CASE REPORT

A Rare Case Report on Dentin Dysplasia Type Ib with Emphasis on Review of Literature.

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Abstract

Introduction: Dentin dysplasia (DD) is a rare autosomal dominant developmental anomaly of dentin characterized by normal enamel but atypical dentin formation with abnormal pulpal morphology. The incidence of this rare disorder is approximately one in every 100,000, 10 times less than that of dentinogenisis imperfecta. It may present with either mobile teeth or pain associated with spontaneous dental abscesses or cyst. The primary and permanent dentitions are affected with equal sex predilection.

Case Presentation: This article describes a rare case of DD-Ib in a 10 year old female patient with a known family history and was confirmed by ground sectioning of the tooth specimen and histopathology examination.

Keywords: Dentin dysplasia, Type Ib,

Key message: Meticulous oral hygiene practice and effective treatment modalities must be established for the retention of the teeth. So dentist play a major role in educating the patient as well as family members regarding the condition and the importance of early intervention.

Introduction

Dentin dysplasia (DD) is a rare autosomal dominant developmental anomaly of dentin characterized by normal enamel but atypical dentin formation with abnormal pulpal morphology. It occurs due to mutation of chromosome 4q21.3 and thus resulting in the disturbance of dentin formation[1]. The incidence of this rare disorder is approximately one in every 100,000, 10 times less than that of dentinogenisis imperfecta[2,3]. It is a rare anomaly of unknown etiology, and usually associated with number of disorders such as Ehlers-Dalons syndrome, calcinosis, and vitaminosis[5]. It may present with either mobile teeth or pain associated with spontaneous dental abscesses or cyst. The primary and permanent dentitions are affected with equal sex predilection.

This article describes a rare case of DD-Ib in a 10 year old female patient with a known family history and was confirmed by ground sectioning of the tooth specimen and histopathologic examination.

Case Presentation

A 10 year old female patient reported to our department with the chief complaint of discolored upper front teeth. Dental history revealed that the eruption of primary teeth was unremarkable in timing and sequence but retention of primary teeth which resulted in ectopic eruption of permanent teeth. Patient has a known family history of dentin dysplasia (DD). Her father and elder sister was previously diagnosed with DD type Ib, earlier from other institution (Figure 1).

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Intraoral examination revealed the patient was in the mixed dentition appropriate for her age with decayed 55, grossly decayed 54, root stump 75, retained 53, and palatally erupting 13. Morphological appearance of all teeth was normal, with a slight yellowish hue. Patient has a midline diastema with high attached frenum and slightly rotated upper central incisors (Figure 2).

Panoramic radiograph showed a mixed dentition with generalized short and malformed roots. Pulp chambers and root canals were seen obliterated in case of anteriors and posteriors showed crescent shaped pulpal remnant at cemento-enamel junction (Figure 3).

Panoramic radiograph of patients elder sister showed a permanent dentition with generalized short and malformed

roots similar to those noted in our case. Anteriors showed obliterated pulp chambers and root canal whereas posteriors showed crescent shaped pulp chambers at cemento enamel junction. 31,32,41,42 were missing. 23 was impacted, and a well-defined periapical radiolucencies were present in association with the apex of the right and left mandibular first molars and maxillary first premolar. So a provisional diagnosis of DD type I was considered. 54 and 55 were extracted and send for histopathologic examination.

Examination of the gross specimen revealed a grossly decayed 54 and dentinal caries with relation to 55. 55 was sectioned mesio-distally into 2 halves. We could even make out macroscopically that the whole pulp chamber was obliterated. One half of the tooth specimen was decalcified using 8% nitric acid and processed for H and E staining, other half of the cut tooth was used for preparing ground sections.

Ground section showed thin layer of enamel with normal dentin seen adjacent to the enamel making up the bulk of the tooth. Almost the whole of the pulp chambers was obliterated except for a crescent shaped pulp in the coronal portion. Below that normal dentinal tubules can be seen to some extent. Beyond that there was abrupt transition of dentin to an abnormal pattern in the deeper layer. The dysplastic dentin composed of multiple foci of calcification surrounded by whorls giving characteristic appearance of lava flowing around the boulders. The radicular portion showed dysplastic dentin but at one area we could appreciate tomes granular layer.

