Case Report

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DENTIN DYSPLASIA TYPE I: CASE SERIES AND REVIEW OF THE LITERATURE

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ABSTRACT:

Dentin dysplasia (DD) type I is a hereditary disturbance in dentin formation. In this anomaly, the teeth generally appear clinically unremarkable with normal crowns, and radiographically it is characterized by normal crown, absence or severe restriction of root formation, obliterated pulp chambers, and periapical radiolucencies without obvious cause of non carious teeth. The teeth characteristically exhibit extreme mobility and are commonly exfoliated prematurely of after only minor trauma .The purpose of this article is to report two cases of dentin dysplasia type I, review the relevant literature and emphasizing the peculiar aspects of this unusual anomaly.

Key words: Dentin dysplasia, rootless teeth, denticles, pulp obliteration.

INTRODUCTION:

Dentin dysplasia (DD) is a rare autosomal dominant hereditary disturbance of dentin formation, characterized by normal enamel formation, but atypical dentin with abnormal pulpal morphology.[1]The condition was first described by Ballschmiede [2-3] but it was Rushton [4] who termed the condition dentinal dysplasia. In 1972, Witkop [5] classified DD into two types, radicular DD as type I and coronal DD as type II. In type I, both the deciduous and permanent dentitions are affected. The crowns of the teeth appear clinically normal in morphology but defects in dentin formation and pulp obliteration are present

Dentin dysplasia type I is divided into four subtypes . Type 1a, there is a complete obliteration of the pulp and usually little or no root development and frequent periapical radiolucencies .[6-8]

Type 1b variation has a horizontal, crescent shaped, radiolucent line, which separates normal coronal dentine from abnormal radicular dentine. The roots are short, conical and rudimentary and frequent periapical radiolucencies.Type 1c variation shows two crescent–shaped horizontal radiolucent lines with their

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concavities toward each other at the cement enamel junction and the roots one half the normal length and variable periapical radiolucencies. Type 1d is characterized by normal root formation, which sometimes may be bulbous in the coronal 3rd. Within the pulpal canal " a stone" may be found ,in this type of DD, the pulp chamber is usually not obliterated and normal root formation occurs and very few periapical radiolucencies. It is not known if DD type I is another allelic disorder of the dentin sialophosphoprotein (DSPP) gene, or a mixed phenotype [9-10].The purpose of this article is to report two cases of dentin dysplasia type I ,highlighting the clinical and radiographic variations of the defect.

CASE DETAIL:

Case 1: A 17 years old patient reported with complaint of missing and mobile anterior teeth and wanted the teeth to be fixed (fig 1). Clinical examination revealed no sigh of gingivitis or periodontitis. There was grade three mobility of lower lateral incisors and lower central incisors were exfoliated spontaneously 5 month back (fig 3) . The radiographic analysis showed that the lower central incisors had no root at all and periapical radiolucency evident and lateral incisors has small rudimantory roots (fig 2) . It appeared as though the crowns of the teeth were placed directly on the bone without any deeper anchorhage. The patient's medical history revealed no evidence of disturbance in general health. The dentition was free of caries, and the patient's oral hygiene was fair with mild plaque. Her family history revealed that there was no such abnormality in other family members except for patient's father ,her father reportedly became edentulous at an early age and had required full maxillary and mandibular dentures.

Case 2: A 20 years female patient reported in our department complaing of loose teeth and asking for possible treatment strategies. Intra-oral examination revealed loose permanent posterior teeth which were normal in shape and size.(Fig 4) The dentition was free of caries, although the patient's oral hygiene was rather poor with a great deal of plaque and calculus. All teeth showed normal response to thermal and electrical stimulus. Radiographic analysis showed that the the molars of upper and lower jaws have total absence of roots or rudimantory roots with normal appearing crown and completely obliterated pulp chamber (Fig 5). Teeth appeared as uniform dense radio opaque bodies without any roots and root canals. The extracted teeth shows excessive resorption of roots (Fig 6). The patient's medical history revealed no evidence of disturbance in general health .Her family history revealed that there was no such abnormality in other family members.

On the basis of the clinical and radiographic appearance, a diagnosis of DD type I, subtype 1a, was suspected.

