

Regional odontodysplasia: A case report

Odontodisplasia regional: relato de caso clínico

Abstract

Purpose: Regional odontodysplasia (RO) is a rare condition characterized by distinctive clinical, radiographic and microscopic findings. It is presumed to be the result of a developmental disturbance that locally affects the odontogenic ectodermal and mesodermal tissues. This report describes the clinical and radiographic findings of RO and its treatment.

Case description: A 15-year-old boy presented with an uncommon case of RO involving two quadrants of the jaws on the right side of his face.

Conclusion: A complex multidisciplinary team is required to fully rehabilitate a patient affected by RO. The treatment of RO is controversial and varies according to the individual patient.

Key words: Regional odontodysplasia; dental dysplasia; dental development; oral surgery

Resumo

Objetivo: Odontodisplasia regional (OR) é uma condição rara caracterizada por achados clínicos, radiográficos e microscópicos bem distintos. É o resultado de um distúrbio de desenvolvimento bem localizado afetando tanto o ectoderma quanto o mesoderma odontogênico. Este trabalho descreveu os achados clínicos e radiográficos de um caso de OR e os tratamentos.

Descrição do caso: Um menino de 15 anos de idade apresentou um caso incomum de OR envolvendo dois quadrantes dos maxilares no lado direito da face.

Conclusão: Há necessidade de uma equipe multidisciplinar complexa para reabilitar inteiramente um paciente com OR. O tratamento de OR é controverso e varia de acordo com as características e resposta do paciente.

Palavras-chave: Odontodisplasia regional; displasia dental; desenvolvimento dental; cirurgia oral

Rafaela Scariot^a
Imara Morosini^b
Cassius Torres Pereira^c
José Miguel Amenabar^c
Nelson Rebellato^d
Renato Gugisch^e

^a School of Dentistry, Pontifícia Universidade Católica do Paraná, Curitiba, PR, Brazil

^b School of Dentistry, Universidade Federal do Paraná, Curitiba, PR, Brazil

^c Discipline of Stomatology, Universidade Federal do Paraná, Curitiba, PR, Brazil

^d Discipline of Oral and Maxillofacial Surgery, Universidade Federal do Paraná, Curitiba, PR, Brazil

^e Discipline of Pediatric Dentistry, Universidade Federal do Paraná, Curitiba, PR, Brazil

Correspondence:

Rafaela Scariot de Moraes
Dr. Brasília Vicente de Castro, 320
Pontifícia Universidade Católica do Paraná
Curitiba, PR – Brazil
81200-340
E-mail: rafaela_scariot@yahoo.com.br

Received: December 26, 2011

Accepted: August 15, 2012

Conflict of Interests: The authors state that there are no financial and personal conflicts of interest that could have inappropriately influenced their work.

Copyright: © 2012 Scariot et al.; licensee EDIPUCRS. This is an Open Access article distributed under the terms of the Creative Commons Attribution-Noncommercial-No Derivative Works 3.0 Unported License.

Introduction

Regional odontodysplasia (RO) is a rare tooth anomaly involving dental tissues of both ectodermal and mesodermal origin, and it usually affects both the primary and permanent dentition (1). It is more frequent in women than in men, and the maxilla is affected twice as often as the mandible (2). Generally, it is limited to only one arch, although it can occasionally cross the midline (2,3).

The etiology of RO is uncertain, although local trauma or infection, teratogenic drug exposure, local circulatory disorders, Rh incompatibility, irradiation, neural damage, hyperpyrexia, metabolic and nutritional disorders and vitamin deficiency have all been discussed as possible contributing factors (4). RO has also been associated with the activation of latent viruses in the odontogenic epithelium and with the presence of nevi, hemangiomas and hydrocephaly; however, most cases of RO have no associated medical disturbances (4,5).

RO is characterized by a distinctive unilateral abnormal growth and maturation of the maxillary bone (primarily within the posterior segment), the teeth and the covering gingiva, resulting in a generalized facial asymmetry (6).

The criteria for diagnosis are mainly clinical and radiographic, although the histological findings are also distinctive (3). Clinical findings often reveal mediolateral expansion of the maxillary posterior alveolar process, extending from the canine to the maxillary tuberosity and their associated gingival, in addition to abnormal spacing, an absence of premolars and/or molars in the affected segment, delayed eruption of adjacent permanent teeth and hypoplasia of the deciduous molars (7).

Affected teeth have an abnormal morphology and irregular surface contours, with pitting and groves on the surface (8). These teeth seem to be discolored, hypocalcified and hypoplastic. The thin enamel is soft on probing, and the teeth are typically discolored, with a yellow or yellowish-brown shade (8). The most frequent clinical symptoms after the eruption of teeth are gingival swelling, periapical infection and abscess formation in the absence of dental caries (4).

Radiographically, the affected bone is dense and sclerotic, with coarse, irregular, and often vertically oriented trabeculae (9). The affected teeth show a "ghostlike" appearance due to the reduced thickness and radiodensity of the enamel and dentin (5,10).

