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Case Report

TYPE I DENTIN DYSPLASIA: A RARE CASE REPORT OF TWO SIBLINGS

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ABSTRACT

Dentin dysplasia (DD) is rare autosomal dominant hereditary disorder related to disturbance in dentin development. The common characteristics of DD are normal enamel, short roots, pulpal obliteration, atypical dentin, early exfoliation and occasional periapical cysts. Two cases of DD are reported and pertinent literature discussed.

KEYWORDS *dentin dysplasia, hereditary, tooth, pulp, tooth*

INTRODUCTION

Dentin dysplasia (DD) is rare autosomal dominant hereditary disorder related to disturbance in dentin development. This rare condition was first described by Ballschmiede as "Rootless teeth" in 1922. The term "Dentin Dysplasia" was coined by Rushton (1). The common characteristics of DD are normal enamel, short roots, pulpal obliteration, atypical dentin, early exfoliation and occasional periapical cysts (2). Shields et al. (3) classified DD into two types. In type I the teeth are clinically normal in size, shape and consistency in both primary and permanent dentition. According to Shields et al. in-type II DD the roots are normal and coronal abnormalities are seen.

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CASE DESCRIPTION

Two siblings (18-year-old female and 19-year-old male), reported to the Dept of Periodontology with the complaint of missing teeth in the lower front region and mobility with upper and lower back teeth. Both patients gave history of exfoliation of few front teeth due to loosening. Both patients medical history was not contributory. There was no history of early teeth exfoliation with family members. Parents of the patients had consanguineous marriage and belonged to lower socio-economic strata.

On clinical examination, both the patients appeared to be well developed and well-nourished without any visible external deformity. All the vital signs were normal in both the patients. Extraoral examination (Fig. 1a and Fig. 1b) in both patients showed normal facial structure and no deformity involving scalp, hands and feet.

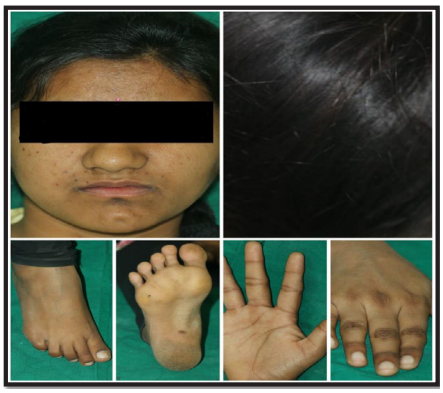

Fig. 1a. *Extraoral features of female patient*

On intraoral examination, (Fig 2a) of the female patient, most of the anterior teeth were missing and the teeth which were present had varying degree of mobility.

The color of the teeth was normal. The oral hygiene status was poor. The mandibular right second and third permanent molar and lower left third permanent molar were cariously destructed. The gingiva had normal colour except for the marginal gingiva which was seen reddish in colour. Ample amount of plaque and calculus was noted in the surrounding region. The contour of the gingiva was scalloped and consistency was firm. Resorbed ridge was seen in the missing teeth region. The size of the gingiva was normal. Generalized bleeding on probing was present. Mulberry molars were seen in mandibular right and left sides of the jaw.

