

Regional odontodysplasia: an unusual case with a conservative approach

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Abstract A case of a 14-year-old male with regional odontodysplasia is reported. In this presentation many atypical clinical and radiographical features of this condition are present. The chief complaint of the patient was the enlargement of the gingiva and, according to the literature, inflammatory processes are the main reason why patients look for care. Moreover, there was no radiographic evidence of unerupted teeth in this report. The functional and psychological benefits of the conservative approach are emphasised.

Regional odontodysplasia is an unusual developmental anomaly in which ectodermal and mesodermal tooth components are affected.¹

The first report of this condition was published by McCall *et al.*, in 1947,² but the term 'odontodysplasia' was introduced by Zegarelli *et al.*,³ in 1963. Since then, various other terms have been used, such as regional odontodysplasia, ghost teeth, odontogenesis imperfecta, localised arrested tooth development, unilateral dental malformation, amelogenesis imperfecta non-hereditaria segmentalis and familial amelodontinal dysplasia.³⁻⁹

Although many possible causes of regional odontodysplasia have been suggested, no completely satisfactory etiologic explanation has been offered for this disturbance until now.^{1,3,4,7-9}

This anomaly tends to affect several adjacent teeth within a particular segment of the jaw, and generally does not cross the midline.^{6,10,11} Frequently, it is located only on one arch, and the maxilla is involved twice as often as the mandible.^{6,11,12} The condition is more common in female than in male patients and is more frequent in the anterior region.^{6,11,12} There is no tendency toward a specific race or ethnic group.⁶ When the primary teeth are affected, the permanent dentition is usually affected also.¹³

Clinically, affected teeth have an abnormal morphology and a rough surface with defective mineralisation.¹⁴ The teeth appear to be discoloured, hypoplastic and hypocalcified. Tooth eruption is delayed or does not occur.⁷

Radiographic aspects show marked reduction of radiodensity and little demarcation between enamel and dentine. These teeth present wide pulp chambers and open apices.¹¹

Histologically, there is a considerable reduction of dentine and the tubules are reduced in number. Areas of clefts can be found within the dentine. The enamel is hypoplastic and hypomineralised



Fig. 1 Clinical aspects of the intra-oral examination. The affected teeth are yellowish with rough irregular surfaces. Note the enlarged alveolar crest that is covered by fibrous tissue.

and contains degenerated globular calcifications. The pulp contains calcifications of varying degrees.^{8,14,15} The dental follicle may contain irregular calcifications.¹²

Case report

A 14-year old black male self-referred patient presented with swelling in the lower left gingiva, with spontaneous bleeding, to the Pathology and Surgery Service at the Federal University of Minas Gerais. The patient reported that the volume increase had appeared for about 6 months prior to his visit. No relevant medical history was found. According to his mother, the deciduous teeth were normal and there were no more cases in the family.

Clinically, the left mandibular teeth were discoloured, hypoplastic and hypocalcified with measurements smaller than the normal, yellowish colour, with rough, irregular surfaces. The adjacent alveolar crest was enlarged and covered by fibrous tissue (fig. 1). The second left inferior molar, as the teeth of other arches, showed normal morphological aspects.

Panoramic and periapical radiographs showed longitudinal shortened teeth with lack of contrast between enamel and dentine. The affected teeth had wide pulp chambers with open apices and were smaller than normal in over-all size. The main radiographic feature was a 'ghost' appearance (fig. 2). There was no evidence of impacted teeth in the affected area. The third left and right inferior molar dental germs were absent (fig. 3). An occlusal radiograph showed a slight bone neoformation in the anterior vestibular area (fig. 4).

Because of the presence of dental vitality and the absence of caries, cosmetic surgery of the fibrous tissue was carried out, and

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Fig. 2 Periapical radiographs showing the affected teeth with a 'ghost appearance': lack of contrast between enamel and dentine, wide pulp chambers with open apices.



Fig. 4 Occlusal radiography. Note a slight bone neoformation in the anterior vestibular area.



the affected teeth were not extracted (fig. 5).

The histopathological examination of the soft tissue revealed the presence of a whorled connective tissue containing numerous rounded calcified bodies that stained densely with hematoxylin. There were many islands of odontogenic epithelium (fig. 6,7).

Based on clinical, radiographic and histopathological features, the diagnosis of regional odontodysplasia was established.

Because of his uncertain prognosis, the patient is attending a preventive programme in order to maintain these teeth in the arch until the complete development of the jaws.

