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Dentine Dysplasia Type II: Report of a Rare Case with Radiographic Criteria of Type I

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Abstract Dentin dysplasia (DD) is a hereditary malformation of dental tissues affecting the dentin during the process of development. The teeth appear clinically normal; however, radigraphically the pulp morphology is abnormal and may be large or obliterated. The roots are diminutive, blunted, deformed or even absent. Periapical radiolucencies may be present without an apparent cause. The affected teeth may exhibit great mobility and exfoliate early even with minor trauma. In this article, we reported clinical and radiographic findings of a rare case of DD that have radiographic features of both DD- type II and DD type I in a 24 years old male patient visited the Out-Patient Dental Clinic of Qassim University, KSA.

Keywords: *dentin dysplasia, hereditary, abnormalities*

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1. Introduction

Dentine Dysplasia (DD) is a rare autosomal dominant hereditary disease affecting primary or mixed dentitions. The incidence of this rare disorder is approximately 10 times less than that of dentinogenesis imperfecta (DI), with an affected patient in every 100,000 individuals [1,2]. This hereditary disease is characterized by disturbance in formation of dentin during tooth development, but does not affect enamel formation [3]. In 1920, this condition was first described as rootless teeth by Ballschmiede [4] who described 7 cases in same family who have short blunt-rooted teeth, hypermobility and subsequent teeth exfoliation. The name DD was introduced by Rushton in 1939 [5]. The first classification of DD was done by Shield et al in 1973 into Typ1 DD (Dentine Dysplasia) and Type 2 (Anomalous dysplasia of dentin), which was confusing during that time [6].

The recent classification of DD was done by Witkop, in1974 who classified DD according to the affected part of the tooth into Radicular Dysplasia, DD type I (DD-I,) and Coronal Dysplasia, DD type II (DD-II) [7].

DD-I affects the roots of both deciduous and permanent dentitions. The roots in DD-I are short, blunt, conical, malformed or even absent. Periapical radiolucencies representing granuloma, abscess or cyst may be present. In permanent teeth, atypical growth of dentin in the pulp chamber leading to decreased pulpal cavity space and incomplete or total obliteration of the pulp chambers [4].

DD-II affects the coronal pulp and characterized by a large pulp chamber. It is described as 'thistle-tube appearance' which means a large pulpal chamber with some extension to a thin root canal. In addition, formation of multiple pulp stones in permanent teeth and obliteration of deciduous teeth are also documented in DD-II [8].

DD-I is further classified into 4 sub categories; DD-Ia, DD-Ib, DD-Ic, DD-Id based on distinctive criteria on radiograph [9]. In DD-1a, there is complete obliteration of pulp chambers and no root development with many periapical radiolucent areas. In DD-1b, the pulp showed a horizontal, crescent shaped, radiolucent remnants and few millimeters of root development with many periapical radiolucent areas. DD-Ic has two horizontal, crescent shaped, radiolucent lines and significant but incomplete root development, with or without periapical radiolucent areas. In DD-Id pulp chambers present with oval pulp stones in the coronal third of the canal. Bulging of the canal around the stone in a normally formed root is usually discernible, with few, if any, periapical radiolucent areas.

Dentin dysplasia type III includes a combination of teeth of type I and type II in the same patient. It was proposed by Ciola et al. [10] as a third group of DD classification. The aim of this study is to report a rare case that possess features of both types of DD and shed highlights on clinical and radiographic findings of such condition.

2. Case Report

A 24 years old male patient visited the Out-Patient Dental Clinic of Qassim University, KSA seeking a construction of a fixed dental prosthesis for a missing anterior tooth for an esthetic reason. The medial history revealed that the patient was healthy and free from any systemic diseases.

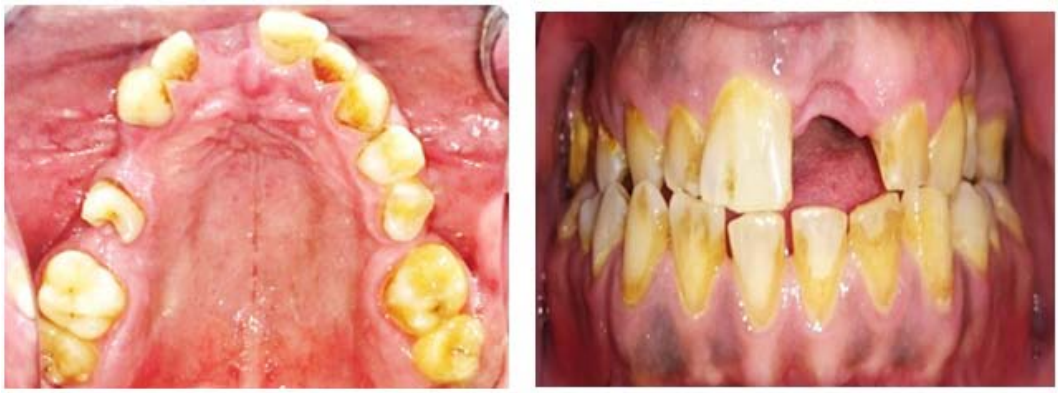

Figure 1. Clinical photograph showing the missing teeth in the upper jaw

On clinical examination, there were missing upper left central incisor, premolar and 1st molar teeth and upper right 1st molar tooth. The oral hygiene was bad with many carious teeth including upper left 2nd premolar, Upper canine and lower right 2nd molar (Figure 1). The crown of the teeth are proportionally normal in size except for the right central incisor which was larger than normal.

