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Case Report

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Dentinogenesis Imperfecta: A Case report with review of literature

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Abstract

Dentinogenesis Imperfecta is an inherited disorder of defective dentin formation, with an autosomal dominant pattern of inheritance. It is characterized by the presence of opalescent dentin, resulting in a dusky blue to brownish discoloration of the teeth. Dentinogenesis imperfecta has been subdivided into three types: type I is associated with osteogenesis imperfecta; in type II there is no associated osteogenesis imperfecta; and when the condition is associated with the brandywine tri-racial isolate and large pulp chambers it is classified as type III. This case report describes a 12-year-old female patient who showed the characteristic dental features of dentinogenesis imperfect type III.

Keywords: *Dentinogenesis imperfect, brownish discolouration, large pulp chamber.*

Introduction

Dentinogenesis imperfecta is a localized mesodermal dysplasia affecting both the primary and permanent dentitions. The disease is inherited in an autosomal dominant fashion with high penetrance and a low mutation rate.[1] It is the most common dental genetic disease, affecting approximately 1 in 8000 births.[2] Clinically, both primary and permanent dentitions are affected. The color of the teeth may vary from brown to blue, and is sometimes described as characteristic "amber or gray".[1] The enamel may show hypoplastic and hypocalcified defect and easily chip off from the underlying dentin.[1]

Case report

A 12 year old female patient reported to the Department of Oral Medicine and Radiology, Bangalore Institute of Dental Sciences, Bengaluru, with a complaint of brownish discolouration of teeth and missing lower front teeth since 2 years. The patient's medical history was non- contributory.

Family history suggested that the patients maternal grandfather was suffering from the similar condition of rapid wearing of teeth and had undergone total extraction at an young age.

The clinical examination revealed extensive tooth loss of upper and lower anterior teeth. The remaining tooth structure appeared brownish in colour. The maxillary anterior incisor teeth on the right side were tender to percussion and with no associated swelling. A detailed history revealed that root canal treatment had previously been attempted on these teeth and not completed. She was previously advised to undergo total extraction and have dental implants. Patient did not consider it as a viable option due to which she discontinued the treatment. Radiographic examination included full mouth IOPAR, maxillary occlusal cross-sectional view and OPG, which showed features characteristic of dentinogenesis imperfecta. Roots were short and crown bulbous with thin enamel which chipped off from some areas as well. The findings of

clinical radiographic examination and of family history were consistent with diagnosis of DI. Osteogenesis imperfecta was excluded from the differential diagnosis since there was no associated medical signs and symptoms.

Therefore a clinical diagnosis of dentinogenesis imperfect was given. Patient was refered to department of pedodontics for treatment and rehabilitation.

Discussion

Dentinogenesis imperfecta is a hereditary developmental disturbance of the dentin originating during the histo-differentiation stage of tooth development. DI may be seen alone or in conjunction with the systemic hereditary disorder of the bone, osteogenesis imperfecta. The incidence of DI is about one in 8000.[3] Two systems, one by Witkop[3] and the other by Shields[4,5], are well accepted classification systems of DI.

Etiology: Type I collagen (product of COL1A1 and COL1A2 genes) is the most abundant dentin protein.[6] The diverse mutations associated with the COL1A1 and COL1A2. genes can cause the DI phenotype in association with osteogenesis imperfecta (DI type I). DI Type II and Type III are autosomal dominant conditions that have been linked to chromosome 4q12-21, suggesting these may be allelic mutations of the DSPP gene encoding dentin phosphoprotein and dentin sialoprotein.[7,8]

Clinical manifestation: In all three DI types, the teeth have a variable blue-gray to yellow-brown discoloration that appears opalescent due to the defective, abnormally-colored dentin shining through the translucent enamel. Due to the lack of support of the poorly mineralized dentin, enamel frequently fractures from the teeth leading to rapid wear and attrition of the teeth. The severity of discoloration and enamel fracturing in all DI types is highly variable, even within the same family. If left untreated, it is not uncommon to see the entire DI-affected dentition worn to the gingiva.[9]

Table 1: Dentinogenesis Imperfecta[10]

Shield's	Clinical presentation	Wiktop
Dentinogenesis Imperfecta I	Osteogenesis imperfecta with opalescent teeth.	Dentinogenesis Imperfecta
Dentinogenesis Imperfecta II	Isolated Dentinogenesis imperfecta	Hereditary opalescent dentin
Dentinogenesis Imperfecta III	Isolated Dentinogenesis imperfecta	Brandywine isolate

