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CASE STUDY

DENTINOGENESIS IMPERFECTA

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ABSTRACT

Dentinogenesis imperfecta is one of the most common hereditary disorders of dentin formation. It follows an autosomal pattern of transmission, affecting both the formation and mineralization of dentin. Either or both primary and permanent dentition is affected by it. This paper describes a case of Dentinogenesis imperfecta which reported for orthodontic treatment.

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INTRODUCTION

Dentinogenesis imperfecta (DI) is an inherited disorder of tooth development. Affected teeth are gray to yellowish-brown and have broad crowns with constriction of the cervical area resulting in a 'tulip' shape. (Roh *et al*., 2010) The disorder is classified into two types: Dentinogenesis imperfecta I - without osteogenesis imperfecta (opalescent dentin), this corresponds to dentinogenesis imperfecta type II of Shields classification; and Dentinogenesis imperfecta II-Brandywine type, this corresponds to dentinogenesis imperfecta type III of Shields classification. (Bhandari and Pannu, 2008) Orthodontic treatment for patients who have dentinogenesis imperfecta is not a routine procedure for orthodontic practitioners. (Crowell, 1998) For the successful treatment of patients with DI, not only is a differential diagnosis between types I and II necessary, but also a multidisciplinary approach should be considered. We report a case of Dentinogenesis Imperfecta I (Shields Type II) who reported to the department for orthodontic treatment.

Case Report

A 17 year old female patient presented with complaints of forwardly placed upper front teeth. She gave a history of discoloured teeth from childhood and history of sensitivity of lower front teeth since the last two months. Also she gave a history of bad breath since the last six months. Her medical history showed no evidence of Osteogenesis Imperfecta, such as a history of frequent fractures of long bones, laxity of joints, blue sclera, hearing loss, or increased bleeding tendency. Patient got a crown restoration irt upper left first molar 3 years back, RCT of lower left central incisor recently and visited a dental college for orthodontic treatment recently. Also, the patient gives a history of her mother and maternal grandfather having translucent teeth and early loss of teeth. Clinical examination revealed a Bluish grey opalescent hue of crowns especially the lower anteriors. Ragged incisal ridges and Enamel chipping were also noticed. (Figure 1) She had a convex profile with posterior divergence with End on molar relation bilaterally, superimposed on a Class II skeletal base with 7mm overjet. The patient had an anterior open bite, proclined upper and lower anteriors, transposition i.r.t 43, 44 (Figure 2) and potentially incompetent lips. The patient had 'tulip' shaped bulbous crowns and tongue tie present. Radiographically she had short narrow roots with the pulp chambers and root canals almost obliterated in the anterior region. In the posterior region the root canals were well formed and the roots showed abnormal morphology. (Figure 3)

DISCUSSION

Dentinogenesis imperfecta is one of the most common hereditary disorders of dentin formation (1:8000).

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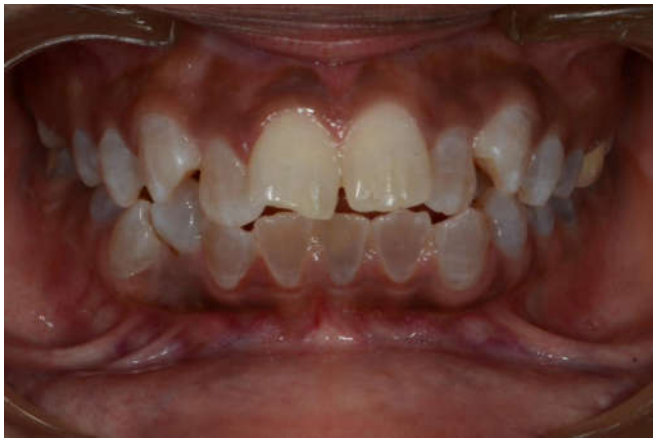

