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Case Report

A Case Report On Dentinogenesis Imperfecta

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ABSTRACT

A 23 Year old female patient came to the department of Oral Pathology, Darshan Dental College, Udaipur with complain of decayed teeth since 7-8 yrs. Onset of dental caries was spontaneous. Progression of caries was gradual in nature. It was not associated with any symptoms. Patient was more concerned about esthetic problem and wanted it to get restored. There were no aggravating and relieving factor associated with it. Dentinogenesis imperfecta is one of the most common hereditary disorders of dentin formation. Dentinogenesis imperfecta type 2 is a disease inherited in a simple autosomal dominant mode. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth and occlusion and improve esthetics. The purpose of this article is to present a case report on Dentinogenesis imperfecta and problems encountered in the treatment of DI with comparison to previous cases.

Key words: autosomal, dentinogenesis imperfecta, dental caries, occlusion.

INTRODUCTION

Dentinogenesis imperfecta (DI) is one of the most common hereditary disorders of dentin formation (1:8000). [1] It follows an autosomal dominant Mandelian trait with a high degree of penetrance. [2] It has a very low incidence of apparent spontaneous mutations, signifying a basic defect in structural and regulatory protein. [3] DI has been classified by Shields and co-workers [1-4] into three types:

1. Type I, DI associated with osteogenesis imperfecta (OI). Both are mesodermal defects, (although OI may occur without DI).
2. Type II, DI without OI.
3. Type III, brandywine type. It is a rare variety characterized by shell teeth, with very little dentin and multiple pulp exposures in the primary teeth. [1,5,6]

Extensive research over the years has proven that DI and OI are two separate and distinct entities, unrelated to each other. Therefore, a revised classification was proposed where DI is classified as 1 and 2. Both types are not associated with OI. DI1 corresponds to DI type II and DI2 corresponds to the DI type III of Shields classification, respectively. There is no substitute for DGI type I in this revised classification. [1]

Clinically, with DI both dentitions are affected. The color of the teeth varies

from brown to blue, sometimes described as amber or gray, with an opalescent shine. [3] The enamel may show hypoplastic or hypocalcified defects in about one-third of the patients and, in an affected patient, tends to crack away from the defective dentin. The exposed dentin may undergo severe and rapid attrition. [3]

Radiographically, the teeth have bulbous crowns with constricted short roots. Initially, pulp chambers may be abnormally wide and resemble "shell teeth," but they will progressively obliterate. [5] Histologically, the enamel, although normal in structure, tends to crack. The dentin-enamel junction is not scalloped. In most cases the structure of the mantle dentin is normal, whereas the dentinal tubules of the circumferential dentin are coarse and branched and the total number of tubules is reduced. [7]

CASE REPORT

A 23 year old female was presented to the department of Oral pathology with the chief complaint of decayed teeth since 7-8 yrs. The onset of teeth decay was spontaneous. Progression of teeth decay was gradual in nature. It was not associated with any symptoms. Patient was more concerned about esthetic problem and wanted it to get restored. There were no aggravating and relieving factor associated with it. There was no Medical History. Dental history revealed decayed teeth in deciduous dentition in a similar manner as it was present in permanent dentition. There was no history of similar pattern of teeth decay in her family. Personal history was brushing of her teeth with paste once in the morning in horizontal direction and diet was mixed in nature.

On clinical examination, there were no abnormality detected related to gait, posture, sclera skin, conjunctiva, hair and extremities and all the vital signs examined were normal. In extraoral examination facial symmetry was present, straight profile, with brachycephalic head. A Solitary submandibular lymph nodes were palpable bilaterally around 1 cm in size. Soft in consistency, mobile (not fixed), no pain on palpation were present. [**fig1**]

