Regional Odontodysplasia: Report of a Case

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ABSTRACT

Regional odontodysplasia is an unusual nonhereditary dental anomaly of uncertain origin. It presents clinically as delayed or failed eruption, abscess formation, early exfoliation, and malformed teeth and radiographically as defective enamel and dentin formation, giving the teeth a fuzzy silhouette. Because teeth are essential for psychological and functional development of the child, appreciation and management of regional odontodysplasia is important for comprehensive dental care. The purpose of this paper was to present the case of a 12-year-old female who presented with an eruption disturbance affecting a single segment of her jaw and characteristic ghost teeth radiographic appearance. The approach toward management should be aimed at preserving the unerupted affected teeth for the development of the alveolar ridge, conserving the erupted teeth (if salvageable) or extraction followed by prosthetic rehabilitation. (**J Dent Child 2012;79:26-9**)

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The first description of arrested tooth development, in 1947, included the following clinical features: delayed or failed eruption; abscess formation; early exfoliation; malformed teeth; radiographic findings of defective enamel, dentin formation, and calcifications within the pulp; and dental follicle[1] affecting several teeth within a particular region of the jaw.[2]

The term "odontodysplasia" was introduced by Zegarelli and Kutscher in 1963.[3] Since then, many cases have been described under various terminologies, such as: localized arrested tooth development; regional odontodysplasia; ghost teeth; odontogenesis imperfecta; unilateral dental malformation; amelogenesis imperfecta nonhereditary segmentalis; and familial amelodentinal dysplasia.[4] The term "regional odontodysplasia" (**RO**), however, has now become the accepted terminology.

RO is a nonhereditary, localized developmental abnormality affecting dental tissues derived from both the ectoderm and mesoderm.[5] Although most are idiopathic, a number of cases have been related to various pathoses,[6] such as: ectodermal dysplasia; epidermal and vascular nevi; hypophosphatasia; hydrocephalus; ipsilateral facial hypoplasia; and neurofibromatosis.

Despite the fact that a definitive cause for RO is uncertain, a variety of etiologies have been proposed,[3,7] including: abnormal migration of neural crest cells; local circulatory disturbance; radiation therapy; latent virus; local trauma or infection; malnutrition; somatic mutation; and hyperpyrexia. RO affects both stages of dentition in either or both jaws, with single or bilateral or multiquadrant involvement[7,8] with a higher predilection for the maxillary anterior teeth.[9,10] It exhibits no racial predilection, a slight female predominance, and a bimodal peak of occurrence correlating with the normal time of eruption of the primary and permanent teeth.[6]

Primary dentition affected by RO can be erupted, hypoplastic, hypocalcified, and discolored, which is commonly associated with periapical inflammatory lesions.[11,12] Primary teeth, when affected, are typically followed by involvement of the succedaneous teeth. The permanent teeth either fail to erupt or erupt partially with inflammatory or fibrous gingival involvement.[9]

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Radiographic examination shows significant reduction in radiodensity of the affected teeth, with lack of contrast between the enamel and dentin when compared to the unaffected counterparts. Also, the enamel and dentin layers tend to be thin, the pulp is enlarged, and the roots are short with open apices, giving the teeth a "ghost-like" appearance.[1,7,13]

Histology[5,14-17] reveals variable thickness of enamel with an irregular prism pattern. There is a marked reduction in the amount of dentin with irregular dentinal tubules and presence of interglobular dentin, 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 oralcavity and the pulp that the affected teeth often present pulp necrosis.[18] The pulp shows varying degrees of calcification, and the follicular tissue contains many irregular enameloid droplet calcifications.

The therapeutic approach is either directed toward conservation of the altered teeth[12,19] or removal with prosthetic rehabilitation,[8-10,20] whichever is possible.

The purpose of this paper was to report a case of regional odontodysplasia in a 12-year-old girl, with details of clinical and radiographic features, thereby facilitating management of this rare anomaly.

CASE REPORT

A 12-year-old female presented to the Department of Oral Medicine and Radiology, Tamil Nadu Government Dental College and Hospital, Chennai, Tamil Nadu, India, with the complaint of noneruption of her permanent maxillary right front teeth. An elaborate history stated that the eruption and exfoliation of primary teeth followed a normal schedule and that all her permanent teeth had erupted normally except for her maxillary right front teeth. The gestation and delivery of the patient was unremarkable, according to her mother, and her past medical and family histories were noncontributory.

