

REGIONAL ODONTODYSPLASIA: CASE REPORT

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ABSTRACT

Regional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth. It affects the primary and permanent dentitions in the maxilla and mandible or both jaws. Generally it is localized in only one arch. The etiology of this dental anomaly is uncertain. Clinically, affected teeth have an abnormal morphology, are soft on probing and typically discolored, yellow or yellowish-brown. Radiographically, the affected teeth show a “ghostlike” appearance. This paper reports the case of a 5-year-old girl presenting this rare anomaly on the left side of the maxillary arch, which crossed the midline. The primary maxillary left teeth (except for the canine) and the primary maxillary right central incisor were missing due to previous extractions. The permanent teeth had a “ghostlike” appearance radiographically. The treatment performed was rehabilitation with temporary partial acrylic denture and periodic controls. In the future, the extraction of affected permanent teeth and rehabilitation with dental implants will be evaluated. The presentation of this case adds valuable information to pediatric dentists to review special clinical and radiographic features of RO, which will facilitate the diagnosis and treatment of patients with this condition.

Uniterms: Regional odontodysplasia; Dental dysplasia; Dentition, primary; Dentition, permanent.

INTRODUCTION

Regional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth⁸. This condition was probably first described by Hitchin¹² (1934). Many authors however, credit to McCall and Wald¹⁴ (1947) the first report of this condition, showing only radiographic features.

The word “odontodysplasia” was coined by Zegarelli, et al.³⁰ (1963). Because this abnormality has a tendency to affect only one quadrant, “regional odontodysplasia” became the most accepted term to define it. The same condition has been described under other denominations, such as “odontogenic dysplasia”²¹, “localized arrest tooth development”²⁴, “ghost teeth”¹⁹, “odontogenesis imperfecta”²⁴, “unilateral dental malformation”²¹ and “familial amelodontal dysplasia”¹⁵.

RO affects both primary and permanent dentitions. Generally, it is limited to only one arch and sometimes crosses the midline. The maxilla is affected twice as often as the mandible⁵, the maxillary left quadrant being most commonly involved^{5,16}. Regarding the teeth, the central and lateral incisors are more frequently affected than the posterior teeth¹⁸. It is also common that in the same quadrant the

teeth are affected in the different degrees¹⁰. It has been suggested that this condition is more common in girls than in boys⁵. There has been no report of tendency towards a particular ethnic group⁹.

The etiology of this dental anomaly is uncertain, although several factors, such as local trauma or infection^{5,19}, teratogenic drugs, local circulatory disorders²⁷, Rh incompatibility, irradiation¹⁷, neural damage¹⁷, hyperpyrexia^{17,27}, metabolic and nutritional disorders¹⁹ and vitamin deficiency^{17,27}, have been discussed. It seems to be non-inherited condition because no cases with affection of family members have been described^{10,16}. RO has been reported in association with vascular nevi²⁷, hemangioma²⁷, epidermal nevus syndrome²², orbital coloboma²⁸, hypoplasia of the affected side of the face²⁹, hypophosphatasia²⁰, ectodermal dysplasia and hydrocephalus⁶.

The criteria for diagnosis of RO are primarily clinical and radiographic¹³. Clinically, affected teeth have an abnormal morphology and an irregular surface contour, with pitting and grooves surface⁷. The teeth appear to be discolored, hypoplastic and hypocalcified. The thin enamel is soft on probing and teeth are typically discolored, yellow or yellowish-brown⁷. It is possible to find some teeth without any alterations in the affected quadrant¹⁰. Affected teeth are more susceptible to caries and are extremely friable,

fracturing at the slightest trauma¹⁰. Tooth eruption is delayed or does not occur. The most frequent clinical symptoms after eruption of teeth with RO are gingival swelling, periapical infection and abscess formation in the absence of caries^{10,13}. There is a range of severity with this condition. In some mild cases, the roots could develop quite well without having any infection or complications²⁵.

