia Type 1 D : A Case Report & Review of Literature

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Abstract

Dentinal dysplasia is a rare condition involving developmental abnormalities of dentin ; it is an autosomal dominant trait that resembles opalescent dentin. We report a case of a 17 year old girl with dentin dysplasia type 1 d.

Keywords : Dentin, dysplasia, pulp stones.

Introduction

Dentin dysplasia (DD) is an autosomal dominant hereditary disturbance in dentin formation, which may present with either mobile teeth or pain associated with spontaneous dental abscesses or cysts[1]. Dentinal dysplasia is generally divided into 2 main classes based on clinical and radiographic appearance. Shields et al proposed a classification for heritable human dentin defects that divided dentinal dysplasia into Type I (DD1), "dentin dysplasia," and Type II (DD2),"anomalous dysplasia of dentin"[2]. Type 1 DD has been classified into type 1 a, 1 b, 1 c and 1 d[3]. We report a case of a 17 year old girl with dentin dysplasia type 1 d with emphasis on the clinical and radiographic features.

Case Report

A 17 year old girl reported to our department with the chief complaint of pain in the right lower back tooth since 15 days. Intraorally, the crowns of all teeth were normal in size, shape and color. Marked attrition of the labial surface of the enamel at the incisal edges was noted, both in the maxillary and mandibular anteriors. The enamel was thin and transparent. Oral hygiene was poor and there were plaque deposits in all the quadrants (figure1). Right and left mandibular first molars were temporarily restored. Tenderness on percussion was present in the right mandibular first molar and vestibular tenderness in the region was also observed. None of the teeth were mobile. A clinical diagnosis of chronic periapical abcess in the right mandibular first molar was given. An intraoral periapical radiograph of the right mandibular first molar region revealed the presence of ill-defined periapical radiolucencies in both the mesial and distal roots, confirming the diagnosis of chronic periapical abcess. In addition, a large pulp stone was noted in the coronal portion of right mandibular first molar. The same radiograph also revealed the presence of two small coronal pulp stones and two large radicular pulp stones in the right mandibular first premolar. Two pulp stones were also noted in the canine. Constriction of the pulp canal was noted apical to the large pulp stones (figure 2). The presence of these multiple pulp stones necessitated a full mouth radiographic survey. On radiographic examination, all tooth roots were normal in length. No pulp stones were seen in the maxillary anterior region. The left mandibular canine showed the presence of two pulpstones. Multple pulp stones were also seen in both right and left maxillary and mandibular premolar-molar region. These pulp stones were round to oval in shape. The smaller pulp stones were located in the coronal pulp while the larger ones extended upto the coronal one third of the radicular pulp. Bulging of the pulp chamber owing to the presence of large pulp stones was noted. Consrtiction of the pulp chamber apical to these stones was also seen. The larger pulp stones especially in the molar region occupied almost the entire coronal pulp chamber (figures 3, 4 and 5). Overall clinical and radiographic features were suggestive of dentin dysplasia type 1d.

Discussion

Dentinal dysplasia (DD), so named by Rushton, is an autosomal dominant hereditary condition affecting dentin formation that is rarely encountered in dental practice. Its prevalence has been recorded as 1 in 100,000[2]. The disorder was first described by Ballschmiede, who called the condition “rootless teeth” because of the short blunted appearance of the roots and spontaneous exfoliation of teeth in 7 children from the same family[2]. In DD the orientation of dentinal tubules is affected resulting in defective root formation[4]. In 1972, Witkop, classified DD into two types, radicular DD as type 1 and coronal DD as type 2[1]. Type 1 affects the root portions of both primary and permanent dentitions. DD type 1 was classified into four subtypes ; 1a, 1b, 1c and 1d. In type 1a, there is a complete obliteration of the pulp and usually little or no root development. Type 1 b variant has a horizontal, crescent shaped radiolucent line which separates normal coronal dentin from abnormal radicular dentin[5]. The roots are short, conical and rudimentary. Teeth affected by 1 c variant show two crescent shaped horizontal radiolucent lines with their concavities towards each other at the cementoenamel junction and the roots one half the normal length. Type 1 d is characterized by normal root formation, which sometimes may be bulbous in the coronal third. Within the pulpal canal, a “stone” may be found[5]. In this type of DD, the pulp chamber is usually not obliterated and normal root formation occurs. In these cases, the pulp around the stones is healthy[5]. These large pulp stones are located in the coronal portion of the canal and create a localized bulging in the canal, as well as root constriction of the pulp canal apical to the stone and few periapical radiolucencies[1] In other cases, the denticle is continuous with dentinal walls[1]. In our case, the lack of mobility of teeth, near normal crown morphology and the presence of multiple pulp stones lead to the diagnosis. Type 1 d is the least severe form of DD[5]. In other forms, first signs of dentin dysplasia type I may be the premature exfoliation of teeth, either spontaneously or with minor trauma, but a delayed dental eruption pattern has also been reported[6].