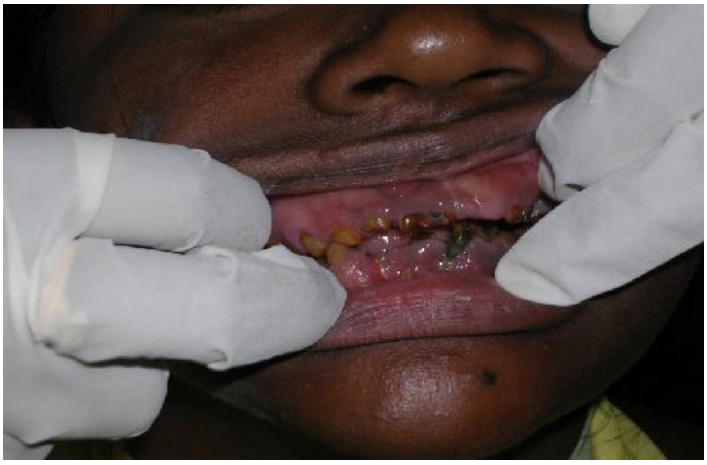

Figure 1: Multiple decayed teeth with brandywine discolouration

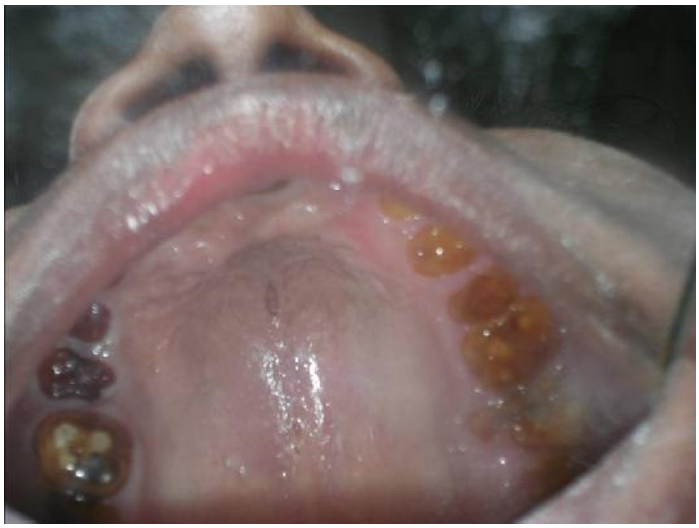

Figure 2: Severely attrited teeth with pulpal exposure

Shields Type I occurs with osteogenesis imperfecta. All teeth in both dentitions are affected. Primary teeth are affected most severely, followed by the permanent incisors and first molars, with the second and third molars being the least altered.

Radiographically, the teeth have bulbous crowns, cervical constriction, thin roots, and early obliteration of the root canal and pulp chambers due to excessive dentin production. Periapical radiolucencies and root fractures are evident. An amber translucent tooth color is common.

Shields Type II is also known as hereditary opalescent dentin. Both primary and permanent dentitions are equally affected, and the characteristics previously described for Type I are the same.

Radiographically, pulp chamber obliteration can begin prior to tooth eruption.

Shields Type III is rare; its predominant characteristic is bell-shaped crowns, especially in the permanent dentition.[9]

Unlike Types I and II, Type III involves teeth with shell-like appearance and multiple pulp exposures. Shell teeth demonstrate normal-thickness enamel in association with extremely thin dentin and dramatically enlarged pulps. The thin dentin may involve the entire tooth or be isolated to the root.[9]

Differential diagnosis: Osteogenesis Imperfecta, other collagen disorders and numerous syndromes have Dentinogenesis Imperfecta like phenotypes associated with them. Dentin Dysplasia Type I clinically has normal appearing crowns, but radiographically the teeth have pulpal obliterations and short blunted roots. Dentin Dysplasia Type II has the same phenotype as Dentinogenesis Imperfecta Type II in the primary dentition but normal to slight blue-gray discoloration in permanent dentition.

Conclusion

Early diagnosis and treatment are essential for obtaining a favorable prognosis, any delay in intervention making the treatment even more complex.[11] One of the greatest challenges for the dentist is to provide an adequate treatment to achieve functional and esthetic restoration in cases of diseases like dentinogenesis imperfecta.

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