

Dentin dysplasia type I

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This paper describes a rare case of genetically determined dentin dysplasia type I in 26-year-old male patient. The paper highlights anatomical and radiological aspects of dental abnormalities and emphasizes the significance of the education of both general practitioners and paediatricians as regards referring patients with diagnosed dentin dysplasia for a multi-specialty therapy. (Folia Morphol 2019; 78, 3: 637–642)

Key words: dental abnormalities, dentin dysplasia, pulp obliteration, computed tomography

INTRODUCTION

Dentin dysplasia is a congenital disease entity which may occur separately or as one of the symptoms of an underlying disease [7]. It is one of dental hard tissue disorders of genetic origin apart from such entities as amelogenesis imperfecta and dentinogenesis imperfecta [7, 11, 22].

The incidence of dysplasia is estimated according to Witkop at 1:100000 cases [2, 7]. It is a genetically determined pathology which occurs with the same frequency in both genders [13, 14, 23]. It is inherited according to the autosomal dominant inheritance pattern. The mutation probably occurs on chromosome 4q 13-21 [6]. The disorder formation mechanism has not been fully explained. Probably as a result of the mutation, abnormal Hertwig's epithelial root sheath cells migrate to the dental papilla which induces an abnormal differentiation of odontoblasts. Pathological dentin is formed as a consequence [15]. However, more recent research suggests that during the incorporation of Hertwig's epithelial root sheath into dental papilla the mutations of the dentin matrix

acidic phosphoprotein gene and the dentin sialophosphoprotein gene may occur [15, 17].

Dentin dysplasia is divided into dysplasia type I, also referred to as root dysplasia, and dysplasia type II, i.e. crown dentin dysplasia. Lesions with symptoms of both type I and type II are referred to as type III, i.e. focal or fibrous dysplasia [24]. Type III occurs least frequently and affects the permanent dentition only [3]. The dysplasia types are diagnosed and differentiated on the basis of the medical history and physical examination as well as a very characteristic X-ray image.

In the case of crown dentin dysplasia (type II according to Shields classification) the symptoms occur in both permanent and deciduous dentition. In primary teeth, normal crown shapes with the colour changed to amber or blue may be observed [7, 20]. The crowns are subject to a quick abrasion, and the roots have normal length and shape [1, 7, 20]. However, permanent teeth may have an only slightly changed colour without an increased susceptibility to abrasion [18]. Both types of dentition are not

prone to caries. The teeth are not prematurely lost because their mobility is not increased. There are also no changes in marginal periodontium. In an X-ray image, permanent teeth have the appearance of so-called shell teeth, and pulp chambers show an abnormal shape depending on the degree of pulp cavity obliteration [14, 23]. The pulp chamber obliteration begins to develop just after the tooth eruption. Incisors, canine and premolar teeth have chambers in the shape of a tube bulging towards the roots, and in the molars they take the shape of a flame or a thistle. Moreover, the occurrence of multiple intra-pulpal calcifications (denticles, pulp stones) may be observed [11, 12]. The occurrence of bone structure density reduction around the root apices is characteristic for this disease entity; however, such changes are not related to caries. The pathomechanism of formation of periapical lesions has not yet been fully explained. According to one of the hypotheses, probably due to a weaker and abnormal bone development, the inflammatory condition penetrates through the weaker mineralisation line in the region of tooth neck or through marginal periodontium. It is also likely that the Hertwig's epithelial root sheath, by not reaching the normal root length, modifies its activity and forms a cyst as a result. A second theory says that the "autointoxication" of the pulp occurs during its obliteration.

In the case of root dentin dysplasia (type I according to Shields classification) the pathological changes may be observed in both deciduous and permanent dentition. Initially, pathologically changed teeth were referred to as rootless teeth. In the clinical examination, no abnormalities in the tooth crown structure are observed. The shape is normal, and the enamel is shiny, hard and does not show any colour changes. In the clinical examination, also a transverse line in the region of dental neck separating the crown portion from the root portion can be seen. Due to a weaker mineralisation at this place, a tooth crown fracture may occur [4, 16]. In the course of further assessment, increased tooth mobility is found, whereas its degree depends on the disease progression. It is associated with shortened or even missing roots [14, 23], which is a reason for early tooth loss in both deciduous and permanent dentition. Also teeth in dysplasia type I are frequently extracted in connection with pain complaints which are not caused by complicated dental caries, but by granulomas or cysts similar to lesions occurring in crown dysplasia [10]. The en-

dodontic treatment is practically impossible due to obliterated pulp chambers and underdeveloped roots. The X-ray image of teeth with root dentin dysplasia is very characteristic. The roots are shortened, have a pointed shape and changed proportions in relation to the pulp chamber. Depending on the progression of underdevelopment of the root and the pulp chambers and canals, Carroll et al. [15] divided the disease into subtypes Ia, Ib, Ic and Id, where the subtype Ia means the mildest and Id the severest form of the disorder. In the case of deciduous teeth, the pulp chamber is subject to a complete obliteration. On the other hand, permanent teeth have a reduced dentin contrast, and the pulp chambers are completely or partially obliterated. In the molars, the pulp chamber frequently takes the form of a crescent. In contrast to crown dysplasia, the obliteration develops yet before the eruption of teeth. In dysplasia type I we may also find denticles which make the treatment even more difficult.

