Regional Odontodysplasia Affecting the Maxilla

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Abstract

Regional odontodysplasia (RO) is a rare dental anomaly of unknown etiology that can affect both deciduous and permanent dentition. RO is characterized by severe hypoplasia of enamel and dentin, and teeth affected are friable and more susceptible to caries and fractures. Most of the lesions occur in the anterior maxilla and correlation with clinical and radiographic features is essential to provide a correct diagnosis. The major criteria for diagnosis are predominantly based on radiography, which shows presence of large pulp chambers and a marked reduction in the radiopacity of enamel and dentin, making the distinction between these mineralized structures difficult. Early diagnosis is important to minimize future sequels and allow preventive or conservative treatment. The therapeutic approach of the RO should be based on the degree of severity of the anomaly and in the individual functional and aesthetic needs of each case. A classic case of RO affecting the maxilla is exemplified in this Sine Qua Non Radiology–Pathology article.

Keywords Regional odontodysplasia · Ghost teeth · Primary dentition · Permanent dentition

History

A 10-year-old female was referred to the stomatology service of the School of Dentistry at Federal University of Sergipe, with a chief complaint of painless swelling in the right maxillary posterior region. During the anamnesis, the patient reported that the teeth of the affected region never erupted. No other symptoms were present.

The patient's mother also denied the use of illicit drugs during pregnancy, however, she took medication for pain

control (Acetaminophen®) in the first two trimesters of gestation. She also stated that the pregnancy and the time of delivery had no intercurrents. Moreover, no congenital or acquired diseases and no history of tooth or genetic anomalies in the family. Previous medical history was not contributory.

Clinical Findings

Extraoral examination was unremarkable (Fig. 1a). The intraoral examination revealed a swelling in the right maxillary, extending from the region of the canine to the tuberosity, covered by normal-colored mucosa, and with approximately 2.5 cm in diameter (Fig. 1b). The tooth 16 was partially erupted with only part of the cusp visible and exhibiting an irregular contour. Additionally, the surrounding gingiva was painful on palpation.

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Fig. 1 Extraoral and intraoral clinical aspects of the patient. **a** No evidence of facial asymmetry. **b** Intraoral view of the right upper arch compromised by regional odontodysplasia, extending from the deciduous canine to the maxillary tuberosity. The tooth 16 is partially

erupted and has a yellowish coloration. **c** Absence of alteration in the dentition of the lower arch. **d** Healthy and normal appearance 2 years after surgical removal of the lesion

Radiographic Features

A panoramic radiograph was requested (Fig. 2) and was observed that deciduous tooth 53 and permanent germs (13, 14, 15, 16 and 17) showed an increase in their radiographic density, characterizing malformation of the mineralized tissues of these teeth. The affected teeth had a fine radiopaque contour, without distinction of the limit between the enamel and the dentin. In addition, the pulp chambers of these teeth were broad suggesting the appearance of “ghost teeth”. The lower dentition showed parameters of normality, as well as teeth of left upper arch.

Diagnosis and Treatment

An incisional biopsy was performed under local anesthesia, and surgical specimen was submitted to the Service of Oral Pathology of the School of Dentistry at Tiradentes

University. Gross appearance revealed multiple soft to hard tan-white fragments. The teeth exhibited altered morphology, yellowish in color, soft or rubbery in consistency, and had short or unformed roots, and wide open apices. Microscopic examination of HE-stained histological slices revealed an unencapsulated loose tissue mass and the presence of small round calcifications associated with spindle cells arranged in a storiform pattern. Small islands of odontogenic epithelium, hyalinized dentinoid-like material, and basophilic psammomatoid calcifications, sometimes referred to as enameloid conglomerates, were also observed. Additionally, scarce lymphocytic inflammatory response was detected (Fig. 3). Based on the combined clinical, radiographic, and morphologic findings, the final diagnosis was RO.

The treatment plan consisted of surgical removal of the affected teeth and subsequent functional rehabilitation. Prosthetic rehabilitation with implants will be performed after the maxillofacial bones attained complete growth. The patient is currently undergoing routine follow-up.

