A case of multiple rootless teeth: A case report and review

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Abstract

Dentin dysplasia is a rare, hereditary disorder affecting the dental hard tissue. It is an autosomal dominant disease of unknown etiology that affects 1:100,000 populations. It may present as affecting only the dental hard tissue or as one of the symptoms of underlying diseases such as calcinosis, Ehlers–Danlos syndrome, rheumatoid arthritis, Vitaminosis D and Branchioskeletogenital syndrome. This was first described by Ballschmiede as rootless teeth in 1920 and termed as dentin dysplasia by Rushton in the year 1939. It is classified into Type I, II and III, in which Type III affects only the secondary dentition. This article reports a rare case of Type I dentin dysplasia in a 26-year-old male patient, and focus on clinical, radiological, ground section and histopathological aspects. It emphasizes the significance of early diagnosis and intervention for the psychological well-being of the individual.

Keywords: Dentin, dysplasia, rootless teeth, tooth anomaly

INTRODUCTION

Dentin dysplasia is a rare hereditary disorder affecting the dental hard tissue. It is an autosomal dominant congenital disease that may present as such affecting only the dental hard tissue or as one of the symptoms of underlying disease such as calcinosis, Ehlers–Danlos syndrome and the branchioskeletogenital syndrome.

Dentin dysplasia occurs due to the mutation of chromosome 4q13–21 and thus resulting in the disturbance of dentin formation. In addition to the above, recent research suggest that there is mutation of DSPP gene and dentin matrix acidic phosphoprotein gene that may occur during the incorporation of Hertwig’s epithelial cells in to the dental papilla. All these findings could suggest dentin dysplasia to be a disease of mixed phenotype.

This condition was first described by Ballschmiede in 1920 as rootless teeth and it was termed as dentin dysplasia by Rushton in the year 1939. It is estimated that the occurrence of this condition is very rare accounting 1:100,000 cases. Both the genders are affected at the same frequency.

Dentin dysplasia is classified by Shields and his associates into Type I and Type II and it was Witkop referred it as...
Radiographic examination is mandatory. In contrary, Sauk et al. have classified Type I dentin dysplasia into four subtypes Ia, Ib, Ic and Id based on the progression of underdevelopment of the root and the pulp chambers and root canals.

- **Type Ia** - No pulp chamber and root formation with frequent periradicular radiolucencies
- **Type Ib** - A single small horizontally oriented and crescent shaped pulp and roots that are only few millimeter in length with frequent periapical radiolucencies
- **Type Ic** - Consist of two horizontal or vertical crescent shaped pulpal remnants surrounding a central island of dentin, with significant but shortened root length and variable periapical radiolucencies
- **Type Id** - Visible pulp chamber 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.\[1,15\]

In the case of Type II dentin dysplasia, the symptoms are identified in both the dentition. The crown of the primary dentition shows normal morphology with notable color change as amber or blue color. The crowns can be easily abraded; however, the roots are of normal length and shape.\[1,2,6,17\] The pulp chambers may become obliterated. The permanent dentition exhibits slight change in color with less susceptibility to abrasion, abnormally large pulp chambers often described as “thistle-tube” appearance.\[18\] Both the dentitions are not affected by caries and does not show any premature loss of teeth. The radiographic image shows the appearance of so-called shell teeth.\[11,12\] The occurrence of multiple intrapulpal calcification may also be observed.\[3,19\]

**CASE REPORT**

A 26 year old male patient reported to the private dental clinic with a chief complaint of multiple mobile teeth. Patient also complains about exfoliation of teeth without any pain. Past dental history reveals that all his deciduous teeth were mobile and exfoliated prematurely. There is no relevant family history. The teeth that were present on clinical examination are shown [Figure 1]. The roentgenographic findings reveal multiple rootless teeth with periapical radioluencies [Figure 2]. Based on the above findings, the case was diagnosed as dentin dysplasia Type I. The treatment plan was explained to the patient and it was initiated. Total extraction was done and the patient was advised for implant supported prosthesis [Figure 3]. The extracted teeth were subjected to the histopathological examination and the diagnosis was confirmed.