CASE REPORT

A 26-year-old male patient came to a dental office due to sensitivity to hot and cold and the episodes of idiopathic pain in right lower second molar (tooth 47). Based on reported symptoms and decreased response to pulp vitality tests, the decision of endodontic treatment was made. However, the trepanation of pulp chamber failed. An intraoral X-ray was taken and a completely obliterated pulp chamber and considerably shortened roots without a visible lumen of root canals were found. Then the patient was referred for an X-ray scan (a panoramic radiograph) and for the specialist endodontic treatment.

At the first visit, following detailed medical and previous treatment history taking, no systemic diseases were found. Patient previously suffered from pain complaints which were initially located mainly within the gums, and then the responses of the teeth to hot and cold appeared. In such cases he was treated only by the oral antibiotic therapy because he was told to have "difficult canals". He also reported that similar symptoms have occurred in the patient's siblings as well as in his father's twin brother and his children.

No abnormalities were found in the extraoral examination. The face was symmetrical, the ostia of the trigeminal nerve painless, the submandibular and submental lymph nodes not enlarged. The intraoral examination showed a normal colour of oral mucosa without visible pathological changes. No abnor-



Figure 1. Clinical picture of left vestibule.



Figure 2. Clinical dental status.



Figure 3. Panoramic radiograph showing the characteristic appearance of teeth with thick and short roots and pulp nodules in dentin dysplasia type I.

malities within the mandibular alveolar region were found. In the maxilla, however, the alveolar process distension (deformation) located between the left maxillary first premolar and left maxillary first molar and painless on pressure was observed (Fig. 1). The midline of maxillary teeth was shifted to the right. The teeth were abnormally positioned in the arch and the mandibular second premolar and first molar teeth were missing. No pathological tooth mobility was found. Clinically, the teeth had a milk-yellow colour, normal crown structure and proportion (Fig. 2). The subject had fillings in numerous teeth and a carious defect in the first right mandibular premolar as well as a temporary filling in the second right mandibular molar. The vitality test showed a lack of response to cold of the left upper lateral teeth and few right lower lateral teeth (teeth no. 24, 25, 26, 44 and 47).

Based on an analysis of the panoramic radiograph, an abnormal structure of pulp cavities and roots as well as the presence of translucent lesion located within the bones at the roots of multiple teeth were

noticed. In the maxilla, in the region of left premolars and molars, a shadowing with the diameter of about 3 cm, situated in the left maxillary sinus was found (Fig. 3). The decision to perform a cone-beam computed tomography (CBCT) scan was taken. The scan was performed using a NewTom 5G device in a resolution with the imaging field of 12 × 8 cm. An analysis of the scans showed that the entire dentition except for the canines was characterised by a shortening of the root length. The changes were mostly visible in the root structure of molar teeth (except for third molars) which were characterised by a pointed shape. In addition, they practically did not show any division into separate roots or such division started in 1/3 of the periapical region (Fig. 4). An almost complete obliteration of pulp cavities of lateral teeth was also found. The canal lumen was visible only within maxillary incisors and canines; a small outline of the pulp cavity could also be seen in the mandibular premolars (35 and 44). In the lumen of pulp cavities of anterior teeth, the presence of numerous spherical well



Figure 4. Sagittal section, left side. Visible characteristic structure of teeth with dysplasia type I and root cyst (an arrow) indenting into the lumen of the left sinus (visible above the first left upper molar tooth).

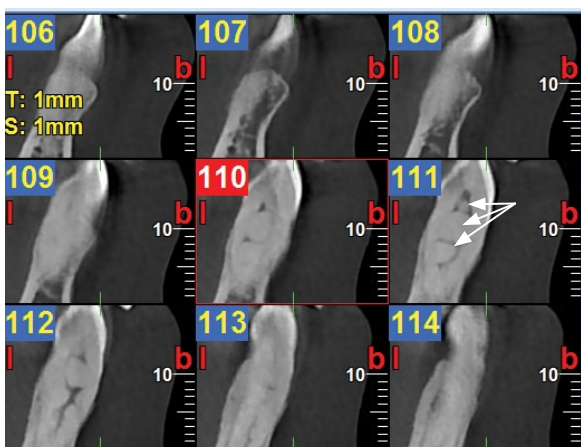


Figure 5. Sagittal sections through the right lower canine tooth. Visible denticles within the pulp cavity (arrows).



