Regional odontodysplasia: A report of three cases

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Three cases of regional odontodysplasia are reported, and the clinical, radiographic, and histopathologic features of the disease in these cases are compared with those of cases previously reported in the literature. (Quintessence Int 1994;25:141–145.)

Introduction

Regional odontodysplasia (RO) is an uncommon developmental anomaly of tooth formation that affects ectodermal and mesodermal dental components. It affects a variable number of teeth and is usually unilateral. The term odontodysplasia was introduced by Zegarelli et al; because the abnormalities tend to affect only one quadrant, the term regional was added to the name. Teeth are defective in both primary and permanent dentitions, and have a unique radiographic appearance.

The etiology of RO remains obscure, although numerous factors—including local trauma or ischemia; infection; irradiation; metabolic and nutritional disturbances; genetic (hereditary) factors; local somatic factors; neural crest migration disorder; and the activation of a latent virus in the odontogenic epithelium during tooth germ development—have been proposed and considered.

The affected teeth tend to be in a consecutive series that does not cross the midline; however, some cases not following this pattern have been reported. Generally, it is localized only on one arch, usually the maxilla (twice as frequently as the mandible) although involvement of both arches is possible. Regional odontodysplasia seems to be slightly more prevalent in females and has no racial predilection.

Clinically, the affected teeth are discolored, hypoplastic, and hypocalcified, showing short roots with wide apices beside wide pulp chambers. The main radiographic feature has been described as a “ghost” appearance. There is a lack of contrast between enamel and dentin, both being less radiopaque than unaffected counterparts. Areas of relatively radiopaque tissue have been noted within the crown. Histologic characteristics of the condition are also well documented.

Regional odontodysplasia has been reported in association with failure of eruption of the affected teeth, gingival swelling, vascular nevi, and hydrocephalus, and also under other names, such as odontogenic dysplasia, localized arrested tooth development, ghost teeth, odontogenesis imperfecta, and unilateral dental malformation.

Case reports

Case 1

A 5-year-old healthy white boy was referred to the Department of Diagnosis and Surgery Department at the Dental School of São José dos Campos, complaining of loss of primary teeth very early in life, without eruption of the corresponding permanent teeth in the left quadrant of the maxilla. The primary teeth, according to his mother’s description, were very different from the others, and had altered morphology and a yellowish color. They
had been quickly destroyed by carious processes often accompanied by abscess formation.

No undisputed cases of RO were found among other family members.

Residual roots of teeth 61, 62, 63, 64, and 65 were still present and were characterized by a yellowish coloration. The remaining dentition was quite normal (Fig 1a).

Radiographic examination of residual roots of the primary teeth showed increased radiolucency. The permanent elements, still inside the bone, showed an altered morphology and retarded developmental age, associated with little demarcation between enamel and dentin (Figs 1b and 1c).

At the histologic examination, demineralized sections of the affected primary teeth showed irregular dentin, with few and tortuous tubules, and globular formations distributed in the area. Closer to the dentino-enamel junction, the dentin was evenly calcified. A layer of globular dentin with few dentinal tubules was observed between the former dentin and the most pulpal dentin where tubules were tortuous and wider, forming large and clear spaces often containing basophilic material. Large, irregular, calcified globules were also noted to some extent in this area as well as in the pulp chamber (Figs 1d and 1e).

Mucosal specimens that accompanied the teeth had parakeratinized lining epithelium showing acanthosis and very hyperplastic rete pegs. The lamina propria, constituted of fibrous connective tissue, was chronically inflamed. The calcified globules inside this tissue were either spherical or deformed and were larger. Odontogenic epithelial rosettes were also observed (Fig 1f). The histopathologic features and the clinical and radiographic aspects were analyzed and the diagnosis of RO was established.

Case 2
A 10-year-old boy complained that his permanent teeth had not erupted in the left mandible.

Through a clinical examination, the following teeth were observed: 11, 21, 41, 42, and 46. The maxillary right permanent lateral incisor was erupting. Primary teeth from the right mandible and both the left and right maxillary quadrants were carious but showed normal features related to morphology and location. In the left mandible some primary teeth had not been exfoliated. Teeth 71 and 72 showed alteration in their color and teeth 74 and 75 were deeply carious. Tooth 73 was absent. The cusps of tooth 36 were appearing in the mouth but already showed hypoplastic aspects (Fig 2a).

An orthopantogram revealed that in the mandibular left quadrant the permanent teeth were still intraosseous and exhibited decreased radiolucency of the enamel and dentin (Fig 2b). Pulp chambers were wider than normal (teeth 31 and 35). Tooth 36, with the crown already extrasosseous, showed the aspects already described, but tooth 37 was normal, considering the radiolucency. Tooth 38 followed the normal chronology (Fig 2c).

