Case Report

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# Dentin Dysplasia Type I: A Rare Case

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## ABSTRACT

Dentin dysplasia is a rare autosomal dominant disturbance of dentin formation which results in either bulbous, discoloured crowns or early exfoliation of the teeth. It has been classified into two types; type I or radicular dentin and type II or coronal dentin dysplasia. Here we present a case of dentin dysplasia type I in a 12 year old patient highlighting the clinical and radiographic features.

KEY WORDS: Dentin dysplasia; Dentin; Rootless teeth.

## INTRODUCTION

Dentin dysplasia (DD) is a rare disturbance that affects dentin formation which is characterised by normal enamel but atypical dentin formation with abnormal pulpal morphology. Initially it was thought to be a single disease entity [1]. It was first categorised in 1939 [2]. The disease first described was that of Ballschmiede in 1920. Here, the spontaneous exfoliation of multiple teeth in seven children of one family was reported. This phenomenon was called 'rootless teeth' [2]. In 1939, Rushton published the first concise description of the disease. He also was the first to designate it as 'dentin dysplasia' [3].

Two major patterns have been described, i.e, type I and type II [1]. DD was separated into type I (dentin dysplasia) and type II (anomalous dysplasia of dentin) by Shields and his associates [4]. Later, Witkop classified this condition as radicular dentin dysplasia (type I) and coronal dentin dysplasia (type II) [5]. Type I has been found to be by far the more common type [2]. According to the definition, DD should have no correlation with systemic disease [1].

Radicular dentin dysplasia or dentin dysplasia type I has been often designated as rootless teeth, because there is loss of organization of the root dentin which leads to shortened root length. Coronal dentin dysplasia or dentin dysplasia type II has numerous features similar to dentinogenesis imperfecta. Here the root length is normal in both dentitions [1]. In type II DD, various forms of dental changes are seen which include bulbous crowns, cervical constriction, thin roots and early obliteration of the pulp [1].

Here, in this article we present a case of dentin dysplasia type I, highlighting its clinical and radiographic features.

## CASE REPORT

A 12 year old female patient reported to the department of Oral Medicine and Radiology with the chief complaint of mobility with respect to the upper and lower teeth since eruption. She said that due to the mobility of the teeth she had difficulty in chewing food. Upper and lower front teeth had fallen off due to extreme mobility. She also gave the history of premature exfoliation of the deciduous teeth due to mobility. Past medical history was non contributory. Patient was younger of the two siblings and the family history was non contributory.

On clinical examination, no abnormalities were detected in relation to soft tissue, 11 21 31 41 were missing and 26 36 46 were decayed. 12 22 32 42 showed grade II mobility and remaining teeth exhibited grade I mobility on palpation of the teeth. There was no associated pain/tenderness. The crowns of the teeth had normal morphologic characteristics but were slightly smaller in size and more yellow in colour than expected for a patient of her age (Fig 1, Fig 2).