**DISCUSSION**

Dentin dysplasia is a rare hereditary autosomal dominant disorder of unknown reason. The possible etiology could be the dental papilla that causes the abnormal root development. The multiple degenerative foci within the papilla become calcified, leading to reduced growth and final obliteration of the pulp space.\[20\] In contrary, Sauk et al. suggested that it was the epithelial root sheath not the dental papilla, that affects the root development. This sheath invaginated too early induced the ectopic dentine formation in the pulp space. However, the most widely accepted etiology as proposed by Wesley et al. is the abnormal interaction of odontoblasts with ameloblasts leading to abnormal differentiation and/or function of these odontoblasts.\[20,21\]

Diagnosis is made based on the history, clinical examination and mandatory radiographic examination. Patients usually report to the dentist with the complaints of pain and mobility of teeth. History of premature loss of deciduous teeth along with different grades of mobility as clinical findings and shortened roots with periapical radioluencies on X-rays suggests the diagnosis of dentin dysplasia type I. The above findings were well correlated to the present case to establish it as DD Type I involving the dentition only. The patient does not exhibit any systemic signs and symptoms which render us to rule out the systemic disorder, as DD Type I might associated with various systemic disorders.

The teeth are extracted due to pain that is caused by periapical granulomas and cyst.\[22\] There is reduction in the bone density at the root apices and it is not related to dental caries. The formation of periapical radioluencies is not clear. However, the probable mechanism could be the penetration of inflammation through the weaker areas of mineralization such as in the neck region of the tooth and or through the
marginal periodontium. Moreover, Hertwig’s Epithelial Root Sheath (HERS) by not reaching the normal root length, its activity is modified, resulting in cyst formation. The other hypothesis also proposed stating that the pulp undergoes “auto-intoxication” during its obliteration.

Histological features of DD type I includes normal enamel and subjacent dentin. Deeper layers of dentin show an atypical

Figure 1: Intra oral image of the patient

Figure 3: Extracted teeth

Figure 4: Steromicroscopic view of incisor and premolar. Ground section of the permanent teeth affected by dentin dysplasia Type I shows globules of irregular dentin showing the features of lava flowing around the boulders and obliteration of pulp chamber

Figure 5: Light microscope (LM) examination of teeth specimens (a and b) Ground section of permanent tooth affected by dentin dysplasia-Type I. Showing normal appearing enamel, globules of dentin showing features of lava flowing around the boulders (×4)

Figure 6: Histological examination of the decalcified teeth showed irregularly placed globules of dentinal structures exhibiting dentinal tubules and obliteration of the pulp chamber

Figure 2: Panoramic radiographic examination revealed multiple rootless teeth with periapical radiolucencies
tubular pattern with irregular organization. Pulpally, normal appearing mantle dentin and globular or nodular masses of abnormal dentin are seen.\[^{23}\] The stromemicroscopic and light microscopic ground section view of the present case shows globules of irregular dentin exhibiting the features of lava flowing around the boulders with minimum amount of dentinal tubules formation and interglobular dentin seen subjacent to enamel [Figures 4 and 5]. The enamel is normal and structures such as enamel lamellae, spindles and cracks are also seen. The H&E-stained decalcified teeth reveals irregularly placed globules of dentinal structures exhibiting dentinal tubules and obliteration of the pulp chamber [Figure 6].

Treatment and management of the patient with dentinal dysplasia is quite difficult. Extraction has been suggested as a treatment of choice for the teeth with pulp necrosis and peri-apical abscesses. However, most of the patients with dentin dysplasia reported in the literature are at younger age as like in our case. Alternate treatment of choice including early diagnosis, awareness of the disorder among the pediatric dentist, pediatricians, physicians and conservative treatment might support the psychological well-being of the individual. Teeth that are with reasonable root length can be treated with peri-apical surgery and retrograde filling.\[^{3}\] In patients where early exfoliation of the teeth has happened can be managed with a combination of onlay bone grafting and a sinus lift technique that can accomplish implant placement successfully.\[^{3}\]

CONCLUSION

In Conclusion Dentin Dysplasia is a rare, inherited abnormality of dentin that may lead to early exfoliation of the teeth at a younger age. Early diagnosis of the condition and comprehensive understanding of the disease might help the overall well being of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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