Light microscopic inspection of a decalcified permanent molar, extracted because of a periodontal abscess, revealed a wide pulp chamber. Dentinal tubules showed a regular course, although some clefts, in addition to small amounts of interglobular dentin, were evident. A small calcified globule was present within the pulp.

Case 3
A 6-year-old boy was brought to the clinic by his mother, who had observed that her son had some difficulties related to speech and mastication because of the absence of some teeth.

It was reported that the child's uncle, fraternal twin of his father, had a son with complete unilateral cleft lip and palate. The same anomaly was also present in another cousin, the son of his father's sister.

The following permanent teeth had erupted and showed normal features: 31, 41, and 46. The remaining maxillary and mandibular primary teeth were also normal, except on the maxillary left side. The left side of the maxilla was toothless, except for the residual roots of teeth 61, 62, 63, 64, and 65. Hence, a loss of vertical dimension was evident on this side (Fig 3a).

The orthopantogram showed ghost teeth in the maxillary left quadrant. A diagnosis of regional odontodysplasia was established (Fig 3b). A localized occlusal view of the region of teeth 62 to 65 showed hypoplasia of the teeth and a detailed aspect of ghost teeth (Fig 3c).

The histopathologic examination of primary residual roots from the left maxilla showed fragments of dentin with normal tubular aspects except for areas of interglobular dentin. The cementum covering the root had areas of resorption and multinucleated cells were present.
Fig 1a  Clinical aspects of the maxilla of the patient in case 1. On the left side, affected by RO, residual roots of teeth 61, 62, 63, 64, and 65 are present.

Fig 1b  Orthopantogram showing increased radiolucency of the permanent teeth in the maxillary left quadrant.

Fig 1c  Radiographic occlusal view showing altered morphology of the teeth, with little demarcation between enamel and dentin on the left side.

Fig 1d  Demineralized section showing outer dentin with tortuous tubules and globular dentin, and the most pulpal dentin with tortuous and wider tubules. In the pulp chamber, calcified globules can be observed.

Fig 1e  Detail of the pulp chamber and the most pulpal dentin shown in Fig 1d.

Fig 1f  Photomicrograph of a mucosal specimen showing calcified globules in the lamina propria, some of which are associated with epithelial tissue.
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Fig 2a Clinical aspects of the mandible of the patient in case 2. On the left side, teeth 74 and 75 are deeply carious. Teeth 71 and 72 show alteration in their color and hypoplastic enamel. Tooth 36 is erupting, and the canines have hypoplastic aspects.

Fig 2b Orthopantogram showing hypoplastic aspects of the teeth in the mandibular left quadrant.

Fig 2c Periapical views are consistent with aspects of hypoplasia of the teeth.

Discussion

Regional odontodysplasia was considered an uncommon disease by Crawford and Aldred in their worldwide literature review. They presented 109 cases from several countries.

The cases presented here may be an additional contribution, not only for their geographic characteristic but also for the gender involvement. The literature states that RO is slightly more prevalent in females, but these three cases occurred in male patients.

The cases presented had several clinical, radiographic, and histopathologic aspects related to the entity RO. Two cases occurred in the maxilla and one in the mandible; i.e., it was slightly more frequent in the maxilla and present just in one of the arches, as is commonly described in the literature. Both dentitions were affected, as expected, and the age of diagnosis was during primary tooth eruption, as well as during the mixed dentition. In cases 1 and 3, primary teeth were lost precociously and, in case 2, eruption of the permanent dentition was delayed, because the patient was already 10 years old and was complaining of absence of permanent teeth in the left mandible.
Clinical common findings were a greater susceptibility of the affected teeth to dental caries in addition to periapical abscess formation. The affected teeth were in a consecutive series and did not cross the midline. Patients also had speech and mastication difficulties as well as facial asymmetry related to the loss of vertical dimension.

The radiographs showed the classic appearance of ghost teeth, with wide pulp chamber and malformed roots associated with little demarcation between enamel and dentin.

Histopathologic features such as tortuous dentinal tubules, interglobular dentin and clefts in the dentin were the most common findings. In case 1, even the fragments of mucosa that accompanied the teeth had spherical calcified masses as have been described previously.

We could not establish any evidence concerning the etiology of the cases presented, although one patient had two cousins with malformations. Therefore, we believe that genetic factors could be associated, although there was a lack of direct association between the features observed in the patient and those of his relatives. The causative factor of this uncommon condition must be active in both the primary and the permanent dentition.

These three cases reported could be included among classic cases of RO. It is important to describe the various aspects of this developmental anomaly to better understand this uncommon condition. More investigation is necessary before all aspects involving this disturbance of tooth formation can be clarified.

References