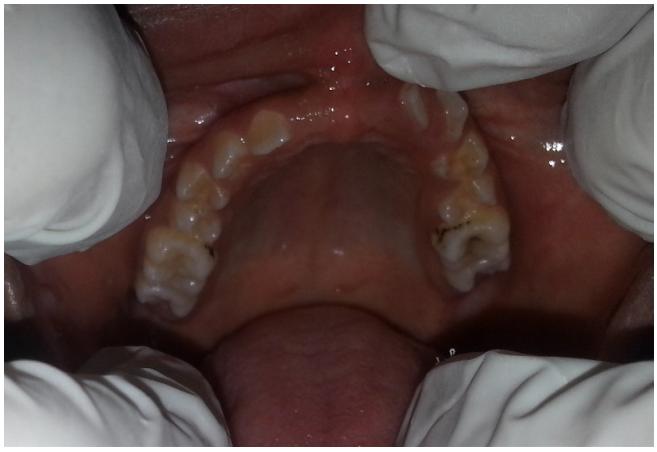

**Figure 1.** Intraoral view of maxillary teeth

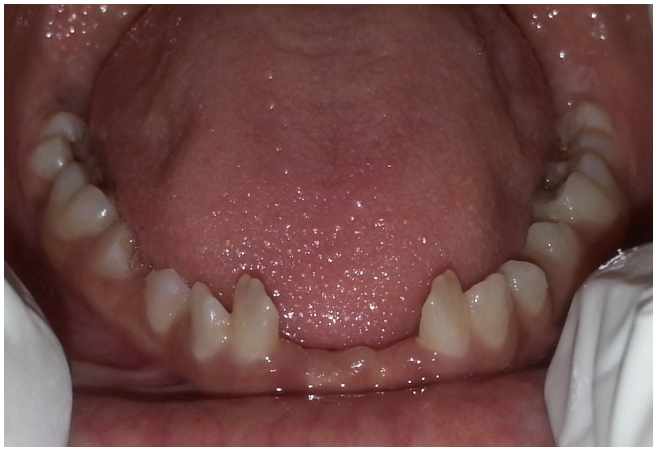

**Figure 2.** Intraoral view of mandibular teeth

Radiographic examination revealed pulpless teeth with no root formation in most teeth and roots of only a few millimeters in some teeth. A single, small horizontally oriented crescent shaped pulp was seen in relation to 16 17 26 36. Erupting tooth buds in relation to 18 28 38 48 were also seen (Fig 3).

Based on the clinical and radiographic features, a diagnosis of dentin dysplasia type I was given. As the patient is 12 year old, she has been kept on a long term follow up.

## DISCUSSION

Dentin dysplasia is an autosomal dominant disturbance of dentin formation [1,6]. It is characterised by normal enamel but atypical dentin formation with either mobile teeth or pain associated with spontaneous dental abscesses or cysts [1,6]. It is a rare anomaly which is seen in approximately one patient in every 100,000 [7]. Shields et al [4] classified it into type I (dentin dysplasia) and type II (anomalous dysplasia of dentin) and Witkop [5] separated DD as radicular dentin dysplasia (type I) and coronal dentin dysplasia (type II).

Both the primary and permanent dentitions are affected in Type I or radicular dentin dysplasia. Morphological appearance of the teeth appear normal. Occasionally there might be an exhibition of slight amber translucency [2]. The radicular dentin loses all organization and subsequently is shortened dramatically. The initial clinical signs that the patient presents with are due to shortened roots which are manifested as extreme tooth mobility and premature exfoliation, either spontaneously or secondary to minor trauma [1], which was seen in the present case. Radiographic presentation in both the dentitions is roots that are short, blunt, conical and malformed. Usually the pulp chambers and root canals are completely obliterated or a crescent shaped pulp remnant may be present in the pulp chamber [1,2]. Our case also presented with these classic features of type I DD.

Further, a subclassification for dentin dysplasia type I was proposed [1]:

DDIa- no pulp chambers, no root formation and frequent peripapical radiolucencies

DDIb- a single small horizontally oriented and crescent shaped pulp, roots only a few millimeters in length and frequent periapical radiolucencies

DDIc- two horizontally oriented and crescent shaped pulpal remnants surrounding a central island of dentin,

