Dentinogenesis Imperfecta: Case Report and Review of Literature

Dr. Jaget Reddy MDS[1], Dr. Christeffi Maybel MDS[2], Dr. P. Sai Archana MDS[3], Dr. Sivansatish MDS[4]

[1] HOD,
[2, 4] Reader,
[3] Lecturer,
Dept. of Oral Medicine & Radiology, Chettinad Dental College and Research Institute, Kelambakkam, Chennai

Abstract

Dentinogenesis imperfecta (DI) is a developmental tooth disorder affecting dentin, characterized by the presence of opalescent dentin caused due to autosomal dominant pattern resulting in a greyish blue to brownish discoloration of the teeth. It can affect both deciduous as well as a permanent dentition. This case report describes a 26-year-old male patient with clinical and radiographical characteristic features of dentinogenesis imperfecta.

Keywords: Autosomal dominant, dentinogenesis imperfecta, dystrophic dentin, pulpal space obliteration.

Introduction

Dentinogenesis imperfecta (DI) or hereditary opalescent dentin, was first described in the late 19th century. It is a localized mesodermal dysplasia affecting both the primary and permanent dentition. The disease is inherited in an autosomal dominant mode with high penetrance and a low mutation rate. Parents seek for dentist's advice and treatment during tooth eruption. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth causing cavity leading to disturbance in the function and in severe cases changes in occlusion. The esthetic concern proves to be a biggest challenge if not diagnosed early and intervened. So the purpose of this article is to make the general dentists and specialists to identify the disease early and be aware of its treatment options present when encountered with a patient suffering from this hereditary disease.

Case Report

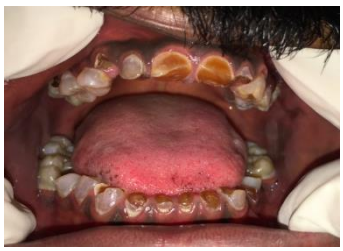

Image - 1

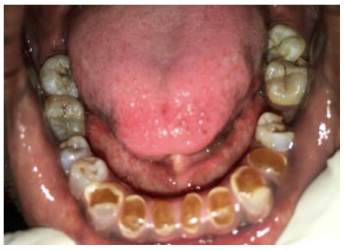

Image - 2

A 26-year-old male, reported to the Department of Oral Medicine with a chief complaint of attrited teeth in his upper and lower jaw and has a desire to place cap on his teeth. Patient has a history of attrited teeth along with yellowish-brown discolouration since childhood. He had neither pain nor sensitivity and claims he never had tooth fracture. His family history revealed the presence of a similar manifestation in his mother, father, grandfather, as well as his child.

Intra oral examination revealed a generalized greyish-bluish opalescence in all the upper and lower teeth along with attrition of teeth , root stumps in 16, 13, 34 and missing teeth in relation to36,46(Image 1,2) . Patient's Intra-oral periapical radiography (Image 3) revealed generalized obliteration of coronal and radicular pulp chamber in teeth. OPG (Image 4) revealed obliteration of the pulp chamber with bulbous crowns. No radiodensity differences are seen between enamel and dentin. Hence on confirming the diagnosis, the patient was reffered to prosthodontics for veneer placement in his teeth.

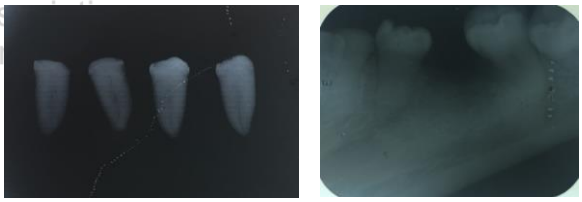

Image – 3 IOPA

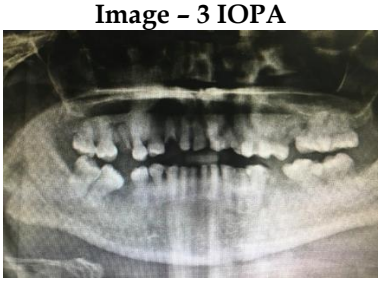

Image – 4

Discussion

Human dentition is subjected to considerable variations in size, form and number of teeth as well as in structure of dental tissues. Disorders in teeth may occur due to various reasons and affects various layers of teeth depending on the time of formation. DI was first recognized by Barret in 1882. The term was coined by Robert and Schour in 1939(1). The first published report describing the disorder as an enamel defect was reported by Talbot as quoted by Witkop. The term ' hereditary opalescent dentin ' was first used by Skillen

, Finn and Hodges to describe the brown translucent enamel that have an opalescent sheen and are lacking in pulp chambers**.** Since DI is inherited in an autosomal dominant disease, there is a 50% chance that a child born to an affected parent will themselves be affected [2]. This also indicates uniform gene expression and complete penetrance for the gene in the family. Mutations in gene or trauma cause dysregulation in formation of structures. If it occurs during formation of enamel it results in the formation of amelogenesis imperfecta and if it occurs during dentin formation it results in dentinogenesis imperfecta. Mutations in the genes encoding the major protein constituents of dentine seem to underlie most hereditary dentine defects. Three distinct protein products are formed from the initially translated polypeptide: dentine sialoprotein (DSP), dentine glycoprotein (DGP) and dentine phosphoprotein (DPP) [3]

Shields et al Classified Dentinogenesis Imperfect into three types [4]
DI type 1 - associated **with osteogenesis imperfecta**.
DI type 2 - has essentially the same clinical radiographic and histological features as DI type but **without osteogenesis imperfecta;**
DI type 3 - is rare and is only found in the triracial Brandywine population of Maryland.

