Multiple Teeth Fractures in Dentinogenesis Imperfecta: A Case Report

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Dentinogenesis imperfecta (DGI) is a hereditary defect consisting of opalescent teeth composed of irregularly formed and hypomineralized dentin. This paper presents the multiple fractures of DGI-affected teeth and suggests the reason of low fracture resistance by observing the dentin microstructures directly using scanning electron microscope (SEM) and by measuring its surface hardness using the Vickers hardness test. **.** *SEM revealed that while the enamel microstructure was similar in the DGI-affected and normal teeth, the microstructure of the DGI-affected dentin was poorly woven and more loosely packed than that of the normal dentin. The Vickers hardness of the DGI-affected dentin was 4.89 times softer than the normal dentin. The low fracture resistance of DGI-affected teeth can be attributed to the poorly woven microstructure of their dentin, which leads to a reduction in hardness.*

Key words: Dentinogenesis imperfecta, Teeth fractures, Scanning electron microscope, Vickers hardness test, children.

INTRODUCTION

Dentinogenesis imperfecta (DGI), which affect both deciduous and permanent dentitions, is a hereditary autosomal dominant disease.[1,2] The incidence of DGI is about 1 in 8,000 and is the most common of the gene-related dentin diseases.[3] Although the enamel of DGI-affected teeth has normal hardness, thickness, and color, the dentin is known to be brownish dark and more fragile than normal dentin, and exhibits obliteration of the pulp chamber and dentinal tubules as a result of increased odontoblast activity.[4-6]

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Several studies have described the microscopic structure of DGI-affected teeth with the published images showing reduced or obliterated dentinal tubules. Although there was a study[7] that found the presence of loose dentin structures in DGI-affected, undemineralized tooth sections, however, there have been no studies of the finer dentin microstructures, for example using high-magnification scanning electron microscopy (SEM). Furthermore, while it is known that the exposed dentin in DGI teeth exhibits severe attrition,[8-10] a quantitative comparison of the hardnesses of DGI-affected and normal dentin has yet to be performed.

Multiple fractures of DGI-affected teeth are presented herein, and an explanation for their low fracture resistance is proposed based on direct observation of the dentin microstructure using SEM and measuring the surface hardness using the Vickers hardness test.

Case Report

An 11-year-old boy visited the Department of Pediatric Dentistry, Yonsei University Dental Hospital (Seoul, Korea) for pain relating to both upper first permanent molars. An oral examination revealed multiple fractures of these molars and of the lower left central incisor, and a sinus tract on the apical area of the lower left central incisor. All of his dentition was a typical yellowish brown color (Figure 1A). The patient's history was devoid of blue sclera, hearing loss, joint laxity, or frequent long-bone fracture, and he had no known systemic diseases or recent history of trauma to the orofacial area.

Panoramic radiography revealed obliteration of the pulp chambers and pulp canals, bulbous crowns, constricted cementoenamel junctions, and roots that were shorter and thinner than found on normal teeth. In addition, radiolucent areas could be observed on

both upper first permanent molars, and a horizontal root fracture line was seen on the coronal one-third of the lower left central incisor root (Figure 1B).

The patient's family members and relatives also had brownish teeth. Given the family tree (Figure 1C), we concluded that this was an autosomal dominant disease. The patient was diagnosed with DGI type II.[1] The upper left first permanent molar was considered to be a hopeless tooth and was extracted and used for analysis.

Figure 1. Intraoral photographs, radiographs, and familial tree of a dentinogenesis imperfecta (DGI) type II patient. **(A)** Intraoral photographs. The arrow indicates a sinus tract due to root fracture of the left mandibular central incisor. **(B)** Panoramic radiograph and periapical radiographs. The arrows indicate fractured teeth. **(C)** Familial tree. The arrow indicates the patient who participated in this study