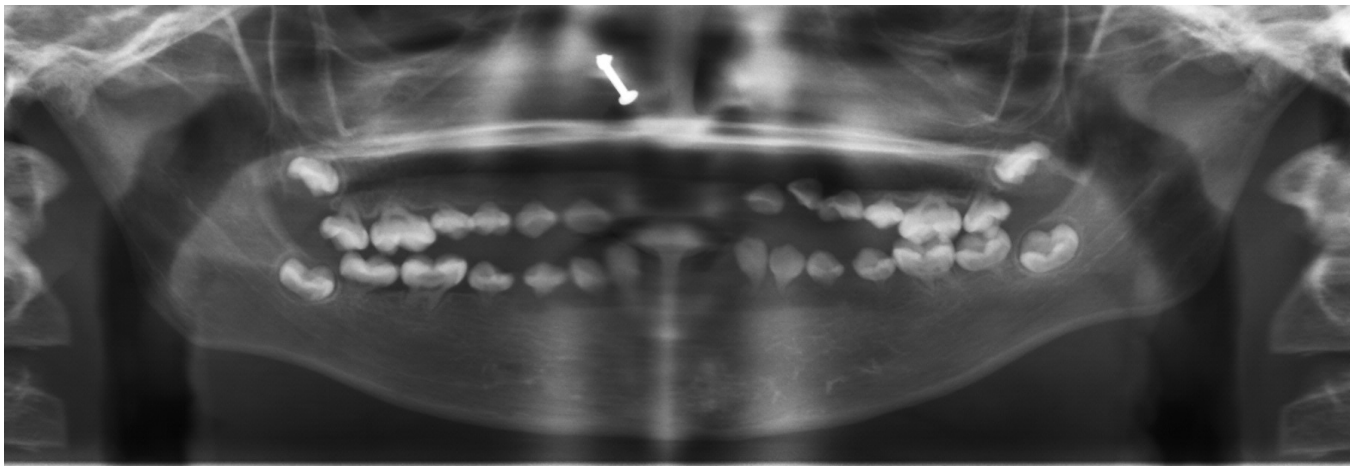

**Figure 3.** Panoramic radiography

significant but shortened root length, and variable periapical radiolucencies

DDId- visible pulp chambers and canals, near normal root length, enlarged pulp stones that are located in the coronal portion of the canal and create a localised bulging of the canal and root, constriction of the pulp canal to the stone, and few periapical radiolucencies

In the present case, based on the radiographic findings it can be considered as a combination of DD subtype Ia and subtype Ib.

Type II or coronal dentin dysplasia also affects both the primary or permanent dentitions. Here the teeth exhibit yellow, brown or bluish grey to amber brown appearance [1,2]. Radiographically, pulp chambers become obliterated or exhibit an abnormally large pulp chamber which is often described as thistle tube in shape. The root length is normal in both dentitions [1,2].

The etiology of DD is still unknown. It was suggested by Logan et al [8] that abnormal calcification of the pulp is related to degeneration of the dental papilla. Another theory was proposed by Wesley et al [9] that the condition is caused by an abnormal interaction of odontoblasts with ameloblasts thus leading to abnormal differentiation and/or function of these odontoblasts. It was postulated by Sauk and co-workers [10] that premature invagination of the epithelial root sheath is associated with abnormal dentin deposition.

In patients with DD type I, preventive care is of foremost importance [1]. Treatment and management of patients with dentin dysplasia has always been a challenge for dentists [6]. Patients have to be kept on continuous follow-up. Routine conservative treatment has to be the part of the treatment plan in DD [11]. Extraction has been advised as a treatment alternative for teeth with pulp necrosis and peri-apical abscess [6]. Another approach for the treatment of teeth with DD has included peri-apical surgery and retrograde filling, which is recommended in teeth with long roots [11,12]. Early exfoliation of the teeth is usually exhibited in these patients, which consequently leads to maxillomandibular bony atrophy. Dental implants should be considered when growth is complete. Ridge augmentation procedures need to be carried out prior to implant placement in maxillo-mandibular alveolar atrophy due to early loss of teeth. Treatment with a combination of onlay bone grafting and a sinus lift technique to accomplish implant placement can be used successfully [13,14,15].

## CONCLUSION

Dentin dysplasia type I is a rare hereditary disorder which results in early exfoliation of teeth. Early diagnosis of this condition is very important. Oral rehabilitation of these patients require elaborate treatment planning. Effective preventive treatment and strategies have to be implemented to help prevent early loss of teeth and prolong retention of the affected teeth.

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