Radiographically, the affected teeth show a “ghostlike” appearance due to reduced thickness and radiodensity of enamel and dentin¹⁹. A demarcation between hypomineralized dentin and hypomineralized enamel is not visible²⁷. The teeth tend to be shorter, have short roots with wide open apices and abnormally wide pulp chambers and canals¹⁰.

The clinical and radiographic characteristics of RO will be discussed in this article, describing this anomaly with certain unique findings and treatment options that need to be considered with specific reference to pediatric patients.

CASE REPORT

A 5-year-old healthy Brazilian girl was referred to the Pediatric Dentistry Clinic at the Dental School of Araçatuba, São Paulo State University (UNESP), for detailed evaluation of her oral condition. According to the report of her mother, the primary maxillary left teeth had erupted differently from the other child’s teeth. The mother also reported that her daughter’s affected teeth had a yellowish color and were rapidly destroyed by caries and fractured often accompanied by pain and gingival swelling.



FIGURE 1- Extraoral aspect of the child with RO



FIGURE 2



FIGURE 3



FIGURE 4



FIGURE 5

FIGURES 2, 3, 4 and 5- Intraoral view of the child with RO

The patient's mother also reported that during pregnancy she had taken medication for high blood pressure control (Enalapril®). Except for this fact, the pregnancy and the birth occurred uneventfully. There was no history of tooth or genetic anomalies in the family. The child's general health was good and no congenital or acquired disease was



FIGURE 6- Intraoral view of the child with RO



FIGURE 8- Rehabilitation of the child with temporary maxillary partial acrylic denture

reported. However, the girl had a urinary infection in the first three days of life, which was treated with Amoxicillin®.

Extraoral examination revealed no facial asymmetry, but a vascular birthmark (haemangioma) on the right side of her face (Figure 1). Intraoral clinical examination revealed a relatively caries-free mouth with normal occlusion, soft tissues and developing dentition except for the maxillary left quadrant (Figures 2-6).

On the left side of the maxilla, the central incisor and the first and second molars were missing as well as the primary right central incisor. According to her mother, the teeth were extracted by a dentist 6 months before the girl was referred to the clinic. The residual root of the left lateral incisor was present and was indicated for extraction. The left canine was previously restored with composite resin and it was less affected compared to the other teeth. It is likely that part of the permanent left first molar had an altered aspect because the primary second molar had been previously extracted. This clinical finding was not presented at the first appointment and it was observed at the third appointment.

The panoramic radiograph, which was taken 6 months before the girl was referred to our Clinic at the Dental School of Araçatuba, revealed the presence of primary teeth and the germs of permanent teeth, including the permanent second molars, except in the maxillary left quadrant. In the affected area, it was possible to observe the primary right central incisor, which was extracted straight away. The root of primary left lateral incisor was also present. In addition, the primary left canine presented reduced radiodensity and showed wide open apex and abnormally wide pulp chambers and canals in comparison to unaffected teeth. Germs of permanent teeth from the maxillary left quadrant and also the germ of the permanent right central incisor presented a "ghost like" appearance, showing that this condition had crossed the midline. Dental development appeared age-



FIGURE 7- Panoramic radiograph showing the teeth with "ghostlike" appearance in maxillary left quadrant and the right permanent central incisor

appropriate. Normal thickness of enamel and dentin in primary and permanent dentitions was observed in the other quadrants, except for the permanent mandibular left first molar, which showed some alterations in its crown, probably a developmental anomaly (Figure 7). This radiographic finding has to be investigated in the future controls.

After extraction of the root of the maxillary left lateral incisor, the affected edentulous quadrant was rehabilitated with a temporary maxillary partial acrylic denture (Figure 8). Prosthetic rehabilitation of patients is important to maintain mastication and phonation, to improve esthetics and to prevent overeruption of opposite teeth. The girl has been followed up periodically (once a month) to observe if there will be eruption of the "affected teeth" and to monitor the growth and development of the maxillary and mandibular arches.

