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Case Report

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DENTINOGENESIS IMPERFECTA: A CASE REPORT WITH FAMILY HISTORY

Dr. Juhi Gupta[1]*, Dr. Anshul Aggarwal[2], Dr. Kauser Jahan Khwaja[3],

[1]*Assistant Professor, [2]Associate Professor and [3]Associate Professor

Deptt. Oral Pathology/ Oral Medicine and Radiology Z. A. Dental College, AMU, Aligarh, India.

***Corresponding Author: Dr. Juhi Gupta**

Ex- Assistant Professor, Deptt. Oral Pathology/ Oral Medicine and Radiology Z. A. Dental College, AMU, Aligarh, India.

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ABSTRACT

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 a simple autosomal dominant mode with high penetrance and a low mutation rate. The reported incidence in the USA is 1:8000 births. Shields et al proposed three types of dentinogenesis imperfecta: DI type 1 is associated with osteogenesis imperfecta. DI type 2 has essentially the same clinical radiographic and histological features as DI type 1 but without osteogenesis imperfecta; DI type 3 is rare and is only found in the tri racial Brandywine population of Maryland. It has been suggested that DI type 2 and DI type 3 are different expressions of the same gene.

KEYWORDS: Opalescent dentin, abnormal dentino-enamel junction, dentin hypoplasia, amber coloured teeth, Bulbous crown.

INTRODUCTION

Dentinogenesis imperfecta is an uncommon defect in the collagen formation that is transmitted as an autosomal dominant trait.[1] This condition causes the teeth to be discolored (most often a blue-gray or yellow-brown color) and translucent. Teeth are also weaker than normal, making them prone to rapid wear, breakage and loss. These problems can affect both primary teeth and permanent teeth.[2] It was probably first recognized by Barret in 1882. The first published report describing the disorder as an enamel defect was by Talbot as quoted by Witkop. The term 'hereditary opalescent dentin' was first used by Skillen, Finn and Hodgesto describe the brown translucent teeth that have an opalescent sheen and are lacking in pulp chambers.[3]

It is a localized mesodermal dysplasia which affects both the primary as well as the permanent dentitions. It is inherited in an autosomal dominant pattern with high penetration and a low mutation rate. It is one of the most prevalent dental genetic disease; affecting approximately 1 in 8000 births.[4]

Patients affected with DGI show a peculiar color deviation from the natural dentition that ranges from gray to brownish violet or yellowish brown, with a characteristic unusual translucent or opalescent hue. This is attributed to the dentinal disturbance, with enamel being normal. These teeth are prone to excessive wear and fracture due to the primary abnormality in the structure and composition of dentin and presumably abnormal dentin-enamel junction that lacks normal scalloping.[9]

However, the caries incidence is low in these patients due to early wear of the fissures and contact points. There is an early attrition of dentin in deciduous teeth with hyperplasia of the residual ridges.[10] with this background we herein present a case of dentinogenesis imperfecta type with family history.

CASE REPORT

10 yr old patient reported to Dental OPD of Z.A. Dental College, AMU, Aligarh with the chief complain of brownish discoloration of teeth. According to the patient's mother the teeth were apparently normal when erupted and eventually turn to brown as child grown up. She had also given history of similar kind of problem with the teeth of her second male child who was just 2 yr. old (Figure 2). In his younger brother discolouration of teeth were evident even though they have not erupted completely. For the age of two years presence of only upper incisors also indicate delayed eruption of teeth. (Figure 2) Family history revealed similar kind of problem in teeth of the father of kid but none of the female members in his father's family was found to be affected. There was no history of frequent bone fracture and sclera of eye appeared normal.

On clinical examination the teeth had typical brown opalescent hue with severe attrition leading to loss of occlusal height. Clinical pulp exposure was evident in

many teeth (Figure1). OPG and IOPA radiological investigations were advised for the patient. OPG (Figure 3& 4) revealed obliteration of pulp chamber of all the deciduous teeth and also partially in first permanent molars. Other developing tooth bud of permanent teeth appeared normal. Bulbous crown of permanent first molars were noted. Attrition in teeth was also noted.

Based on clinical examination, family history and radiological findings we arrived to the diagnosis of Dentinogenesis Imperfecta.