Fig. 2 Panoramic radiograph findings in the teeth of the right maxillary segment. **a** The appearance of “ghost teeth” contrasting with the normal aspect in lower dentition. **b** Details of the “ghost teeth” showing large pulp chamber and incomplete root formation (arrow) without distinction of the limit between the enamel and the dentin

Fig. 3 Hematoxylin and eosin-stained sections. **a** Pulp tissue exhibiting calcifications. **b** Detail of pulp calcifications. **c** Odontoblast degeneration characterized by decreased amounts of cytoplasm, disorganization of the epithelial cell layers and lack of nuclear polarity

in cells. **d** Detail of dentin showing tubular structures with tortuous format and presence of areas of interglobular dentin. **e, f** Fibrous tissue containing small islands of odontogenic epithelium. **g** Basophilic calcifications sometimes referred to as enameloid conglomerates

Discussion

Regional odontodysplasia is a rare nonhereditary anomaly that affects the development of enamel and dentin of the deciduous and/or permanent dentition [1–3]. The first description of RO was probably made by Hitchin in 1934 [4], however, some authors credit to McCall and Wald (1947) [5] the first report of this condition. The term “odontodysplasia” was suggested by Zegarelli et al. only in 1963 [6]. Nowadays, the term “regional odontodysplasia” has become the accepted terminology because this anomaly has segmental involvement and a tendency to affect only one quadrant of the jaw bones [3, 7].

RO tends to occur during the eruption of the deciduous teeth or during the mixed dentition [1]. Most of the lesions occur only in the maxilla, as in our case, with a female predominance and there is no tendency to occur in a specific ethnic group [1, 2, 7]. Rarely more than one quadrant may be affected or the midline is compromised [2, 7]. Additionally, once the deciduous teeth are affected, usually the same malformation occurs with the permanent teeth [1, 2], data also coinciding with our findings.

The etiology of RO is still unknown [1–3]. However, several factors have been suggested including local circulatory disorders, infections, drug use during pregnancy, trauma, metabolic and nutritional disorders, and somatic mutations [2, 3, 7]. However, the condition is not hereditary, and this suggests one or more local factors affecting the formation of dental tissues during development [3]. Considering the mechanism involved in the RO development in our case, we can not speculate that local circulatory disturbances created local ischemia affecting the odontogenesis, once no alteration was mentioned to occur during the pregnancy by the mother. No infections, trauma or other disorders also experimented. Although the use of Acetaminophen was mentioned as analgesic by patient's mother, it should not be the cause; once according to the Food and Drug Administration (FDA), this medicament is listed in category B of pregnancy risk, being the safest analgesic for use during gestation [8]. Also, additional studies are necessary to confirm or exclude a possible association. We could not rule out the occurrence of somatic mutation in genes of tooth development, once it was not investigated. Failure in the embryogenesis during the migration of neural crest cells which contribute to pulp and dentin formation are also factors involved in the RO development [9], and could only speculate that it may have contributed to RO in our case.

The cases of RO are diagnosed by the combination of clinical and radiographic characteristics [2, 3]. Clinically, the teeth affected by RO are yellow or yellowish-brown in color, hypocalcified, and hypoplastic [2, 10]. Because

of the structural defects in enamel and dentin, these teeth are usually small and more susceptible to dental caries [1, 2, 10]. The alveolar ridge in the affected region is usually enlarged and covered by fibrous tissue [3, 7]. The eruption of the teeth can be delayed or not occur [1, 7]. Other signs and symptoms include non-inflammatory gingival enlargement, and some patients may report pain and develop abscesses in the affected area due to the bacterial access to the pulp through the cracks in the defective enamel and dentin [2, 3, 7].

Radiographically, the affected tooth presents a tenuous contour creating the appearance of a ghost tooth due to the marked reduction in radiodensity of the thin layers of enamel and dentin [1, 2]. The demarcation between enamel and dentin is poorly visible, the pulp chambers and root canals are broad, and short roots with open apices may occur [2], as demonstrated in this case report. Often, pulpal calcifications are identified in the coronary pulp and chronic or acute periapical lesions may develop in association with affected teeth [2, 3, 7].

In our case, the radiographic features consistently demonstrated teeth restricted to the posterior region of the maxilla with a “ghost-like” appearance, without distinction of the limit between the enamel and the dentin, pulp chambers and canals enlarged, and incomplete root formation. Therefore, our diagnostic hypothesis included mainly dental developmental anomalies with segmental distribution commonly observed in young patients such as RO. Conditions such as amelogenesis imperfecta (AI), dentinogenesis imperfecta (DI), hypophosphatasia (HPP), and dentin dysplasia (DD) show similarities to RO and should be differentiated [10, 11]. Furthermore, many cases of RO are misdiagnosed as malformed teeth or odontomas [3].