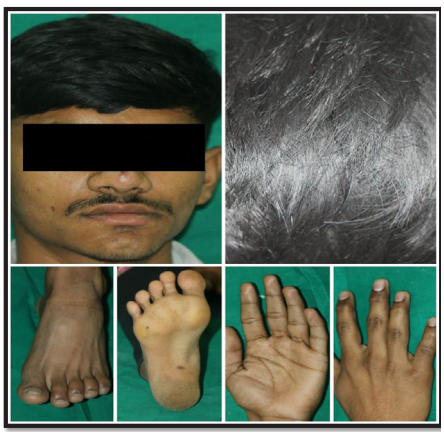

Fig. 1b. *Extraoral features of male patient*

Teeth present –

8 7 6 5 4	3 4 5 6 7 8
8 6 5 4 3	3 4 5 7 8

Grade I mobility –

7 6 5 4	4 5 6 7 8
6 5 4	4 7

Grade II mobility -

	3
	5

Grade III mobility -

3	3

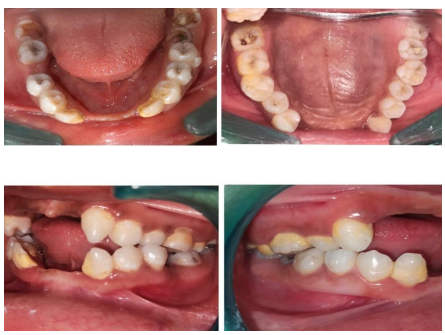

Fig 2a. *Intraoral features of female patient*

In the male patient, intraorally, (Fig 2b) the color of the teeth was normal.

The oral hygiene status was fair. The gingiva was pale pink colour, scalloped contour and firm consistency. All incisors were missing and mobility was present in upper right premolar and lower left canine. Ridge resorption was seen in the missing teeth region. The size of the gingiva was normal. Mulberry molar were seen in lower right and left sides of the jaw.

Teeth present -

8 7 6 5 4 3	3 4 5 6 7 8
8 7 6 5 4 3	3 4 5 6 7 8

Grade I mobility -

4 3	
3	3

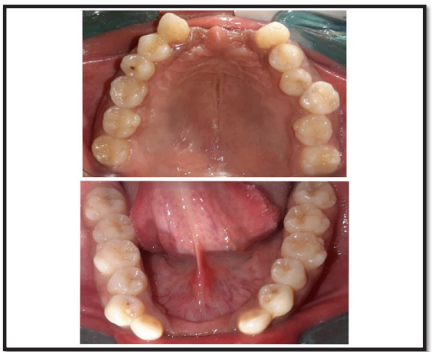

Fig. 2b. *Intraoral featurs of male patient.*

Investigation

Extraoral radiographs (Fig 3a, 3b), hand-wrist radiograph (Fig 4a, 4b) and intraoral radiographs (Fig 5a, 5b) were taken.

