Dentin dysplasia type I – a diagnostic challenge both for the dentist and paleodontologist

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Abstract
The last research of calcified tissue like bones and teeth has provided new information with regard to the genetic factors that control the formation of these tissues. The environmental factors like stress, different diet, long range mobility, and health of an individual can be also recorded in the skeleton and teeth. Dentin may be affected by morbid conditions present during development and, like enamel, may be affected by processes after eruption. Dentin dysplasia is one of the congenital disorders of mineralized tooth tissues. It is characterized by dentin abnormality. The pathological conditions affecting teeth have for reconstructing important aspects of the health of past human groups. The aim of the paper was to present the clinical manifestation of dentin dysplasia to get better acquainted with characteristic abnormalities in dentin which can be also used in paleodontology research. Despite many reports and descriptions of cases of patients with dentin dysplasia, etiology, diagnosis and treatment of this disease remain unclear so far but with a help of radiological characteristic image the correct differentiation and recognition is crucial. Another important point is proper and detailed documentation which can be also helpful in the future long term observation.

Keywords: dentin dysplasia; mineralized tissues disorders; paleodontology
Introduction

The last decade of calcified tissue research on mineralized tissues like bones and teeth has provided new information with regard to the genetic factors that control the formation of these tissues. The regulatory factors are now being studied in light of their effects on mineralized matrices, both in-vivo and in-vitro conditions. The use of molecular approaches in paleodontology is still a small field, but the combined information available in the proteins and the DNA from archeological bones and teeth has become a stable application in recent years. Unlike bone and dentin, enamel does not turn over, and once it is secreted it must last the lifetime of the individual. In the permanent dentition enamel bonds in the protein and carbonate generated in the pre- and early postnatal period. At death, a human adult skeleton and mineralized tissue consists of mineral and matrix that was formed at different times during the lifetime of the individual (1, 2). The stress, different diet, long range mobility, and health of an individual can be recorded in the skeleton and teeth, often in ways that are difficult to assess, but potentially of great import to paleodontology. Among hard tissues dentin has greater potential than enamel to respond to pathological processes. Odontoblasts fill the inner surface of dentin, and cellular extensions pass through the dentin but stop at the border with enamel. Blood and nerve supply, as the major components of the pulp cavity, are in close contact with dentin. Odontoblasts have limited potential to form secondary dentin, and this is often seen in worn teeth. Dentin may be affected by morbid conditions present during development and, like enamel, may be affected by processes after eruption. The pathological conditions affecting teeth indicate the potential that data on these conditions in archeological human remains have for reconstructing important aspects of the health of past human groups. Despite a substantial literature on paleodontology, a full understanding of dental paleopathology and its implications awaits further research. One of the very rare congenital disorders of mineralized tooth tissues is dentin dysplasia. It is a diagnostic challenge both for clinician and paleontologist. In archaeological samples the material is more or less damaged and it is hard to define the disease but results can provide information about potential dentin disorders (1, 2). In the clinic, patient with dentin dysplasia also provides a diagnostic problem.

So before paleodontology assessment it is worth to gain all clinical knowledge based on radiological and clinical signs and symptoms.

Objective

The aim of this paper was to present to the reader the clinical manifestation of dentin dysplasia based on detailed description of the patient affected with this disorder to get better acquainted with characteristic abnormalities in dentin which can be also used in paleodontology research.

Dentin dysplasia - general characteristics

Dentin dysplasia is one of the congenital disorders of mineralized tooth tissues. It is characterized by dentin abnormality. The changes related to dentin dysplasia may occur in other systemic diseases, such as generalized and nodular calcifications, rheumatoid arthritis, hypervitaminosis of Vitamin D, bone sclerosis and bone anomalies (4).

In 1973, Shields while classifying congenital anomalies of dentin, proposed the division of the abnormality into two types: dentin dysplasia type I and dentin dysplasia type II (3, 6). Witkop in 1975 suggested that dentin dysplasia type I (DD I) should be called radicular dysplasia and dysplasia type II (DD II) - coronal dentin dysplasia (3-8). Ciola et al., when describing changes in dentition having dysplasia type I and II, identified additional dentin dysplasia type III, also called focal or fibrous dysplasia. Dysplasia type III is described as a disorder occurring only in the permanent dentition (3, 4). Dentin dysplasia is a very rare disease entity, with the incidence estimated at 1:100,000 patients (3, 4, 9-11). This hereditary abnormality of the dentin structure occurs in both sexes with equal seriousness. The defect is inherited in autosomal dominant way. It may be associated with a disorder located on chromosome 4q 13-21 (3, 8, 9).

