Dental Research Journal

Case Report

Dentinogenesis imperfecta associated with osteogenesis imperfecta

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Received: October 2011
Accepted: February 2012

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ABSTRACT

This paper presents a case with dentinogenesis imperfecta (DI) associated with osteogenesis imperfecta. Systemic and dental manifestations of OI and its medical and dental treatments are discussed in this paper. A 5-year-old child with the diagnosis of OI was referred to the Dental School of Shaid Beheshti University of Medical Sciences. On clinical examination yellow/brown discoloration of primary teeth with the attrition of the exposed dentin and class III malocclusion was observed. Enamel of first permanent molars was hypoplastic. Radiographic examinations confirmed the diagnosis of DI. A histological study was performed on one of the exfoliating teeth, which showed abnormal dentin. Primary teeth with DI were more severely affected compared to permanent teeth; enamel disintegration occurred in teeth with DI, demonstrating the need for restricts recalls for these patients.

Key Words: Brittle bone disease, dental anomalies, dentinogenesis imperfecta, osteogenesis imperfecta

INTRODUCTION

Osteogenesis imperfecta (OI) or brittle bone disease considered as a genetically heterogeneous connective tissue disorder which is characterized by bone fragility and thus repeated bone fractures.[1,2] Consequently skeletal deformities may arise as a result of reduced bone mass and frequent bone fractures.[3] The incidence of OI varies between 6 and 20 in 100,000 newborns and its prevalence is 4-10 in 100,000 individuals.[4] According to Sillence,[5] OI is classified based on clinical, genetically, and radiographic features in four groups [Table 1]. Type I is a mild form of OI. Type II is the lethal form of OI, even during the prenatal and perinatal period. Type III patients show progressive limb deformation. Patients with type IV of OI are those who show moderate to severe phenotypes and do not fit into any of the first three categories. Some of them demonstrate heterogeneous features, which are not even according to Sillence[6] classification. There are three other new but uncommon types of OI. Hence, patients affected by these types do not demonstrate DI and blue sclera;[6] OI types V-VIII are called syndromes resembling OI.[7]

A reduction in collagen type I synthesis results in type I of OI, while quantitative or qualitative alterations in type I collagen leads to types II, III, and IV. Therefore all tissues rich in type I collagen may be affected in these patients as a result of impaired collagen synthesis. Some clinical signs and symptoms may arise including DI, blue sclera, hearing loss, growth deficiency, and joint laxity.[2,7,8]

Dentinogenesis imperfecta (DI) has been reported in more than 50% of patients suffering from OI.[9] DI is a hereditary disorder of dentin formation, which exhibits mostly an autosomal dominant (AD) trait.[10] Although DI type I is the oral manifestation of deficient collagen formation and is mainly associated with OI, DI types II and III are related to a mutation in the dentin sialophosphoprotein (DSPP) gene.[10] Primary and permanent dentitions are both involved in DI type I, though primary teeth are more severely

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