

CASE REPORT

Regional odontodysplasia on the left side of the mandible: report of an unusual case

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Abstract

Regional odontodysplasia (RO) is a rare developmental abnormality of epithelial and mesenchymal dental tissues. Due to its poorly understood etiology, assessing and discussing related clinical cases of this dental anomaly is crucial to guide professionals in improving its treatment and outcomes. This article aimed to report the case of a 9-year-old male patient who presented to our department with the main complaint of absent eruption of permanent left mandibular quadrant teeth. This is the first case reported in China from a patient with multiple cutaneous nevi on the face and neck, and based on the retrieved clinical and radiographic features, we described and discussed the treatment and etiology of RO.

Keywords

Regional odontodysplasia; Ghost teeth; Etiology

1. Introduction

Regional odontodysplasia (RO) is a sporadic dental dysplasia of unknown etiology. It tends to affect several adjacent teeth within one quadrant of the jaw bones, although it might occasionally cross the midline. The number of teeth affected may range from one tooth to all teeth [1], involving all dental tissue. Radiographically the affected teeth are characterized by a “ghost-like” appearance because the enamel and dentin are hypomineralized and hypoplastic, reducing radiodensity with wide pulp chambers and short roots with open apices [2]. Due to the rarity of this condition or missing documentation, its prevalence remains unclear. However, it was found that this anomaly has no ethnic tendency. The mandible is less frequently affected than the maxilla [1, 3]. A recent systematic review of published RO cases by Nijakowski *et al.* [4] reported no significant gender predilection. Conversely, other literature showed that women might be slightly more affected than men [3].

One of the major challenges of RO is determining its etiology to reduce the possibility of disease occurrence. Suggested factors include local trauma and ischemia caused by vascular defects or disorders, irradiation, metabolic disturbances and nutritional deficiencies, activation of a latent virus during tooth germ development, hyperpyrexia accompanying severe childhood diseases, local somatic mutations, neural crest migration disorder [5, 6], and syndromal involvement [7]. Presently, the etiology of RO is considered to be multifactorial, comprising local and systemic factors. Koskinen *et al.* [8] first reported the *PAX9* gene mutation in a 6-year-old girl patient with RO. Magalhães *et al.* [9] described a case of RO with hemangioma and suggested that the possible etiologic

factors could be ischemia or vascular morphological defects. Local trauma is also considered a possible causative factor. A previous study reported the occurrence of RO in a 2-year-old boy with a history of intrusive trauma in the lesion area [10]. Thus, complete patient history and clinical symptoms should be recorded and assessed to clarify the underlying etiological factors.

Since RO is a rare disease with limited reported literature, achieving evidence-based treatment is challenging, and its treatment remains controversial as to whether affected teeth should be kept or extracted. Many authors [11–13] have proposed early extraction of the impacted teeth, which could otherwise develop into more severe dental pathology. Poorly mineralized enamel and dentin or cleft could allow microorganisms to enter the dental pulp even in the absence of caries [14, 15]. This strategy aimed to minimize the number of treatments and avoid pain caused by the affected teeth. Some authors suggest that keeping the affected teeth promotes jaw development and reduces psychological trauma associated with premature tooth loss [16].

2. Case Report

A 9-year-old boy, accompanied by his mother, came to our department with the main complaint that his permanent teeth in the lower left quadrant of the mandible did not erupt. The condition was painless, and his mother reported no abnormalities in the eruption of deciduous teeth, but the deciduous teeth in the affected region were prematurely lost due to “early childhood caries”. The boy was born at term by normal delivery. His medical history was unremarkable, he had no history of local trauma, and no other family member had dental

or maxillofacial abnormalities.

Clinical examination: Extraoral examination revealed no facial asymmetry, but multiple cutaneous nevi were observed on his bilateral facial and neck skin (Fig. 1). Intraoral study revealed a mixed dentition, and the mucosa had normal texture and color. There was a reduction in the alveolar bone in the lower left quadrant, and dental chronology was abnormal because only the left mandibular first permanent molar was present (Fig. 2A). In the lower right quadrant, we observed erupted permanent central incisor, lateral incisor, primary canine, partially erupted the first premolar, primary second molar, and the first permanent molar. In the maxillary arch, teeth eruption was even on both sides and presented as follows: 16, 15, 14, 53, 12, 11, 21, 22, 24, 65 and 26, and all teeth had normal shape and color.