Discussion

Since regional odontodysplasia is a rather unknown entity, many cases are probably misdiagnosed as malformed teeth or odontomas.⁸ Although numerous theories have been proposed, the pathogenesis of this condition remains uncertain.⁸

Other conditions such as dentinal dysplasia, shell teeth, hypophosphatasia, dentinogenesis imperfecta or amelogenesis imperfecta can mimic some features of regional odontodysplasia. However, these disorders tend to affect the entire dentition.⁸

The patient in the above report exhibits several aspects of the common clinical, radiographic and histopathologic features related to regional odontodysplasia. However, some of these important features are absent. According to the literature, abscess formation is the main reason for extraction of affected teeth,^{1,16} but local gingival enlargement was the chief complaint of our patient. This is not usual, as emphasised by some authors.¹²

It is interesting that, although the maxillary arch is normally more affected than the mandibular arch,^{6,11,12} the alterations in the present case were only in the mandibular arch. Moreover, the patient is a male and according to the literature this condition



Fig. 5 Clinical features after cosmetic surgery.

seems to be more prevalent in females.¹² Therefore, further studies with a greater number of cases are necessary to confirm this tendency.

Eruption of the affected teeth is delayed or does not occur.⁷ According to the patient's mother there was no evidence of this disturbance in his deciduous teeth, but this information may not be reliable since the patient was not examined at that time. However, as the patient had no unerupted teeth, it seems probable that, in some cases, deciduous teeth are lost according to the normal chronology and permanent teeth can erupt.

Treatment of regional odontodysplasia has given rise to controversy.⁸ The main question is whether to remove the affected teeth or not, but we believe that this procedure is caused by the presence of inflammatory disorders and/or associated unerupted teeth. Most clinicians opt for extracting the affected teeth as soon as possible. In this case, it is necessary to insert a prosthetic replacement. Other professionals elect to use restorative procedures, whenever possible, to protect the affected erupted teeth.¹⁷ In the present report these restorative procedures were not indicated because of the thickness and hypocalcification of the mineralised structures as well as the enlarged pulp chambers. In our opinion the affected teeth should be preserved for as long as possible in order to provide normal jaw development and to lessen the psychological effects caused by premature loss of the teeth. Because of the young age of our patient and as long as clinical signs and radiographic aspects indicate no infection, the short-term-treatment philosophy was to maintain the affected teeth. We believe that the developmental and psychological benefits outweigh immediate extraction.



Fig. 3 Panoramic view showing no impacted teeth and absence of inferior third molars germs.

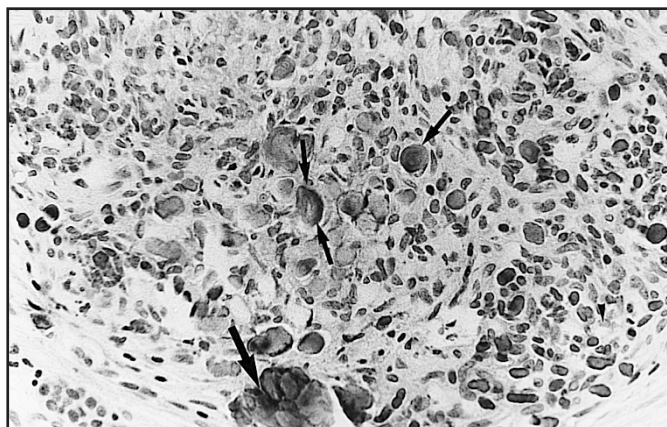


Fig 6 Histopathological features: whorled connective tissue with calcified bodies (arrows).

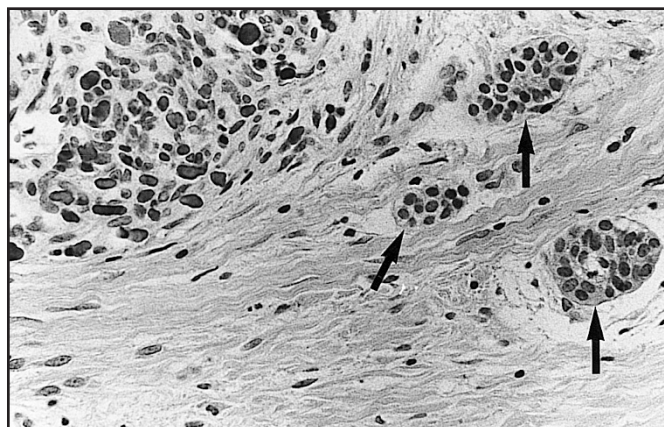


Fig 7 Histopathological features showing islands of odontogenic epithelium (arrows).

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