In H&E stained section, coronal portion of the root lined by a thin layer of enamel and below that normal dentinal tubules can be seen. A crescent shaped obliterated pulp chamber was noted containing eosinophilic round substance resembling dentinoid. A layer of normal dentinal tubules can be seen to some extend immediately below the pulpal remnant. At some areas vascular spaces resembling pulp chamber can be seen. Radicular portion surrounded by cementum at the periphery and apically it is lined by osteodentin. Periodontal ligament can be seen lining the extra radicular portion of the tooth.

On the basis of clinical, radiographic and histological finding a diagnosis of DD type Ib was confirmed.

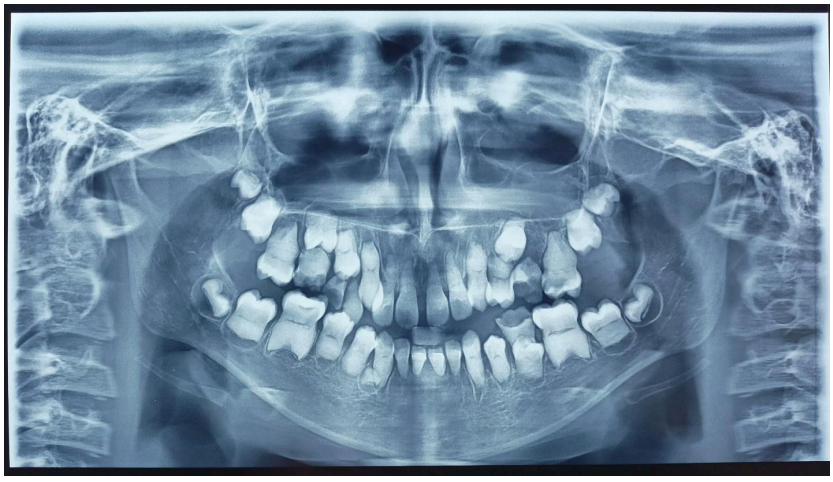

Fig. 1: Extra oral photograph of patient

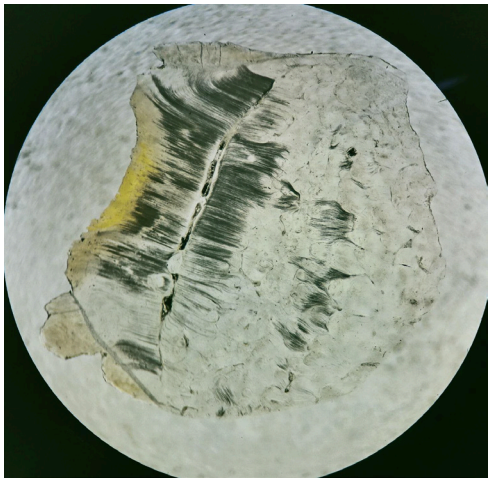

Fig. 2: Intra oral photograph showing rotated upper central incisor with a slight yellowish hue.

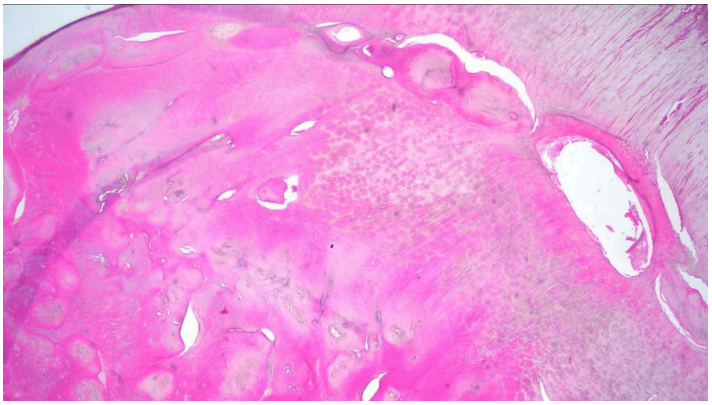

Fig. 3: Orthopantomogram showing generalized short and malformed roots with obliterated pulp chambers in case of anteriors and crescent shaped pulpal remnants in posteriors.

Discussion

Dentin dysplasia is a rare inherited dominant hereditary disturbance mainly affecting dentin formation. It manifests as short rooted tooth with tooth mobility and early exfoliation and there is frequent periapical cyst formation. In most of the cases pulp is obliterated. Enamel formation is normal in these cases. Our patient has a positive family history, her father and one of her sibling is being affected.

