ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of Development Research
Vol. 09, Issue, 08, pp. 29390-29393, August, 2019

CASE REPORT OPEN ACCESS

DENTINOGENESIS IMPERFECTA TYPE II: A CASE REPORT

***[1]Dr. Somasingh R Lamani, [2]Dr. Ranjani Shetty and [3]Dr Ashok L.**

[1]Assistant Professor, Dept of oral Medicine and Radiology, Govt dental College & Research Institute, Ballari – 583104, Karnataka, India
[2]Professor, Dept of oral Medicine and Radiology, Bapuji Dental College and Hospital, Davangere – 577004, Karnataka, India
[3]Professor and HoD, Dept of oral Medicine and Radiology, Bapuji Dental College and Hospital, Davangere – 577004, Karnataka, India

ARTICLE INFO

Article History:
Received 03rd May, 2019
Received in revised form
26th June, 2019
Accepted 04th July, 2019
Published online 30th August, 2019

Key Words:
Dentin, Dentinogenesis Imperfecta Type II, Teeth.

ABSTRACT

Dentinogenesis Imperfecta is one of the most common autosomal dominant hereditary disorders of dentin formation. It is classified as, type I which is associated with osteogenesis imperfecta; type II not associated with osteogenesis imperfecta; and Type III is associated with the Brandywine triracial isolate.Clinically the teeth shows discolouration and often results in shearing of the overlying enamel which results in attrition and fracture. Radiographically it shows structural defects such as bulbous crowns and small pulp chambers. Early diagnosis and treatment can achieve better functional and aesthetic results and psychological benefit. Inthis case report we present a 27 year old female with generalized brownish discolouration of the teethand severe attrition with radiographic features of bulbous crown and spike shaped roots suggestive of Dentinogenesis Imperfecta type II and the literature including etiology and management is briefly reviewed.

Citation: Dr. Somasingh R Lamani, Dr. Ranjani Shetty and Dr Ashok L. 2019. "Dentinogenesis imperfecta type II: a case report", *International Journal of Development Research*, 09, (08), 29390-29393.

INTRODUCTION

The most common hereditary disorders of dentin formationis Dentinogenesis imperfecta (DGI). It is an autosomal dominant Mandelian trait and has a very low incidence of spontaneous mutations, thus signifying a basic defect in structural and regulatory protein (Bhandari, 2008). It isalso known as hereditary opalescent dentin or Capdepont dysplasia. The dental papilla of either or both primary and secondary dentin will be abnormalas it is a localised mesodermal defect (Rajendran, 2006). In 1908, it was diagnosed as a defect, predominantly due to abnormal dentin (Subramaniam, 2008). Mutations in the dentin sialophosphoprotein gene, located at locus 4q12q21, results in defect in the dentin sialophosphoprotein and dentin phosphoprotein, 50% of the noncollagenous structure of dentin is formed by these proteins (Akhlaghi, 2016). Shields *et al* classified Dentinogensis imperfecta into three categories: Type I is associated with osteogenesis imperfecta where primary teeth are more severely affected than permanent. In Type II, also known as heredity opalescent dentin, primary and permanent dentition both are affectedequally. Type III is rare gives them a shell-like appearanceand affects the permanent dentitionmost commonly (Akhlaghi, 2016). This case report describes the clinical manifestations and radiographic features of a 27 year old female patient with Type II Dentinogensis imperfecta.

Corresponding author: **Dr. Somasingh R Lamani**
Assistant Professor, Dept of oral Medicine and Radiology, Govt dental College & Research Institute, Ballari – 583104, Karnataka, India

CASE REPORT

A 27 year old female patient (Fig: 1) reported to the department of Oral Medicine and Radiology with a chief complaint of pain in the lower right back tooth since 1 month. Pain was sudden in onset, intermittent, moderate, aching type, localised, aggravated on chewing food and relieved by taking medication. She gave a history ofbrownish discoloration and chipping of both milk and permanent teeth since childhood. Her medical history, past dental history and family history was non-contributory.