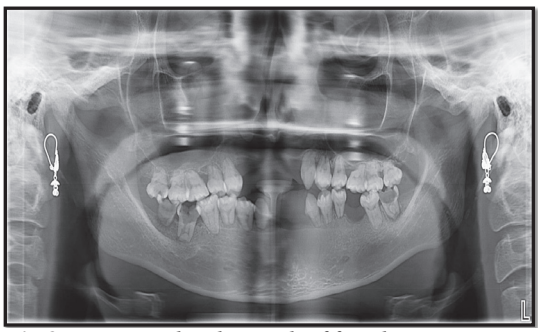

Fig 3a. *Extraoral radiograph of female patient*

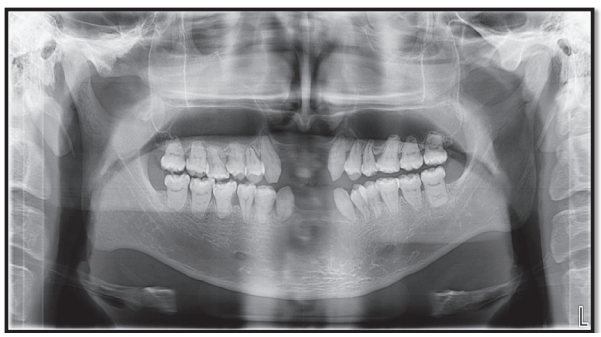

Fig 3b. *Extraoral radiograph of male patient.*

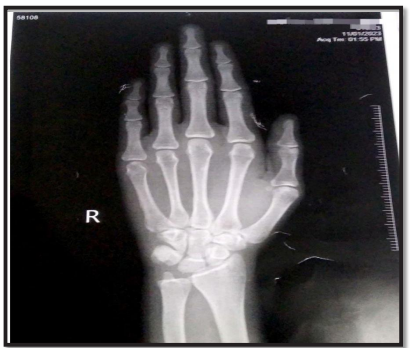

Fig 4a. *Hand – wrist radiograph of female patientfemale patient*

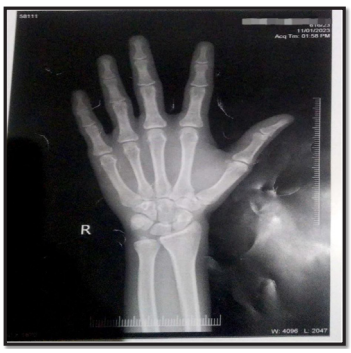

Fig 4b. *Hand – wrist radiograph of male patient*

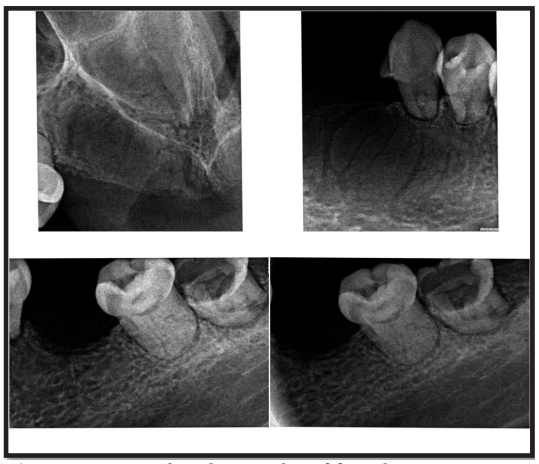

Fig 5a. *Intraoral radiographs of female patient*

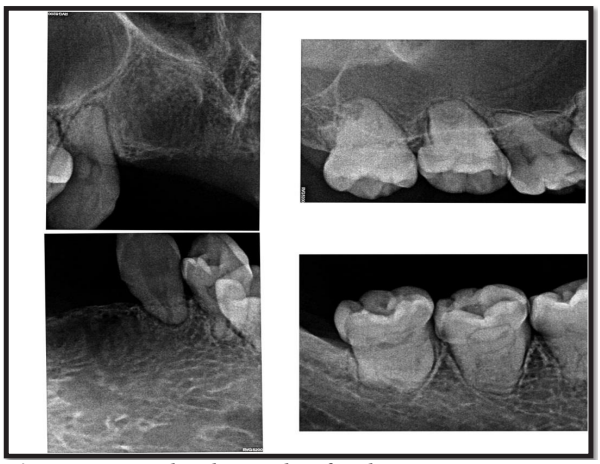

Fig 5b. *Intraoral radiographs of male patients*

Radiographically, the teeth appeared to have normal enamel and shorter roots. Crescent shaped pulpal remnants were seen. Pulpal obliteration was clearly visible and open apices were seen. (Fig 3a, 3b) Taurodontism was noted with all molars. Periapical radiolucencies were noted in female patient with 38 and 48. Tests like complete blood count, serum phosphorous, serum calcium, blood sugar levels, bleeding and clotting time were performed. All the lab investigations were within the normal range for both the patients.

Differential diagnosis

Generally, diagnosis is based on history and the clinical and radiographic features of the patient (4). The features of DD I most commonly resemble with DD II and dentinogenesis imperfecta. Some systemic disorders like calcium deficiency, rheumatoid arthritis, vitamin D deficiency, sclerotic bone and skeletal anomalies (5) also needs to be differentiated. In patients with dentinogenesis imperfect, the root appears to be constricted and short (3) with translucent crown. The crown fractures and enamel chips off easily. In patients with sclerotic bone and skeletal anomalies, along with DD I features the sclerotic bone is dense and skeletal anomalies are seen in wrists and hand bones. Defective calcification of mineralized structures is seen in patients with Vitamin D dependent rickets (VDDR). Along with large pulp chambers and short roots VDDR patients also exhibit features like short stature, open fontanels, muscle weakness and convulsions (6).

Diagnosis

After careful evaluation of the clinical feature, radiographic features, lab investigations, and differential diagnosis, both patients were diagnosed with Dentin Dysplasia type I c.

