Case report

DENTINOGENESIS IMPERFECTA: A Case Report and Review of Literature

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ABSTRACT

BACKGROUND: One of the commonest hereditary disorders of dentine formation is dentinogenesis imperfecta (DI). It is inherited in an autosomal dominant fashion thus affecting the formation and mineralization of dentine, which makes the teeth prone to attrition and fracture. Early diagnosis and treatment is recommended to prevent or mitigate further deterioration of occlusion and teeth, and to improve esthetics.

OBJECTIVE: A report of a case of dentinogenesis imperfecta type II who presented on account of sequelae of the dental anomaly, to highlight the importance of early diagnosis and treatment of the condition.

CASE REPORT: A case report of a 14-year-old female presenting with dentinogenesis imperfecta with clinical and radiological evidence. There were associated (i) acute apical periodontitis of tooth 46, (ii) longitudinal crown root fracture of tooth 36 secondary to occlusal trauma. Both teeth were extracted; her 26, 27, 37 and 47 were billed for stainless steel crown placement and acrylic jacket crown for 11, 21, 31 and 41.

CONCLUSION: Early diagnosis and treatment of DI is recommended to prevent or intercept deterioration of the teeth and occlusion, and thus improve aesthetics. Long term follow-up is imperative to prevent developing complications and adjust the treatment to the changes in the dentition and occlusion.

Keywords: Dentinogenesis imperfecta, Sequelae, Early Diagnosis, Treatment.

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INTRODUCTION

Dentinogenesis Imperfecta (DI), also known as, Hereditary Opalescent Dentine, was first described in the late 19th century. It is an autosomal dominant disorder with variable expressivity. Mutations in dentine sialophosphoprotein have been implicated and it typically affects the dentine of both primary and permanent dentition (Regezi et al, 2017). Shields et al (1973) proposed three types of DI: Type I is associated with osteogenesis imperfecta, type II is found in patients with dentition abnormalities alone without bone disease, while type III is rare and only found in the tri-racial Brandywine

population of Maryland. Non-Syndromic DI is reported to have an incidence of 1:6000 to 1: 8000 births (Kim and Simmer, 2007) and has no sex predilection. The teeth of both dentitions are amber and translucent and show significant attrition; radiographically the teeth have bulbous crowns with marked cervical constriction and small pulp chambers (Barron et al, 2008). There is dentine hypertrophy leading to pulpal obliteration before or just after eruption. The roots are short and constricted.

In general, patients with DI require complex treatment that should take into account the degree of tooth destruction, age and cooperation of the patient (Sapir and Shapira, 2001). This involves a multidisciplinary approach including restorative, prosthodontics, and orthodontic treatments. Therefore, early diagnosis and treatment is key to preventing and/or intercepting its many sequelae. This is a report of a case of dentinogenesis imperfecta type II who presented on account of sequelae of the dental anomaly, to highlight the importance of early diagnosis and treatment of the condition.

CASE REPORT:

A 14-year old female presented at the Paediatric Dentistry Clinic of the University of Benin Teaching Hospital, with chief complaint of toothache in the mandibular right and left quadrants of 3 days duration and generalized tooth discoloration. The pain on the mandibular right quadrant was said to be worse than that on the left. Pain was spontaneous, sharp, disturbed sleep, aggravated by mastication and transiently relieved by analgesics prescribed over the counter. Pain from the mandibular left quadrant was dull, and intermittent, with no known aggravating or relieving factor. She gave a history of similar tooth discoloration in her primary dentition. She had no known underlying medical condition and medical history was essentially non-contributory. Family history revealed similar features in her mother.

Oral examination showed generalized opalescent teeth with grey to yellowish brown discoloration (figure 1). There was generalized chipping of enamel of anterior teeth with the dentine on the labial surface of 31 and 41 exposed (figure 2 and 3). Complete enamel breakdown of 46 with an opalescent sheen and was tender to percussion. Tooth 36 had a longitudinal undisplaced crown root fracture communicating with the pulp in a periapical radiograph (figure 4). Fractured cusps were also seen on multiple teeth. Oral examination also revealed simplified oral hygiene index of 2.

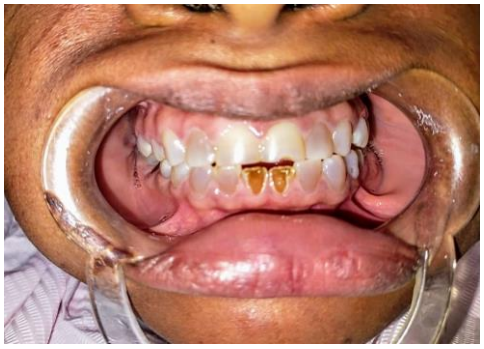

Figure 1: showed generalized opalescent teeth with grey to yellowish brown discoloration.

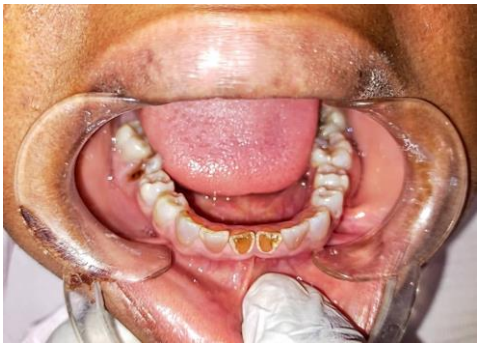

Figure 2: Show chipping of enamel of lower anterior teeth with the dentine on the labial surface of 31 and 41 exposed.