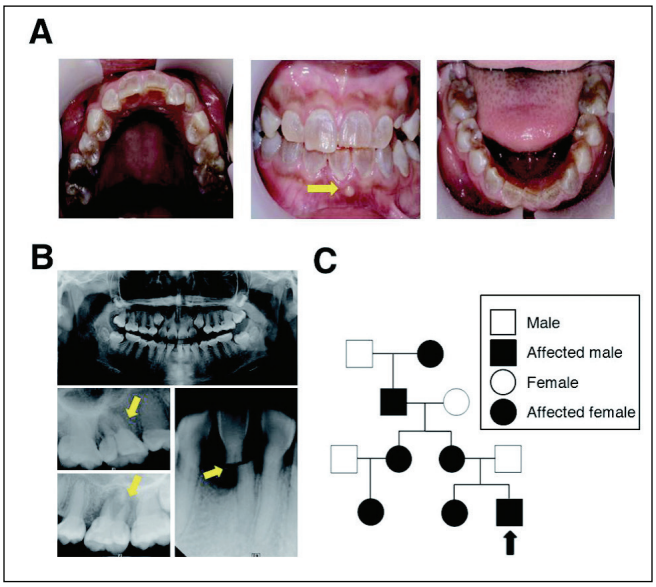
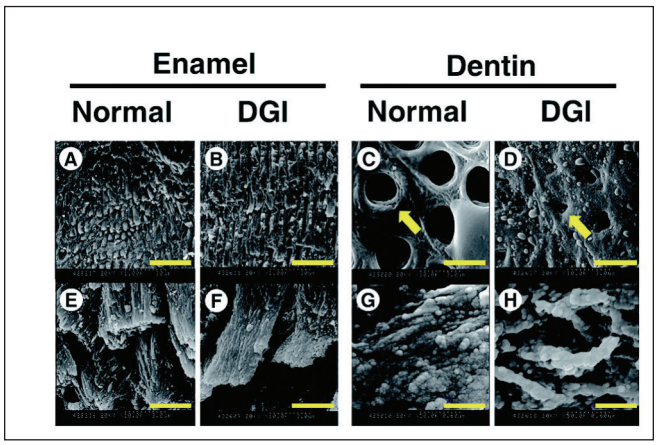


Figure 2. Scanning electron microscopy (SEM) images of normal and DGI-affected teeth. Normal enamel. (**A**, **E**) DGI-affected enamel. (**B**, **F**) Normal dentin. (**C**, **G**) DGI-affected dentin. (**D**, **H**) Arrows point to the dentinal tubules. Scale bars (original magnifications): 30 μm (×1,000) in **A** and **B**, 3 μm (×10,000) in **C**–**F**, 0.6 μm (×50,000) in **G** and **H**.



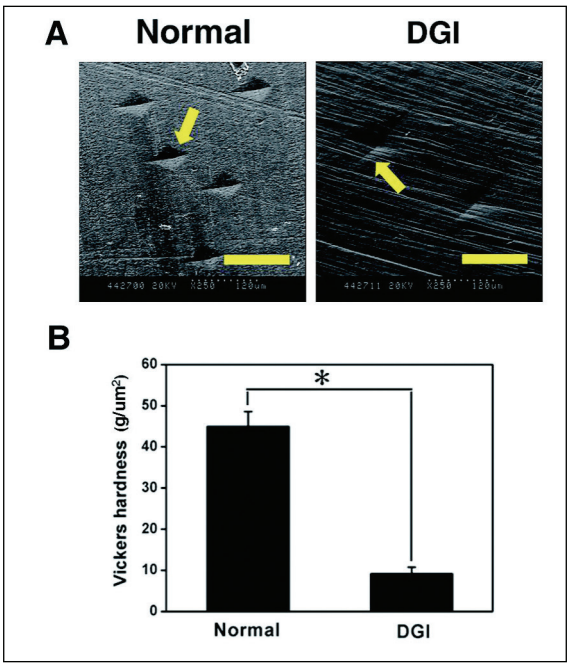
Analysis of SEM images

All procedures were approved by the Institutional Review Board of the Dental Hospital, Yonsei University, and informed consent to participate was obtained from both of the subjects (and their parents; approval #2-2011-0061). We studied the upper left first permanent molar from our patient and an upper right lateral incisor that was extracted for orthodontic reasons from an 11-year-old boy with no known relevant medical history who was not affected with DGI, as a control.

We fractured the palatal fragment of the extracted upper left first permanent molar and the control tooth in half after saline irrigation. The organic substrates of the prepared specimens were removed by soaking them for 1 week in a 5% solution of sodium hypochlorite (Duksan Pure Chemicals, Asan City, Korea). The remaining inorganic substance of each tooth specimens was irrigated with saline and then fixed for over 6 hours in Karnovsky fixation solution (2% glutaraldehyde, 2% paraformaldehyde, and 0.5%$CaCl_2$; all purchased from Merck, Frankfurt, Germany). The resulting specimens were observed using SEM (S-800, Hitachi, Tokyo, Japan).

SEM images revealed crystalline hydroxyapatite in both the DGI-affected and normal enamel, with no significant qualitative difference between the two groups (Figure 2A, B, E, and F). However, the dentin microstructure differed between the DGI-affected and normal teeth; the dentinal tubule diameter was

Figure 3. Vickers hardness test in normal dentin and DGI-affected dentin. **(A)** SEM images made after the Vickers hardness test. Arrows indicate the indentations that were formed with a loading time of 10 seconds and a loading force of 200 g. Scale bars: 120 μm. **(B)** Vickers hardness values with the loading conditions of 10 seconds and 50 g. *Significant at $p<0.05$ (Mann-Whitney U test, $n=10$).



smaller and the intertubular area thicker in the former than in the latter. That is, obliteration of the dentinal tubules and a reduction in the dentinal tubule opening size was observed in the DGI-affected dentin (Figure 2C, G). Higher-magnification examination disclosed many gaps between the mineralization crystals, giving the dentin a loose crystal pattern, while the structure of the normal dentin was dense and packed closely (Figure 2D, H).