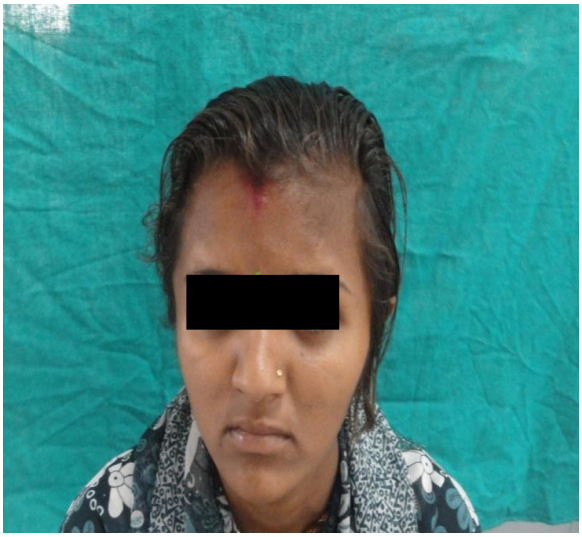

Figure 1. Patient with Dentinogenesis imperfect II

Intraoral examination revealed of Mouth opening of 40mm.Vermillion border, labial mucosa, buccal mucosa, tongue, floor of mouth , soft palate were normal and no signs of gingival inflammation present. On hard tissue examination, dentition present was permanent. 26,36,37,46,47 were decayed. Grade 1 mobility with 46, Parulis present with 46.Absence of enamel in 16,15, 14, 13, 12, 11, 21, 22 ,31, 32, 33, 41, 42, 43, 46.Dentin was amber color in 11,12,13,14,15,16,21,22,31,32 33, 41, 42, 43, 46.There was generalized loss of tooth contact, anterior open bite and Posterior open bite was present on right side owing to loss of tooth structure.

The detailed examination of enamel pattern showed Smooth margins of enamel present in relation to 11 21. Translucent opalescent hue present at cervical one third of tooth in relation to 11 21. Also present in respect to 23 32 33.There was complete

absence of enamel on labial surface in respect to 12 13 22 31 42 and opaque white enamel in respect to 24 25 34 35 44 45. [**fig2**]

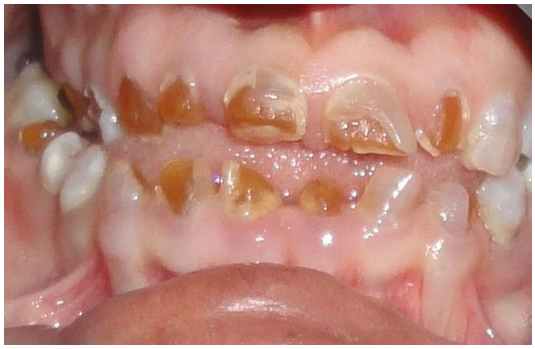

Figure 2 Intraoral examination shows pattern dental caries

Radiographic investigations with Panoramic imaging revealed obliteration of pulp chamber, loss of enamel in respect to 11 12 13 14 16 21 22 31 32 33 41 42 43. cervical constriction of teeth and generalized open contacts . Periradicular radiolucency and loss of bone present in respect to 36 37 47. Mandibular canal is wide on both sides and bifid on left side. Pulp stone present in relation to 47. There was no thinning of cortical bone and trabeculae. Well formed mandibular angular cortex.**[fig3]**

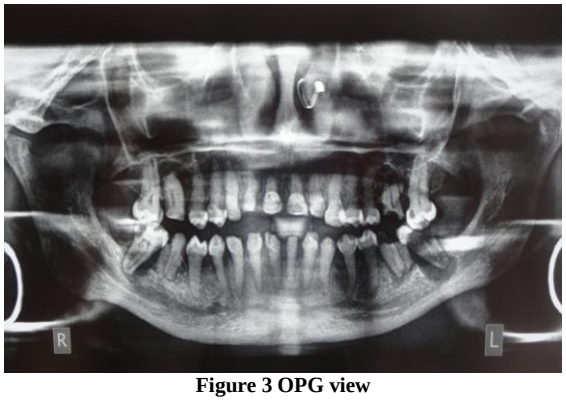

Figure 3 OPG view

IOPA revealed loss of enamel in 11 12 21 22. obliterated pulp chamber. IOPA revealed intact enamel in 44, 45 but completely obliterated pulp chamber.**[fig 4a &b]**