Clinically with this disorder, both dentitions are affected. The color of the teeth varies from brown to blue, sometimes described as amber or gray, with an opalescent sheen. 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 [5] . The exposed dentin may undergo severe and rapid attrition. Radiographically the teeth affected shows obliterated pulpal chambers in coronal and radicular pulp. Bulbous crowns can be seen with short roots.

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 tu- bules is reduced. The presence of an atubular area in the dentin with reduced mineralization and a reduced number of odontoblasts are consistent findings. Pulpal inclusions and much interglobular dentin are also frequent. The biochemical characteristics of the dentin include a col- lagen defect and a primary defect in the calcifying matrix [6].

Syndromes that are associated with dentinogenesis imperfect are Ehlers Danlos syndrome [7], Goldblatt syndrome, Brachio-skeletogenital syndrome, Osteodysplastic primordial short stature with severe microdontia, opalescent teeth, and rootless molars

Differential diagnosis

Dentin dysplasia (DD) **Type I** clinically has normal appearing crowns, but radiographically the teeth have pulpal obliterations and short blunted roots **DD 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. Both DD and DI have amber tooth coloration and obliterated or occluded pulp chambers. However, the pulp chambers do not fill in before eruption in DD Type II. A finding of a thistle-tube shaped pulp chamber in a single-rooted tooth increases the likelihood of DD diagnosis. The crowns in DD are usually are normal in size, shape, and proportion while the crowns in DI typically are bell-shaped with a cervical constriction. The roots in DD usually are not present or appear normal while the roots in DI typically are short and narrow. Association of periapical radiolucencies with non-carious teeth and without obvious cause is an important characteristic of DD Type I.

Fluorosis and non-fluoride-induced opacities needs to establish differences between symmetrical and asymmetrical and/or discrete patterns of opaque defects [8].

Congenital erythropoietic porphyria is a condition resulting from an inborn error of porphyrin metabolism. This deficiency leads to haemolytic anaemia, photosensitivity, blistering of the skin, and deposition of red-brown pigments in the bones and teeth.

Rhesus incompatibility- The discolouration which ranges from yellow through to green, brown and grey to black is usually found at the necks of teeth and the enamel hypoplasias are usually located in the coronal third of the teeth.

Tetracyclines have the ability to chelate calcium ions and to be incorporated into developing teeth, cartilage and bone, resulting in discolouration of both the primary and permanent dentitions. This permanent discolouration varies from yellow or grey to brown depending on the dose or the type of the drug received in relation to body weight.

Hence after diagnosis of disease, the treatment modalities for dentogenesis imperfect are as follows

The Dental approach for managing DI [9] –

- Vary depending on the severity of the clinical expression
- General considerations and principles of management Providing optimal oral health treatment for DI

Aims:

- Preventing severe attrition associated with enamel loss
- Rapid wear of the poorly mineralized dentin
- Rehabilitating dentitions that have undergone severe wear,
- Optimizing esthetics
- Preventing caries and periodontal disease

The clinician must be cautious in treating individuals with osteogensis imperfecta as there is increased risk of bone fracture in the case of dentinogenesis imperfect associated with osteogenesis imperfecta.

Restorative care

Routine restorative techniques often can be used effectively to treat mild to moderate DI in more severe cases with significant enamel fracturing and rapid dental wear, the treatment of choice is full coverage restorations in both the primary and permanent dentitions. In children the treatment will range from composites in anterior teeth and stainless steel crowns in posteriors whereas it's a mixture of treatments which may range from bleaching to veneering of teeth. The success of full coverage is greatest in teeth with crowns and roots that exhibit close to a normal shape and size, minimizing the risk of cervical fracture and loss of vertical dimension .Cases having severe loss of coronal tooth structure and vertical dimension may be considered candidates for overdenture therapy. Different types of veneers can be used to improve the esthetics and mask the opalescent blue-gray discoloration of the anterior teeth

Bleaching has been reported to lighten the color of DI teeth with some success; however, because the discoloration is caused primarily by the underlying yellow-brown dentin, bleaching alone is unlikely to produce normal appearance in cases of significant discoloration

Endodontic considerations: Some patients with dentinogenesis imperfecta will suffer from multiple periapical abscesses from pulp exposure due to extensive coronal wear. The potential for periapical abscesses is an indication for periodic radiographic surveys on individuals with DI. Because of pulpal obliteration, apical surgery may be required to maintain the abscessed teeth. Periapical curettage and retrograde amalgam seals have demonstrated short-term success in teeth with short roots

Occlusion: Class III malocclussion with high incidences of posterior crossbites and openbites occur in DI Type I and should be evaluated [10].

Multidisciplinary approaches are essential in addressing the complex needs of the individuals affected with DI.

Preventive care

Regular periodic examinations can identify teeth needing care as they erupt. Meticulous oral hygiene, calculus removal, and oral rinses can improve periodontal health. Fluoride applications and desensitizing agents may diminish tooth sensitivity.

Discussion

Early identification and preventive interventions are critical for individuals with DI in order to avoid the negative social and functional consequences of the disorder.

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