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Case report

Prosthetic treatment in dentinogenesis imperfecta type II: a case report

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Abstract

Introduction: Dentinogenesis imperfecta (DI) or hereditary opalescent dentin is an autosomal dominant disorder affecting both primary and permanent dentition. Early diagnosis and treatment of DI is important for normal facial growth and esthetic continuity by preserving occlusion and tooth structure. It also provides psychological motivation by increasing the patient's quality of life. Providing functional dentition in DI patients prevents loss of the vertical dimension, while enabling normal growth of the facial bones and jaw joint.

Case report: A 20-year-old male with DI was referred to our clinic with chewing difficulty and esthetic and speech problems. His brother also had this disease. Oral examination showed the loss of many teeth and the absence of enamel on most of the remaining teeth, causing discoloration and exposing soft dentinal tissue with calcification disorder. Despite widespread attrition of the teeth, pulp chambers were not exposed. The tip of the lower jaw was prominent in the patient's profile. Placing metal-ceramic fixed dentures in the lower jaw and an overdenture prosthesis in the upper jaw improved the patient's psychological state as well as his function, phonation, and esthetics.

Conclusion: This case report presents the intraoral findings in a patient with DI, including the histopathological findings, and the prosthetic treatment approach and the treatment outcome.

Keywords: Dentinogenesis imperfecta; prosthodontics; hereditary opalescent dentin

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Introduction

Dentinogenesis imperfecta (DI) is an autosomal dominant disease that especially affects the dentin. It occurs equally in both sexes and can affect both the primary and permanent dentition. The incidence of the disease is 1:8000.[1,2] DI is classified into three types: type I is associated with osteogenesis imperfecta; type II has essentially the same clinical, radiological, and histological findings as DI type I, but is not seen with osteogenesis imperfecta; and type III is a rare type seen in the Brandywine tri-racial population from Maryland and Washington DC, in the United States of America.[3]

Clinically, in DI, the teeth are of normal size, but the crowns are bulb-shaped. When the enamel does not have sufficient support, it is easily broken and removed, and the teeth appear opalescent, changing in color from brown to blue and sometimes amber or gray. Radiographically, the pulp chambers and root canals are narrowed or completely mineralized. The roots can be shorter than usual.[1-3] The enamel is normal or thinner than normal. There are hypoplastic or hypocalcific defects in the enamel in one third of DI patients. The exposed dentin may be subject to severe, rapid attrition.[3] Radiographically, narrow short bulbous crowns can be seen, and the pulp chambers can initially be abnormally large, forming "shell teeth." The root canal system clogs over time. Although the enamel structure is histologically normal, it tends to break. The dentin-enamel junction is not interwoven to the normal extent. Although the mantle dentin structure is normal in most cases, the environmental dentin tubules are irregular and decreased in number. Intertubular dentin mineralization and the number of odontoblasts are decreased. There is a collagen disorder of the dentin and primary defects in the calcified matrix. The cementum, periodontal membrane, and supporting bone are normal in appearance.[5-10]

Because DI patients need complex treatment, patient age, the degree of tooth destruction, and the patient's cooperation must all be considered. Benefits

of treatment include protecting the vitality, form, and size of the teeth by maintaining tooth health, protecting the esthetic appearance at an early age to avoid psychological problems, preventing loss of the vertical dimension to support functional dentition, and ensuring the normal growth of the facial bones and jaw joint via the permanent dentition. It is necessary to begin treatment as early as possible to ensure these benefits. However, poor patient cooperation at an early age may adversely affect treatment.[9,10]

In DI, primary teeth are influenced more markedly owing to their structural characteristics and the vertical dimension is lost quickly. Because problems occur more quickly with the primary dentition, to prevent loss in the vertical dimension, treatment should make use of overdenture prostheses or stainless steel crowns.[9-12]

Dentinogenesis imperfecta may require a multidisciplinary approach involving restorative, prosthodontic, and orthodontic treatments. The options for dental treatment include a dental prosthesis, faceting or stainless steel crowns on the front teeth, a simple removable appliance, metal–ceramic restorations, jacket crowns, and adhesive castings alone or in combination.[8-12]

This case report presents the oral and histopathological findings of a patient with DI and the prosthetic treatment.