Legends for Figures
Figure 1: Deciduous of teeth showing severe attrition with yellowish brown discoloration
Figure 2: Brown discoloration erupting teeth with no attrition in his younger brother
Figure 3: OPG showing obliteration of pulp chamber with bulbous crown of posterior deciduous teeth.
Figure 4: IOPA showing obliteration of pulp chamber and severe attrition of upper anterior deciduous teeth

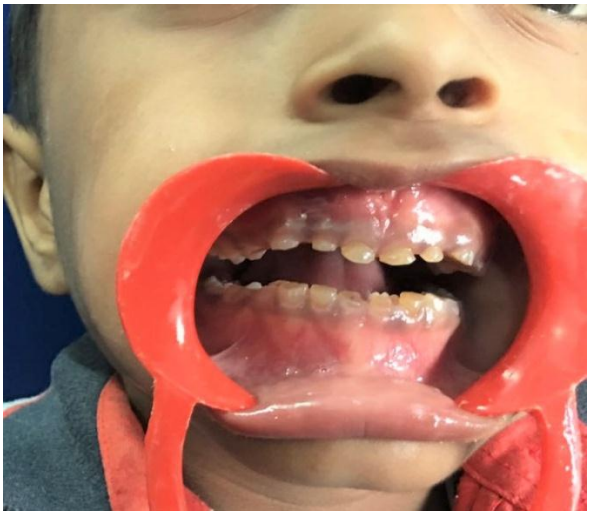

Figure 1

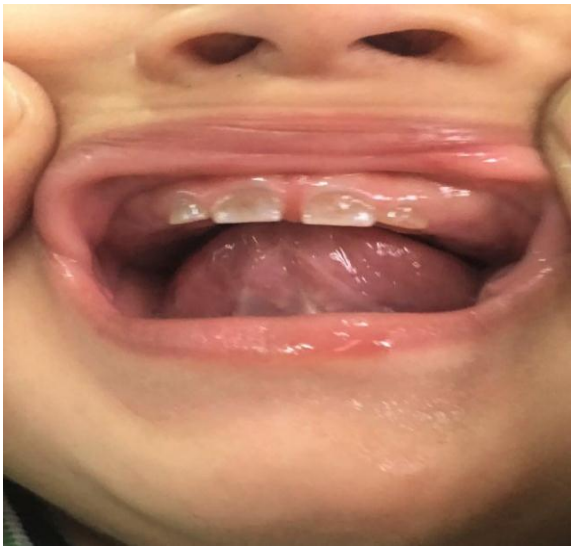

Figure 2

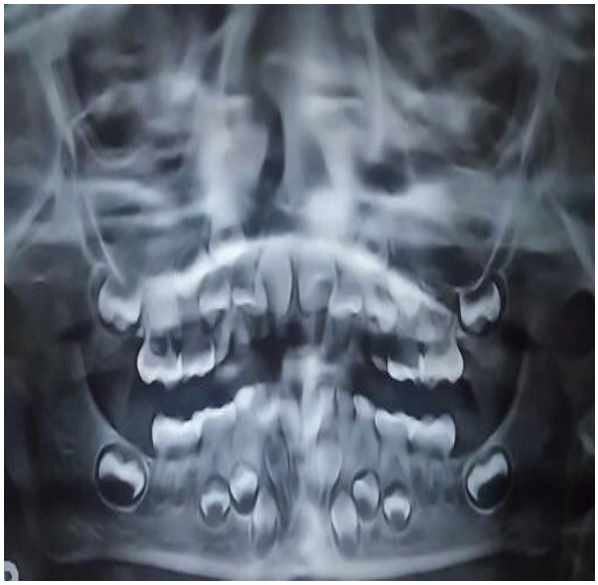

Figure 3

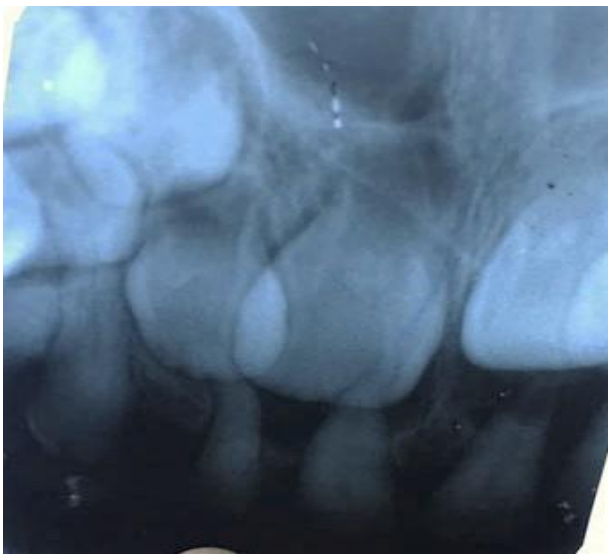

figure 4

DISCUSSION

Dentinogenesis is a highly ordered process in which the organic predentine matrix is progressively mineralized by ectomesenchymally-derived cells called odontoblasts.[6]

During the bell stage of tooth development odontoblast differentiate and form a single cell line outlining the pulp cavity. It secretes organic predentin in the underlying space.