Vickers hardness test

The two specimens were embedded in an acrylic resin mold (polymethylmethacrylate, Dentaurum, Ispringen, Germany). The observation point, which was set as the middle one-third of the root, was exposed and polished flat using 600-, 800-, 1200-, and 2000-grit silicone points (Shofu, Kyoto, Japan). The surface hardness was then measured using a Vickers hardness tester (**DMH-2, Matsuzawa** Seiki, Tokyo, Japan). Ten indentations were made randomly in the observation area, with loading conditions of 50 g and 10 seconds. The data collected from the samples were recorded, and the Mann-Whitney U test ($p<0.05$) was performed using SPSS for Windows version 12.0 (SPSS, Chicago, IL, USA).

The indentations of both samples were diamond-shaped. The diagonal lines of the indentations were much longer in the DGI-affected dentin than in the normal dentin (Figure 3A). The surface of the normal dentin was 4.89 times stronger ($p<0.05$) than that of the DGI-affected dentin (Figure 3B).

DISCUSSION

DGI-affected teeth are generally known to have low wear resistance[8] and tend to fracture early.[11] Preiswerk[12] reported early crown fracture, severe dentin attrition, and brownish discoloration in DGI-affected patients with osteogenesis imperfecta, and Lee *et al* reported early crown fractures in DGI-affected patients without osteogenesis imperfecta.[13] Similar to the patient in the present study, a class II crown fracture of the upper central incisor was reported in a 7-year-old boy who had no history of trauma.[14] The patient in the present study was unable to recall any particular history of trauma, which means that the force that had fractured his teeth would have been minor. Thus, we inferred that DGI-affected teeth have a low fracture resistance. The etiology of this low fracture resistance could be attributable to two aspects, macroscopic and microscopic. At the macroscopic level, characteristics such as bulbous crowns, cervical constriction, and short and thin roots[15] that are unable to completely support the tooth against external forces are thought to be responsible for the low fracture resistance of DGI-affected teeth. In addition, at the microscopic level, the loose dentin structure and lower level of hardness revealed herein could also explain the low fracture resistance.

Other studies have found differences in dentin structure between DGI-affected and normal dentin, usually based on histological observations made with the aid of an optical microscope. One study[16] found that the dentinal tubules in DGI-affected dentin were interrupted, another[17] found an irregular dentin structure in DGI-affected teeth. However, it is difficult to accurately reveal the microstructure of DGI-affected dentin when using optical microscopy.

SEM images of DGI-affected teeth have also been published previously. In one study the presence of dentinal tubule obliteration was observed at magnifications of ×1,000 and ×4,000,[9] and in another, SEM images revealed few or no dentinal tubules at a magnification of ×10,000[18] and that the structure of dentinal tubules in DGI-affected teeth was irregular at a magnification of ×7,000.[19] Thus, although SEM images produced at magnifications of lower than ×10,000 have been reported previously, the present study is the first to provide SEM images taken at a magnification of ×50,000, which can disclose the loose dentin microstructure at the intertubular dentin level but not at the dentinal tubule level.

DGI-affected dentin is known for its low mineral content. Kinney et al.[20] measured the mineral concentration in DGI-affected and normal dentin using synchrotron radiation computed tomography, and found it to be 26.5% (by volume) and 39.4% (by volume), respectively, providing quantitative evidence of the low fracture resistance of DGI-affected dentin. The presence of irregular dentinal tubules and mineralization defects of DGI-affected dentin on undemineralized ground sections has also been reported previously.[7]

The finding of exposed dentin in DGI-affected teeth being easily worn down after chipping away of the enamel layer has been reported previously,[8-10] but no previous studies have measured the surface hardness of DGI-affected dentin. We have established herein that the DGI-affected dentin in the present patient's extracted tooth was almost five times softer than that of the normal, control dentin. This low hardness is thought to be attributable to the poorly woven microstructure and mineralization defect in DGI-affected dentin.[20] Clinically, stainless-steel crown restorations have traditionally been recommended to prevent fracture and wearing of exposed dentin.[4,18] In addition, patients and parents should be advised that special care will be required to prevent trauma to avoid future fracture.

Despite these meaningful results, this report has a limitation: we used only one sample of each type (DGI-affected and control), and our single specimen may not be representative of all DGI-affected teeth. Thus, further study with many more subjects is needed to confirm our findings. Regardless of this limitation, the findings of the present study demonstrate that high-magnification SEM imaging and quantitative measurements of hardness, as performed herein, are required in order to understand the low fracture resistance of DGI-affected tooth, and this study may thus become the foundation for more advanced research.

CONCLUSION

The looser microstructure of DGI-affected dentin may be responsible for the low resistance to fracture in these teeth.

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