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Premature Teeth Loss During Childhood – a Diagnostic Challenge

Paula Silva Braga,[1] Lorrany de Barros Del Piero,[1] Luciana Faria Sanglard,[2] Marcelo Massaroni Peçanha,[3] Maria Helena Monteiro de Barros Miotto,[2] Sérgio Lins de-Azevedo-Vaz,[2] Liliana Aparecida Pimenta de Barros,[2] Tânia Regina Grão Velloso,[2] Danielle Resende Camisasca[2]

[1]Graduate Program in Clinical Dentistry, Universidade Federal do Espírito Santo (UFES), Vitória, ES, Brazil
[2]Department of Clinical Dentistry, School of Dentistry , Universidade Federal do Espírito Santo (UFES), Vitória, ES, Brazil
[3]Department of Prosthetic Dentistry, School of Dentistry , Universidade Federal do Espírito Santo (UFES), Vitória, ES, Brazil

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Dear Editor,

Type I dentinal dysplasia (DD-I) is a condition that affects 1:100,000 patients; its etiology is unknown, although admittedly associated with a recessive mode of autosomal dominant inheritance.[1] This is a rare anomaly, first described by Ballschmiede in 1930,[2] characterized by a defect in dentin formation and classified by Witkop[3] in 1972 as Type I and II Dentinal Dysplasia. Type I presents normal clinical aspect of teeth, both in the primary and permanent dentition; radiographically, however, we can observe radicular malformation or even absence of the root, which justifies premature tooth loss. Type II presents dysplasia in the coronary dentin characterized by yellow, brown or grayish amber crown coloration, in addition to translucent deciduous teeth with complete or incomplete pulp obliteration.

The premature exfoliation of the teeth may be related to several causes, from the simplest ones (e.g., trauma), to vitamin C deficiency, syndromes, malignant neoplasms such as leukemia and Langerhans cell histiocytosis, aggressive periodontitis, hypophosphatasia and as mentioned, type I dentinal dysplasia.[4] When there is no evidence of periodontal disease or trauma, or evident biochemical changes, the final diagnosis can be challenging.

Thus, a severe case of premature exfoliation of both dentitions is described.

Clinical Case Report

A 6-year-old Caucasian boy, born in the state of Espírito Santo, was referred by his dentist to the Interdisciplinary Clinic for Child Patients of the School of dentistry of Universidade Federal do Espírito Santo (UFES), due to premature loss of deciduous teeth.

The patient is the only affected in the family. He is a second child of non-consanguineous parents, and the mother has diabetes. The mother reported difficulty of the patient in social interaction, precisely due to the absence of teeth.

Weight and height were compatible with his age both in neonatal examinations and throughout childhood. At three years of age, his skin was well-hydrated and regular in appearance, his hair was curly and voluminous, with visible eyebrows (Figure 1); there were no complaints related to sweating. He reported no history of fractures, bone pain, or delayed walking pace.

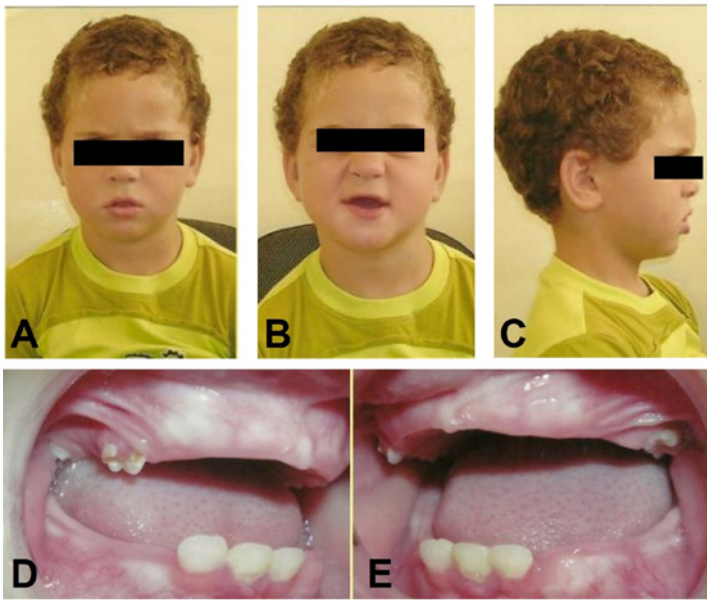

Figure 1. Extra- and intraoral photographs of the patient at 3 years of age. 'A-C': Hydrated skin, curly and voluminous hair, presence of eyebrows. 'D and E': Absence of deciduous teeth, with only the first molars and lower incisors present, with a crown smaller than usual. The presence of biofilm in the incisors and lesion suggestive of caries in the first deciduous molar is observed.