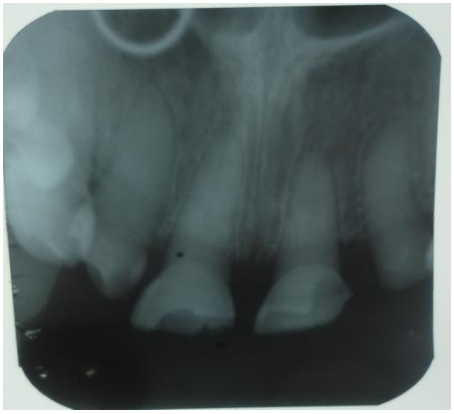

Figure 4a IOPA with 11,1 2,21,22

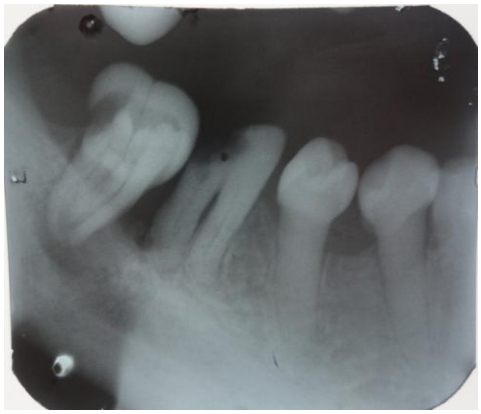

Figure 4b IOPA with 44,45

Provisional diagnosis of Dentinogenesis imperfecta and dental fluorosis in respect to 24 25 34 35 44 45

DIFFERENTIAL DIAGNOSIS

Amelogenesis imperfecta, Hypocalcified Amelogenesis imperfecta, Hypomaturation pigmented pattern, Dentin dysplasia type II (coronal type), Osteogenesis Imperfecta.

FINAL DIAGNOSIS

The clinical and radiographic examinations suggested of Dentinogenesis Type II as Final Diagnosis

In this case Restoration of the teeth with metal crown for posterior teeth and jacket crown for anterior teeth was done and extraction with 36, 37, 47.

DISCUSSION

DI was first reported by Talbot [8] (1893), however, Roberts, Schour [9] (1939) coined the term Dentinogenesis Imperfecta in 1939. Many cases of DI have been investigated in the last decades. [10]

The presented DI type II case showed generalized amber colored discoloration distributed in all the four quadrants as compared to a more intense discoloration in the mandibular incisors teeth compared to the maxillary incisors. It was observed that the reported proband showed cross bite with vertical dimension loss due to the attrition of the crowns, dentin exposure and fast abrasion due to the lower dentin mineral content whereas in the present case anterior open bite and Posterior open bite was present on right side owing to loss of tooth structure.

There are no guidelines on restorative treatment in teeth affected by DI. The treatment should begin as early as possible, considering the degree of tooth destruction and patient cooperation. The importance of restoring dental defects associated with DI is obvious. There is no clear answer as to the best time for initiating treatment. The literature abounds with case reports and treatment modalities for DI; in general, the recommendation is to start treatment as early as possible. [11] The rational use of general anesthesia or pharmacotherapeutic regimens at an early age enables us to safely utilize state of the art of restorative techniques in the treatment of young patients suffering from hereditary dental anomalies. However, there are few case reports describing treatments of the adult patients. [12,13]

Daugaard-Jensen and Nielsen described 3 cases, the youngest being a 19-month-old with DI type. The initial treatment was with stainless steel crowns on the molars which were replaced later with cast gold crowns due to severe attrition. The cast gold crowns were not retentive and parapulpar pins were prepared to improve retention. The anterior teeth were restored in one case with polycarbonate crowns and with stainless steel crowns in another case. [14] Whereas in the present case of patient aged 23 restoration of the teeth with metal crown for posterior teeth and jacket crown for anterior teeth was done and extraction with 36, 37, 47.

Considering that DI-II is an uncommon dental anomaly, the authors believe that the case described illustrates the occurrence of this disease in different parts of the world, affecting people of different ethnicity. Furthermore, as the dental treatment of individuals with DI is very difficult, [15] presenting the dental treatment with this disease could be useful to other clinicians.

CONCLUSION

Total rehabilitation of the patient with dentinogenesis imperfecta is a challenge to the dental surgeon which requires the active involvement of various branches of dentistry. Oral rehabilitation of this type of condition should be oriented towards functional and aesthetic rehabilitation. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth and occlusion and improve esthetics. Long term follow up is imperative in order to intercept complications and adjust the treatment to the changes of the dentition and occlusion. Timely diagnosis and appropriate treatment is of paramount significance to prevent psychological and functional morbidity to the patient. However, the most important factor, and one which is beyond the control of the dentist, is the time the patient reports to the clinic for treatment. With advancing age, providing the patient with optimal treatment is diminished. Most of the cases affected with DGI require a comprehensive

interdisciplinary planning dictated by the age at the time of presentation, clinical presentation, amount of morbidity, patient's expectations, and resources.

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