DISCUSSION:

Dentin dysplasia type I is a very rare condition affecting the primary and permanent dentition , in which the teeth

present with normal appearing crowns but no or only rudimentary root development, incomplete or total obliteration of the pulp chamber and periapical radiolucent areas or cysts.[11,12] The etiology of DD is still unknown; Logan et al suggested that the dentinal papilla is responsible for the abnormalities in root development [13]. Sauk et al postulated an invagination of the epithelial root sheath resulting in abnormal dentin formation [14]. Wesley et al proposed that the condition is caused by an abnormal interaction of odontoblasts with ameloblasts leading to abnormal differentiation and/or function of these odontoblasts[15] Clearly, the precise nature of the defect is yet to be determined. Of significant interest is the discovery of periapical radiolucency representing granulomas , cysts or abscess involving apparently otherwise intact teeth [1] The cause of periapical radiolucencies in DD type I is not understood. DD is recognized as a genetic disorder and is possibly thought to be a single gene mutation. Multiple family members having a similar pattern of pathosis supports the diagnosis of a hereditary condition [17].Teeth with features of DD occur in a number of disorders such as *calcinosis*, *Ehlers-Danlos syndrome,* and the *brachioskeletogenital syndrome*[18]. Some association has also been reported between dentine dysplasia and osseous changes in addition to sclerotic bone formation.

Management of patients with dentinal dysplasia has presented dentists with many problems.

In case 1 extraction of lower lateral incisors was done and a cast partial denture was given, fixed partial denture was not advisible due to the decreased root surface area of abutment teeth for support.

In case 2 extraction of affected teeth was done (Fig 6) and treatment with a combination of onlay bone grafting and a sinus lift technique to accomplish implant placement is planned.

CONCLUSION:

Dentin dysplasia type I is a rare inherited abnormality of the dentin that leads to premature exfoliation of the primary and permanent teeth. Early diagnosis of the condition is important for initiation of effective preventive treatment. In this regard, the pediatric dentist has an important role in the early diagnosis of this disorder and in guiding patients in the selection of measures to prolong the retention of affected teeth.

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FIGURES:

CASE 1:

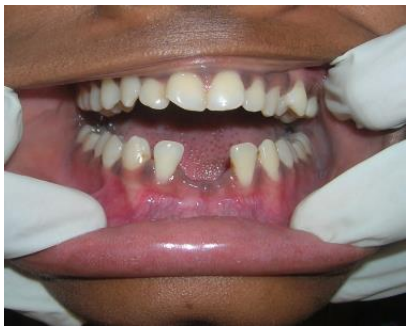

Fig 1.Intraoral view:-shows missing Central incisors and mobile lateral Incisors with fair oral hygiene.

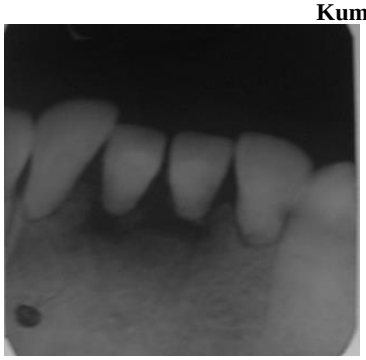

Fig 2:- IOPA shows central incisors with no roots and lateral incisors with small rudimentary roots

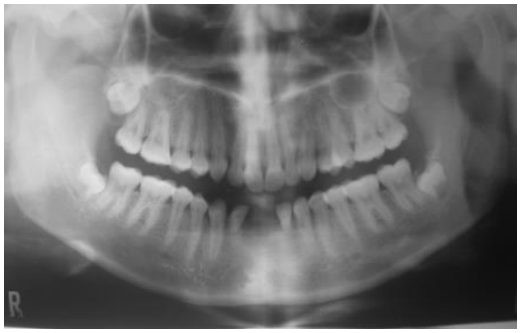

Fig 3:-OPG shows exfoliated central incisors and lateral incisors with rudimentary roots

CASE 2:

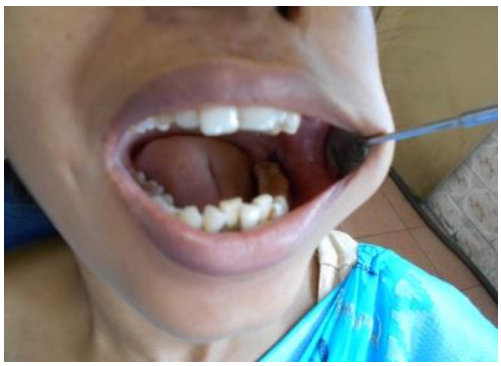

Fig 4:-Intra-oral examination revealed loose permanent posterior teeth which were normal in shape and size

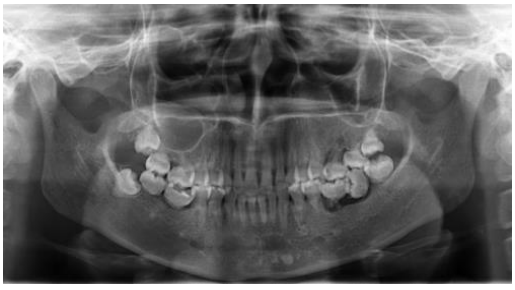

Fig 5:- OPG shows total absence of roots or rudimantory roots with normal appearing crown and completely obliterated pulp chamber in upper and lower jaw

Fig 6:- Extracted teeth with marked resorption of the roots