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Case Report

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Capdepont's Teeth : A Case Report And A Review

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Abstract
Dentinogenesis Imperfecta or capdepont dysplasia also known as hereditary opalescent dentin is one of the most common hereditary disorder of dentin formation. It is inherited in simple autosomal dominant pattern with high penetrance and low mutation rate. It affects either or both primary and permanent dentitions. DGI type II corresponds to a localized form of mesodermal dysplasia, observed in histodifferentiation. Early diagnosis and treatment are fundamental, aiming at making favourable prognosis since late interventions make treatment more complex. This report describes a 16-year-old male patient who showed the characteristic clinical, radiographic and morphological dental features of DGI Type II (DGI I as per latest classification). The aetiology and prevalence of the disorder, and a comprehensive treatment plan are briefly reviewed.

Key Words
Dentinogenesis Imperfecta, Hereditary Opalescent Dentin, Dentin Dysplasia, Mesodermal Defect, capdepont dysplasia

Introduction

Dentinogenesis imperfecta is the most common dental genetic diseasecharacterised by abnormal dentin structure affecting either the primary or both the primary and secondary dentitions in absence of any other systemic disorder. DI affects approximately 1 in 8000 births[1]. The disease is inherited in an autosomal dominant pattern with 100% penetrance and a low mutation rate[2]. Commonly known as hereditary opalescent dentin or capdepont dysplasia, it was first recognized by Barret in 1882. The first published report describing the disorder as an enamel defect was by Talbot in 1893[3]. The fact that the defect is primarily due to abnormal dentin was first recognised by Frgin Foyelle and Mallassez in 1908. The term was coined by Robert and Schour in 1939. The term 'hereditary opalescent dentin' was first used by Skillen[4], Finn[5] and Hodges[6] to describe the brown translucent teeth that have an opalescent sheen and are lacking in pulp chambers. Some authors suggest that the term 'hereditary opalescent dentin' should be preferred over 'dentinogenesis imperfecta' as it describes the general appearance better. Dentinogenesis imperfecta is a localized mesodermal dysplasia affecting both the primary and permanent dentitions. Witkop reported that it is the most common autosomal dominant disease affecting westerners[3]. The color of the teeth varies from brown to yellowish brown or amber blue, with a characteristic transluscent or opalescent hue. The exposed dentin may undergo severe and rapid attrition[2]. Radiographically, the teeth have bulbous crowns with cervical constriction and short roots giving the teeth, a typical tulip appearance. Initially, the pulp chambers may be abnormally wide, giving the appearance of "shell teeth", but they progressively get obliterated[7]. Periapical radiolucent areas may be visible radiographically, in the absence of any clinically obvious pathology.

Case Report

A 16yr old male patient reported with a complaint of discoloured and small teeth since last 4 -5 years **(Fig.2)**. The patient also complained of sensitivity to hot and cold food since past few months. Patient further revealed that the discolouration was present since the time of eruption of permanent teeth but has been gradually increasing since last 3 -4 years along with gradual wearing away of the tooth surface. Medical history revealed typhoid 3-4 years back. There was no history of unusual bone brittleness or unexplained hearing loss. Family history revealed that maternal side of the family also had similar problem. The medical examination (including blood tests and full body X-rays) of the child did not reveal any systemic abnormality or specifically symptomatic of Osteogenesis Imperfecta. Extra-oral examination revealed a bluish hue in the sclera **(Fig.1)**. Clinically intraoral examination revealed generalized occlusal wearing of the teeth. There was generalized yellowish-brown discoloration of the teeth along with enamel hypoplasia. Mulberry molars were also present. Rootstumps of maxillary right first molar were present **(Fig. 2)**.

Intra-oral periapical radiograph **(Fig. 3)** of 12, 22, 31, 32, 35, 36, 37, 41, 42 revealed obliterated pulp canals and pulp chambers along with attrited crown

Fig.1 : Extraoral Profile Showing Blue Sclera

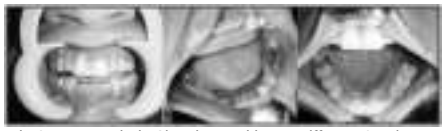

Fig.2 : Intra-oral Pic Showing Attrition In Different Quadrants

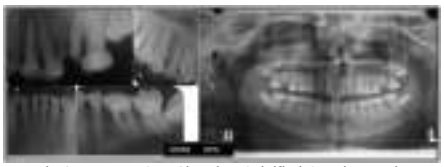

Fig.3 : Iopar & Opg Showing Calcified Canals & Pulp Chambers

portions. Radiograph also revealed bone loss and periapical pathology associated with root stumps of maxillary first molar.

Orthopentomograph (OPG) **(Fig. 3)** showed bulbous crown portions along with cervical constriction .Partial or complete obliteration of root canals of almost all teeth, spike like roots which were short and slender and periodontal ligament widening of teeth.

