Dental Management of Hypophosphatemic Vitamin D Resistant Rickets

Hipofosfatemik Vitamin D Dirençli Rikets’in Dental Tedavisi

Akif Demirel¹, Ayşe Tuba Altuğ², Esra Erdemli³, Firdevs Tulga Öz¹
¹Ankara University Faculty of Dentistry, Department of Pediatric Dentistry, Ankara, Turkey
²Ankara University Faculty of Dentistry, Department of Orthodontics, Ankara, Turkey
³Ankara University Faculty of Medicine, Department of Histology-Embryology, Ankara, Turkey

ABSTRACT
Vitamin D resistant rickets (VDRR) is an X-linked disease, causing mineralization disturbances of hard tissues such as bones and deciduous and permanent dentition. The dental findings of VDRR are enlarged pulp horns and chambers, defective enamel/dentin tissue, pulpitis, pulp necrosis, periapical recurrent abscesses and periapical complications without dental caries or trauma. The treatment options of this condition are extraction, endodontic approaches such as pulpotomy/pulpectomy, restorative and preventive applications. The aim of this case report is to summarize dental clinical, radiographical, histopathological findings and treatment options of VDRR in a 4.5-year-old girl.
Keywords: Vitamin D resistant rickets, dentinal clefts, dental abscess, histopathology, primary teeth

ÖZ
Vitamin D dirençli rikets (VDDR), X’e bağlı kalıtılan, daimi diş, süt dişi ya da kemik gibi sert dokularda mineralizasyon bozukluğuna sebep olan bir hastalıktır. VDDR’innin dental bulguları genişlemiş pulpa boynuzları ve odası, defektif mine ve dentin, pulpitis, pulpa nekroz ve travma ya da diş çırığı olmaksızın tekrarlayan periapikal apseler ve periapikal komplikasyonlardır. Bu durumun tedavisi diş çekimi, pulpa amputasyonu ya da kanal tedavisi gibi endodontic yaklaşım, restoratif ve koruyucu yaklaşımlardır. Bu olgu sunumunun amacı 4,5 yaşında VDDR kız hastanın dental klinik, radyografik, histopatolojik bulguları ve tedavi seçeneklerini özetlemektir.

Anahtar Kelimeler: Vitamin D dirençli rikets, dentinal clefts, dental abscess, histopathology, primary teeth

Introduction
Vitamin D plays a vital role in the absorption of calcium and phosphate. Low levels of vitamin D can trigger the body to release hormones that lead to the eventual loss of calcium and phosphate from bones, which causes insufficient bone mineralization. Vitamin D resistant rickets (VDRR) is also known as X-linked hypophosphatemia with a prevalence of 1:20000 people (1-4). It is characterized by a calcium and phosphate metabolism disorder affecting mineralized tissues bone and teeth (1,5). VDRR was first documented by Albright et al. (6) and mineralization defects of hard tissues in these cases are derived from renal transepithelial transport disturbance resulting in decreased tubular reabsorption of phosphate and hypophosphatemia (1,4,7). Physical findings are growth failure, bowing of the legs, short stature and walking disturbances (4,7,8). VDRR has been related to several primary and permanent teeth alterations. Dental findings of this condition are enlarged pulp chambers, pulp horns which spread outside the dentino-enamel junction, enamel and dentine defects, poorly defined lamina-dura, short roots and hypoplastic alveolar ridge (4,9-11).

Address for Correspondence/Yazışma Adresi
Akif Demirel MD, Ankara University Faculty of Dentistry, Department of Pediatric Dentistry, Ankara, Turkey
Phone: +90 506 287 33 80 E-mail: akifdemirel@ankara.edu.tr ORCID ID: orcid.org/0000-0002-1433-0452
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11. Histological analysis point to clefts in dental hard tissues especially dentine, marked globular dentine and increased predentine width (2,7,9). The combination of all these factors contribute to recurrent dental abscesses in VDRR cases. Bacteria and their toxins which come from the oral cavity to the pulp cause pulpitis, pulp necrosis, periapical recurrent abscess or periapical complications without dental caries (1,2,4,5,7,9-14). In VDRR cases, in spite of impairment of dentine mineralization, it is known that the odontoblastic cell function is normal (15,16). Additionally, dentine tissue is more affected than the enamel (4). Furthermore, because enamel and dentine formation occur between 4. months in-utero and 11. months of age, deciduous teeth defects cannot be avoided. However, permanent dentition development could be improved by medical therapy after birth. Therefore, mineralized tissue defects are more common in primary dentition than permanent dentition (3,4,16). This case report aims to present dental clinical, radiographic and histological features of VDRR case at our clinic.