The predentine (10–40 μm thickness) is an unmineralised region containing type I collagen which separates the odontoblast cell bodies from the mineralisation front. At the mineralization front, the collagenous component of the matrix is thought to provide the correct three-dimensional structure into which the mineral component of dentine is deposited while dentine phosphoprotein, which is secreted from cellular processes extending from the odontoblasts.[7] Dentinogenesis imperfecta is a

developmental defect of mesodermal component of developing tooth bud.

Shields et al proposed three types of dentinogenesis imperfecta: DI type I is associated with osteogenesis imperfecta. DI type II has essentially the same clinical radiographic and histological features as DI type I but without osteogenesis imperfecta; DI type III is rare and is only found in the tri racial Brandywine population of Maryland. It has been suggested that DI type 2 and DI type 3 are different expressions of the same gene.[5]

The Shields' system is increasingly out of date as it does not account for the molecular etiologies of the hereditary dentine defects elucidated so far, for example, those underlying osteogenesis imperfecta and other syndromes manifesting defective dentine formation.[11]

Mendelian Inheritance of Man (MIM) is the most current classification for molecular genetics of human pathosis. In the MIM system, Dentinognesis Imperfecta type 1 as described in original Shield's classification (DGI1) has been removed from the list of DGI and was classified properly as osteogenesis imperfecta. DGI II has become DGI I. However DGI III, dentin dysplasia type I (DD-1) and dentin dysplasia type II have retained their previous names. Current evidence strongly suggests that DGI III simply represents a variable expression of dentinogenesis imperfecta. Identical mutations of DSSP (dentin sialophosphoprotein) have been shown to manifest as DGI II and DGI III in different families. It seems very clear that dentinogenesis imperfecta type 2 and 3 represents single disease with variable expression.[8]

Modified Classification of Hereditary Disorders Affecting Dentin[8]

Disorder	Inheritance	Involved gene or genes
Osteogenesis imperfecta with opalescent dentin	Autosomal Dominant or Recessive	COL1A1 and COL1A2
DSPP-associated dentin disorder Dentin Dysplasia type II (DD-II) Dentinogenesis Imperfecta (includes old DGI II and III)	Autosomal Dominant	DSSP
Dentin Dysplasia type II (DD-II)	Autosomal Dominant	??

Dentinogenesis imperfecta affects both primary and permanent dentitions and usually the deciduous dentition is more severely affected. There could be severe attrition of teeth and obliteration of pulp chamber. As in our case the teeth when erupted were of normal color but with few months there was change in color and loss of teeth structure due to attrition of teeth. Even delayed eruption of teeth was also reported in our case.

There was a definite family history but the case is unique in sense that only the male members were involved. Usually in autosomal dominant trait both male and female off spring had equal chance to inherent the trait as the mode of transmission is vertical with very less chance of mutation.

Teeth suffering with dentinogenesis imperfecta suffer with severe mechanical attrition under normal occlusal force. Normal scalloping at dentino enamel junction acts as mechanical inter locking between enamel and dentin and help it to resist the mechanical wear and tear due to normal occlusal forces. Due to defective dentin formation and lack of normal scalloping at dentino enamel junction leads to fracture and early loss of enamel leading to loss of massive crown structure despite presence of healthy roots.

Rehabilitation of pediatric children with dentinogenesis imperfecta is really challenging. Usually restoration of such teeth is usually impossible and extraction of teeth and prosthetic rehabilitation remains the only way to restore the restoration.

But diagnosis of condition at the early stage may help the dentist to treat and provide better prosthetic rehabilitation of teeth. It is deemed necessary that prosthetic rehabilitation must be started prior to loss of teeth structure due to attrition of teeth or development of any periapical pathology in the affected teeth.

In our case the patient was very young and the permanent teeth were yet to erupt. There was no periapical pathology in relation to any teeth and patient also did not complain of any tooth ache (except discoloration of teeth) so he was kept on follow up. However patient was advised to report back in case he develops any tooth ache.

CONCLUSION

Dentinogenesis imperfecta is an inheritable disease with a progressive clinical course. Due to defective dentin formation and abnormal dentino enamel junction enamel undergoes rapid attrition leading to sever loss of crown structure following few years of eruption of teeth. Management of teeth suffering with dentinogenesis imperfecta is directed towards saving the natural crown structure as much as possible. So early diagnosis of condition is very necessary to intervene the progression of disease at the earliest.

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