Regional Odontodysplasia: Report of Two Cases

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ABSTRACT
Regional odontodysplasia is a rare localized disorder of tissues of dental origin resulting in characteristically bizarre clinical and radiographic appearances. It most commonly affects the maxillary teeth of both the primary and permanent dentitions. This developmental anomaly is characterized by defective dentin and enamel formation leading to the radiographic appearance of ‘ghost teeth’ and sometimes by calcifications within the pulp and dental follicle. We report two cases of regional odontodysplasia affecting the maxilla on the right side. In one case, the regional odontodysplasia affected the maxillary left central incisor which is considered to be rare. The pathogenesis, clinical and radiographic features are discussed hereunder.

Keywords: Regional odontodysplasia, Ghost teeth.

INTRODUCTION
Regional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth. This condition was probably first described by Hitchin (1934). Many authors, however, credit to McCall and Wald (1947) the earliest report of this condition, showing only radiographic features. Zegarelli and Kutcher introduced the term ‘odontodysplasia’ in 1963. As the condition tends to affect several adjacent teeth within a particular segment or region of the jaw, it was prefixed ‘regional’ by Pindborg and regional odontodysplasia has become the accepted terminology. The same condition has been described under other denominations, such as ‘odontogenic dysplasia’, ‘localized arrested tooth development’, ‘ghost teeth’ or ‘odontogenesis imperfecta’.

This dental abnormality involves the hard tissues that are derived from both epithelial (enamel) and mesenchymal (dentin and cementum) components of the tooth forming apparatus. RO affects both primary and permanent dentitions. Generally, it is limited to one arch and sometimes crosses the midline. The maxilla is affected twice as often as the mandible; the maxillary left quadrant being most commonly involved. Regarding the teeth, the central and lateral incisors are more frequently affected than the posterior teeth. It is also common that in the same quadrant the teeth are affected in different degrees. It has been suggested that this condition is more common in girls than in boys. There has been no report of tendency towards a particular ethnic group. The etiology of this dental anomaly is uncertain, although several factors, such as local trauma or infection, teratogenic drugs, local circulatory disorders, Rh incompatibility, irradiation, neural damage, hyperpyrexia, metabolic and nutritional disorders and vitamin deficiency have been described.

The criteria for diagnosis of RO are primarily clinical and radiographic. Clinically, affected teeth have an abnormal morphology and irregular surface contour, with pitting grooves on the surface. The teeth appear to be discolored, hypoplastic and hypocalcified. The thin enamel is soft on probing and teeth are typically colored yellow or yellowish brown. It is also common to find some teeth without any alterations in the affected quadrant. Affected teeth are more susceptible to caries and are extremely friable, fracturing at the slightest trauma. Tooth eruption is delayed or may even be absent. The most frequent clinical symptoms after eruption of teeth with RO are gingival swelling, periapical infection and abscess formation in the absence of caries.

Radiographically, the affected teeth show a ‘ghostlike’ appearance due to reduced thickness and radiodensity of the enamel and dentin. The demarcation between hypomeralized dentin and hypominalized enamel is not clear. The teeth tend to be shorter have short roots with wide-open apices and abnormally wide pulp chambers and canals.

Histologically, all dental tissues were altered. The enamel was characterized as hypoplastic and hypocalcified, with variable thickness and irregular surfaces. The enamel prisms were irregularly distributed and could include aprismatic regions with degenerated globular calcifications. The dentin layer was reduced and presented a decreased quantity of tubules that were also irregularly distributed. Other observations included extensive interglobular and amorphous dentin areas, predentin layer enlargement, and clefts that could establish communication between the pulp and oral cavity. It is due to the existence of
these clefts between the oral cavity and the pulp that the RO teeth often present pulp necrosis in the absence of caries and trauma.\textsuperscript{17,18} This article reports two cases of RO involving the maxilla on the right side, one crossing the midline.

**CASE REPORTS**

**Case 1**

An 11-year-old female patient complained of three missing upper right front teeth. There was a history of trauma at the age of three years which resulted in loss of the upper right deciduous central incisor, lateral incisor and canine.

Extraorally, the face was grossly asymmetrical (Fig. 1). On intraoral examination 11, 12 and 13 were missing (Figs 2 and 3). Twenty-one was partially erupted and hypoplastic. There was calculus in relation to the right upper posterior teeth. The gingiva in the maxillary right quadrant was edematous and erythematous. Alveolar ridge in the region of 11, 12 and 13 was narrow and small.