An extraoral examination revealed no noticeable problems, and an intraoral examination (Figure 1) was significant only for the absence of the permanent maxillary right central incisor, lateral incisor, canine, first premolar, and second premolar and the presence of the primary maxillary right canine. All other permanent teeth for her age group had erupted.

An intraoral periapical radiograph (Figure 2) and maxillary cross sectional radiograph (Figure 3) revealed the presence of the maxillary right central incisor, lateral incisor, canine, and first premolar and the absence of a second premolar. These teeth showed a delayed development in comparison to the patient's homologous permanent teeth and primary canine. They had thin enamel and dentin walls with reduced radiodensity and lack of contrast as well as wide pulp spaces lacking root formation. These features suggested the appearance of ghost teeth, and a diagnosis of RO was made.

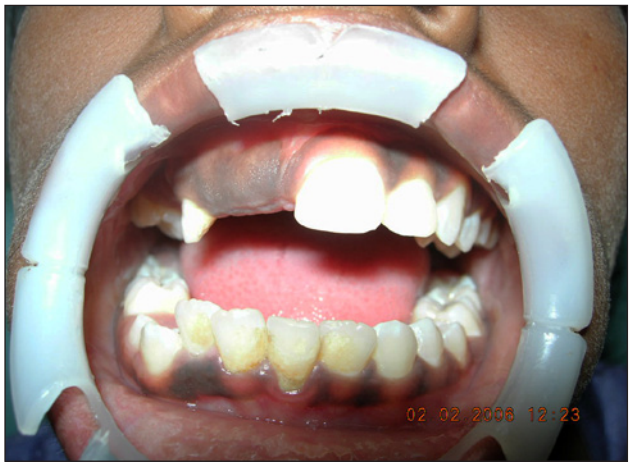

Figure 1. Intraoral view.

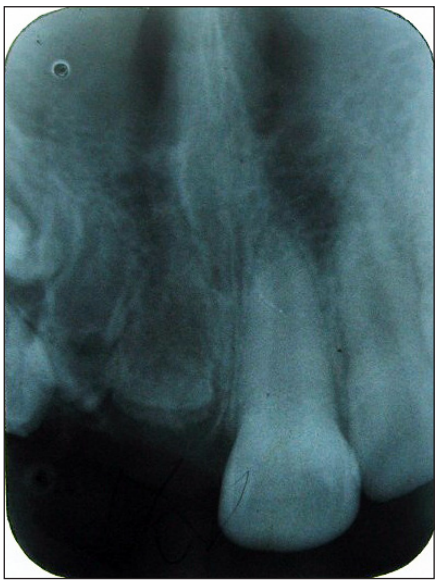

Figure 2. Periapical radiograph showing the characteristic ghost teeth appearance.

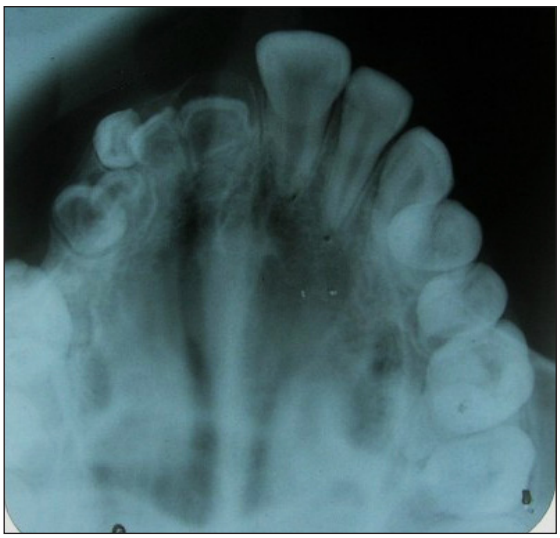

Figure 3. Maxillary occlusal radiograph showing the teeth with a ghost–like appearance in the maxillary right quadrant.

