Regional Odontodysplasia: Report of a case

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Regional odontodysplasia is a rare developmental anomaly involving both mesodermal and ectodermal dental components in groups of contigous teeth. RO affects the primary and permanent dentition in the maxilla and mandible or both jaws. Generally, it is localized in only one arch. The maxillary arch is affected more often than the mandibular arch. The affected teeth tend to be in a consecutive series that does not cross the midline, although some cases do not follow this pattern, as in the present case, have been documented. Radiogaphically wide pulp chambers and thin poorly defined hard tissue outlines described as a "ghost teeth" appearance, are typical features. A case of regional odontodysplasia in a 5 year old male patient is presented. The clinical and radiogaphical findings of this developmental anomaly and treatment are described.
J Clin Pediatr Dent 299(1): 45-48, 2004

INTRODUCTION

Regional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in group of contigous.[1,2] The first report of this condition was published by McCall and Walk[3] in 1947. Since that time, a number of cases have been described under a variety of names; such as localized arrested tooth development,[4] odontogenic dysplasia,[5] odontogenesis imperfecta,[6] unilateral dental malformation,[7] ghost teeth.[8] The term "odontodysplasia" was introduced by Zagarelli *et al*.[9] in 1963 to describe " .. a peculiar dental anomaly of unknown cause", then Pindborg[10] used the term "regional odontodysplasia" in 1970, to describe the segmental and localized nature of this condition. The term RO has now become the accepted terminolgy for this condition.

RO affects the primary and permanent dentition in the maxilla and mandible or both jaws. Generally, it is localized to only one arch. The maxillary arch is affected more often than the mandibular arch (twice as frequently as the mandible) with the maxillary left quadrant being most commonly involved.[11] Though the condition most often affects only one quadrant, cases with bilateral or multiquadrant involvement have also been reported.[12-17]

The etiology of this dental anomaly is uncertain, although several factors such as local trauma, infection, local ischemia, local vascular defects, Rh incompatibility, irridation, neural damage, hyperpyrexia, metabolic and nutritional disturbances and vitamin deficiency have been proposed and considered.[18-23] No one factor has been able to completely explain this abnormality.

Clinically, affected teeth have an abnormal morphology and an irregular surface contour with pitting and groves[24] and a rough surface with defective mineralisation.[25] The teeth appear to be discolored, hypoplastic, and hypocalcified. The thin enamel is soft on probing, and teeth are typically discolored, yellow or yellowish-brown.[25] Affected teeth are more susceptible to caries and are extremely friable, fracturing at the slighest trauma.[16] Tooth eruption is delayed or does not occur.[13,19,26]

Teeth of both dentitions may be affected; when the primary teeth are involved, the permanent successors are prone to present a similar condition.[8,11,15,27] The affected teeth tend to be in a consecutive series that does not cross the midline, although some cases not following this pattern have been documented.[11,13,22,26, 28,29]

The main radiographic feature has been described as a "ghost" appearance because of the reduced thickness of hard tissues.[8] Radiographically, enamel and dentin layers are thin and show the same radiodensity making it hard to distinguish between the two structures so both are less radioapaque than unaffected counterparts.[15] The teeth tend to be shorter, have short roots with wide open apices and abnormally wide pulp chambers and canals.[2,6,17,28,30]

The purpose of this report was to describe a case of regional odontodysplasia which had crossed the midline and, in which the main concern in the treatmant was to rehabilitate the patient's appearance.

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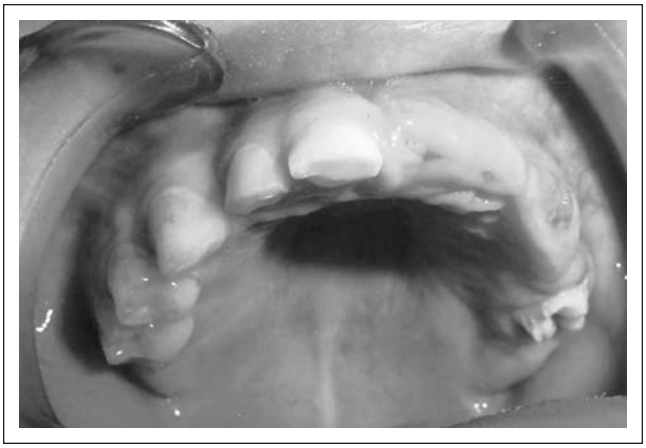

Figure 1. Intraoral view of upper left area with regional odontodysplasia.The maxillary left quadrant is toothless except for the tooth 65.