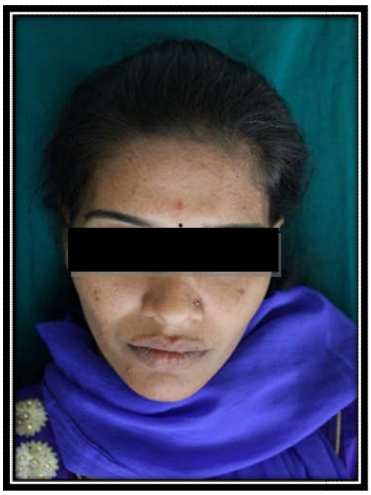

Fig. 1. 27 year old female patient.

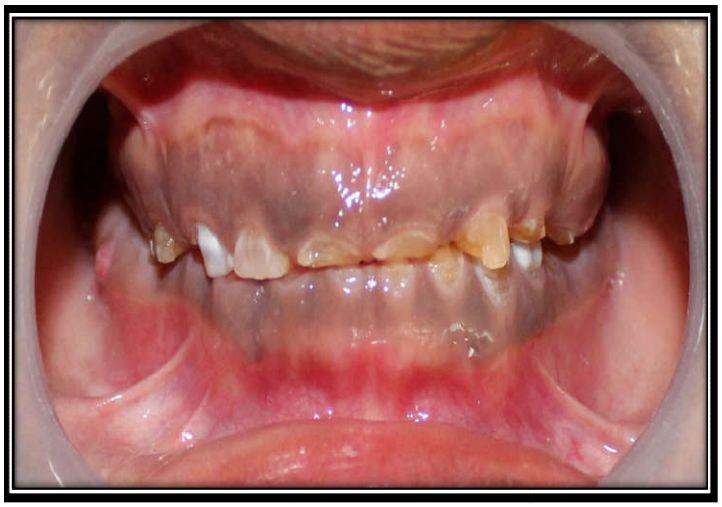

Fig. 2. Teeth small in size and shows severe attrition, with complete loss of enamel in most of the teeth and coronal height of the teeth reduced