DISCUSSION

DD is commonly associated with autosomal dominant inheritance (7). Dental papilla is associated with the abnormalities seen in root formation. Reduced growth and obliteration of pulp space is seen due to calcification of the multiple degenerative foci in the papilla (8). The condition is mainly caused due to abnormal interaction in ameloblasts and odontoblast and causing abnormal differentiation and function of odontoblast (9). Witkop (10) suggested that the dysplasia resulted from breaking off and migrating of the epithelial cells from the sheath of Hertwig into dental papilla, where they induce odontoblast differentiation and dentin formation. It is classified into two types i.e., Type I dentin dysplasia and Type II dentin dysplasia. In type I DD the colour, shape and consistency of the crown portion is seen normal in both deciduous and permanent teeth. Radiographically the primary dentition shows complete pulp obliteration (7). According to Carroll et al., in permanent dentition four slightly different radiographic appearances have been seen and are further classified based on those differences (7). DD1a has complete root obliteration, no root formation. In DD1b minimal remnants of pulp at CEJ are seen with short root and no visible canals. DD1c is seen with two crescent shaped pulp remnants and intermediate root length. DD1d has pulp chambers near CEJ, widened canals with pulp stones. Root bulge in anterior teeth is seen and normal root length with visible canals (7). In type II DD the roots appear to be normal in size and shape. The crown portion appears to be translucent and discoloured in nature. Radiographically in DDII the tooth is normal sized, but with thistle tube shaped canals where pulp chambers suddenly constrict at base into narrow canal. Pulp stones are seen in the chamber and no bulging of root is seen (7). Dentinogenesis Imperfecta is also associated with genetic inheritance like dentin dysplasia in which both deciduous and permanent teeth are affected (2). In this the crown appears to be amber and are translucent. The crown structure is more prone to fracture and the enamel readily chips off. The roots and the pulp chamber appear to be smaller than the normal size, and due to deposition of irregular dentin pulpal obliteration is seen (2).

Treatment of patients with dentin dysplasia is symptomatic and is mainly focused on retention of the teeth present rather than replacement. Extraction is suggested for the teeth with hypermobility, pulpal necrosis and periapical abscess (5). Procedures which reduce discomfort during mastication and prevent premature loss of the teeth are carried out (11). Patient should be recalled for regular follow ups due to unfavourable prognosis of affected teeth because of short roots and associated periapical radiolucency and conservative treatment such as prevention of caries should be done (12). For aesthetic purposes, composite crowns can be adde4d (13). Periapical surgery and retrograde filling can be done in teeth having longer roots (12, 13). Placement of removable dentures or space maintainers can be done if necessary. Due to

pulpal obliteration and presence of pulp stones, endodontic therapy is difficult (12). Treatment combining onlay bone grafting and sinus lift procedure can be done for successful placement of the implants (14). The severity and the outcome of the treatment depends upon the patient's age. Acceptable results can be achieved with early diagnosis and proper treatment (7). Recently, three mutant genes have been detected in three affected pedigrees (15-17).

Oral prophylaxis was performed in both the patients. Dietary and oral hygiene instructions were given. Extraction was done in female patient for teeth which were cariously destructed and having grade III mobility. For the male patient, removable prosthesis was delivered in Dept of Prosthodontics with maxillary and mandibular anterior teeth. As for the female patient since there was mobility involved with maximum number of teeth, prosthesis could not be given. No comprehensive treatment including implants could be done due to the socio-economic status of the parents. Both patients were kept on regular maintenance protocol.

CONCLUSION

DD-1 is a rare anomaly related to genetic inheritance. It possesses various difficulties during the treatment of such patients. This abnormality of dentin is associated with early exfoliation of the primary as well as permanent teeth, rootless teeth, hypermobility etc. In patients with dentin dysplasia preventive care is most effective. Maintenance of oral hygiene and meticulous diet habits should be established for retaining the natural teeth for longer duration of time. In such cases the dentist plays a very critical role in early diagnosis and effective treatment for the patients. Although a series of pathogenic genes have been identified, detailed pathogenic mechanisms remain unclear. Further exploration of genetic function and signaling pathways is needed to acquire a comprehensive understanding of these diseases.

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