Figure 1. Bluish grey opalescent hue of crowns

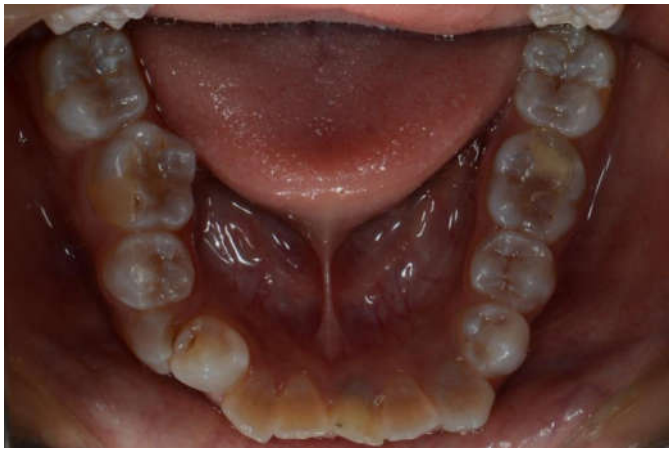

Figure 2. Transposition irt 43 and 44

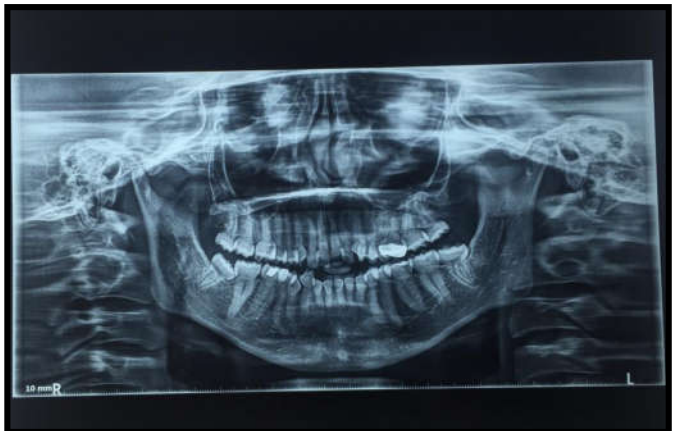

Figure 3. Radiograph showing short roots with obliterated pulp chambers

It follows an autosomal dominant Mendelian trait with a high degree of penetrance. (Bhandari and Pannu, 2008) It was first reported by Tolbot as an autosomal dominant trait. Roberts and Schonr were the first to use the term DI, describing it as a condition similar to that found in OI. (Devaraju *et al*., 2014) Treatment of both the dentitions is exigent and often demands a multidisciplinary approach to prevent psychological morbidity to the patient. The gene maps to chromosome number 4. It encodes a protein called dentin sialophosphoprotein (DSPP). This protein constitutes about 50% of the noncollagenous component of dentin matrix. It is not known how the mutant protein causes near obliteration of the pulp. (Maciejewska and Chomik, 2012) Dentin defect associated with osteogenesis imperfecta was earlier listed as dentinogenesis imperfecta type I (Shields classification). Extensive studies have proven that dentinogenesis imperfecta is clearly a disorder distinct from osteogenesis imperfecta, and hence the classification was revised. (Bhandari and Pannu, 2008) Dentinogenesis imperfecta I (Levins classification) is also known as Opalescent dentin, DI without OI, Opalescent teeth without OI, DI Shields type II or Capdepont teeth. This condition is caused by mutation in the DSPP gene (gene map locus 4q21.3), encoding dentin phosphoprotein and dentin sialoprotein. (Orsini *et al*., 2014) The teeth appear to be blue-gray or amber brown and opalescent. On dental radiographs, the teeth have bulbous crowns, roots that are narrower than normal, and pulp chambers and root canals that are smaller than normal or completely obliterated. The enamel may split readily from the dentin when subjected to occlusal stress. (Devaraju *et al*., 2014)

Dentinogenesis imperfecta II is also known as DI Shields type III or Brandywine type dentinogenesis imperfecta. This disorder was found in the Brandywine triracial isolate in southern Maryland. The crowns of the deciduous and permanent teeth wear readily after eruption and multiple pulp exposures may occur. The dentin is amber and smooth. Radiographs of the deciduous dentition show very large pulp chambers and root canals, at least during the first few years, although they may become reduced in size with age. The permanent teeth have pulpal spaces that are either smaller than normal or completely obliterated. (Dure-Molla *et al*., 2015) Recent studies are consistent with the hypothesis that DI I and DI II are allelic or the result of mutations in two tightly linked genes. MacDougall (1998) provided information on the intervals separating four genes that map to this same region, all four of which are involved in dental development: DSPP, DMP-1, IBSP, and SPP1. MacDougall *et al* (1999) stated that the manifestations of DI II can differ from those of DI I by the presence of multiple pulp exposures, normal nonmineralized pulp chambers and canals, and a general appearance of 'shell teeth'. (Maciejewska and Chomik, 2012) The case presented here was diagnosed as Dentinogenesis Imperfecta I (Shields type II) as the patient had a positive family history. Also the patient had blue gray opalescent & few deep yellow teeth, bulbous/Tulip shaped crowns and showed involvement of her 1st molars and incisors. Another contributing evidence was the presence of prominent cervical constriction and partially obliterated pulp chambers and root canals, visible radiographically.

Histologically, dentin is composed of irregular tubules, with large areas of uncalcified matrix in patients with this condition. Tubules tend to be larger in diameter and less numerous than normal in given volume of dentin. In some patients, there is complete absence of the tubules. Odontoblasts have only limited ability to form well organized dentin matrix, and they appear to degenerate readily, becoming entrapped in this matrix. (Orsini *et al*., 2014) The pulp chamber is usually obliterated by continued deposition of dentin. The dentinoenamel junction (DEJ) is not scalloped and appears flattened although it appears qualitatively normal. Enamel appears defective with subtle hypocalcification defects in enamel rods just above the DEJ. In most cases, the structure of the mantle dentin is normal, whereas the dentinal tubules of the circumferential dentin are coarse and branched. The presence of an atubular area in the dentin with reduced mineralization and a reduced number of odontoblasts are important findings. Pulpal inclusions & much interglobular dentin are also

frequent. (Teixeira *et al*., 2008) This condition may have syndromic associations which include osteogenesis imperfecta, Ehlers Danlos sydrome, Brachio-skeleto-genital syndrome and osteodysplastic primordial short stature with severe microdontia, opalescent teeth, and rootless molars. (Devaraju *et al*., 2014)

Conclusion

Timely diagnosis and appropriate treatment is of paramount significance to prevent psychological and functional morbidity. The treatment is directed primarily towards preventing the loss of enamel and subsequent loss of dentin through attrition. Treatment of mixed and permanent dentition is challenging and frequently demands a multidisciplinary approach. The treatment objectives include :

- Preserving vitality, form and size of the dentition;
- Maintaining esthetic appearance at early age to prevent psychological problems;
- Providing a functional dentition;
- Preventing loss of vertical dimension;
- Avoiding interference with eruption of the remaining permanent teeth;
- Allowing normal growth of the facial bones and TMJ.

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