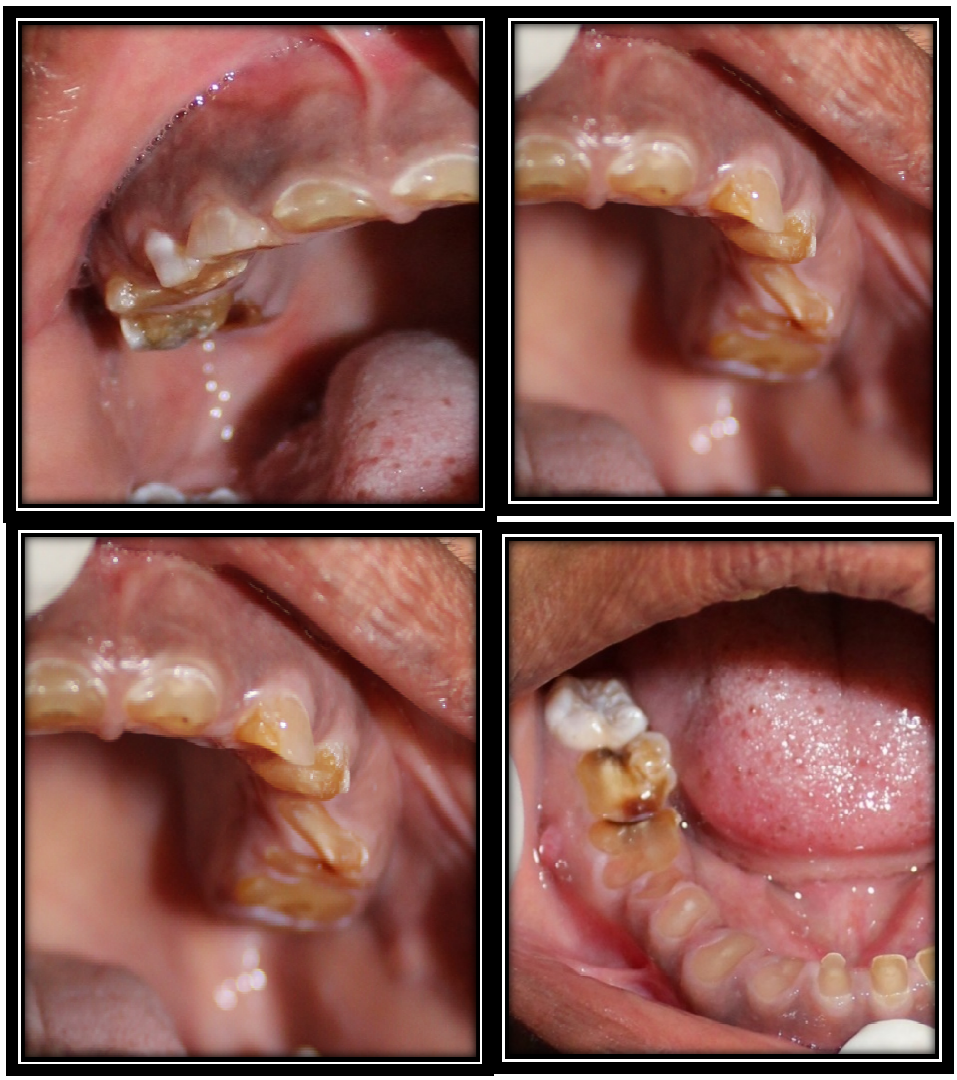

Fig. 3. Class two cavity is evident in disto-occlusal aspect of 36 and mesio-occlusal aspect of 46with sinus opening on the attached gingiva. Bell shaped crown is seen with respect to 34, 38 and 48

General physical examination revealed she was moderately build, well nourished, well oriented to time, place and person. Extra oral examination revealed right submandibular lymphnodes were palpable roughly oval in shape, size approximately 1x1 cm in diameter, soft in consistency, mobile and tender. On intraoral examination, complete compliment of permanent teeth were present except for 18 and 28.

Generalized brownish discolouration of the teeth with a translucent hue was evident. The teeth were small in size and showed severe attrition, with complete loss of enamel in most of the teeth (Fig: 2). Coronal height of teeth was reduced. Exposed dentin as well as pulp in the various teeth were seen. Class two cavity was evident in disto-occlusal aspect of 36 and mesio-occlusal aspect of 46 with sinus opening on the attached

gingiva. Bell shaped crown was seen with respect to 34, 38 and 48 (Fig: 3). On probing of the teeth, generalized surface softness, chipping of the enamel and dentin was evident. 46 was tender on percussion with grade one mobility. Based on the history and clinical findings a provisional diagnosis of chronic apical periodontitis wrt 36, chronic periapical abscess wrt 46, and dentinogenesis imperfecta type II was considered.Patient was subjected to investigations like intraoral periapical radiographs (IOPA) w.r.t upper and lower incisors andmolars and Digital OPG.IOPA Radiographs (Fig: 4) revealed, altered morphology of crown, with decreased density of enamel and dentin and obliteratioon of the root canals.

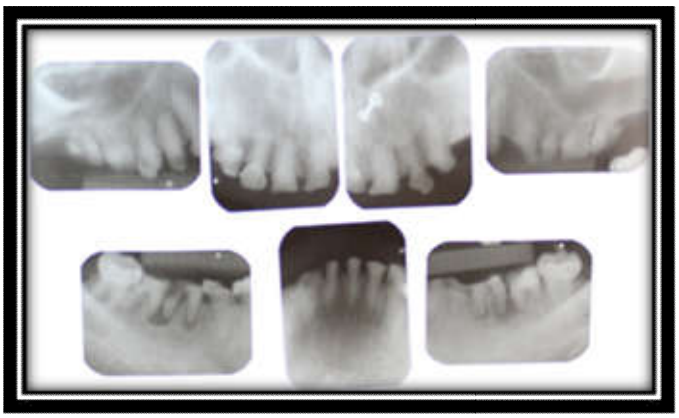

Fig. 4. IOPA Radiographs shows altered morphology of crown, with decreased density of enamel and dentin and obliteratioon of the root canals.Well defined radiolucency evident at apical one-third of both root with 36 suggestive of periapical granulama.46 shows diffuse periapical radiolucency suggestive of periapical abscess