DISCUSSION

Regional odontodysplasia is a rare developmental anomaly of dental hard tissues. According to a previous literature review²⁵, 138 cases of RO had been published from 1934 up to the end of the year 2002. After their publication in 2004, other 6 cases were retrieved from a Pubmed database search until the present moment. The patient of this case report exhibited many of the common clinical and radiographic features consistent with the diagnosis of RO. The clinical and radiographic characteristics involving the permanent dentition in the maxillary left quadrant (and also including the right permanent central incisor) strongly supported the diagnosis of this condition.

In this case, RO affected only one quadrant in the maxilla, but it crossed the midline, as previously reported^{11,28}. RO seems to be more prevalent in females and to occur in both dentitions²⁵, as found in this case. In addition, the finding of possible taurodontism in an unaffected quadrant (mandibular left first molar) suggests that RO might occur in association with other disorders²⁶.

The etiology of RO remains unknown¹⁰. Several factors have been associated to its occurrence¹¹. In this case, the history of maternal use of medication during pregnancy, as well as the urinary infection in the first three days of the child's life may have not been related to the condition. Otherwise, the prevalence of this anomaly would be higher. Some authors have suggested a strong correlation between vascular nevi and the pathogenesis of RO²⁷. In this case, the haemangioma was observed upon examination, but on the right side of her face (not affected by RO) in accordance to Fearn, et al.⁷ (1986). Although the haemangioma is localized in the opposite site to that affected by odontodysplasia, it could be an indication of abnormal vasculature, probably due to local ischemia or to morphological defects in blood vessels, perhaps following somatic mutation⁷. Unfortunately the cause of the present case remains unknown. In order to confirm possible etiological factors, further detailed investigation of this anomaly through the histological analysis must be reported¹³. Being RO a rare disease, it seems difficult to get

a significant number of patients to study this anomaly in a more detailed manner, for clinical, microradiographic and histopathological analyses. Microradiography is indicated to evaluate the mineral content of affected teeth because in conventional radiography the density of enamel is not evident due to the thinness of enamel layer in these teeth³.

Several factors must be considered to determine the best treatment option for a child with RO, such as age of the patient, any relevant medical history, previous dental experience, child's and parental attitude regarding dental treatment and number of affected teeth^{2,11}. There has been much debate as to whether affected teeth (with or without abscesses) should be extracted or saved^{5,10}. In this case, the residual root of the left lateral incisor was indicated for extraction because it could not be restored. In contrast, the left canine had been previously restored with composite resin because it was less affected by the anomaly. Other teeth probably were in same condition of that the left lateral incisor and had probably been extracted by another dentist.

The partial denture was fabricated to provide function, phonation and esthetics in this critical period. It was not possible to place posterior teeth in this denture yet because there is not space available. The early primary molars loss during the growth phase leads to reduction of vertical occlusal dimension. Because of this, it has to be considered the use of an appliance with screw to compensate maxillary growth. Another treatment option for patients with RO was described by Cahuana, et al.² (2005), who consider autotransplantation as an alternative during this phase. However, it is limited by the availability of suitable donor teeth^{3,13}.

Although the child and her mother are satisfied with treatment, the rehabilitation in this case cannot be considered ideal because the patient's oral esthetic and function were not totally restored. For the time being, the patient is under control visits to observe the growth and development of the maxilla and mandible. During the appointments, the appliance has been adjusted or changed for a new one. The permanent teeth will probably not erupt or will have altered eruption pattern. The noninfected permanent teeth will be not extracted before eruption because these teeth help maintaining the alveolar bone³. The presence of teeth is important during the skeletal growth phase. However, the prognosis of the affected permanent teeth is poor. Because of this, in the future, the extraction of permanent teeth and the rehabilitation with dental implants must be evaluated^{3,23}.

CONCLUSION

The presentation of this case helps pediatric dentists to review special clinical and radiographic features of RO. The therapeutic considerations of RO should be based on the degree of the anomaly, the functional and esthetical needs of each case.

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