Histological examination of affected teeth reveals a fibrous enlargement of the pulp and an irregular pulp/dentin interface, with pseudoinclusions and pulp stones. Tubular defects are found in the coronal dentin from the pulp horn to the cuspid tip, an irregular tubular structure is found in the circumpulpal dentin of the apical half, and a focally deficient odontoblast layer and widespread external resorption are also of note (11). A dense collagenous fibrous tissue with focal aggregations of calcified bodies and many islands of odontogenic epithelium can also be identified in the gingival histology (10).

The optimal treatment for this rare anomaly is controversial. Some authors advocate a more conservative approach, including the use of restorations to protect the permanent teeth or simply long-term follow-up (1,4), while others argue in favor of immediate extraction of affected teeth followed by prosthetic rehabilitation (5,6,12).

This paper reports a case of RO affecting the permanent teeth of the right side that is associated with agenesis and an eruption disorder.

Case Description

A 15-year-old boy was referred to the Oral and Maxillo-facial Surgery outpatient clinic of the School of Dentistry at Universidade Federal do Paraná (UFPR), Curitiba, PR, Brazil, because some of his permanent teeth had not yet erupted. No other symptoms were present, and the patient reported no pain. The past medical history was non-contributory. No congenital or acquired diseases were reported.

No facial asymmetry was noted on extraoral examination. Intraoral examination revealed an adequate left maxillary relation and a right maxillary overgrowth mainly on the buccal side, extending from the region of the canine to the tuberosity. There was a hypoplastic central incisor in the right quadrant of the maxillary dental arch that was associated with a fistula. In the right mandibular quadrant, both the central and the lateral incisors were hypoplastic (Fig. 1 and 2).

A panoramic radiograph (Fig. 3) revealed a dysplastic central incisor with an important root dilaceration on the affected maxillary side, with a well-circumscribed radiolucent area of 10 mm in diameter adjacent to the tooth. A sclerotic and ground-glass appearance to the bone was noted in the upper right maxillary arch. On the right side of the mandible, the lateral and central incisors had a striking "ghostlike" appearance. These teeth had very thin dentin and enamel layers. Demarcations between them were not observed, and their pulp chambers were wide.

In mandibular right quadrant, the dental development was not age-appropriate. The canine was unerupted and its germ was associated with a radiolucent area that was suggestive of a cyst. The first premolar and the second molar were normal. There was a second premolar germ, but no first or third molar was observed.

Given the observed delay in dental eruption and the fragile hard tissue of the affected teeth, the treatment plan consisted of two phases: an initial biopsy followed by extraction, with subsequent functional rehabilitation through orthodontics, orthognathic surgery and implants.

The "ghost" central incisor and the associated lesion were removed. The excised tissue was fixed in 10% formalin and was histologically processed. In a second procedure, the right mandibular lateral and central incisors were removed along with the unerupted germs. Alveoloplasty was performed. Histological examination of the removed teeth showed irregular dentin with areas of interglobular dentin. Immature odontogenic epithelium was observed in the connective tissue.



Fig. 1. Intraoral view of the patient with regional odontodysplasia.



Fig. 4. Intraoral view of the patient with regional odontodysplasia after the initial treatment.



Fig. 2. Intraoral view of the right quadrant of the patient with regional odontodysplasia.



Fig. 5. Intraoral view of right quadrants of the patient with regional odontodysplasia regional after the initial treatment.



Fig. 3. Panoramic radiograph showing a dysplastic maxillary central incisor and teeth with a "ghostlike" appearance in the mandibular right quadrant.



Fig. 6. Panoramic radiograph after initial treatment.

The surgical procedures improved the patient's overall oral health status, as observed both clinically (Fig. 4 and Fig. 5) and radiographically (Fig. 6). However, it was still necessary to rehabilitate the patient with implants. The patient was therefore referred for orthodontic treatment prior to the orthognathic surgery in order to impact the maxilla on the right side by aligning and leveling the other teeth to improve both their function and aesthetics.

Discussion

RO is a rare tooth anomaly that is generally limited to only one arch (1,3). In this reported case, the patient exhibited many of the common clinical and radiographic features consistent with the diagnosis of this condition, but this case was notable due to the uncommon involvement of both the right maxillary and mandibular quadrants. In 2010, Quinderé (1) described a case of regional odontodysplasia involving three quadrants of the jaws. To date, only six cases of generalized odontodysplasia have been described in the literature (13).

The etiology of regional odontodysplasia is not well understood. Local, systemic, and genetic factors, such as circulatory disorders, infections, teratogenic drugs, defects or trauma to cells of the neural crest have been implicated (4). The cause of the present case remains unknown because the patient's past medical history was non-contributory and no congenital or acquired diseases were reported.

RO usually affects both the primary and permanent dentition (1), but in this case, the patient and his parents were unable to provide clear information on the patient's primary dentition history.

Interestingly, this patient described in this report is a male, but RO is known to be more prevalent in females (3), although a recent study described three cases in males (14). Thus, further studies will be necessary to confirm this putative gender bias.

Intraoral examination revealed a right maxillary overgrowth mainly on the buccal side and in addition to gingival hyperplasia; both of these findings are consistent with the majority of cases reported previously (1,6,10).