Case report

Clinical findings

A 20-year-old man with DI presented to the prosthodontics clinic complaining of esthetic and chewing problems. His brother also had DI. The oral examination showed the loss of many teeth, and there was no enamel on most of the remaining teeth, which were discolored, exposing soft dentin tissue with calcification disorder. Despite the marked tooth loss, the pulp chambers were not exposed. In profile, the tip of the patient's lower jaw was prominent (Figures 1 and 2). Radiographically, he had impacted teeth and teeth with short roots and large pulp chambers (Figure 3).

Histopathological findings

Immediately after extraction, the teeth were placed in formalin fixative for 24 h. Following fixation, the teeth were washed overnight. Subsequently, they were decalcified in nitric acid, dehydrated, and embedded in paraffin. Then, 5 µm-thick sections were cut and stained with Masson Trichrome. The sections were evaluated under an Olympus BH-2 light microscope. The histology of the tooth specimens was assessed by a researcher blind to the case history. There were many irregular tubules within the dentin and many non-decalcified areas were observed (Figure 4).

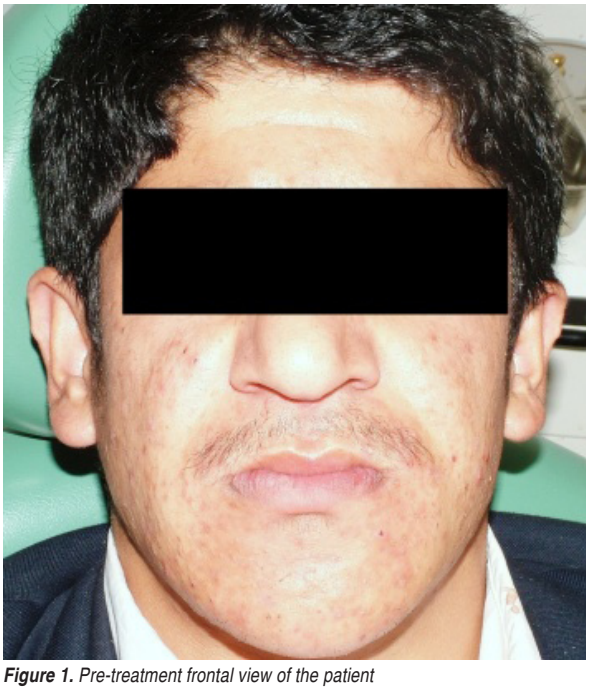

Figure 1. *Pre-treatment frontal view of the patient*

Clinical procedure

The impacted teeth were removed surgically, as were the teeth that would cause problems from prosthetic and periodontal perspectives. After healing was completed, endodontic treatment was performed on teeth numbers 13, 14, 15, 24, and 45. Then, the patient underwent periodontal treatment and was given oral hygiene education.

To restore the lost vertical size and the esthetic appearance, we decided to use an overdenture prosthesis for the upper jaw and metal-ceramic restorations for the lower jaw due to the lack of tooth support.

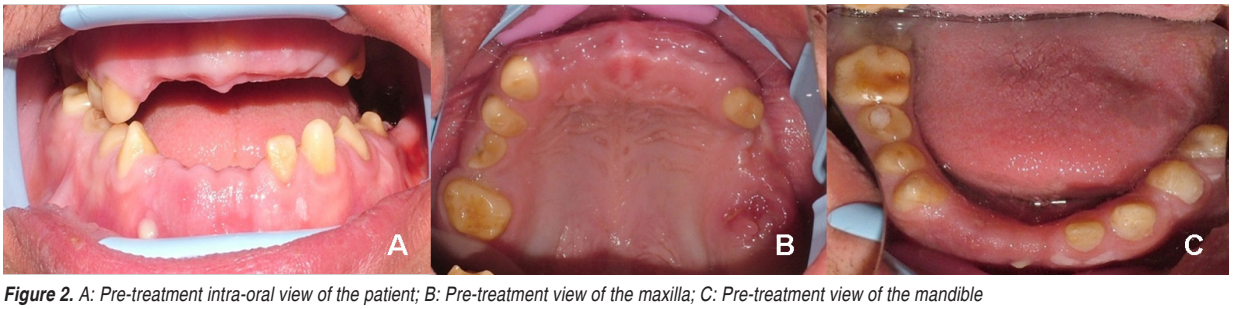

Figure 2. *A: Pre-treatment intra-oral view of the patient; B: Pre-treatment view of the maxilla; C: Pre-treatment view of the mandible*