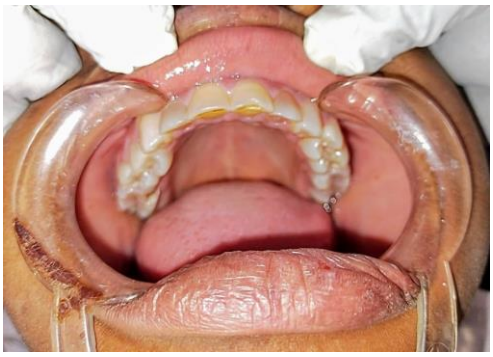

Figure 3: Show chipping of enamel of upper anterior teeth.

Intraoral periapical radiograph of 35, 36, 37 and 46, 47, 48 revealed markedly bulbous crowns, slender roots and obliterated pulp chambers. 36 revealed fracture of the distal root in its middle third (figure 5). Orthopantomogram showing the various affectations of teeth (figure 6).

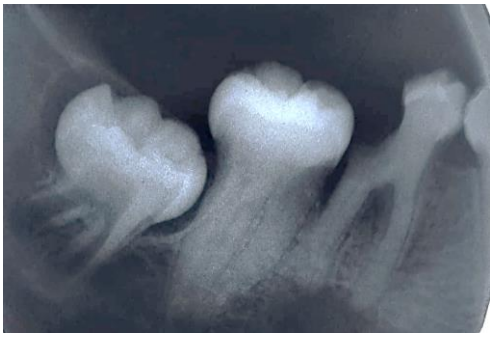

Figure 4: Tooth 36 had a longitudinal undisplaced crown root fracture communicating with the pulp.

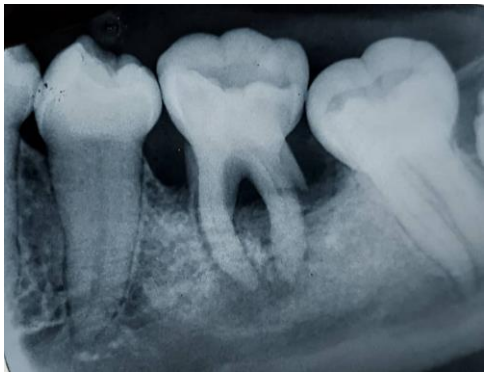

Figure 5: Show markedly bulbous crowns, slender roots and obliterated pulp chambers. 36 revealed fracture of the distal root in its middle third.

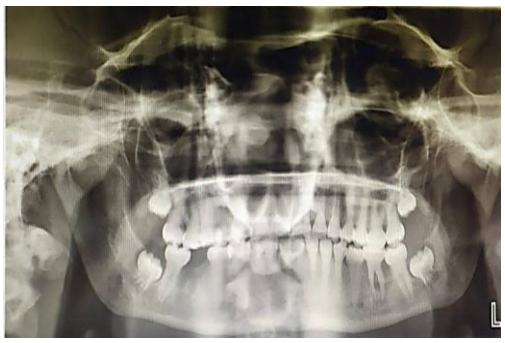

Figure 6: Showing the various affectations of teeth.

Based on the history, clinical examination and radiological investigations, a diagnosis of dentinogenesis imperfecta type II was made with the following sequalae viz: (i) acute apical periodontitis of 46, (ii) longitudinal crown root fracture of tooth 36 secondary to occlusal trauma. Both teeth were extracted; her teeth 26, 27, 37 and 47 were billed for stainless steel crown placement and acrylic jacket crown for 11, 21, 31 and 41.

DISCUSSION

Providing adequate functional and restorative treatment to children with dentinogenesis imperfecta can be very challenging for the paediatric dentist. Of importance is the early diagnosis and treatment which is an essential ingredient for obtaining favourable outcomes. Delay in intervention could result in more complex procedures and poorer prognosis (Subramaniam et al, 2008).

Different treatment options have been suggested in the management of dentinogenesis imperfecta; some of which include use of stainless-steel crowns, acrylic jacket crowns, over-dentures, pin retained crowns, crowns with aesthetic facing, removable appliances such as partial dentures or a combination of these (McDonald, 2004). The use of stainless-steel crowns in posterior teeth to prevent further tooth-wear from attrition is advised in primary and young permanent dentition and should be done immediately the tooth erupts (Wei, 1988). Even orthodontic treatment has been documented to be successfully done on patients with various degrees of dentinogenesis imperfecta (McDonald, 2004). Emphasis has been placed on restorations not being permanent due to the diminished hardness of dentine, thus when fractures occur at the gingival or below the gingiva, extraction is indicated (Shafer et al, 1993), as seen in the lower first permanent molars in the case reported.

A family history was noted in this report (the mother had a similar condition), although further investigation could not be done without consent. Such findings have been reported in a previous study with cases spanning generations, exhibiting various degrees of attrition attributed to age and degree of penetrance of the genetic defect (Bhandari and Pannu, 2008).

CONCLUSION

This present case report describes the findings in an adolescent with dentinogenesis imperfecta. Early diagnosis and treatment of DI is recommended to prevent or intercept deterioration of the teeth and occlusion, and thus improve aesthetics. Long term follow-up is imperative to prevent developing complications and adjust the treatment to the changes in the dentition and occlusion.

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