Dentin Dysplasia Type I: A Rare Case Report and Management Protocol

Shimasadat Miri1, Mahsasadat Miri2, Parisa Soltani3*, Amirhossein Moaddabi4

1Department of Endodontics, School of Dentistry, Kermanshah University of Medical Sciences, Kermanshah, Iran
2Department of Obstetrics and Gynecology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran
3Department of Oral and Maxillofacial Radiology, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, Iran
4Department of Oral and Maxillofacial Surgery, School of Dentistry, Mazandaran University of Medical Sciences, Sari, Iran

*Corresponding author: parisa.soltani@live.com

Abstract Dentin Dysplasia (DD) is a rare autosomal dominant anomaly that disturbs the formation of dentin in primary and/or permanent dentitions. This condition is classified into types I (radicular) and II (coronal) based on radiological findings. A case of DD type I in an 11-year old Iranian boy is presented and the clinical and radiological findings and selected treatment plan are discussed.

Keywords: dentin dysplasia, radicular, pediatric dentistry

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1. Introduction

Dentin Dysplasia (DD) is a rare autosomal dominant anomaly that disturbs the formation of dentin either in the primary or both primary and permanent dentitions. Prevalence of this condition is reported to be approximately 1 in 100,000 patients [1]. Ballschmiede was the first one reporting this malformation as ‘rootless teeth’ in 1920 [2]. In 1939 Rushton suggested the term DD for this condition [3]. In 1972 Witkop classified DD into two types based on radiological findings: type I (DDI: radicular dysplasia) and type II (DDII: coronal dysplasia) [4].

DDI is characterized by presence of primary and permanent teeth with morphologically normal or slightly amber colored crown with no or only rudimentary root development, improper growth of dentin resulting in incomplete or total obliteration of the pulp chamber, and periapical radiolucent areas or cysts which can lead to early loss of tooth [5,6,7]. Although delayed eruption of teeth is reported in DDI, generally teeth erupt in the normal time [8]. In DDI teeth appear to have greater resistance to caries than normal teeth [9]. In histologic examination, the enamel and immediately subjacent dentin appear normal. Deeper layers of dentin show atypical tubular patterns with amorphous, atubular areas and irregular organization. Globular or nodular masses of abnormal dentin are seen pulpally to the normal appearing dentin [10].

The present report describes a case of DDI in an 11-year-old boy attending a private office in Iran.

2. Case Report

An 11-year-old boy attended to a private office for dental examination. Extraoral examination and medical history did not indicate any abnormal finding. Intraoral examination revealed poor oral hygiene of the patient, class I occlusion, and gingivitis. The teeth did not show pathological mobility or infection or abnormal appearance. Figure 1 depicts the intraoral photograph of the patient. Teeth number 15 and 31 had caries and the upper canines were not completely erupted. Radiographic examinations revealed no cystic or radiolucent lesion. Panoramic and intraoral radiographs of the case are presented in Figure 2. All teeth had short and malformed roots with closed pulp chambers. The patient was diagnosed with DDI based on clinical and radiologic examinations. This condition was not observed in other family members of the patient.

Figure 1. Intraoral photograph of the patient
3. Discussion

As mentioned DD is a disturbance in dentin formation. The etiological and pathogenesis basis of DD is not exactly determined yet. Extracellular matrix of the dentin is the result of the differentiation of neural crest ectomesenchymal cells into odontoblasts. Odontoblasts express specific genes products which form the collagenous dentin extracellular matrix. This matrix consists of mostly type I, type I trimer, type III, type V, and type VI collagens and several noncollagenous proteins also found in bone extracellular matrix, such as osteonectin, osteocalcin, osteopontin, bone sialoprotein, and dentin matrix protein 1. However, two dentin matrix proteins, dentin sialoprotein (DSP) and dentin phosphoprotein (DPP), are expressed by odontoblasts and transiently by ameloblasts [11]. These noncollagenous proteins are believed to be essential for initiation and control of mineralization in the transition of predentin to dentin [12]. DSP and DPP are cleavage products expressed from a single transcript encoded by dentin sialophosphoprotein (DSPP) gene on human chromosome 4 [13]. Mutations in this gene are detected in several disorders of dentin formation process including DD and dentinogenesis imperfecta [14,15,16]. Wesley et al suggested that abnormal interactions of odontoblasts with ameloblasts resulting in abnormal differentiation and function of odontoblasts can cause DD [17]. Logan et al commented that multiple degenerative foci in the dental papilla become calcified and it can cause obliteration and reduced growth of the pulp space [18]. However Sauk et al proposed that early invagination of the epithelial root sheath can induce ectopic dentin formation in the pulp space and can obliterate the pulp chamber [19].

Coronal morphology in DDI is usually normal [5,20,21]. In our case the teeth had normal size and shape but were slightly amber colored. Moreover, malocclusion is not a striking feature in DDI [22]. However, it can be present in some cases. Our patient had Class 1 occlusion with posterior open bite.

Management of DD patients can be problematic in some cases. Follow up and routine conservative treatment is the desired treatment plan in DD cases [23]. Extraction of infected teeth with periapical abscess can be considered as the final choice [24]. Teeth with relatively long roots can undergo endodontic treatment [9] if the pulp chamber is not obliterated. Early exfoliation of teeth and associated bone loss can necessitate techniques such as onlay bone grafting and sinus lift for placement of dental implants in adults [25]. In our case oral hygiene improvement was suggested to prevent from further dental complications. Restoration of teeth with carious lesions was considered. As endodontic treatment of tooth number 15 was not possible due to pulpal obliteration, it was sealed with MTA and restored. One-year follow-up revealed no complications in this case.

4. Conclusion

Dentin dysplasia type I is a rare genetic anomaly in dentin formation which can lead to early exfoliation of
primary and/or permanent dentition. Early diagnosis and preventive measures to avoid further complications is of considerable importance in management of patients with DDI.

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