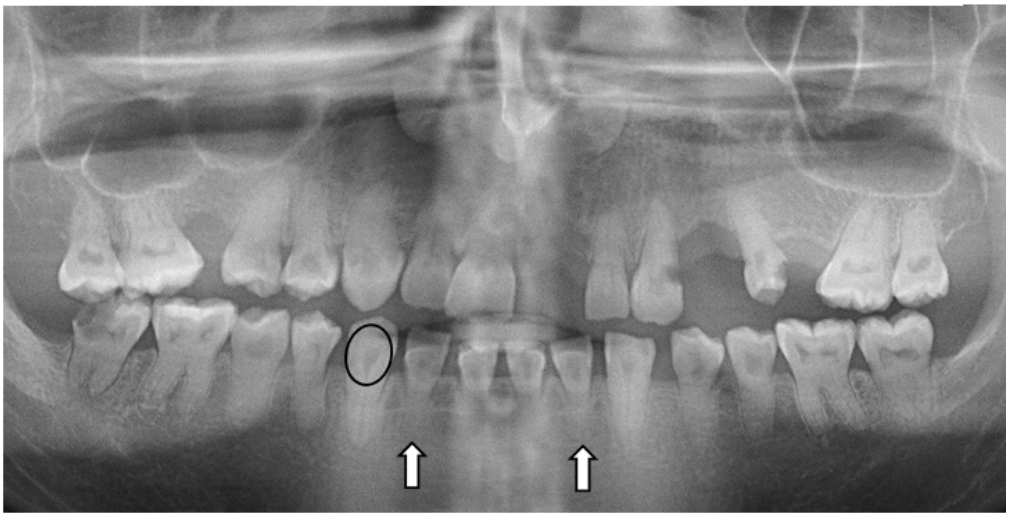

Figure 2. Panoramic radiograph showing short blunted or conical roots of teeth and large pulp chambers

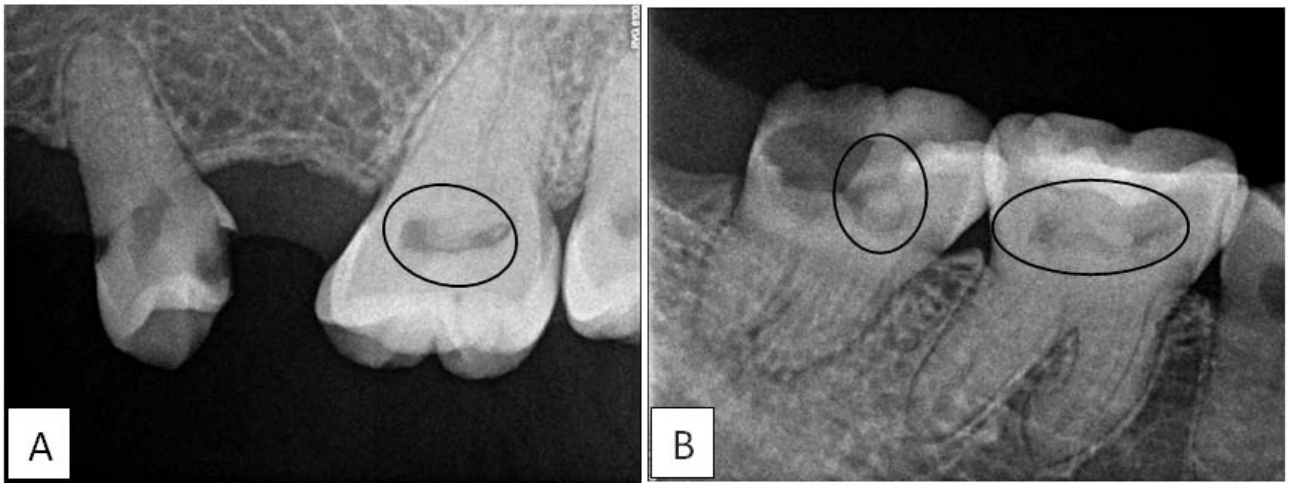

Figure 3. Periapical radiograph of the A) upper left molars and B) lower right molars showing pulp chambers occluded with radiopaque globules and reduction of pulp size

3. Radiographic Findings

Periapical and panoramic x-ray films were performed in order to evaluate the remaining teeth for construction of prosthesis. Radiographic examination of OPG revealed large pulp chambers (thistle shaped appearance) in some of teeth, which was evident in single rooted teeth. Other teeth were obliterated with radiopaque globules (denticles). The roots of some teeth are short, blunted or conical in shape, appearantly in lower incisors (Figure 2). In

periapical radiographs of upper molars (Figure 3A) and lower molars (Figure 3B), there were large radiopaque globules obliterating the pulp chamber and decreasing the sized of root canals. In periapical radiograph of upper central and lateral incisors, the pulp chambers were large extending to radicular pulp giving the '**thistle tube appearance**'. Radiopaque globules were observed within the pulp chamber leaving remnants of pulp spaces (Figure 4). Based on the clinical and radiographic appearance, a diagnosis of DD-II with some features of DD-I was rendered.

