Case Report
Journal of Pierre Fauchard Academy (India Section), *Volume 38(1-2), DOI: 10.18311/jpfa/2024/36550, January-June 2024 p. 21-25*
ISSN (Print) : 0970-2199
ISSN (Online) : 2405-772X

Regional Odontodysplasia: A Rare Developmental Anomaly

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Abstract

Regional Odontodyplasia (RO) is a rare abnormality of unknown aetiology affecting dental tissue derived from mesoderm and ectoderm. It occurs more commonly in women, with no racial predilection and often in maxillary dentition restricting to a single quadrant. These odonto-dysplastic teeth radiographically appear as Ghost teeth with hypoplastic enamel, large areas of hypo-mineralised interglobular dentin with an irregular arrangement of dentinal tubules and denticles in the enlarged pulp cavity. This case report discusses clinical, radiological and histological findings for accurate diagnosis of RO.

Keywords: Anomaly, Ghost Teeth, Regional Odontodysplasia (RO)

1. Introduction

Regional odontodysplasia is a developmental dental anomaly in which one or several teeth in a localised area are affected in an unusual manner[1]. Hitchin (1934) first described regional odontogenic dysplasia. The term odontodysplasia was coined by Zegaralli *et al.* and he described it as a developmental anomaly affecting tooth formation involving ectodermal and endodermal tissue[2]. Pindborg (1970) added the term 'Regional' with odontodysplasia to describe its nature of affecting one quadrant of the jaw[1-4].

The etiology of RO remains uncertain, with potential causes including trauma, infection, local ischemia, vascular defects, RH incompatibility, irradiation, neural damage, hyperpyrexia, metabolic and nutritional disturbances, as well as vitamin deficiencies[1-3,5,6]. It has a low incidence rate of < 1 case per 10,000,000 live births[3]. RO affects both primary and permanent dentitions. Generally, it is limited to only one arch but sometimes crosses the midline[6]. The maxilla is affected twice that of the mandible, with a higher prevalence of the left jaw quadrant being affected compared to the right, and instances of crossing the midline being rare[3-9].

The characteristics of this anomaly are discoloured and soft teeth that can be accompanied by swelling or abscess[3-6,8]. In this anomaly, enamel and dentin are thin and hypoplastic, therefore, in the radiograph the teeth give the impression of Ghost teeth. In radiography, the delineation between enamel and dentin is not clear and the pulp chamber is wide[3,5-8]. Histologically, areas of hypocalcified enamel are observed and the enamel prisms appear to be irregular in direction. There is a disturbance in dentin formation and dentinal tubules are reduced in number[3,4,7,8].

Management of these cases should be based on the aesthetics, functional needs and the degree of involvement. This report presents rare and unusual findings showcased in two cases of RO, which were identified through clinical, and radiographic examinations and histologically confirmed.

2. Case Report

Two cases (Case 1- 17-year female and Case 2- 20-year male) were reported to the Department of Oral Medicine and Radiology, Case 1 complained of non-eruption of teeth in the right maxillary quadrant region and Case 2 reported pus discharge in the upper anterior region since two months. Both cases were the product of a nonconsanguineous marriage and had a normal birth following an uncomplicated pregnancy, no notable medical history/ medication during pregnancy or any significant events post-birth, or history of trauma was

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Article Received on: 12.02.2024 Revised on: 12.07.2024 Accepted on: 10.08.2024

reported. History of normal eruption and exfoliation of deciduous teeth with normal size and shape were reported in both patients. Intraorally, both cases exhibited a normal pattern of permanent dentition eruption except in the maxillary right quadrant crossing midline and involving the opposite side central incisor (Figure 1).

In Case 1, teeth #11, #12, #13, #14, #16, and #21 were missing. Tooth #15 and #17 were partially erupted with a chalky white appearance and soft piercing enamel (Figure 1A, B). Gingiva in the affected quadrant was enlarged, firm, non-tender and lobulated in appearance. The teeth in the opposing right mandibular jaw were supraerupted along with remarkable gingival enlargement.