At 3 years of age, an intraoral examination detected the absence of deciduous teeth in the maxilla, with only the first molars present and a smaller than usual crown. The teeth present in the mandible were 31, 41, 42; the gums showed no signs of gingivitis or periodontitis, but there was the presence of microbial biofilm as shown in Figure 1. In periapical radiography of lower incisors, short and malformed roots were observed, with the presence of diffuse periapical bone rarefaction in teeth 31, 41 and 42 (Figure 2).

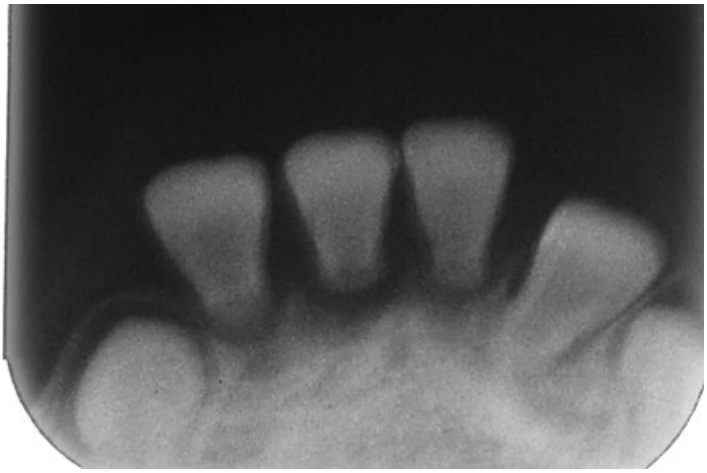

Figure 2. Periapical radiography: At the age of three. Presence of incisors and alteration in root formation.

For diagnostic clarification, the mother had already taken the child to a geneticist evaluation at 5 years of age. In the dysmorphological evaluation at first, syndromes and evident bone alterations were excluded. At this age, bone age was examined for the development of skull and thorax bones, and the hand and wrist ossification centers; the values obtained were compatible with the patient's age.

In a panoramic radiography performed at age 5, there was absence of lower incisors and permanent molars with incomplete rhizogenesis and taurodontism (Figure 3A). In a new panoramic radiography performed at age 6, the absence of teeth 11, 21 and 43 was noted (Figure 3B).

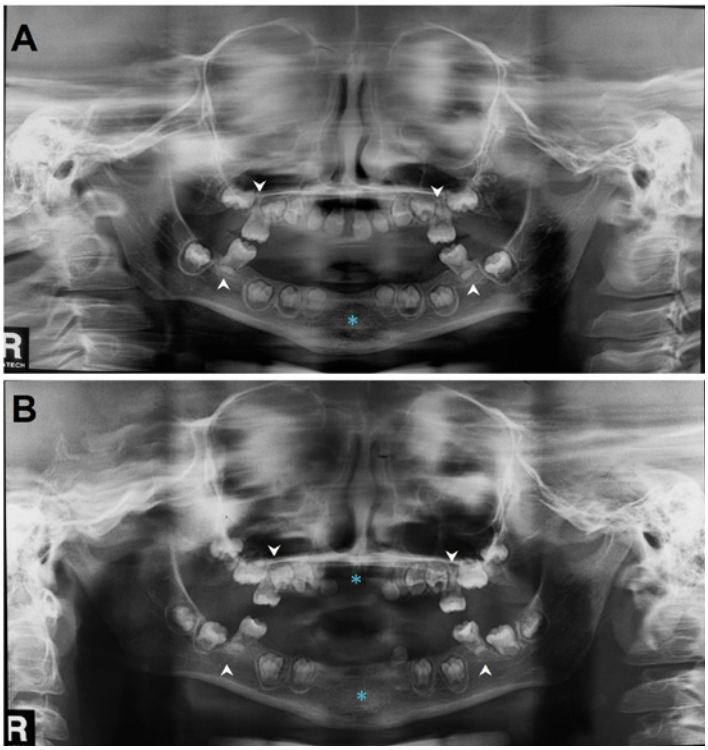

Figure 3. Panoramic X-rays. 'A': At age five. 'B': At age six. Absence of deciduous and permanent teeth in the region of the upper and lower incisors (*) and alteration in root formation and tooth size, as well as taurodontism (arrowhead).