In addition, normal appearing crowns with severe hypermobility of teeth and spontaneous dental abcesses or

cysts may be seen[1]. Histologically, the enamel and the immediately subjacent dentin appear normal. Deeper layers of dentin show an atypical tubular pattern with an amorphous, atubular area and irregular organization. Pulpally to normal appearing mantle dentin, and globular or nodular masses of abnormal dentin are seen[1]. In our case, since none of the teeth were indicated for extraction, histopathological examination could not be performed. DD type 2 produces a grayish or brownish opalescent coloration in primary teeth; these have normal morphologic character. After eruption of primary dentition, continued deposition of abnormal dentin causes obliteration of the pulp chamber and root canals. Both crowns and roots of permanent teeth appear clinically normal. Radiographic examination, however, reveals abnormally large pulp chambers, which have been described as "thistle tube" shaped[4]. Differential diagnosis includes dentinogenesis imperfecta. In particular, the following differences between the two conditions were noted: dentinogenesis imperfecta was found to genetically present as a single dominant characteristic that was not sex-linked, whereas dentinal dysplasia was found to have a hereditary association in a single family (i.e., it was an autosomal dominant trait). In cases of dentinogenesis imperfecta, the crown was found to be bell-shaped, and the teeth had short, narrow roots[3]. Treatment options in primary dentition include stainless steel crowns on the molars to prevent tooth wear and maintain the occlusal vertical dimension. The esthetics may be improved using composite facings or composite strip crowns. If, however, the child presents late, the teeth may have undergone attrition to the level of the gingiva and the only treatment option then is to provide over-dentures[7]. Extraction, should be performed in cases involving radiolucent areas at the periapical region, mobility, or both radiolucency and mobility; such cases would necessitate replacement prostheses[3]. Some patients present with severe tooth wear in the permanent dentition. As in the primary dentition, one of the options is over-dentures. Those with DD-I have mobile teeth due to very short roots and as a result tend to lose teeth early in the primary and permanent dentition. Until growth is complete, the treatment of choice for the replacement of missing teeth is dentures. Dental implants may be considered when growth is complete at about 18 years of age. Maxillo-mandibular atrophy is a consequence of no or rudimentary root development and early tooth loss. Ridge augmentation prior to implants is often required[7]. Other treatment strategies include conventional endodontic therapy, periapical curettage or preventive regimen to maintain the teeth as long as possible[8]. Successful endodontic treatment of teeth with pulp stones can be challenging. Chelating agents are employed to overcome this obstacle. The most commonly employed such agent is Ethylene Diamine Tetracetic acid, which was advocated first by Nygaard and Ostby in 1957[9].

To conclude, there are still many inconclusive issues in the diagnosis and management of patients with dentin dysplasia. The diagnostic features of this rare disturbance will remain incompletely defined until additional cases have been described. Early diagnosis of the condition and initiation of effective regular dental treatments may help these patients to prevent or delay loss of dentition[1].

References

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Legands

Fig. 1 Intraoral picture showing normal crown morphology with attrition of the labial surface of the enamel in maxillary and mandibular anterior teeth.

Fig. 2 Intraoral periapical radiograph showing four pulpstones in the right mandibular second premolar. Apical constriction is present apical to the large pulp stone. Pulp stones can also be seen in the canine, first premolar and first molar.

Fig. 3 Pulpstones in the right maxillary second premolar and second molar.

Fig. 4 An intraoral periapical radiograph showing pulpstones in the left mandibular canine, first premolar.

Fig. 5 Pulpstones in the left maxillary second premolar, first and second molar.

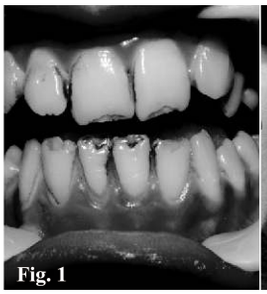
Fig. 1

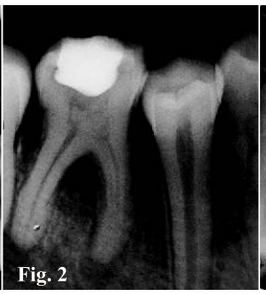
Fig. 2

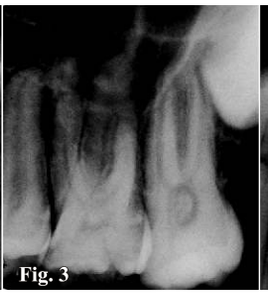
Fig. 3

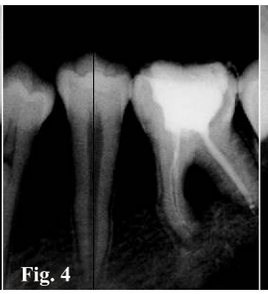
Fig. 4

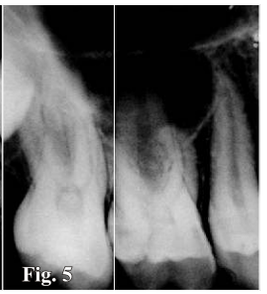
Fig. 5