The mechanism of dentin-inherited disorders has not yet been fully elucidated (12). Dentin dysplasia may be caused by abnormal proliferation of Hertwig epithelial root sheath to the dental papilla. These cells induce abnormal differentiation of odontoblasts, resulting in the deposition of pathological dentin (3, 6, 8, 13). The formation of abnormal dentine according to the latest research is thought to be the result of
mutation in the dentin matrix acidic phosphoprotein gene (DMP) and dentin sialophosphoprotein gene (DSP) when epithelial cells from the Hertwig sheath break off and migrate into the dental papilla, where they produce ectopic dentin formation (11). Attempts have also been made to clarify the mechanism of periapical lesions in teeth with dentin dysplasia. It has been noted that Hertwig epithelial root sheath without the expected correct root length probably changes its activity, creating a cyst (3, 4).

Radicular dentin dysplasia (DD I)

In the radiographic analysis the dentition does not present abnormalities in relation to the teeth crowns that have normal size, shape and color, and are covered with hard, smooth, glossy enamel. The abnormal teeth present some resistance to decay (10, 11, 16). Clinical examination of dysplasia type I in permanent and deciduous dentition confirms that the degree of pathological mobility of teeth is related to the length of the roots conditioned by the progression of the disease (2, 5, 6, 8, 12). Also a marginal periodontal inflammation of varying severity has been observed (3). In some patients, the premature loss of deciduous and permanent teeth dependent on the length of the roots has also been noted. Examination of reactions of the pulp in patients affected by this anomaly gives a negative result or in some teeth - uncharacteristic response (10, 11, 16). Within the dental cervix, we can see a horizontal line indicating a weaker mineralization of dental hard tissues separating the crown from the root. At this line, dental fracture often occurs (12).

Radiographs of permanent dentition present periapical radiolucent areas (3, 10). Radiograph is very characteristic in patients with DD I. In primary teeth there is complete obliteration of the pulp chambers and root canals, and in permanent teeth, obliteration of the pulp chambers and the root canals is incomplete or complete depending on the severity of lesions (3, 5, 6, 9, 10, 13). Obliteration of pulp chambers appears before the eruption of teeth (6, 11). Abnormalities in radicular dentin dysplasia relate mainly to disorders in the structure of the tooth roots, which have a characteristic of the disease shortening and pointed shape and modified crown-root proportions (5, 6, 8, 9). Based on the analysis of radiographs of various cases of dysplasia dentine type I, Carroll et al. proposed 4 subtypes of DD- I, depending on the stage of malformation of the root, which they designated as DDI a-d (4, 7, 10, 11, 17, 18). In DD Ia, the most advanced form, there is complete obliteration of pulp chambers and absence of roots, or the roots are very small and short. In DD Ib, in cementoenamel junction projection there is a horizontal crescent-shaped line separating normal dentin from the atypical dentin within the tooth crown and short, conical roots. DD Ic shows 2 horizontal crescent-shaped radiolucent lines, concave towards each other. They are located at the cementoenamel junction, and the roots are shorter by half than normal and generally do not have the root canals. DD Id is the least advanced form characterized by visible reduced pulp chamber localized within cementoenamel junction. The roots have a near-normal length with visible canals, and large pulp stones that are located in the coronal third portion of the canal and create a localised bulging in the canal. In the anterior teeth, dental roots are dilated in the pulp stone projection (4, 11, 14, 15, 17, 18).