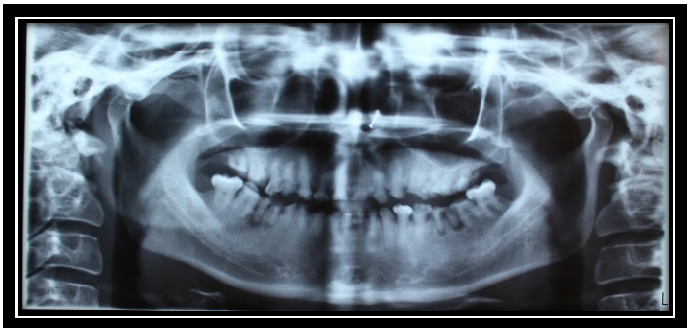

Fig. 5. Digital OPG shows features of generalized marked attrition of theocclusal surface with reduced density of enamel, dentin thinning and generalized bulbous crown with small spike shaped roots having marked cervical constriction and obliterations of root canals

Disto-occlusal radiolucency was evident involving enamel, dentin and pulp wrt 46. Radiolucency involving enamel, dentin and pulp was presentin relation to 47. Well defined radiolucency was evident at apical one-third of both root with 36 was suggestive of periapical granulama. 46 showed diffuse periapical radiolucency suggestive of periapical abscess. Digital OPG (Fig: 5) revealed, features of generalized marked attrition of theocclusal surface with reduced density of enamel, dentin thinning and irregular surface of the incisal and occlusal aspect of teeth. Generalized bulbous crown with small spike shaped roots having marked cervical constriction and obliterations of root canals. Based on the radiographic findings, a final diagnosis of chronic periapical abscess w.r.t 46, periapical granuloma w.r.t 36 and dentinogenesis imperfecta type II was considered.

DISCUSSION

DGI is a group of autosomal dominant genetic conditions characterised by abnormal dentin structure affecting either the primary or both the primary and secondary dentitions. The teeth appear amber, brown/ blue or opalescent brown while radiographically the crowns may appear bulbous, pulp chambers are often small or obliterated and the roots are often narrow with small or obliterated root canals (Barron, 2008). Non-syndromic DGI is reported to have an incidence of 1 in 6,000 to 1 in 8,000 (Barron, 2008 and Abukabbos, 2013). DGI has been classified by Shields and co-workers into three types:Type I, DGI associated with Osteogenesis imperfecta (OI). Both are mesodermal defects, (although OI may occur without DGI).Type II, DGI without OI.Type III, Brandywine type. It is a rare variety characterised by shell teeth, with very little dentin and multiple pulp exposure in the primary teeth (Bhandari, 2008). Dentin is that component of the tooth which encloses the dental pulp and is itself enclosed, above the gingival margin, by the enamel. Structurally, dentine is composed of a mineral phase of hydroxyapatite (70%), an organic phase (20%) and water (10%). The organic phase is primarily of type I collagen (85%) and the remaining, non-collagenous protein is dominated by dentine phosphoprotein (15%) (Nanci, 2008). Kim and Simmer (Kim, 2007) described the etiology of dentinogenesis imperfecta as a defect in the gene that codes for most dentin proteins, including collagen type I. The enamel appears to be normal; however, it tends to detach and fracture because of occlusal stress, thus exposing the dentine, which exhibits a soft consistency and therefore, wears quickly and lead to the alveolar process (Witkop, 2007). Some authors have reported that attrition reduces the incidence of carious lesions; however, periapical lesions are very common because of the quick exposure of the pulp due to attrition (Wieczorek, 2013). The most common clinical manifestations of DI are tooth discoloration (varying from amber-like translucent gray or brownish purple to yellowish brown) (Biria, 2012), and excessive tooth wear. The enamel might peel off to leave dentin exposed, making it susceptible to severe and rapid decay. Radiographically, teeth have normal enamel radiodensity and thickness (Shields, 1973).