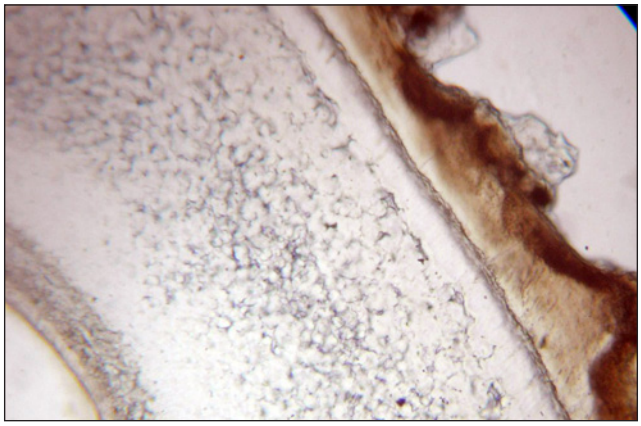

Figure 4. Ground section reveals an irregular enamel surface lacking a lamellated appearance and scalloped dentoenamel junction.

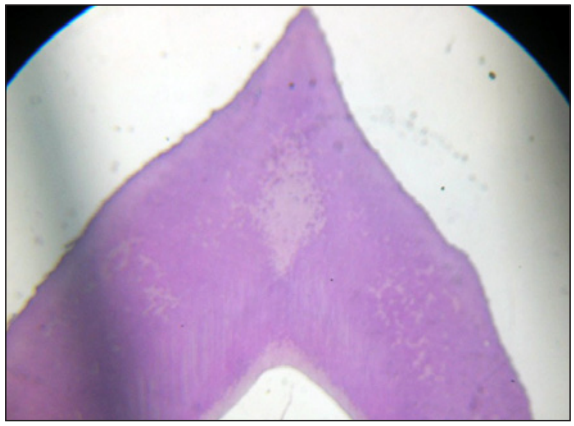

Figure 5. H&E section showing irregular dentinal tubules; interglobular dentin; and amorphous material with a widened predentinal zone.

Though dentinogenesis imperfecta was considered in the differential diagnosis, it was excluded due to the lack of history of familial occurrence and generalized involvement. Initial treatment planning called for extraction of the primary canine and retention of the affected teeth to favor the development of the alveolar ridge, followed by prosthetic rehabilitation for psychologic and functional support.

Considering the radiographic details, which showed the teeth edges nearly at the level of the alveolar crest, covered only by soft tissue and the possibility of an infection, the affected teeth were removed surgically and prosthetic rehabilitation was planned, which proved successful.

The extracted teeth were submitted for ground sectioning and histologic analysis (Figures 4 and 5). This revealed: the features of an irregular enamel surface lacking a lamellated appearance; a scalloped dentoenamel junction; dentin showing irregular dentinal tubules; interglobular dentin; and amorphous material with a widened predentinal zone.

The patient was placed on periodic recall to monitor the growth and development of her dental arch.

DISCUSSION

RO is a relatively rare, nonhereditary anomaly in which both the enamel and dentin are hypoplastic and hypocalcified, resulting in a localized arrest in tooth development. Multiple etiologies have been proposed, and various pathoses are associated with RO. In this case, it was noted that the patient did not present with any of these associated conditions; thus, the etiology was unknown. The dental literature suggests that this anomaly has no racial predilection but has a slight female predominance, as supported by this case.

RO typically affects a focal area of dentition involving several contiguous teeth, occasionally intermixed with an unaffected tooth. There is a maxillary predominance with predilection for anterior teeth. RO is a rare finding that occurs in both dentitions. In this case only the permanent maxillary right anterior teeth and premolars were affected and the primary dentition was unaffected, as evidenced by the patient's history and clinical and radiographic features.

Abscess formation, delayed or failed eruption, early exfoliation, malformed teeth, and noninflammatory gingival enlargement were the most common presenting complaints in past reported cases.[21] In our case, failure of eruption was the presenting symptom, with no sign of abscess formation. The radiographic and histologic features in our case included all the characteristic features of RO.

Other developmental alterations affecting the structure of teeth, such as amelogenesis imperfecta, dentinogenesis imperfecta, and dentin dysplasia, however, are similar to RO; a nonhereditary pattern and localized involvement excludes them from the differential list.