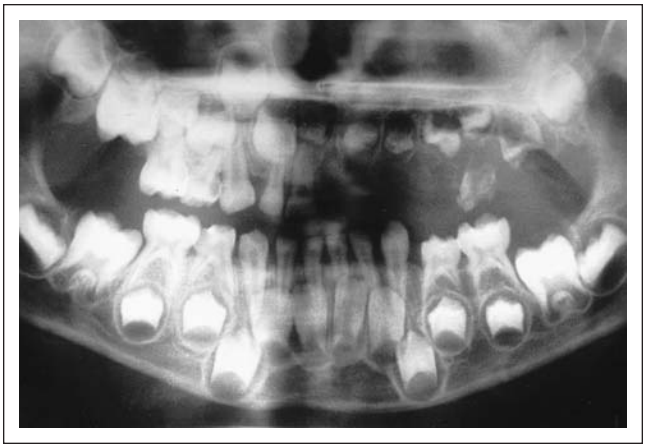

Figure 2. Panoramic radiograph shows the teeth with "ghostly" appearance in maxillary left quadrant. All teeth are normal except in left maxillary quadrant.

CASE REPORT

A 5-year-old healthy Turkish boy was referred to the Department of Pedodontics at the University of Ankara, Faculty of Dentistry with chief complaint of pain originating from the upper left second primary molar tooth and loss of primary teeth very early in life in the left quadrant of the maxilla. According to the description by his mother, the primary teeth had been different from the others and a yellowish color. They had become quickly destroyed by carious processes often accompanied by an abscess formation.

His mother reported having no fevers or taking any medication during pregnancy. The patient was the result of a normal pregnancy and birth. There was no history of similar cases in the family. The general health of the patient was good and no congenital or acquired disease (systemic, methabolic or inherited condition) was reported. However, the medical history of the child included interesting events. When he was one year old he developed pneumonia and asthma like symptoms, and spent one week in hospital under observation during which time antibiotics were given. He was breast fed until the age of one when he developed an allergy to milk, including his mother's milk and cows milk. Between one and three years of age he took soya-bean based products. After the age of 3, the patient again drank milk and no allergic reaction was observed.

Extra-oral examination revealed no significant abnormalities. There was no facial asymmetry or any vascular neoplasia like birthmark on his face. Intraoral clinical examination revealed a relatively caries-free mouth with normal occlusion, soft tissues and developing dentition except for the upper left quadrant (Figure 1). All the other primary teeth that belong to the other quadrant were present, normally formed and healthy, but there were carious lesions going deep into the dentin on the mandibular right first and second primary molars and maxillary right first primary molar. In the maxillary arch on the left side, the central incisor, the lateral incisor, the canine and the first molar were missing. The second molar showed rough, hypoplastic yellow enamel with pitting and chalky areas.

The panoramic radiographs revealed that the present primary tooth in the upper left quadrant and the unerupted germs of permanent teeth (including the unerupted permanent second molar) were all reduced in radiodensity in comparison to the unaffected teeth as is seen in RO patients (Figure 2). In addition to this, however the germ of the right permanent central incisor was also affected in the same manner showing that the condition had crossed the midline. All teeth in the mandible and right maxilla were radiographically of normal appearance. All the primary teeth and the germs of the permanent teeth in the other quadrants were normal in radiodensity. The radiographs indicated that all permanent tooth germs in the upper left quadrant and also the germ of the right permanent central incisor were showing "ghost teeth" appearance. All permanent teeth germs were showing the anomally because of deficiency in tooth structure, and no demarcation between enamel and dentin was visible. The pulp chambers of the primary right first central incisor and primary left second molar were large. The root of the primary left second molar was short with open apices. The root of the left upper primary central and lateral incisors were examined. Dental development from radiographs appeared age-appropriate and showed normal thickness of enamel and dentin in primary and permanent dentitions in the other quadrants.