Case Report

A 4.5-year-old female patient was referred to our clinic with the complaint of a speech problem, including delayed speech and difficulty in making some sounds. As a result of questioning her medical history, it was discovered that she had VDRR and its physical findings (Figure 1). Clinical dental examination revealed that it was fistula formation at the periapical area of the left maxillary primary central incisor without caries and a dental trauma history (Figure 2) and as a result of radiographic examination that a related tooth had a periapical lesion (Figure 3). Additionally, the right maxillary primary central incisor had colour changes but a cold vitality test showed a positive response. Furthermore, caries of mandibular primary first molars was diagnosed both clinically and radiologically. In addition, in speech examination, there were problems with the z/s sound and consultation with the orthodontics department showed that the patient had a “deep palatal vault”. The parents of the patient were informed about the treatment and their consent was obtained. Initially, the left maxillary primary central incisor was extracted and the mandibular first primary molars were restored with compomer (Dyract XP, Dentsply, DeTrey GmbH). After that, to avoid infantile swallowing, improve speech ability and for aesthetic purposes, a removable partial child prosthesis was applied (Figure 4). Histological evaluation was carried out in order to diagnose dental manifestation of VDRR accurately. In the histological assessment, a healthy primary incisor tooth which exfoliated physiologically used to compare with the patient’s left upper primary central incisor. After fixing the dental tissues of the extracted teeth with 10% buffered formalin, they were decalcified with 8% formic acid.
acid/8% hydrogen chloride. Following the routine histological preparations, the teeth were embedded paraffin sectioned to obtain sections 4 µm in thickness. These sections were stained using hematoxylin-eosin and examined under a light microscope (Zeiss AxioScope A1, Carl Zeiss, Germany).

Histological assessment of the teeth showed that, although the odontoblastic cell layer was intact, the predentin layer width increased when comparing with healthy primary incisor tooth (Figure 5). Additionally, the evaluation revealed that the specimens had increased predentine, marked globular dentine with hipomineralized areas, dentinal clefts and dilated dentinal tubules (Figures 6, 7). Finally, in order to provide oral health care, oral hygiene procedures were given to the patient and her parents. Topical fluoride varnish (Duraphat, Colgate-Palmolive, GmbH, Hamburg, Germany) was applied to all the teeth to avoid probable pulp pathologies and the patient had follow-ups periodically.

**Discussion**

VDRR is a disease which characterized by defective reabsorption of phosphate from the proximal renal tubule (1,7,9). Hypophosphathemic VDRR may be less severe when it occurs in the late stages of life and does not cause bone deformity. In a study of the orthodontic treatment of 9-year-old girl with VDRR, Kawakami and Takano-Yamamoto (17) reported that VDRR can be diagnosed by dental examination. In this VDRR case, histological examination of the extracted tooth showed dentinal cleft/dilated dentin tubules and these conditions agreed with the dental findings of VDRR. Furthermore, as noted in some cases (1,7,9), marked globular dentine, increased predentine width and intact odontoblastic cell layer were seen in the present case. The enamel in these cases was reported to be normal but thinner contrary to defective dentin tissue (4,11). However, sometimes, enamel cracks (18,19) and enamel hypoplasia (3,18,20) can be observed in patients. Prophylactic pulpal/endodontic treatments, conservative treatment and tooth extraction may be applied in these cases (4,5,21). Although the prophylactic pulpotomy is one of the treatment options of VDRR cases, it is not recommended due to lack of...
sufficient evidence of a good prognosis (21). In this case, tooth #61 was extracted because of fistula formation and periradicular lesion. Following extraction, dentin caries were restored and removable denture was applied to avoid infantile swallowing, physiological and speech problems (22). Additionally, dentin caries of the mandibular first primary molars were restored with polyacid-modified composite resin (compomer) to prevent probable pulpal disease. In addition to progressive caries and abscess formation, attrition of the dental mineralized tissues is also seen in VDRR cases (9). At this point, to avoid attrition and microfractures of enamel and dentine, prefabricated metal or polycarbonate resin crowns can be applied (23). However, in the present case, this application was not carried out because there was no defect in the enamel and the perforation risk of the pulp chambers (16). Finally, following the therapeutic, restorative and preventive applications, the patient was recalled for the periodical controls of the prosthetic appliance, general dental conditions, orthodontic examination and caries preventing procedures.

**Ethics**

Informed Consent: Consent form was filled out by the patient’s parents.

**Author Contributions**


**References**