The best treatment option for RO depends on the time of diagnosis, presenting symptoms and signs, functional and esthetic needs, and available treatment modalities. The affected unerupted teeth may result in the delay or failure of the process of eruption in addition to structural defects, and the possibility of bacterial infection is frequent. Because addressing the psychological and functional needs is more important in this group of patients, less invasive therapeutic approaches have been suggested. We opted to surgically remove the involved teeth, execute prosthetic rehabilitation, and provide regular follow-up care.

REFERENCES

1. Langlais RP, Langland OE, Nortje CJ. Diagnostic Imaging of the Jaws. 1st ed. Philadelphia, Pa: Lippincott Williams and Wilkins; 1995:144-6.
2. McCall JO, Wald SS. Clinical Dental Roentgenology. 3rd ed. Philadelphia, Pa: WB Saunders; 1952:170.
3. Zegarelli EV, Kutscher AH, Applebaum E, et al. Odontodysplasia. Oral Surg Oral Med Oral Pathol 1963;16:187-93.

4. Pandis N, Polido C, Bell WH. Regional odontodysplasia: A case associated with asymmetric maxillary and mandibular development. Oral Surg Oral Med Oral Pathol 1991;72:492-6.
5. Hamdan MA, Sawair FA, Rajab LD, et al. Regional odontodysplasia: A review of the literature and report of a case. Int J Paediatr Dent 2004;14:363-70.
6. Neville BW, Damm DD, Allen CM, et al. Oral and Maxillofacial Pathology. 2nd ed. Philadelphia, Pa: WB Saunders; 2002:99-100.
7. Ozer L, Cetiner S, Ersoy E. Regional odontodysplasia: Report of a case. J Clin Pediatr Dent 2004; 29:45-8.
8. Lustmann J, Klein H, Ulmansky M. Odontodysplasia: Report of two cases and a review of the literature. Oral Surg Oral Med Oral Pathol 1975; 39:781-93.
9. Vaikuntam J, Tatum NB, McGuff HS. Regional odontodysplasia: Review of the literature and report of a case. J Clin Pediatr Dent 1996;21:35-40.
10. Sabah E, Eden E, Unal T. Odontodysplasia: Report of a case. J Clin Pediatr Dent 1992;16:115-8.
11. Crawford PJ, Aldred MJ. Regional odontodysplasia: A bibliography. J Oral Pathol Med 1989;18:251-63.
12. Yuan SH, Liu PR, Childers NK. An alternative restorative method for regional odontodysplasia: Case report. Pediatr Dent 1997;19:421-4.
13. Rushton MA. Odontodysplasia: Ghost teeth. Br Dent J 1965;119:109-13.
14. Kerebel B, Kerebel LM. Structural, ultrastructural, microradiographic, and electron-probe studies of an unusual case of regional odontodysplasia. J Dent Res 1982;61:1056-62.
15. Kahn MA, Hinson RL. Regional odontodysplasia: Case report with etiologic and treatment considerations. Oral Surg Oral Med Oral Pathol 1991;72: 462-7.
16. Tervonen SA, Stratmann U, Mokrys K, et al. Regional odontodysplasia: A review of the literature and report of four cases. Clin Oral Investig 2004; 8:45-51.
17. Courson F, Bdeoui F, Danan M, et al. Regional odontodysplasia: Expression of matrix metalloproteinases and their natural inhibitors. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003;95:60-6.
18. Juliana OG, Hermes P, Ramalho Lizeti TO, et al. Regional odontodysplasia in early childhood: A clinical and histological study. J Indian Soc Pedod Prev Dent 2009;27:175-8.
19. Oncag O, Eronat C, Sen BH. Regional odontodysplasia: A case report. J Clin Pediatr Dent 1996;21: 41-6.
20. Cabral LA, Carvalho YR, Moraes E, et al. Regional odontodysplasia: A report of three cases. Quintessence Int 1994;25:141-5.
21. Steiman HR, Cullen CL, Geist JR. Bilateral mandibular regional odontodysplasia with vascular nevus. Pediatr Dent 1991 Sep-Oct;13(5):303-6.

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