Figure 6. Odontogenic epithelial root cyst.

mineralised formations tightly filling the space, suggesting the presence of denticles, was found (Fig. 5).

In the projection of root apices of the right upper lateral teeth and in the mandible around the roots of the teeth 37, 35 and 44 the bone structure density reductions with a considerable degree of progression were present. At the level of the left maxillary lateral teeth (24, 25 and 26) a shadowing with the diameter of 3 cm indenting into the lumen of the left sinus was found.

Based on clinical and radiological examination, the patient was diagnosed with the dentin dysplasia type I with a considerable degree of progression.

The interdisciplinary treatment was started with oral hygiene instructions. Oral hygiene procedures as well as a closed curettage within the right lower second molar (47) were performed in order to reduce the inflammation around the root apex. The endodontic treatment was not possible due to the degree of pulp cavity obliteration. After the pain subsided, the defect in the tooth 47 was filled with the composite material. Furthermore, the patient was referred to the Maxillofacial and Plastic Surgery Clinic to have the cyst within the left maxillary sinus enucleated. Based on the maxillary computed tomography scan the range of the lesion was determined and its enucleation together with the extraction of the teeth 24, 25 and 26 was scheduled. The cystectomy (Fig. 6) and the root resection of the teeth 24 and 25 with the retrograde filling of root canals of these teeth were performed. The tooth 26 was not affected by the lesion. A pathomorphological examination of the specimen revealed an odontogenic epithelial root cyst. Due to the patient's young age, during the procedure the decision to leave the teeth 24 and 25 in the oral cavity was taken. Half year observation showed good oral tissues healing. At a later stage, an orthodontic consultation and a prosthetic reconstruction of existing dental defects were scheduled. The patient still remains under the care of a specialist team.

DISCUSSION

The dentin dysplasia still remains a disease entity with an unexplained aetiology. It may also cause difficulties at both diagnostics and treatment stages. Due to the lack of changes in crown shape and colour, the root dentin dysplasia (type I) is still in most cases discovered by accident. Patients most frequently decide to see a dentist because of pain complaints, and the difficulties in locating the canal ostia force dentists to consult specialists who diagnose a dysplasia based on a characteristic X-ray image (panoramic

radiograph). Previously, such extraoral images were the main source of information about the status and the anatomy of patient's teeth; however, at present, in each case of suspected complex structure of pulp cavities it is recommended to perform a CBCT scan [8]. The CBCT scan makes it possible to precisely determine the degree of dental canal obliteration and the place where denticles occur, which increases the chance of proper treatment. In spite of clear indications for a CBCT scan, no case study of root dentin dysplasia with the use of such imaging was found in the available literature.

The significance of multi-specialty therapy to which each patient with diagnosed dentin dysplasia should be subjected already at the stage of deciduous dentition is to be emphasised. Unfortunately, due to both parents' and dentists' insufficient knowledge and awareness of hereditary origin of this disease, a premature loss of teeth rarely induces them to perform an extended diagnostics in family members. This is why the education of both general practitioners and paediatricians who frequently are primary care physicians is so significant.

The aim of the therapy is to make sure that the teeth remain in the oral cavity for as long as possible. It comprises complex restorative, orthodontic, periodontal, surgical as well as prosthetic treatment. It is necessary to put emphasis on the prevention which consists not only in removing dental plaque at a dental office with the use of fluoride preparations, but also in providing appropriate patient education. Professional oral hygiene instructions including also proper dietary habits and frequent follow-up visits allow reducing the risk of occurrence of dental caries and periodontal diseases which may further weaken the periodontal apparatus. In many cases, like in the case described above, the endodontic treatment is not possible, which is why it is so important to prevent pulp complications [5]. It is frequently possible to suppress the inflammation around root apices by curettage procedures with the application of medicines into gingival pockets [9, 19, 21].

The orthodontic treatment in dentin dysplasia is quite controversial because of the action of excessive forces which may lead to a faster bone resorption, and consequently to an increased tooth mobility. However, it is important to eliminate excessive occlusal overloads or occlusal disease symptoms and to restore normal occlusion. The surgical treatment consists not only in dental extractions, but also, due

to resection or hemisection procedures, enables the diseased teeth to survive longer despite the lack of possibility to carry out proper endodontic treatment. The described case indicates the possibility of tooth retention even in the event of occurrence of an extensive root cyst. Unfortunately, in spite of the use of different treatment methods, a tooth loss frequently occurs in root dentin dysplasia patients. Then it is necessary to provide a prosthetic rehabilitation in order to restore the chewing function and improve the aesthetics of the patient's dentition.

CONCLUSIONS

In spite of an increasing number of reports of diagnosed new dentin dysplasia cases, the treatment of this disease entity is difficult and unpredictable. Therefore, it is important to keep records including not only case studies or photographs, but also a long-term patient observation, in order to choose the best possible therapy which should be individual for each patient.

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