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Case Report

Type I Dentinal Dysplasia—Proposal of A New Classification System Combined with A Variant Case Presentation

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ABSTRACT

Dentine dysplasia is an extremely rare autosomal dominant hereditary condition primarily characterized by defective dentine formation affecting both the deciduous and permanent dentitions. The aetiology of dentinal dysplasia remains imprecise to date, in spite of the numerous hypotheses put forward by various authors. Also, of late, new case reports of type I dentine dysplasias are emerging with clinical and radiographic findings which are unique and variant from the classical characteristic features of this anomaly, rendering it difficult to fit into the previously established classification systems. In this article, after a thorough review and understanding of the drawbacks of the previous classification systems and the cumulative findings from the published case reports in literature, we propose a new classification system for dentine dysplasia type I. We also present a case report which exhibited an absence of familial hereditary pattern, the absence of periradicular radiolucent lesions and osseous pathologies as well as atypical morphological defects of the molar roots which were diverse from the classical findings of the various sub types of dentine dysplasia type I reported to date. Early diagnosis and implementation of preventive and curative therapy is crucial for avoiding premature exfoliation of deciduous and permanent dentition and associated residual ridge resorption, thereby overcoming functional and aesthetic deficits. The new classification proposed in this article will help clinicians to diagnose and categorize the patients effectively, facilitating proper treatment and supportive care for all.

Keywords: Classification systems, Dentine dysplasia, Radiograph, Periradicular lesions, Dental papilla

INTRODUCTION

Dentine dysplasia is an extremely rare autosomal dominant hereditary disturbance primarily characterized by defective dentine formation. The pathogenesis and contributing etiological factors of this atypical anomaly is still unknown in the dental literature. This condition affects approximately one in every 1,00,000 patients (Kim 2007). This pathology was first described by Ballschmiede in 1920 and later the

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condition was termed as dentinal dysplasia (Rushton 1939). Depending on the parts of the teeth affected, in 1975, Witkop classified dentinal dysplasia (DD into two broad categories, radicular dentine dysplasia as type I and coronal dentine dysplasia as type II. Type I DD is characterized by the presence of primary and permanent teeth with normal appearance of the crown portions but absent or rudimentary root development, incomplete or total obliteration of the pulp chamber and periapical radiolucent areas or cysts. Type II DD is characterized by deciduous teeth with complete pulpal obliteration and brown or amber bluish discolouration of crown structures of teeth. The permanent teeth have a normal appearance or slight amber discolouration, the roots are normal in size and shape with a thistle tube shaped pulp chamber with pulp stones. A third type of dentine dysplasia or focal odontoblastic dysplasia with radiographic features of both DD I and DD II, has also been described (Eastman 1977). Later, four subtypes of DD I was identified (Neville 2008). In type 1a, there is no pulp chamber and root formation and there are frequent periradicular radiolucencies; type 1b has a single small horizontally oriented and crescent shaped pulp and roots are only a few millimetres in length and there are frequent periapical radiolucencies; in type 1c, there are two horizontal or vertical and crescent-shaped pulpal remnants surrounding a central island of dentine and with significant but shortened root length and variable periapical radiolucencies; in type 1d, there is a 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 peri-apical radiolucencies. In 1987, Scola and Watts proposed a subclassification 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 and subtotal DD I characterized by permanent teeth with roots of intermediate length. O' Carrol in 1991, presented another subclassification based on radiographic findings referring to the severity of pulp chamber obliteration, root length and periapical radiolucencies. Four subtypes were proposed as the first type presenting with total pulp chamber obliteration, no root development and multiple periapical radiolucencies, in the second and third types, the pulp chambers are less obliterated and present crescent shaped radiolucent areas, minimal root formation with less frequent periapical radiolucencies, in the fourth subtype, there are radiographically visible and delimited chambers, pulpal nodules found in the coronal third of the canal with the roots having significant development but no or few radiolucencies observed.

Also, an association between dentine dysplasia and osseous changes including sclerotic bone deposition were reported (Morris 1977) indicating the possibility of occurrence of bone related pathologies in some patients with dentine dysplasia type I. The diversities in the clinical and radiographic presentations of this disease in each and every affected patient has made it difficult to sub categorize the findings of each new case into the currently available classification systems. Taking into consideration the drawbacks of the previous classification systems and the cumulative findings from the published case reports, we propose a new classification system for dentine dysplasia type I in this article. We also present a case report which painted a mixed picture with classical and atypical findings of DD I which could not be fitted into any of the subtypes of the established classification systems.

CASE REPORT

A 17-year-old female patient reported to the Department of Periodontics, Faculty of Dentistry with a chief complaint of severe mobility of her upper and lower front teeth. This was the first dental visit for the patient and the past medical history was non-contributory. There was no relevant familial history of the disease and hence the patient was considered to be a first generation sufferer. Clinical intraoral examination revealed the presence of apparently normal colour and morphology of crowns of all the teeth. Upper and lower anterior teeth and all the four first premolars demonstrated grade III mobility on examination. The molar teeth were immobile. The patient demonstrated satisfactory oral hygiene and there were no other relevant intraoral

findings noted. Orthopantogram of the patient revealed rudimentary and nearly absent root structures in relation to the upper and lower anterior teeth and the premolars (Fig. 1).