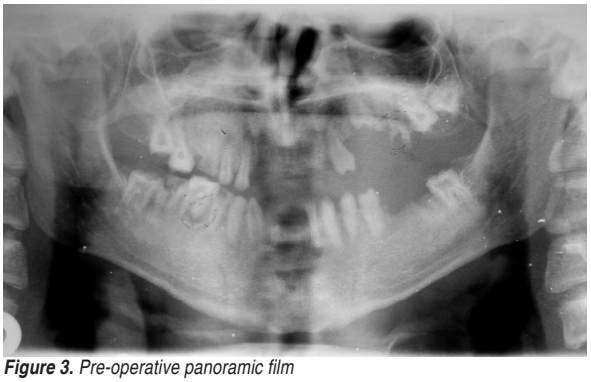

Figure 3. *Pre-operative panoramic film*

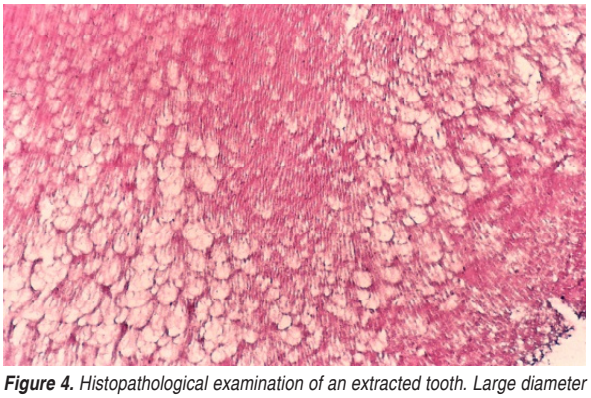

Figure 4. *Histopathological examination of an extracted tooth. Large diameter of irregular shaped tubules are observed within the non-decalcified matrix. (Masson-Trichrome, original magnification ×200)*

To determine the occlusal relationship, impressions were taken by using alginate. The resulting plaster models were placed in an articulator based on the patient's vertical dimension. In the prosthetic treatment phase, the teeth were prepared according to the principles for metal-ceramic restorations using the chamfer marginal termination design (Figure 5). The tooth preparations were measured using additional silicone impression material. Next, the mandible metal substructures and upper jaw copings were made from a metal alloy using a casting method (Figure 5), and the color properties of the teeth were determined with the traditional method using the Vita 3D Master color scale (Vita Zahnfabrik, Bad Säckingen, Germany). After porcelain was applied on the metal substructure for the lower jaw restorations, the necessary esthetic and functional evaluations were performed. The upper jaw copings were polished and the lower jaw restoration was glazed, and the cementation was done by using a zinc polycarboxylate cement.

After the restorations of the upper jaw copings were cemented, an individual tray was made from acrylic resin for the upper jaw. Then, an impression was taken by using hydrocolloid impression material applied into the individual tray. The overdenture prostheses were prepared using standard methods and delivered to the patient (Figures 6 and 7). We ensured that the vertical dimension was normal with the prosthesis. After the prosthetic treatment, the patient had no problems with the dentures, which restored the lost function and