Laboratory tests were required at ages 3, 4, 5, 6 and 7 of the child. Alkaline phosphatase increased at 3 years of age at 190 U/L (reference up to 115 U/L) and at 7 years of age it was with 160 U/L (reference up to 350 U/L). The values of urinary phosphoethanolamine (PEA) and pyridoxal-5'-phosphate in serum (PLP) were within the expected values. Total calcium levels in the serum at 6 years of age were at 10.4 mg/mL (reference 8.6 to 10.3 mg/mL), whereas serum values of phosphorus, potassium, ferritin, chloride, and magnesium were within normal limits.

The values for erythrocyte sedimentation rate (ESR) were 13 mm/h (reference up to 8 mm/h) at 5 years of age and for lactate dehydrogenase (LDH) 536 u/L (reference: 207 to 414 u/L), both above the normal reference values; in addition, in the erythrogram, the VCM and HCM were below the normal values at 3, 4 and 5 years of age and the leukogram showed a discrete eosinophilia (667 microliter) at 4, 5 and 6 years of age. Creatinine at 6 years of age of the patient was slightly elevated at 0.5 mg/dL (reference 0.29 to 0.48 mg/dL)

The tests of thrombrogram, glucose, uremia, creatinine, alanine aminotransferase (ALT), aspartate aminotransferase (AST), total bilirubin and fractions, feces, folic acid, protein electrophoresis, hemoglobin electrophoresis and vitamin B6 presented values within normal intervals in all tests performed at different times of the child's life.

The mother provided one of the deciduous teeth that underwent premature exfoliation for histopathological analysis, which, after decalcification, revealed the presence of dysplastic dentin (Figures 4 and 5).

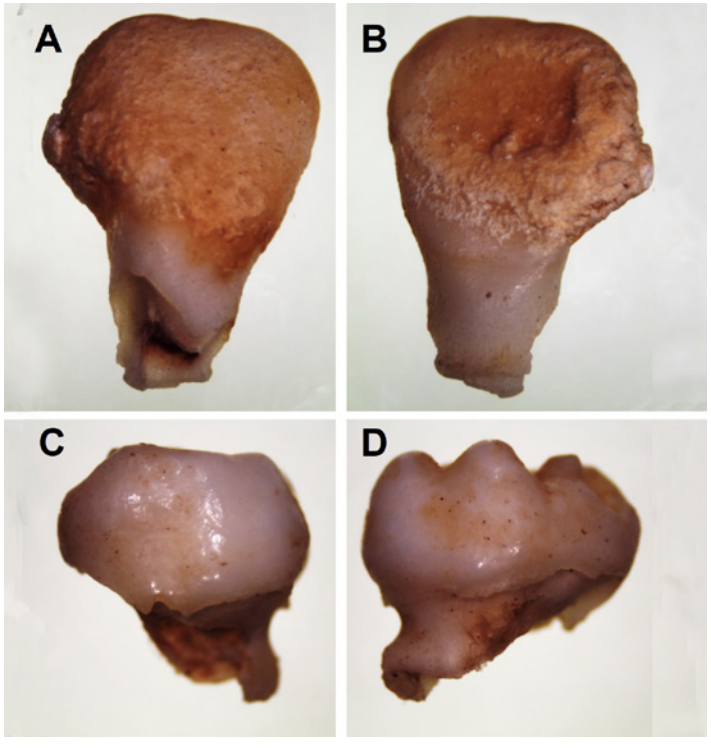

Figure 4. 'A e B': Lower incisor deciduous teeth, vestibular ('A') and lingual ('B'). 'C and D': Lower molar deciduous teeth, vestibular ('A') and lingual ('B'). Teeth given by the mother, after exfoliation, that were sent for histopathological analysis (stereomicroscopic view).