TREATMENT

After the extraction of the left upper second primary molar, the remaining root of the left upper primary central and lateral incisors were also extracted. The affected edentulous quadrant was rehabilitated with temporary acrylic maxillary partial denture (Figure 3). Prosthetic restoration with partial denture is important for these patients to provide mastication and

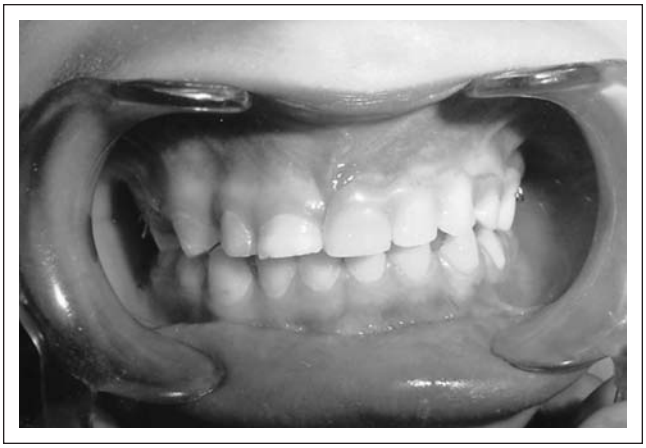

Figure 3. Frontal view of the maxillary partial denture in occlusion.

phonation, normal vertical dimension, space preservation, and improved esthetics. The patient was placed on periodic recall (4- to 6- month follow-up appointments) to observe progressive eruptive changes of the "ghost teeth" and to monitor the growth and development of the maxillary and mandibular dental arches.

DISCUSSION

RO is a rare developmental anomaly that affects both the dentin and enamel of a group of contiguous teeth. The patient in this report exhibited many of the common clinical and radiographic features consistant with the diagnosis of regional odontodysplasia. Intraoral findings were insufficient to support the diagnosis, as the primary central and lateral incisors, canine and first molar had been lost because of caries. In addition to this, only the maxillary left second primary molar tooth remained with enamel hypoplasia and excess caries in the affected quadrant. However, the panoramic examination revealed the distinct characteristics of RO. The generalized "ghost teeth" features involving the permanent dentition (including the right permanent central incisor) in maxillary left quadrant strongly supported the diagnosis of RO.

This case presents several aspects of the common clinical picture related to regional odontodysplasia. According to the Lustmann *et al*.,[11] teeth in the maxillary arch is affected more frequently by regional odontodysplasia, and this condition rarely crosses the midline. As demonstrated in this case, regional odontodysplasia occured in the maxilla, and not only all the teeth on the left side were affected, but also the right permanent central incisor was affected in the same manner, showing that the condition had crossed the midline. The same feature has been reported in a minority of other cases.[11,13,22,26,28,29] Although according to the literature, RO seems to be more prevalent in females,[24] it was seen in this case that males also can exhibit regional odontodysplasia. Therefore, further studies with a greater number of cases are necessary to confirm this tendency.

The etiology of RO has not been determined, although numerous theories have been proposed.[14,30] Rushton[8] suggested that a viral infection could permanently damage the dental organ in the early stages of development. With respect to the cause of the present case, a history of having developed pneumonia and asthma like symptoms, and having taken antibiotic when he was at the age of one may be the cause of his condition. In addition to this, a history of allergy to milk can cause a deficiency in nutrition. The allergic reaction against milk may suggest nutritional disturbances as a possible causal factor in the development of RO, but it is still difficult to accept as a cause in this case. Authors have suggested that there is a strong correlation between vascular nevi and the pathogenesis of regional odontodysplasia.[15,17,31,32] However, no facial nevi were noted upon examination, and there had not been any nevi at birth. Neither did heredity appear to be a factor as no family members were identified with dental anomalies. In conclusion, the cause of the present case seems to be unknown.

The care and treatment of a child with regional odontodysplasia requires a multidisciplinary approach. Consultation with specialist will greatly enhance function and psychological development of an affected child.[1] Treatment goals should take into account the need for future prosthetic and esthetic treatment.[26]

There has been much debate as to whether affected teeth (with or without abscesses) should be extracted or saved.[26,30,33] The appropriate treatment modality for RO is still controversial. Most clinicians advocate extracting the affected teeth as soon as possible and inserting a prosthetic replacement. Other clinicians have emphasized restorative procedure, if possible, to protect the erupted affected teeth.[28,33] However, selection of method and timing appear to be critical factors in the treatment of RO.[26] Although in very young children, teeth in the arch should be kept, teeth associated with abscesses cannot be restored, and need to be extracted. On the other hand, in older children, abscessed permanent teeth should be extracted with others retained until final rehabilitation with implants and /or fixed protheses can be constructed.[30]

In our case, all primary teeth on the upper left quadrant except the second primary molar had been extracted previously. The upper left second primary molar tooth with abscess formation was extracted as mentioned above and a partial denture was made to provide function, phonation, space preservation and esthetics in this critical period. The patient was placed on periodic recall to observe the growth and development of the maxillary and mandibular dental arches.

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