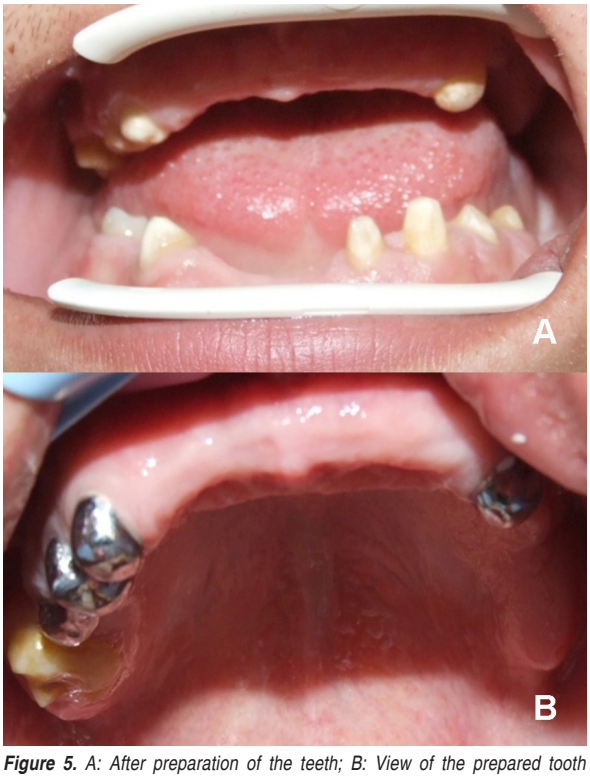

Figure 5. *A: After preparation of the teeth; B: View of the prepared tooth copings*

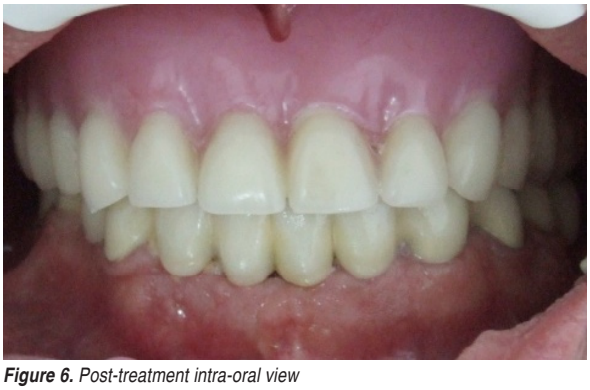

Figure 6. *Post-treatment intra-oral view*

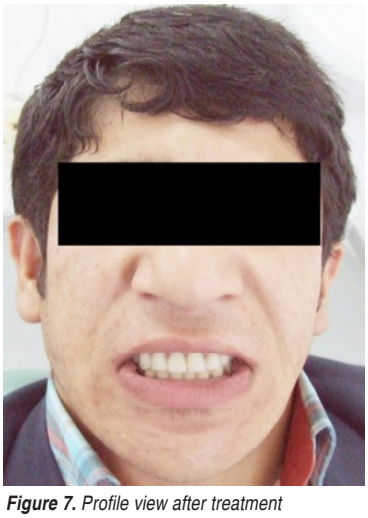

Figure 7. *Profile view after treatment*

esthetics. The patient is being followed every 6 months to check the prosthesis and the oral hygiene.

Discussion

As the patient had no findings of osteogenesis imperfecta, he was diagnosed with DI type II based on the clinical, radiological, morphological, and histopathological examination. DI or localized mesodermal dysplasia is an inherited disease that affects the dentin and both the primary and permanent dentition. DI types I, II, and III are caused by the dentin sialophosphoprotein (DSPP) mutation.[13] As a result of defective dentin formation, the teeth are prone to break. The main oral problems seen in DI are residual roots, a reduced vertical dimension, hypodontia, and extensive decay. Early diagnosis and treatment are necessary to protect occlusion and esthetics in patients with DI. The need for invasive procedures can be avoided by preventing dental caries, periapical lesions, and premature tooth loss by proper oral hygiene, diet, and restoration.[13-16]

The patient's age, degree of tooth destruction, patient cooperation, and dentition period are the most important criteria in the selection of therapy for treating the oral problems in DI. Dentinogenesis imperfecta patients can develop severe esthetic and functional problems. For the primary and permanent dentition, crown protectors are recommended to minimize problems due to excessive tooth wear.[14,15] Stainless steel crowns on the anterior and posterior teeth are one restorative treatment used to restore the reduced vertical dimension as a result of tooth attrition; however, this can create problems both esthetically and due to the presence of large pulp chambers.[10]