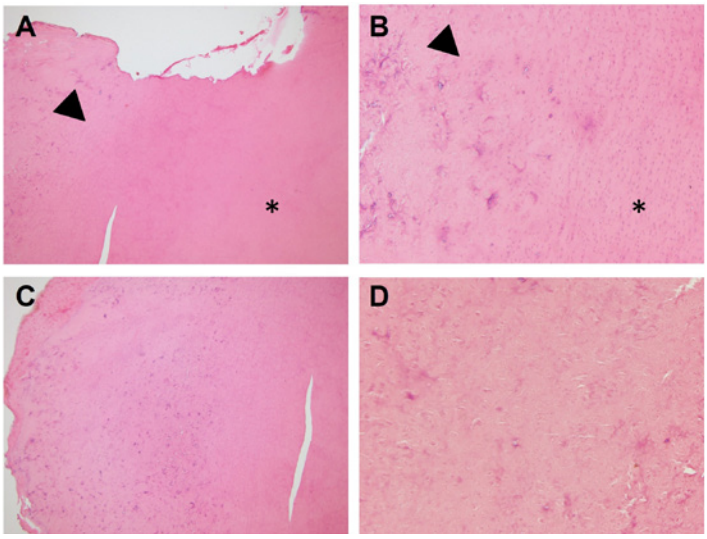

Figure 5. Histopathological aspect of dysplastic dentin. 'A e B': Normal (*) and altered dentin (arrowhead), with an absorption area ('A') (Coloration: Hematoxylin and Eosin (HE), magnification of 10x in 'A' and 40x in 'B'). C e D: Abnormal dentin with formation of globules and circles when approaching the apex (Coloring: Hematoxylin and Eosin (HE), magnification of 10x in 'A' and 40x in 'B').

Based on clinical, radiographic, biochemical, imaging and histopathological data, type I dentinal dysplasia was diagnosed.

Upper and lower removable prostheses were made for oral rehabilitation of the patient, to facilitate his social interaction and quality of life (Figure 6).

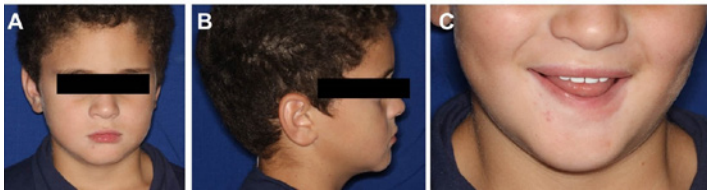

Figure 6. Rehabilitated 6-year-old patient carrying upper and lower removable prosthesis. 'A': Front view. 'B': Side view 'C': Patient smiling with his prosthesis.

Discussion

Type-1 dentinal dysplasia (DD) is an autosomal dominant hereditary condition,[5] however, in the analysis of the patient's family history, no cases of this disease were reported, and the patient was considered primary affected carrier.[6] Both deciduous and permanent dentition may be affected in type I DD,[3] as observed in our case.

Premature exfoliation of deciduous teeth may occur as clinical expression of both dentinal dysplasia (type I and type II) and hypophosphatasis.[7] Hypophosphatasia is a hereditary disorder characterized by defective mineralization of bones and teeth and low systemic values of alkaline phosphatase (ALP), an enzyme that participates in the process of formation of mineralized tissues[7]. Since the clinical pictures in hypophosphatasis are variable, it can range from stillbirths delivered without mineralized bone structure to only early loss of teeth without other bone symptoms. However, laboratory trials performed in our patient showed transient elevation of ALP and no decrease. At this time, the hypothesis of pseudohypophosphosfatasia was raised,[8] in which there would be no alteration of the ALP and that would be proven by the dosages of phosphoethanolamine and pyrideoxal 5'-phosphate (PEA and PLP) which are natural substrates for the ALP enzyme, however there were no changes in the markers PEA and PLP, which excluded this hypothesis.

Type 1 DD histologically shows apparently normal enamel while deeper layers of dentin present atypical tubular patterns and irregular organization with amorphous dentin, which has atubular organization.[9] When performing histopathological analysis of exfoliated teeth, the aspect of dysplastic dentin was well evidenced, which contributed to the diagnosis by associating it with previously observed clinical-radiographic information. Although hypophosphatasis also presents an atypical dentin formation there may be a hypoplasia of the enamel[10] and defective cementogenesis, which is a thin hypoplastic layer[7] that can be easily lost; however, the cement layer may also have been lost during manipulation of exfoliated deciduous teeth.