The occlusal radiograph, OPG showed there were missing tooth germs of tooth #12, #13, and #16. The partially erupted teeth #15 and #17 and impacted teeth #11, #14, and #21 exhibited reduced radiodensity of enamel, dentin, and cementum compared to teeth in the unaffected region. Affected teeth were smaller in size and had a short slender root. The radiodense round structure seen at the position of tooth #11 had a density similar to affected teeth with a radiolucent rim (Figure 2A). CBCT findings confirmed the above findings and showed an ill-defined hypodense region in the periapical region of tooth #17 with loss of lamina dura and impacted #11 with altered shape, irregular enamel surface and widened pulp chamber (Figure 1C).

In Case 2, a diffuse swelling was seen in the maxillary depth of the vestibule, in the region of teeth #11 and #12, accompanied by sinus tract formation and pus discharge. Discolouration of tooth #11 and #21 with irregular enamel surface, malformation of the crown and soft porous enamel were noted (Figure 1B). These clinical findings of partial anodontia with enamel hypoplasia affecting the maxillary anterior region with chronic apical abscess were present.

The crown and roots of affected teeth #11, 12, 13 and 21 were smaller and malformed with reduced density of enamel and dentin. Ghost teeth appearance was seen specifically in this region due to enlarged pulp chamber and open apices. The affected canine had a smaller size, altered shape and absence of roots giving the appearance of Rootless teeth. An Ill-defined radiolucency was seen in the periapical region about tooth #21 with loss of lamina dura, suggesting apical abscesses (Figure 2A, B). The CBCT evaluation depicted an irregular enamel surface, decreased radiodensity between enamel and dentin and widened pulp chamber of tooth #11 (Figure 2C).

Provisional Diagnosis: After radiographic evaluation in both cases regional odontodysplasia was suspected with differential diagnosis of dentin dysplasia. To confirm the diagnosis gingival biopsy was performed alongside the extraction of affected teeth and samples were sent for laboratory investigation.

The histopathological findings from the gingival biopsy revealed para-keratinized stratified squamous epithelium with hyperplastic gingiva. Ground section analysis of the teeth showed irregular enamel deposition and enamel spindles. Dentin displayed large areas of interglobular dentin with an irregular arrangement of dentinal tubules. Denticles were observed in the pulp cavity for Case 1 (Figure 4).

Based on the comprehensive clinical, radiological, and histological findings, a diagnosis of Regional Odontodysplasia was established for both cases. A Removable Partial Denture (RPD) with occlusal adjustment to increase vertical dimension was advised as an interim treatment to case 1 for six months (Figure 5A, B) followed by prosthetic rehabilitation using an implant. For case 2 extraction of teeth #11 and #13, endodontic treatment for 12 and 21 followed by prosthetic rehabilitation was advised.

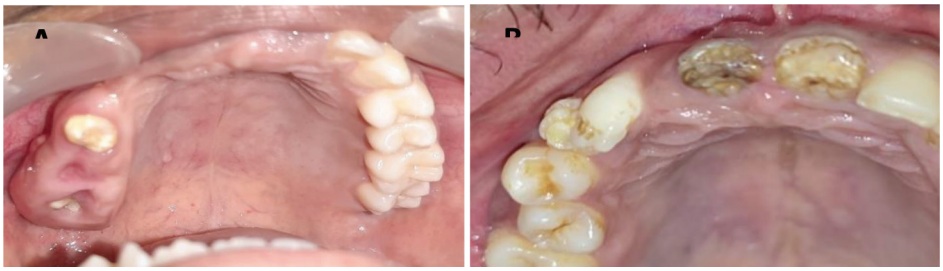


Figure 1. **(A)** Case 1: Oligodontia of right maxillary arch crossing the midline with partially erupted tooth #15 and #17, (chalky white appearance) and associated gingival enlargement; **(B)** Case 2: Malformed tooth #11 and #21 (altered size and shape with irregular enamel surface) associated with spacing.