RO is a non-hereditary disorder, contrasting with the pattern of inheritance of AI, DI, HPP and DD [12–18]. Dentin dysplasia affects dentin formation of both dentitions [13, 14] and radiographically may show small or no roots (rootless teeth) and obliteration of the pulp chambers. In addition, the affected teeth often exhibit marked mobility [14–17], which was not detected in our case. Hypophosphatasia affects the development of bones and teeth [18, 19], with reduced levels of serum alkaline phosphatase (ALP) and clinical signs range since dental abnormalities as hypocalcification, short roots, enlarged pulp chambers, to alveolar bone loss and premature loss of the primary dentition (typically incisors) with intact roots [18–20]. In our case, no information about premature dental loss or childhood rickets suggesting HPP was mentioned, and (ALP) levels were within a normal range.

The characteristics of our case also rule out the diagnosis of AI and DI. In AI, the defective tooth structure is limited to enamel showing a thin layer on occlusal and interproximal surfaces [12], with no alteration of dentin, pulp chambers, and root, and all teeth of both dentitions are affected

[12, 21]. Differing from AI, in DI only the dentin is altered but with a varied translucency, which can give teeth a gray-blue tinge or brownish-yellow. In addition, DI radiographs show slender roots with ribbon-like pulp canals and bulbous crowns, cervical constriction, and obliteration of the root canals and pulp chambers [17], which were not observed in our case. Thus, adequate clinical and radiologic correlation is required to provide a correct diagnosis.

Histologically, it is observed an alteration of all the structures that compose the dental germ [2, 10, 22]. The enamel layer has a variable thickness, with irregular prismatic structures, as well as hypoplastic and hypocalcified areas [22]. In relation to the dentin, a thin layer and a reduced number of tubular structures with tortuous format are observed [2, 10]. The most significant feature is the presence of areas of amorphous dentin, interglobular dentin, and globular masses permeating the dentin tubules. The pulp tissue may also contain tubular or laminated calcifications [2, 13].

Carlos et al. (2008) have evaluated the ultrastructural characteristics of the affected teeth as well as the immunohistochemical features of the soft tissue associated with two cases of RO and observed that both enamel and dentin showed alterations, including alterations on the prismatic enamel surface, dentinal grooves, interglobular dentin, and hypoplastic and hypocalcified dental hard tissues. In addition, the soft tissue surrounding and covering the affected teeth showed calcifications and small islands of odontogenic epithelium positive for cytokeratin 19 [22]. In our case, many small islands of odontogenic epithelium and two types of calcifications were observed: (i) small round calcifications associated with spindle cells arranged in a storiform pattern, and (ii) eosinophilic calcifications with globular purple calcified material described as enameloid conglomerates. Several cases published in the literature show similar characteristics [2, 7, 10, 22, 23].

The treatment of RO is controversial [10, 23] varying from a more conservative approach, including the use of restorations to protect permanent teeth or simply long-term follow-up until the skeletal growth is complete, to immediate extraction of affected teeth, followed by prosthetic rehabilitation [3, 7, 23]. In the present case, due to delayed development and eruption of teeth and considering the susceptibility of affected teeth to infection, it was decided to extract the teeth compromised by RO and proceed with prosthetics rehabilitation with implants in the future after the complete development of the maxilla. The prosthetic rehabilitation besides the aesthetic improvement contributes greatly to the socialization of the child in the family and school environment. In both situations, the dentists should take into consideration the patient's age, medical history, number of teeth affected, presence or absence of any disease, as well as attitudes and desires of patients [2]. The purpose of the treatment should include measures to correct mastication

and phonation, promoting normal growth and development, reduce emotional impacts, and, if possible, protect some affected erupted tooth [2, 10, 23].

In conclusion, this case illustrates the classical clinical and radiographic features of RO. Although several cases have been reported, the literature is still limited to reports of individual cases. As a result, there is no consensus on the best therapeutic approach for this condition. Dentists should be familiar with dental development disorders so that a proper diagnosis and appropriate therapeutic strategy are performed, and thus minimize the clinical implications for the patient's dentition, improving the quality of life. Additionally, the presentation of this case helps general and pediatric dentists to review the clinical and radiographic characteristics of RO.

Compliance with Ethical Standards

Conflict of interest No conflicts of interest to disclose.

Ethical Approval For this type of retrospective case report, formal consent is not required.

Informed Consent No identifier information is included in the case report, and the study meets the waiver criteria for the institutional review board of Tiradentes University.

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