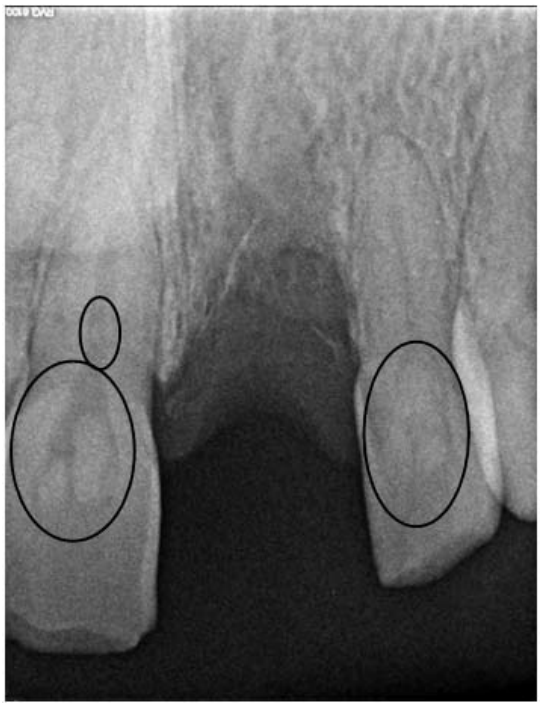

Figure 4. A periapocal radiograph of upper central and lateral incisors showing large pulp chambers (thistle-tube appearance) containing oval shaped radiopaque pulp globules

4. Discussion

DD and DI are developmental autosomal dominant conditions affecting dentine formation. DD is a rare condition when compared to DI with an incidence one-tenth that of DI [1,2,11]. The genetic mutations for DD- II and Non-syndromatic DI type II and III were reported in dentin sialophosphoprotein gene (DSPP) which is carried on chromosome 4q21.3 encoding the major non-collagenous proteins. This gene is involved in initiation and growth of crystals of hydroxyapatite and the process of mineralization [11]. Genetic disturbance in DD-I have not been clarified yet [11,12,13].

It was reported in one study a genetic mutation in Sparc Related Modular Calcium Binding Protein-2 gene (SMOC-2) in 2 affectedfirst cousin relatives' in Turkish family who have DD-I associated with microdontia and malformed teeth [14].

Although DD-I is inherited as an autosomal dominant disorder, a rare cases of autosomal recessive mode of inheritance were reported in two separate studies [13,14]. The first study of DD-I was reported in sons and daughters of healthy parents who were relatives [14]. The second study of an autosomal recessive case of DD-I was reported in 4 affected individuals born from a healthy Moroccan parents who were also relatives [15].

There are several theories in the previous dental literature regarding the pathogenesis of DD, however, the exact etiology need to be elucidated [16,17]. The theory proposed by *Logan et al* [16] suggested that foci of degenerative changes occur in dental papillae leading to delayed growth and abnormal calcification with subsequent obliteration of the pulp chamber. Another theory suggested by *Wesley et al* [17] proposed that DD is due to an abnormal differentiation and/or function of odontoblasts resulting from unusual interaction between odontoblasts and ameloblasts. Moreover, it was suggested that ectopic dentin is formed within the dental papilla either due to the occurrence of an earlier ingrowth of the radicular epithelial sheath [18] or due to displacement and proliferation of the internal cells of the developing dental organ in the dental papilla [7].

Microscopically, the enamel and the immediately subjacent dentin appear normal. Deeper layers of dentin, however, show atypical atubular patterns, with amorphous and irregular organization that lacks tubules. On the pulpal side of the normal-appearing mantle of dentin, globular or nodular masses of abnormal dentin are seen [1].

In the present case the teeth appeared clinically normal in shape and size except for the crown of upper 1st incisors which appears lager than normal. The radiographic findings include atypical pulp morphology that extended to radicular pulp. These changes were evident in single rooted teeth. Presence of radiopaque globules within the pulp chambers supports the diagnosis of DD-II (coronal type). However, other findings in the present study such as short, blunted or conical roots of lower incisors and obliteration of pulp chamber are features of DD-I (radicular dysplasia). Such findings supported our diagnosis of DD-II which also possess radiographic features of DD-I. To the best of our knowledge, this is the second case in English literature. The first case was reported in 1978 by Clio, et al, which was in accordance with our findings [10]. This report suggested that DD type III may be considered for future classification of DD. Further studies are recommended in order to get more features of such cases.

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