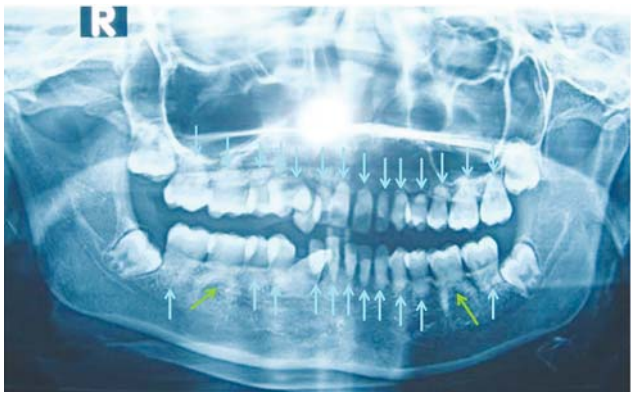


Figure 1: Orthopantamogram revealing the pathological changes. Blue arrows indicate the generalized rudimentary or nearly absent root structures. Green arrows indicates the strikingly flared inverted crescent-shaped appearance of roots of the mandibular first molars. Note the absence of periapical radiolucencies and osseous pathogies.

The roots of all the molar teeth showed defective morphology and the mandibular first molars demonstrated a strikingly flared inverted–crescent-shaped appearance of the roots (Fig. 2).

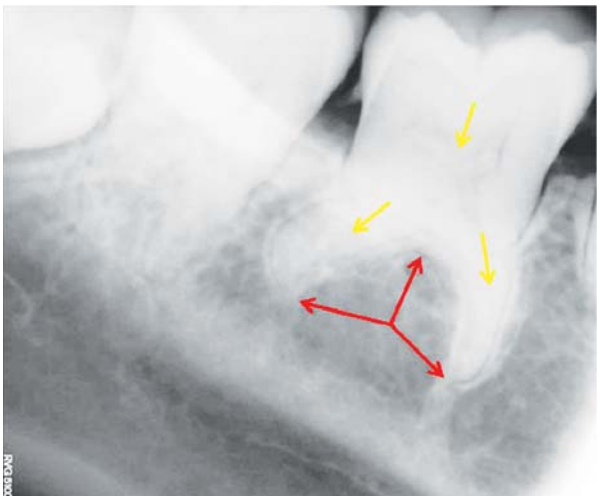
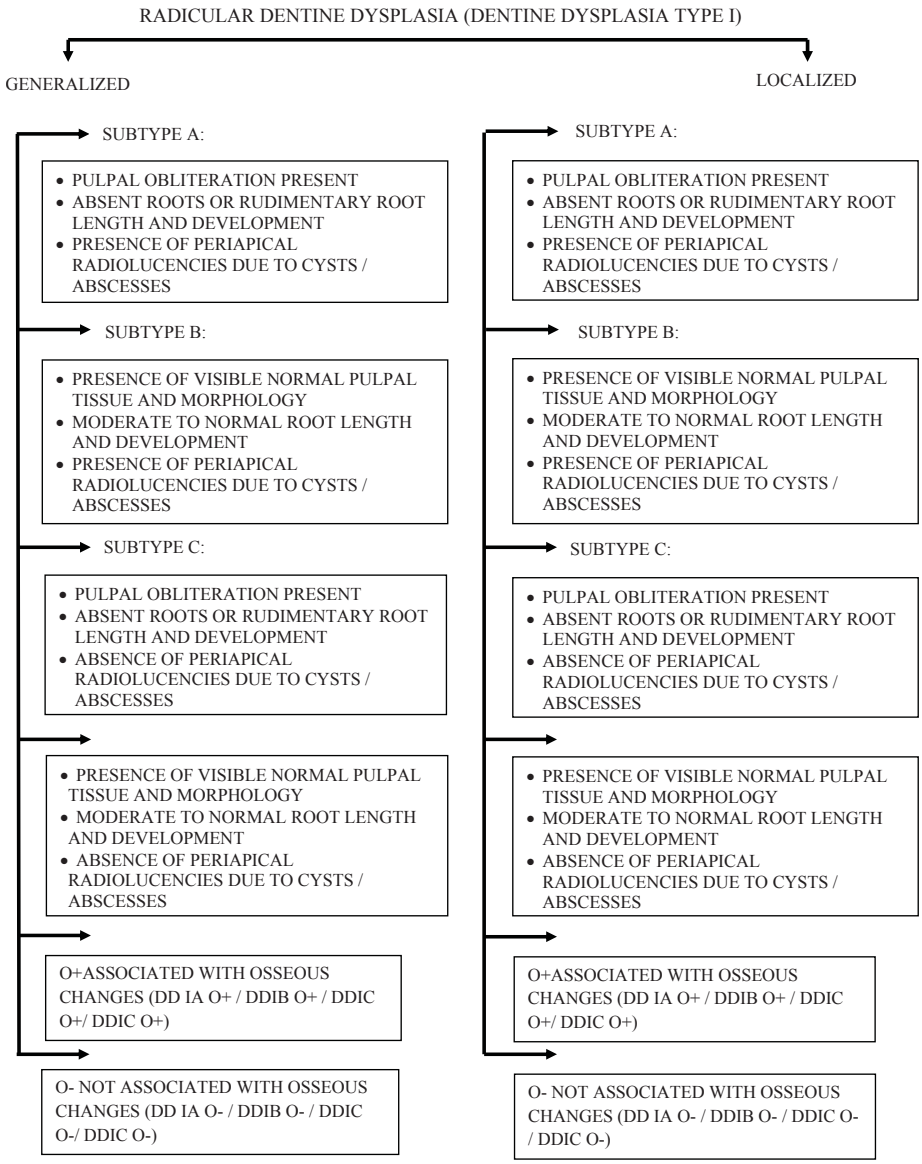