DD can affect both primary and permanent dentition with an Incidence of one patient in every 100,000. Researchers recently discovered three pathogenicity genes in case of DD I and the genes were VpS4B, SSUH2 & SMOC2 found in three affected family from different countries[5]. Mutation of DSPP gene mapped in chromosome 4q 21.3 encodes the dentin sialophosphoprotein, a major non collagenous protein is responsible for isolated cases of dentinogenisis imperfecta II, III and dentin dysplasia II[5].

When reviewing the literature two main theory of pathogenesis are proposed which lead to the defect in root formation. Logan et al proposed that it is the dental papilla that is responsible for the abnormalities in the root development, suggested multiple degenerative foci within the papilla become calcified, leading to reduced growth and final obliteration of the pulp space[6]. Wesley et al proposed that the condition is caused by an abnormal interaction of odontoblast with ameloblast leading to abnormal differentiation and function of these odontoblast[7].

This condition was first described as rootless teeth in1920 by Ballschmide but it was Rushton in 1939 termed the condition dentin dysplasia[8]. In 1973 Shield classified DD into type I (dentin dysplasia) and type II (anomalous dysplasia of dentin) .Witkop referred it as radicular dentin dysplasia as type I and coronal dentin dysplasia for type II. Type III dentin dysplasia occurs least frequently with the symptoms of both type I and type II affecting only permanent dentitions.

Scola and Watts proposed a sub classification of DD I as total DD I characterized by the presence of teeth with significantly narrowed or obliterated pulp spaces and permanent teeth with short roots. Subtotal DD I characterized by permanent teeth with root of intermediate length[9]. Carroll et al subclassified dentin dysplasia into four types[10]. In type 1a, there are no pulp chambers and root formation, and there are frequent periapical radiolucencies; type 1b has a single small horizontally oriented and crescent shaped pulp, and roots are only a few millimeters in length and there are frequent periapical radiolucencies; in type 1c, there are two horizontal or vertical and crescent shaped pulpal remnants surrounded by a central island of dentin and with significant but shortened root length and variable periapical radiolucencies; in type 1d, there are visible pulp chambers and canal with near normal root length and large pulp stones that 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. Our patient exhibit the classical feature of type Ib .

Clinically in DD I, teeth exhibit extreme mobility and gradually premature exfoliation as a result of conical or absent root. But in our case these features were not present and also gave a history of retained deciduous tooth and ectopic eruption of permanent dentition. Other frequent symptoms and complaints such as delayed dental eruption pattern, opaque incisal margin, spontaneous exfoliation and discomfort caused by severe tooth mobility especially after meals were also noted in some cases.

Radiography reveals that the roots are short, blunt, conical or similarly malformed in both the dentitions. In the deciduous teeth the pulp chambers and root canals are usually obliterated, while in the permanent dentition, a crescent shaped pulpal remnant may still be seen in the pulp chamber. This obliteration in the permanent teeth commonly occurs preeruptively. Periapical radiolucencies representing granulomas, cysts or abscesses involving otherwise intact teeth[11].

Histologically a portion of coronal dentin is usually normal. Apical to this may be areas of tubular dentin, but most of that which obliterates the pulp is calcified tubular dentin, osteodentin and fused denticles. Normal dentinal tubule formation appears to have been blocked so that new dentin forms around obstacles and takes on the characteristic appearance described as lava flowing around the boulders. Electron microscopic studies suggested that this pattern of cascades of dentin results from repetitive attempts to form root structure. Interestingly, the dentin itself is histologically normal but is simply disoriented. These findings were consistent with our present case[12]

Management of DD has always been a challenging task to the dentist. Follow up and routine conservative treatments are often advised. Maintenance of a good oral hygiene for the survival of mobile tooth is also recommended. Endodontic treatment is contraindicated in teeth with obliteration of root canals and pulp chambers. Periapical surgery and retrograde filling is recommended in some cases. Orthodontic treatment is suggested however further resorption of roots, loosening of tooth, and premature exfoliation may occur[12]. Successful oral rehabilitation with complete denture after exfoliation of all teeth is proposed as a main treatment modality[13].

Conclusion

DD is a rare hereditary disease with a low incidence rate. It often leads to premature exfoliation of primary and permanent teeth. Early diagnosis and intervention is important to prevent or delay the loss of dentition. Meticulous oral hygiene practice and effective treatment modalities must be established for the retention of the teeth. So dentist play a major role in educating the patient as well as family members regarding the condition and the importance of early intervention.

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