In both the deciduous and permanent teeth, dentinogenesis imperfecta presents root canals and pulp chambers with progressive obliteration due to continuous and disordered dentin deposition by ondontoblasts (De La Dure-Molla, 2015). The tooth crowns are short and have a bulb or signet form, while the roots, in addition to being short, are constricted (Devaraju, 2014), Other features of DI include bulbous crowns with marked cervical constrictions and partial or total precocious obliteration of pulpal space. Significant decay can be seen over a short period of time. Histologically, dentinal tubules are sparse, irregular and larger in diameter, and often have large areas of uncalcified matrix (Akhlaghi, 2016). A medical history should aim to establish if the dental condition is a ‘syndromic’ form of DGI as this is a variable feature of a number of heritable conditions including Osteogenesis imperfect (Martin, 2007). Ehlers Danlos syndrome, Goldblatt syndrome, Schimke immunoosseous dysplasia, Brachio-skeleto-genital syndrome, and osteodysplatic and primordial short stature with severe microdontia, opalescent teeth, and rootless molars.[5] The aims of treatment are to remove sources

of infection or pain, restore aesthetics and protect posterior teeth from wear. Treatment varies according to the age of the patient, severity of the problem and presenting complaint. The outcome of a diagnosis of DGI largely depends upon the age at which the diagnosis was given and the speed and quality of the treatment provided. Where diagnosis occurs early in the life of the patient and treatment follows the recommendations outlined above, good aesthetics and function can be obtained thereby minimising nutritional deficits and psychological distress.

REFERENCES

Abukabbos H, Al-Sineedi F. Clinical manifestations and dental management of dentinogenesis imperfecta associated with osteogenesis imperfecta: Case report. *The Saudi Dental Journal*. 2013; 25(4):159-165.

Akhlaghi N, EshghiAR, Mohamadpour M. Dental Management of a Child with Dentinogenesis Imperfecta:*J Dent (Tehran)*. 2016; 13(2):133-138.

Barron MJ, McDonnell ST, MacKie I, Dixon MJ. Hereditary dentine disorders: dentinogenesis imperfecta and dentine dysplasia. *Orphanet Journal of Rare Diseases*. 2008; 3:31.

Bhandari S, Pannu K. Dentinogenesis imperfecta: A review and case report of a family over four generations. *Indian J Dent*. 2008; 19:357-61.

Biria M, Abbas FM, Mozaffar S, Ahmadi R. Dentinogenesis imperfecta associated with osteogenesis imperfecta. *Dent Res J (Isfahan)*. 2012; 9(4):489-94.

De La Dure-Molla M, Philippe Fournier B, Berdal A. Isolated dentinogenesis imperfecta and dentin dysplasia: revision of the classification. *European Journal of Human Genetics*. 2015; 23(4):445-451.

Devaraju D, Devi BY, Vasudevan V, Manjunath V. Dentinogenesis imperfecta type I: A case report with literature review on nomenclature system. Journal of Oral and Maxillofacial Pathology : JOMFP. 2014; 18(1):S131-S134.

Kim JW, Simmer JP. Hereditary dentin defects. *J Dent Res.* 2007; 86:392-9.

Martin E, Shapiro JR. Osteogenesis imperfecta: Epidemiology and pathophysiology. CurrOsteoporos Rep. 2007; 5:91-97.

Nanci A: Dentin-Pulp Complex. In Ten Cate's Oral Histology: Development, Structure and Function 7th edition. Edited by: Nanci A. St. Louis, Missouri, USA: Mosby Elsevier. 2008:191-238.

Rajendran R. Developmental disturbances of oral and paraoral structures. Shafer's textbook of oral pathology. Elsevier; 2006.p.75-7.

Shields ED, Bixler D, El-Kafrawy AM. A proposed classification for heritable human dentine defect with a description of a new entity. *Arch Oral Biol.* 1973; 18(4):543-53.

Subramaniam P, Mathew S, Sugnani SN. Dentinogenesis imperfecta: A case report. *J Indian SocPedodoPrev Dent.* 2008; 26(2):85-7.

Wieczorek A, Loster j. Dentinogenesis imperfecta type II: Ultrastructure of teeth sagittal sections. Folia HistochemCytobiol. 2013; 51:244-7.

Witkop CJ Jr. Hereditary defects of dentin. *J Dent Clin North Am.* 1975; 19:25-45.

****