In DI, the dentinal tubules are less affected by caries, as they are often clogged. However, enamel wear, fractures, and changes in the vertical dimension can result in a complex situation.[3,16] Another treatment to restore the lost vertical dimension is an overdenture prosthesis, which is often sufficient.[8-12]

A wide range of treatments are available for the permanent teeth in patients with DI: carbamide peroxide bleaching, veneers, dental implants, overdenture prostheses, faceting or stainless steel crowns on the anterior teeth, a simple removable appliance, metal-ceramic restorations, crowns jackets, and adhesive castings.[8-17] In the presented case, we used metal-ceramic fixed dentures in the lower jaw and an overdenture prosthesis in the upper jaw due to multiple tooth loss, with the aim of restoring the patient's vertical dimension.

In DI, the deterioration of the enamel-dentin junction results in the ready loss of enamel. The main goals of DI treatment are to protect the dentin from caries and to prevent tooth attrition, abrasion, and erosion. The dental treatment of patients with DI should protect the structure, esthetics, and tooth function. According to Gallusi *et al.*[18] bonding systems cannot form a true hybrid layer. However, there are no contraindications to composite resin restorations. Treatment usually involves prosthetic crown restoration. This should be considered in esthetic treatment planning in young patients.[19-21]

Conclusion

Patients are affected by DI to different degrees, and the treatment methods depend on the degree and severity of DI. In this case, after periodontal treatment of the abutment teeth in a patient with DI type II, the patient's function, phonation, and esthetic requirements were met with metal-ceramic fixed dentures in the lower jaw and an overdenture prosthesis in the upper jaw, resulting in a marked improvement in the patient's psychological well-being.

Conflict of interest disclosure: The authors declare no conflict of interest related to this study.

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Tip II dentinogenezis imperfekta'nın protetik tedavisi: olgu bildirimi

Özet

Tanitim: Dentinogenezis imperfekta (Dİ), herediter opalesan dentin olarak da tanımlanan hem süt ve hem de sürekli dentisyonu etkileyen otozomal dominant geçiş gösteren kalıtsal bir hastalıktır. Oklüzyonun ve diş yapılarının korunarak yüz gelişimlerinin normal devam edebilmesi ve estetik devamlılık için Dİ'nin erken teşhis ve tedavi edilmesi çok önemlidir. Özellikle teşhis ve tedavinin erken yapılması, hastaların yaşam kalitesini arttırarak psikolojik olarak motivasyonlarını sağlar. Ayrıca Dİ hastalarında fonksiyonel dişlenme sağlanarak dikey boyut kaybının önlenmesi, yüz kemikleri ve çene ekleminin normal büyümesi gerçekleştirilir.

Olgu bildirimi: Yirmi yaşındaki Dİ'li erkek hasta, estetik görünüm, konuşma problemi ve çiğneme güçlüğü şikayetleri ile kliniğimize başvurmuştur. Aile hikayesinde bu hastalıktan kardeşinin de etkilendiği öğrenildi. Hastanın ağız içi muayenesinde diş kayıplarının çok olduğu ve kalan dişlerinin çoğunda mine tabakasının tamamen ortadan kalktığı ve renklenmelerin olduğu, yumuşak ve kalsifikasyon bozukluğu gösteren dentin dokusunun açığa çıktığı görüldü. Dişlerdeki yaygın atrizyona rağmen pulpa odaları açığa çıkmamıştı. Hastanın profil görünümünde alt çene ucu çok belirgindi. Bu hastada, alt çenede seramik köprüler ve üst çenede overdenture protez uygulanarak, hastanın fonksiyon, fonasyon ve estetik gereksinimleri karşılanarak psikolojik olarak kendini daha iyi hissetmesi sağlanmıştır.

Sonuç: Bu olgu bildiriminde, Dİ'li bir hastanın ağız içi bulguları, histopatolojik incelenmesi ve vakaya uygulanan protetik tedavi yaklaşımı sunulmuştur.

Anahtar Kelimeler: Dentinojenez imperfekta; prostodonti; herediter opalesan dentin