FIGURE 1. Extraoral clinical aspects of the reported case: multiple cutaneous nevi on both facial and neck.

Radiographic examination: Panoramic radiograph revealed that the maxillary dentition and the right mandibular teeth were expected. Permanent incisors, canine and premolars were unerupted in the left mandibular region and gave the typical “ghost-like” appearance (Fig. 3). The teeth were tiny and hypoplastic. The large radiolucent areas surrounded the crowns. It was also observed that tooth germs number 31, 32 and 33 only had a small amount of root formation, and the small degree of tooth germ development of number 34 (Nolla’s stage 5) and 35 (Nolla’s stage 4) could be seen radiographically. A low alveolar bone density was detected around the affected teeth. Thus, based on the clinical and radiographic findings, the patient was diagnosed with RO.

Treatment: Considering his age, we estimated that removing the affected dental germs could impair mandible development and did not extract the affected teeth. A temporary removable partial mandibular denture was made for esthetics and repair mastication in the period of jaw growth (Fig. 2B). Prosthetic rehabilitation was essential to prevent a mesial inclination of tooth 36, overeruption of opposite teeth and achieve normal vertical dimension. The patient was given proper oral hygiene guidance. Since timing is critical in the treatment of RO, the patient was regularly followed up at our department to monitor the development of permanent tooth germs and assess

the severity of odontodysplasia, based on which subsequent therapeutic approaches are to be determined. During the later follow-up, the pros and cons of available treatments will be weighed to choose the optimal treatment strategies *via* a multidisciplinary team.

3. Discussion

Early diagnosis is essential to establish a therapeutic scheme and minimize future concomitant complications. The clinical manifestations of RO are diverse. If the affected teeth erupt, the teeth might become hypoplastic, hypocalcified, fragile, easy to wear, prone to caries, yellowish or brown discoloration and demonstrate abnormal morphology [17]. Abnormal eruption of the affected teeth is the most typical clinical symptom of RO [3], similar to our presented case. The affected gingival tissue is often enlarged, fibrous or hyperemic and might contain a fistula [5, 13, 18]. However, the patient in our case had a normal gingiva texture and color. Additionally, although the maxilla is usually more affected than the mandible [3, 13, 19], the changes observed in this boy occurred only in the mandibular arch.

Based on present literature, most RO involves both primary and permanent dentition [4]. In the current case, the deciduous teeth in the affected region were prematurely lost due to “early childhood caries”. Since it is unusual that all the patient’s primary teeth were without caries and fillings, we highly suspected that the deciduous teeth were prematurely lost due to odontodysplasia.

Extraoral examination is also important. A brief examination of the head and face features is recommended to record indications of abnormal development. Inspection of the skin, hair and nails may, in some cases, provide useful information since they develop from an ectoderm-like enamel. Upon extraoral examination, multiple cutaneous nevi on both facial and neck skin were observed in our case. Tooth morphogenesis depends on neural crest cells called ectomesenchyme. Neural crest cells also contribute to dermal and epidermal structures. In this case, we found the co-existence of tooth malformation and nevoid alterations in the overlying maxillofacial neck skin. Although the etiology of our presented case was unclear, failure of the neural crest cell migration and differentiation might explain this phenomenon. Most reported cases had not been attributed to any particular cause [17, 19–21], and considering that the potential etiopathogenesis of this condition has not been established to date, further epidemiological and experimental research are urgently needed.

Radiographically, affected teeth present a “ghost-like” appearance owing to a noticeable reduction in radiodensity of the thin enamel and dentin layers. Additionally, shortened roots with open apices could be visible [22], with almost no boundary between enamel and dentin. Enlarged pulp chambers and root canals often have diffuse calcification [13]. Cone-beam computed tomography may indicate a decrease in the fractal size of the trabecular bone region affected by RO [2]. In this present case, the radiographic features showed a typical “ghost-like” appearance, and large radiolucent areas surrounded the crown of the unerupted tooth, similar to that reported by Matsuyama *et al.* [23]. The diagnostic criteria