Figure 2: Red arrows indicates the strikingly flared inverted crescent-shaped root morphology in relation to mandibular first molar, Yellow arrow indicates the complete obliteration of pulps tissue in the pulp chamber and root canals.

There was a generalized total obliteration of pulp tissues within the pulp chambers and canals. An interesting feature was an absence of periapical radiolucencies and periradicular lesions. Also, there was no associated osseous changes or bone pathologies detected. Based on the clinical and radiographic findings of the disease, a diagnosis was established as dentine dysplasia type I. However, the findings of this case did not fit into any subtypes of the existing subclassifications of the disease. The observations of an absence of familial hereditary pattern, the absence of periradicular radiolucent lesions and osseous pathologies and the morphological defects of the molar roots were diverse from the classical findings of the various sub types of dentine dysplasia type I reported to date.

DISCUSSION

The aetiology of dentinal dysplasia still remains blurred in spite of the numerous hypotheses put forward by various authors. In 1962, Logan hypothesized that abnormal degeneration and calcification would occur in the dental papilla resulting in reduced growth and final obliteration of pulpal space. Sauk in 1972 suggested the occurrence of an earlier ingrowth of the radicular epithelial sheath resulting in ectopic dentin formation. Witkop in 1975 proposed that internal cells of the developing dental organ would be displaced and proliferate in the dental papilla producing ectopic dentin formation. According to Wesley in 1976, a failure in the interactions between odontoblasts and ameloblasts would occur, causing differentiation and/or abnormal function of the odontoblasts. Since there is no consensus in the dental literature regarding the aetiology of dentine dysplasia and also considering the rarity of this disease, case reports can serve as useful tools to elucidate and understand the different aspects of this condition. Dentine dysplasia is usually an autosomal dominant hereditary condition but in the present case report, the patient showed no signs of family heritage and these findings are in concurrence with three other reports (Cristiane 2011, Lida 2010, Vieira 1998). Also, in our patient, there was no report of formation of any painful, spontaneous abscesses and cysts and radiograph confirmed the absence of



Flowchart 1 New classification system for dentine dysplasia type I

periapical radiolucencies, which was consistent with the findings of only one other recent case report (Cristiane 2011). An association was also noted between dentine dysplasia and osseous changes including sclerotic bone deposition in a case report (Morris 1977) but the present case had no signs of any bone related pathologies. The diversities in the clinical and radiographic presentations of this disease in each and every affected patient has made it difficult to sub categorize the findings of each new case into the currently available classification systems. Taking into consideration the drawbacks of the previous classification systems and the cumulative findings from the published case reports, we propose a new classification system for dentine dysplasia type I as shown in Flowchart 1.

Proposed new classification

According to this classification, dentine dysplasia type I can be broadly classified into generalized and localized types depending on the percentage of the remaining dentition affected. If greater than 30 percentage of the dentition is affected, the condition is considered as generalized and if less than 30 percentage of the dentition is affected, then it is considered as a localized lesion. Each category is again subdivided into four subgroups namely subgroup A, B, C, D depending on the presence or absence of the clinical and radiographic features of the condition. Furthermore, O+ denotes the presence of associated osseous changes and O- denotes the absence of associated osseous changes, either of which should be added to the relevant subgroup categories depending on the patient findings. Hence, for example, if a patient presents with a generalized form of dentine dysplasia type I with osseous changes as well as findings of subgroup B, then the diagnosis is written as Generalized DD I BO+. Or, if a patient has a single tooth involvement with no osseous changes and characteristic features of subtype C, the diagnosis is made as Localized DD I CO- and so on.

CONCLUSION

Over the years, the management of patients with this rare genetic anomaly has presented dentists with numerous problems. This is mainly due to the variant forms of expressions of this disease among patients, making the diagnosis and treatment planning a challenge. The outcome of a diagnosis largely depends upon the age at which the diagnosis was made and the speed and quality of the treatment provided (Martin 2008). Early diagnosis and intervention is imperative for avoiding premature exfoliation of deciduous and permanent dentition and associated residual ridge resorption, thereby preventing further functional and esthetic problems. The new classification proposed in this article will help clinicians to diagnose and categorize the patients effectively, facilitating proper treatment and supportive care for the affected.

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