There is a characteristic occurrence of the bone structure thinning around root apices, described as cysts or granulomas. The presence of cysts or granulomas around the teeth without carious lesions is an important distinctive feature of this disease (3, 5, 7, 9, 11). The cause of their formation may be a weaker and defective development of bone and penetration of inflammation through a transverse line of weaker mineralization around the tooth cervix or through marginal periodontium (3, 16). Morphological picture of teeth with dentin dysplasia is very distinctive. Significant deviations from the norm seen in images under an optical and scanning electron microscope (SEM) relate to a small extent to enamel and mainly concern dentin. Within enamel we found interprismatic widening and in a limited section of enamel-dentine line - no close contact of enamel with the dentine. The dentine, being located circumferentially closest to the enamel, presented normal image. However, in the more inward-located dentin, there was a significant number of mineralized globules. It is an indication of improperly running processes in dentin mineralization. Proliferative changes were observed in the form of budding papillae, limited irregularly by wide canals, in which there were arranged in a disordered way crystal elements - this indicates a disturbed mineralization process of the tooth (8).
Clinical description of dentin dysplasia

A 26-year-old patient M. B. visited the dental clinic to supplement the missing tooth No. 24. Intraoral examination revealed missing tooth 24, single foci of dental decay, periodontal disease of chronic nature, and open bite with coexisting teeth crowding. Teeth were having amber color corresponding to the color A3.5 (Vita shade guide), with the correct shape. Response to stimuli of the pulp was incorrect (Figure 1).

The radiographs revealed abnormal structure of the roots of teeth in mandible and maxilla. They were significantly shortened, with changed shape, with absent root canal and obliteration of the pulp chambers. In the maxilla, premolars and molars had similar structure. Incisors and canines had normal structure of the root. Canines of the jaw showed the presence of pulp stones located in the root canal. Both in the maxilla and mandible there were single foci of the bone structure thinning formed without an obvious reason (Figure 2).

Medical history did not reveal such diseases as rickets resistant to vitamin D, Albight osteodystrophy, hypophosphatasia, brachio-skeletal-genital syndrome, Ehlers-Danlos syndrome. In the late childhood, fractures in the wrist bones and the bones in the ankle of the leg had occurred. The patient also reported premature loss of deciduous teeth probably due to decay. There were not confirmed vitamin deficiencies, anemia, hormonal disorders, liver and kidney disease, infections (chicken pox, mumps, syphilis, tetanus, measles, pneumonia), irradiation, genetic defects, allergies, and neurological diseases. Pregnancy proceeded well without signs of hypoxia. There was a breech birth. In the patient, heart defect in the form of pulmonary valve regurgitation was found.

Since dentin dysplasia is inherited as an autosomal dominant trait and appears in family members, the attention was put on the search for other cases among family. It was found that in the family history, the patient’s mother had been treated surgically for existing cysts of the jaws and periodontal diseases. Currently she uses dentures, suffers from rheumatoid arthritis. The mother’s brother had also been treated surgically within the jawbones during childhood. The daughters of the mother’s brother suffer from rheumatoid arthritis, also had been treated surgically because of cysts in the bones of the jaws. The patient’s father suffers from celiac disease. The patient’s younger brother also suffers from celiac disease. In the early childhood, he broke his leg. He had been treated surgically (tooth extractions) – Figure 3.

Based on dental extraoral and intraoral examination, medical history and characteristic radiological picture, type I dentin dysplasia was diagnosed finally.

Treatment plan accepted by the patient included instruction on oral hygiene, management of periodontal disease, treatment of individual carious lesions, replenishing the missing tooth 24 with single unit bridge. At a later stage, whitening in the anterior teeth was made using overlay method (Opalescence 16%) and tooth 27 was extracted of because of intensifying pain. The patient was satisfied with the treatment and informed about the prognosis. She was encouraged for control visit after 3 months and to continue preventive oral care.

Summary

Treatment of patients with dentin dysplasia requires multidisciplinary approach. Often, despite the conservative, periodontal and orthodontic treatment, the expected results cannot be achieved, and consequently there is a loss of permanent teeth in patients at a very young age. Then there is a need for prosthetic treatment, adjusted to patient’s age and occlusal conditions.

Despite many reports and descriptions of cases of patients with dentin dysplasia, etiology, diagnosis and treatment of this disease remain unclear so far but with a help of radiological characteristic image the correct differentiation and recognition is crucial. Another important point is proper and detailed documentation which can be also helpful in the future long term observation.
Figure 1. Clinical picture of patient with dentin dysplasia.

Figure 2. Panoramic X-ray of the patient with dentin dysplasia.
Figure 3. Panoramic X-ray of the patient’s brother – visible characteristic short roots.

References