Based on the clinical and radiographic features, the case was diagnosed as DGI Type II (DGI I as per latest classification)

Discussion

Dentin, the most abundant tissue in teeth, is produced by odontoblasts, which differentiate from mesenchymal cells of the dental papilla.

It was recognized first time by Barret in1882. In 1887, the condition was first described in a case involving a completely normal boy with dark staining on the teeth[8]. The term 'Dentinogenesis imperfecta' was coined by Robert and Schour in 1939.[9]

DGI is an inherited mesodermal condition affecting the primary and permanent dentition.Inheritance of dentin defects is typically autosomal dominant, although autosomal recessive and X-linked cases of dentin defects associated with syndromes are reported. Dentin defects occur as a feature in a number of syndromes, including Osteogenesis Imperfecta, Ehlers-Danlos syndrome EDS, tumoral calcinosis and hypophosphatemic rickets as well as potentially newly described syndromes.

Witkop named the types as Dentinogenesis imperfecta, hereditary opalescent dentin and brandywine isolate[7]. Schields et al divided Dentinogenesis imperfect into DGI type I, II and III

1. Type 1: DGI associated with Osteogenesis Imperfecta (OI)
2. Type 2: DGI without Osteogenesis Imperfecta (OI)
3. Type 3: Brandywine Type, It is a very rare variety, characterized by Shell teeth, with very little dentin and multiple pulp exposures in primary teeth.Patients with DGI type III appear to be limited, in large measure, to a population in the region around Brandywine in southern Maryland[7].

Extensive research over the years has proven that Dentinogenesis imperfecta and Osteogenesis Imperfecta are two different and separate entities, unrelated to each other. Therefore, a revised classification was proposed where DGI is classified as I and II. Both types are not associated with Osteogenesis Imperfecta. DGI I corresponds to DGI Type II and DGI II corresponds to DGI Type III of shields classification respectively. There is no substitute for DGI Type I in this classification. DGI is an inherited disorder affecting dentin. Mutation in DSPP is the cause for this defect. The DSPP gene is located at 4q21.3 in a cluster of dentin and bone matrix genes. DSPP encodes both dentin sialoprotein (DSP) and dentin phosphoprotein (DPP) as one precursor protein that is cleaved before secretion. DSP and DPP have different roles in DGI. DPP severs as a nucleator of mineralization and induces apatite formation. DGI-I clinically presents with amber-brown or bluegrey coloration of teeth with opalescence. The opalescence color is not due to the pulp showing through but to the collagen structural defect within the dentin, which may reflect either a bluish light through the enamel or a brown color. The enamel tends to chip away from the incisal rim of the anterior teeth and from the occlusal surface of posterior teeth.

The enamel fracturing is believed to be due to the poor support provided by the abnormal dentin and possibly in part to the absence of the scalloping normally seen between dentin and enamel that is believed to help mechanically lock the two hard tissues together [10].

The enamel does not actually fall off the Dentinoenamel junction (DEJ), as some believe; the DEJ in DGI seems to be structurally and functional normal. The pre-odontoblasts within the dental papilla seem to undergo normal cellular differentiation and form nearly normal mantle dentin. These odontoblasts die and are replaced by newly differentiated odontoblasts from the dental papillae that never mature into fully functional odontoblasts. These defective odontoblasts secrete an abnormal collagen, which is undermineralized and fails to form odontoblastic tubules. The result is the imperfect dentin that reflects light in the brown or blue spectrum but has formed a near normal DEJ because of the less affected preodontoblasts and the mantle dentin they produce. Some enamel wears excessively because of hypoplastic hypocalcified areas within the enamel rods. This is presumably due to fact that amelogenesis is induced and initiated by the initial mantle dentin deposition in normal tooth formation. The abnormality in dentin formation results secondarily in enamel that is also microscopically somewhat defective. The exposed dentin, which undergoes quick attrition, to such an extent that the dentin becomes smooth and continuous with the gingival tissue. In DI, water content of teeth is greatly increased as much as 60% of the normal while the inorganic content is less than that of normal dentin that takes its hardness close to cementum, thus explaining the rapid attrition of affected teeth[11].

Radiographic feature include bulbous crown with characteristic cervical constriction and short roots, giving a tulip like appearance, obliteration of pulp chamber and multiple pseudo periapical radiolucency with the absence of pulpal exposure or pulpal necrosis. Although the pulp is usually obliterated by excess dentin production, some teeth may show normal sized pulp or pulp enlargement. Cementum and alveolar bone are normal[12].

The histological features include complete absence or reduction in the number of Dentinal tubules. They are irregular in shape, size and course. Hypo mineralized interglobular dentin is increased and cementum is normal. Odontoblasts entrapment may be seen within the dentinal matrix. Large areas of unmineralized dentin and irregular border between the unmineralized and mineralized dentin is seen. The enamel appears to be normal; however, there are subtle hypocalcification defects in the enamel rods just above the DEJ.