Typical RO features seen in this case include the delayed eruption of adjacent permanent teeth, the absence of premolars and/or molars in the affected segment (7), teeth with an unusual "ghost-like" appearance (4,5,10) and

delayed dental development. The teeth had very thin dentin and enamel layers with poor demarcation and wide pulp chambers.

The treatment of RO is controversial and requires a continuous and multidisciplinary approach. This condition affects children and adolescents, which hinders definitive rehabilitation due to the continued potential for growth. Removal of some teeth has been proposed by several authors (5,13) due to abscess formation and restoration difficulties (5) secondary to serious structural defects (13).

Due to this patient's age, all affected teeth were extracted, including the two permanent germs. The ghost teeth and the maxillary central incisor were both infected. The decision to extract the germs was based in their developmental stages, which were classified as stage 4 (2/3 of crown formed) and stage 7 (crown formed and 1/3 root formed) for the canine and premolar, respectively, according to Nolla's staging criteria. The relationship between these results and the mineralization chronology of the permanent teeth (15) shows the chronological age of the canine between 2.5 and 4 years and the premolar between 9 and 11 years. As noted above, these chronological ages are incompatible with the patient's actual age, indicating a significant developmental delay of the dentition.

Uninfected teeth, such as the second premolar germ in this case, should not be extracted before eruption because their presence helps maintain the alveolar bone height and width during the skeletal growth phase. However, the prognosis of the affected permanent tooth is poor (4). This patient will undergo rehabilitation through an ortho-surgical approach, and his initial treatment plan was based on the expectation of his final rehabilitation. The use of a partial denture during the transition phase was not feasible given the lack of space in the posterior segment.

Conclusions

This case reviews the unique clinical and radiographic features of RO.

The treatment of RO is controversial and varies according to the individual patient. The ideal treatment remains unknown given the lack of sufficient cases and case follow-up in the current literature.

A complex multidisciplinary team is required to fully rehabilitate a patient affected by RO.

References

1. Quinderé LB, Cavalcante RB, Nonaka CF, Miguel MC, de Souza LB. Regional odontodysplasia involving three quadrants of the jaws: A case report. *Quintessence Int* 2010;41:13-6.
2. Canela AH, Rezende KM, Benitez M, Bönecker M. Early diagnosis of regional odontodysplasia in an infant. *J Craniofac Surg* 2012;23:e134-6.
3. Crawford PJM, Aldred MJ. Regional odontodysplasia: a bibliography. *J Oral Pathol Med* 1989;18:251-63.
4. Magalhães AC, Pessan JP, Cunha RF, Delbem ACB. Regional odontodysplasia: A case report. *J Appl Oral Sci* 2007;15:465-9.

5. Gündüz K, Zengin Z, Çelenk P, Özden B, Kurt M, Gunhan O. Regional odontodysplasia of the deciduous and permanent teeth associated with eruption disorders: A case report. *Med Oral Patol Oral Cir Bucal* 2008;13:E563-6.
6. Özpınar B; Gökçe B; Uzel G; Çömlekoglu ME. Prosthetic rehabilitation of segmental odontomaxillary dysplasia: seven-year follow-up. *Cleft Palate Craniofac J* 2009;46:103-7.
7. Beckett KB, Reibel J, Vedel B, Kjør I. Segmental odontomaxillary dysplasia: clinical, radiological and histological aspects of four cases. *Oral Dis* 2002;8:106-10.
8. Fearne J, Williams DM, Brook AH. Regional odontodysplasia: a clinical and histological evaluation. *J Int Assoc Dent Child* 1986;17:21-5.
9. Packota GV, Pharoah MJ, Petrikowski CG. Radiographic features of segmental odontomaxillary dysplasia. A study of 12 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996;82:577-84.
10. Gerlach RF, Jorge Jr J, Almeida OP, Coletta RD, Zaia AA. Regional odontodysplasia: report of two cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1998;85:308-13.
11. Armstrong C, Napier SS, Boyd RC, Gregg TA. Histopathology of the teeth in segmental odontomaxillary dysplasia: new findings. *J Oral Pathol Med* 2004;33:246-8.
12. Ferguson F, Schlissel E, Kucine A, Alexander S, DeSantis A, Hendricks R, Xu L. Long-term management of a child with regional odontodysplasia: A case report. *Pediatr Dent* 2009;31:346-9.
13. Juliana O, Hermes P, Lizeti TR, Santos-Pinto L, Elisa MG. Regional odontodysplasia in early childhood: a clinical and histological study (case report). *J Indian Soc Ped Prev Dent* 2009;27:175-8.
14. Barbería E, Sanz Coarasa A, Hernández A, Cardoso-Silva C. Regional Odontodysplasia. A literature review and three case reports. *Eur J Paediatr Dent* 2012;13:161-6.
15. Nicodemo RA, Moraes LC, Medice Filho E. Tabela cronológica da mineralização dos dentes permanentes entre brasileiros. *Rev Fac Odontol São José dos Campos* 1974; 3:55-6.