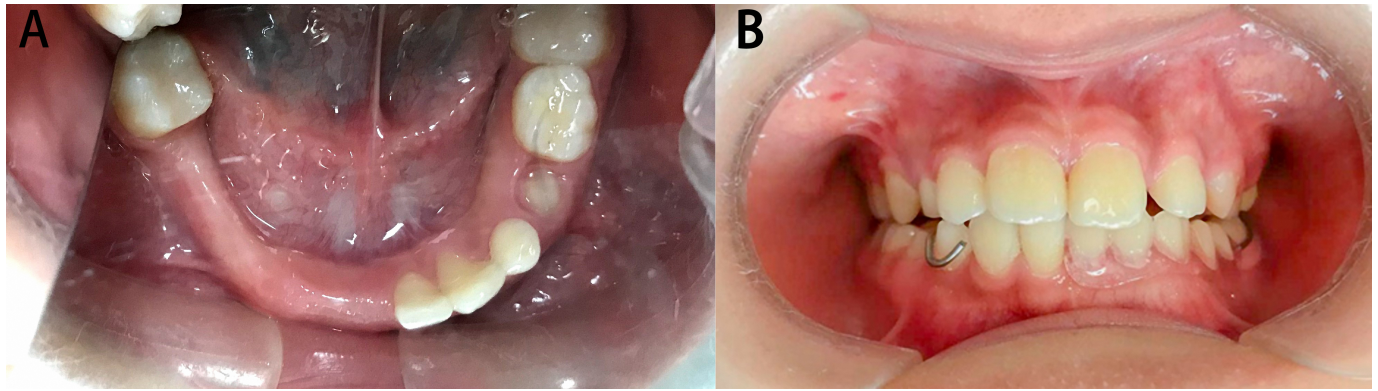


FIGURE 2. Intraoral view of the mandibular arch. A: Before rehabilitation. B: After partial removable denture restoration.



FIGURE 3. Panoramic radiograph of the affected teeth showing a “ghost-like” appearance.

of RO are based on characteristic clinical and radiographic features.

Differential diagnosis: Amelogenesis imperfecta (AI) and Dentinogenesis imperfecta (DI) may display similar appearances. AI is a genetic disease characterized by clinical and genetic heterogeneity, which affects the structure and clinical appearance of the enamel of all or nearly all teeth [24]. DI has a genetic predisposition and presents with normal enamel mineralization of the affected teeth. RO involves all dental tissues [25], is nonhereditary and tends to affect several adjacent teeth within the same quadrant. In this case, the abnormal teeth eruption and radiological features of the affected area supported the diagnosis of RO.

Once diagnosed, decisions on whether to extract the affected tooth should be based on the severity of odontodysplasia, patient age and cooperation, personal characteristics and aesthetic needs, preferably *via* a multidisciplinary approach. In this case, there were no signs of gingival swelling or abscess formation; thus, a conservative approach was considered. Early teeth extraction can greatly reduce the height and quality of the alveolar ridge, and subsequent defects could cause huge

issues for future repairs. It is advantageous to preserve noninfected teeth to promote mandible development and remove the psychological impact of early tooth loss. A partial removable denture was used to repair the edentulous area to maintain masticatory function and to preserve space and normal vertical dimensions. We recognized that this case would benefit from a multidisciplinary team approach, and the treatment planning would be long and complex. Therefore, a long-term follow-up program is conducted regularly to monitor the development of permanent tooth germs and assess the severity of odontodysplasia. The patient’s management will depend on the severity of the odontodysplasia. Koruyucu *et al.* [16] described a case of preserving RO-affected teeth after orthodontic, endodontic, periodontal and restorative treatments. However, extractions of the affected teeth could be inevitable in conditions with acute abscesses and pain. Other treatment options include auto-transplantation [26] or implant treatment when the patient reaches adulthood [27]. The long-term treatment goals include maintaining masticatory function and esthetic improvement, facilitating jaw development, protecting some affected erupted teeth, and reducing the psychological impact.

4. Conclusion

RO is a rare dental developmental anomaly, and discussing clinical cases of this dental anomaly might help general and pediatric dentists review the clinical and radiographic characteristics of RO to formulate better therapeutic approaches. As the potential etiopathogenesis of this condition has not been established to date, further research, including epidemiological and experimental studies, is required.

AUTHOR CONTRIBUTIONS

YJ—performed the treatment, analysis and interpretation of data, original draft preparation, and manuscript review and editing. YW, SH—analyzed the data and manuscript editing. FZ—designed the research study and contributed to manuscript review and editing. All authors read and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Patient information and data collection for this study took place at The Children's Hospital, School of Medicine, Zhejiang University, China. This study was conducted in accordance with the Medical Ethics Committee of Zhejiang University (ID: 2021-IRB-069). The patient's parents gave written consent for inclusion before he participated in the study and gave written informed consent for publication of this study.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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