Intraoral periapical radiographs (Fig. 4) showed ghost teeth in the region of 11, 12 and 13. Shadows of crowns of teeth with less radiopacity were seen in relation to 11 and 12. Enamel and dentin could not be differentiated from each other. Shadows of the crown and root with reduced radiopacity were seen in relation to 13. The pulp chambers were wide with open apices. Orthopantomograph (Fig. 5) revealed ghost teeth in 11, 12 and 13 region. The remaining teeth were normal. Based on the above said findings, a diagnosis of RO was arrived at.

**Case 2**

A 22-year-old male patient complained of incomplete eruption of upper right front teeth. The prenatal, postnatal, medical, dental and family histories were unremarkable.

Extraorally, the face was grossly symmetrical. Intraoral examination showed a thin alveolar ridge in relation to 11, 12, 13 and 21 when compared with the contralateral side (Fig. 6). Eleven, 13, 14 and 21 were partially erupted, yellowish brown in color and hypoplastic. Minimal calculus was seen in the affected region and the gingiva was swollen, edematous and erythematous. The remaining dentition was normal and caries free. Radiographs showed a remarkable reduction in the radiodensity of enamel and dentin in relation to 11, 12, 13 and 21 when compared to the other teeth on the radiograph (Fig. 7). The demarcation between hypomineralized dentin and hypomineralized enamel was not visible. The affected teeth were shorter with short roots and open apices suggesting the appearance of ‘ghost teeth’ (Fig. 8).

**DISCUSSION**

RO is a relatively rare localized developmental anomaly that affects both the enamel and dentin of a group of contiguous teeth. It occurs in both deciduous and permanent dentition, but has a marked prevalence for the maxilla.\textsuperscript{19} The etiology of RO is unknown, although factors, such as local trauma, irradiation, hypophosphatasia, hypocalcemia and hyperpyrexia\textsuperscript{1,10,20} were considered. Though the etiology in the first case reported is attributed to local trauma, the etiology in the second case seems to be unknown. Diagnosis was based on the clinical and radiographic findings characteristic of RO.
Females are more often affected than males (1.4:1).\textsuperscript{20,9} The maxilla is involved twice as much as the mandible. It is usually unilateral and rarely crosses the midline.\textsuperscript{21} Though equal gender predilection was observed in the present article, large sample size is needed to infer on gender predilection. Both the cases affected the maxilla on right side and one case crossed the midline which is considered to be interesting and rare. The clinical presentation in first case was missing teeth and in second case it was incomplete eruption. Though a thin alveolar ridge was seen in both the cases, gingival swellings were also seen in the second case supporting the previous reports.\textsuperscript{1,12} Presence of thin alveolar ridge in both the cases could be implied as underdeveloped maxilla on the right side could be due to the teeth being affected by odontodysplasia. The partially erupted teeth in both the cases were yellowish brown and hypoplastic supporting the previous reports.\textsuperscript{12,13,16,17,19} In both cases, only one quadrant in the maxilla was affected. In case 2, along with the affected right side, left maxillary central incisor was involved thus, crossing the midline which was considered to be interesting and rare as cited by previous reports.\textsuperscript{16,22,23}

Radiographically, the affected teeth in both the cases showed a ghost-like appearance (ghost teeth), marked reduction in the radiodensity of enamel and dentin, absence of demarcation between hypomineralized enamel and hypomineralized dentin, wide pulp chambers and open apices. These findings were in accordance with the previous reports.\textsuperscript{7,12,14,18} The criteria for diagnosis of RO are primarily clinical and radiographic.\textsuperscript{15} Kaan Gunduz et al\textsuperscript{24} stated the conditions which show same similarities to RO included amelogenesis imperfecta, dentinogenesis imperfecta, dentin dysplasia types I and II, shell teeth and hypophosphatasia. Apart from the characteristic features of each condition, these anomalies tend to affect the entire dentition instead of showing segmental distribution. There has been much debate as to whether affected teeth (with or without abscesses) should be extracted or saved.\textsuperscript{9,12} Accepted treatment modalities of RO include removal of the affected teeth and fabrication of prosthesis to provide function, phonation and esthetics and sometimes restoring the affected teeth by endodontic and esthetic therapies. Several factors must be considered to determine the best treatment option for a patient with RO, such as age of the patient, patient’s attitude regarding the dental treatment, number of teeth affected and degree of odontodysplasia. Cahuana et al\textsuperscript{16} considered autotransplantation.
as an alternative treatment option for patients with RO. However, it is limited by the availability of suitable donor teeth.\textsuperscript{15,21} The proposed treatment in the second case was removal of the affected teeth and fabrication of prosthesis to provide function, phonation and esthetics. Removable prosthesis was advised in the first case as she was under mixed dentition stage.

REFERENCES