Changes in the roots may be present throughout the dentition or in only a few teeth, showing the variable expression of type I DD[11,12] differently from type II DD, in which the roots are present in normal size and shapes.[13] The present clinical case reports a condition characterized by teeth that have normal crown color, unlike cases of imperfect dentinogenesis, where there is an altered color aspect of the crown added to long and narrow roots.[14] However, our case portrays the absence of root formation and, when present, had a short, conical aspect with only a few millimeters, which distinguishes the case from both imperfect dentinogenesis and type II DD.

Furthermore, type II DD presents the yellowish, brown or amber gray appearance of the crown, as well as a translucent appearance, with complete obliteration of the pulp.[14] These characteristics highlight the disparity with our case in which the patient presented normal appearance and coloration of his teeth crowns during intraoral examination by visual inspection.

The physical examination performed at 3 years of age showing well-hydrated and regular-looking skin, ruled out Papillon-Lefevre Syndrome, which presents palmoplantar hyperkeratosis and even other skin lesions,[15] the patient also presented curly and voluminous hair and visible eyebrows, showing the possible absence of nutritional deficiency and incompatibility with Ectodermal Dysplasia, in which the hairs are sparse and cutaneous appendages may be altered, also with hypodontia and/or conical teeth.[16]

In the dysmorphological evaluation presented by the geneticist at 5 years of age, syndromes and evident bone

alterations (e.g., skeletal dysplasias) were discarded, suggesting aggressive periodontitis, which would be compatible with the values for erythrocyte sedimentation rate (ESR) 13 mm/h (reference up to 8 mm/h) and lactate dehydrogenase (LDH) 536 u/L (reference: 207 to 414). Such high values of inflammatory markers suggest aggressive periodontitis, which contrasts with the clinical findings of the patient's periodontal condition, i.e., the gum within normal standards.

The occurrence of periapical radiolucent areas associated with caries-free teeth is a finding described in type I DD,[17] verified on radiographic images in the apex region of the lower incisors. Most cases of DD in the literature interpret these images as root cysts,[18] however, they may represent granulomas or periapical abscesses. Periapical curettage and retrograde filling are alternative treatments but not recommended in teeth with very short roots.[18] Complete mouth rehabilitation is considered with prosthesis, as recommended in this case (Figure 6). Such prosthesis should be changed frequently, until body growth is complete and other forms of rehabilitation are possible, e.g., the use of implants and prosthesis on implants.

Conclusion

The diagnosis of type I dentinal dysplasia challenges the involved professionals, with histopathological examination being a fundamental tool as a diagnostic criterion associated with clinical-radiographic examinations. Type I DD patients requires a multidisciplinary approach and the treatment aims to reduce psychosocial and nutritional damage to the affected patient. This disease has severe social impact due to tooth loss, requiring rehabilitative treatment to improve the patient's quality of life.

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Mini Curriculum and Author's Contribution

1. Paula Silva Braga – DDS; MsC. Contribution: Effective scientific and intellectual participation for the study; data interpretation; preparation and draft of the manuscript; critical review and final approval. ORCID: 0000-0002-2911-869X
2. Lorrany de Barros Del Piero – DDS; MsC. Contribution: Effective scientific and intellectual participation for the study; data interpretation; preparation and draft of the manuscript; critical review and final approval. ORCID:0000-0002-6409-8807.
3. Luciana Faria Sanglard – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0002-2520-7062
4. Marcelo Massaroni Peçanha – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0002-1336-1839
5. Maria Helena Monteiro de Barros Miotto – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0002-3227-7608
6. Sérgio Lins de-Azevedo-Vaz – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0001-5578-5309
7. Liliana Aparecida Pimenta de Barros – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0002-1649-3116
8. Tânia Regina Grão Velloso – DDS; PhD. Contribution: effective scientific and intellectual participation for the study; data acquisition, data interpretation; critical review and final approval. ORCID: 0000-0001-6865-7955
9. Danielle Resende Camisasca – DDS; PhD. Contribution: Effective scientific and intellectual participation for the study; data acquisition, data interpretation; preparation and draft of the manuscript; critical review and final approval. ORCID: 0000-0003-1193-2113

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Corresponding author:
Danielle Resende Camisasca
E-mail: daniellecamisasca@yahoo.com.br