Syndromes associated with dentinogenesis imperfecta are Osteogenesis imperfecta, Ehlers Danlos syndrome[13], Schimkeimmuno osseous

dysplasia, Goldblatt syndrome, Brachio-skeletogenital syndrome, Osteodys plastic primordial short stature with severe microdontia, opalescent teeth, and rootless molars[14]

Differntial Diagnosis

The differential diagnosis for DGI includes Dentin Dysplasia, amelogenesis imperfecta, dental fluorosis, congenital erythropoetic porphyria, tetracycline staining.

In dentin dysplasia: As these two conditions seem to form a continuum, with similarities including the potential to affect the color of the deciduous and permanent teeth, and both initiating about the same time, their differentiation may be difficult. Both entities may have occluded pulp chambers, but at least in Type II Dentin Dysplasia the pulp chambers are never filled before eruption. Also, if Thistle –tube shaped pulp chambers are observed in the single rooted teeth, the probability of dentin dysplasia is strengthened. In addition, the size will help distinguish between the two: the teeth in opalescent dentin have typical "BELL- SHAPED "crowns with a constriction in the cervical region, whereas the crowns in Dentin Dysplasia are usually normal in shape, size, and properties. It the roots are short and narrow, the condition is likely to be Opalescent Dentin. On the other hand, normal appearing roots or practically no roots at all should suggest dentin dysplasia. Periapical rarefaction in association with non carious teeth are strong presumptive evidence that the condition is dentin dysplasia: they are seldom found in opalescent dentin. Dentin dysplasia type I, has normal coloration of both the primary and permanent dentition, although their pulps are almost completely obliterated giving a typical crescent shape.

Amelogenesis Imperfecta can be recognized by its more clinically apparent enamel defect. In AI, teeth are usually sensitive and on radiographs enamel is less radio-dense and thinner than dentin. Pulp chambers and root canals are usually not sclerosed.[15]

Fluorosis is common in areas of increased fluoride content in water (fluoride belt) with patterns of discolouration that are bilateral, diffuse (not sharply demarcated), opaque, and white striations that run horizontally across the enamel [16]

In congenital erythropoeitic porphyria-It is a rare condition resulting from an inborn error of porphyrin metabolism. Abnormally high levels of porphyrin pigments are incorporated into teeth during their formation. The entire primary and secondary dentitions the discolouration ranges from yellow through to green, brown and grey to black with no pulp sclerosis and is usually found at the necks of teeth with the enamel hypoplasias usually located in the coronal third of the teeth [17].

Tetracycline staining-The erupting affected teeth have a bright yellow band-like appearance that fluoresces under ultraviolet light. On exposure to sunlight, the colour gradually changes to grey or redbrown. Radiographically there is no pulp sclerosis in tetracycline staining while in Dentinogenesis imperfecta pulp sclerosis is present.

Treatment

The foremost treatment regimen of this Autosomal dominant pattern is the genetic counseling to the parents as there could be 50% chances that the child born would also be affected with the same[17]. Many treatment modalities have been suggested, eg, over-dentures, stainless steel crowns, jacket crowns, pin retained cast-gold 'thimbles' under acrylic resin crowns, stainless steel crowns with acrylic facing, and simple removable appliances[9]. The use of a combination of partial dentures and prosthetic crowns on the anterior teeth has also been described. Orthodontic treatment has been successfully performed in patients with different degrees of dentinogenesis imperfect[18]. Steel crowns used to prevent attrition of the dental structure can be used in deciduous teeth and in young permanent posterior teeth, where esthetics is not an issue. According to Wei such a procedure must be undertaken as soon as the tooth erupts[19]. Shafer et al. emphasize that restorations cannot be permanent owing to the diminished hardness of dentin[20]. Consequently, when fractures occur at the gingival level or below the gum, exodontia is indicated, as in the case of teeth that exhibit periapical rarefaction and root fracture[18],[19]. When dealing with deciduous and young permanent anterior teeth, celluloid crowns are recommended; permanent molars should receive full cast crowns; metalloceramic restorations are recommended for premolars; and permanent anterior teeth should be restored with esthetic facets[18]. The goal of the treatment is to establish a more favorable prognosis for such a complex anomaly, and to insure the integrity of the erupting dentition along with functional and esthetic restoration, thus improving the self esteem and quality of life for these individuals. The treatment should aim to prevent the abrasion of the erupted teeth and to establish the proper vertical dimension. Timely diagnosis and appropriate treatment is of paramount significance to prevent psychological and functional morbidity to the patient. Most of the cases presented with DGII require a comprehensive interdisciplinary planning dictated by the age at the time of presentation, clinical presentation, amount of morbidity, patients expectations and resources. Early diagnosis and treatment are essential for obtaining a favorable prognosis; any delay in intervention makes the treatment even more complex[21]. The case has been reported as an attempt on our part to create awareness among general dentists so that they make common masses aware of these genetic oral disease. This will help the disease to be curbed at its onset in deciduous dentition and pave the way for a healthier permanent dentition and reinforces the importance of proper history taking of the patients.

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