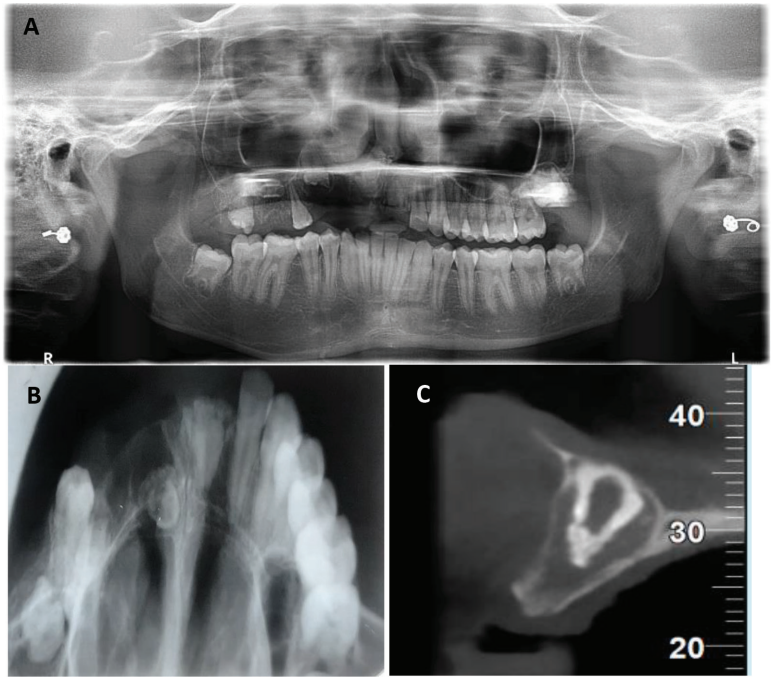


Figure 2. Case 1: **(A)** Orthopantogram, **(B)** Occlusal radiograph depicting multiple missing and malformed teeth with winded pulp chamber with reduced radiodensity of enamel and dentin giving Ghost tooth appearance involving right maxillary arch crossing midline, canine was notably smaller, misshapen, and appeared rootless; **(C)** CBCT image displaying affected tooth with malformed smaller crowns and roots, reduced enamel and dentin density, and a "ghost teeth" appearance due to enlarged pulp chambers and open apices.

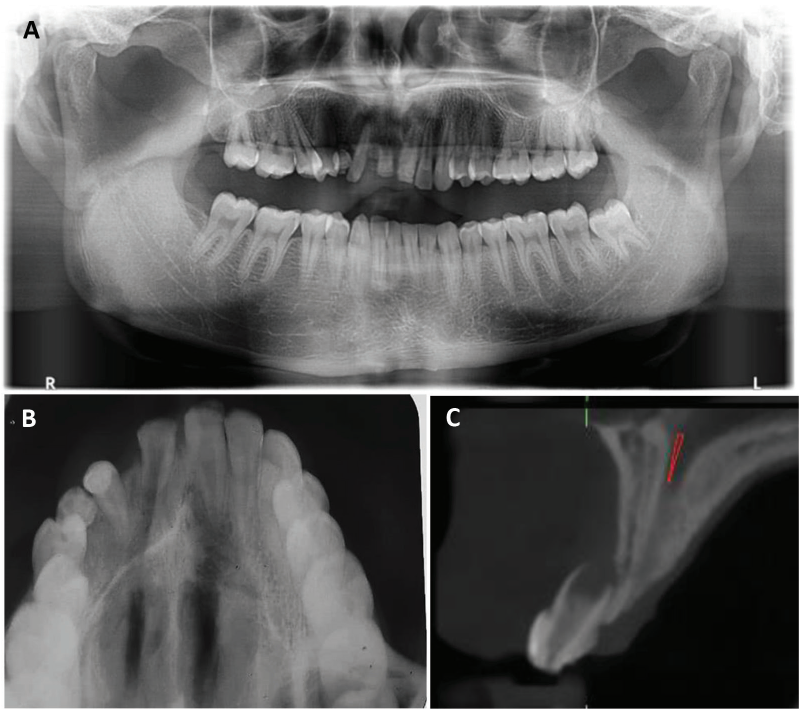


Figure 3. Case 2: **(A)** Orthopantogram, **(B)** Occlusal radiograph short roots of tooth #11 and #13; **(C)** CBCT section shows irregular enamel surface smaller size, reduced radiodensity involving enamel and dentin and winded pulp chamber with open apices involving affected teeth giving Ghost tooth appearance.

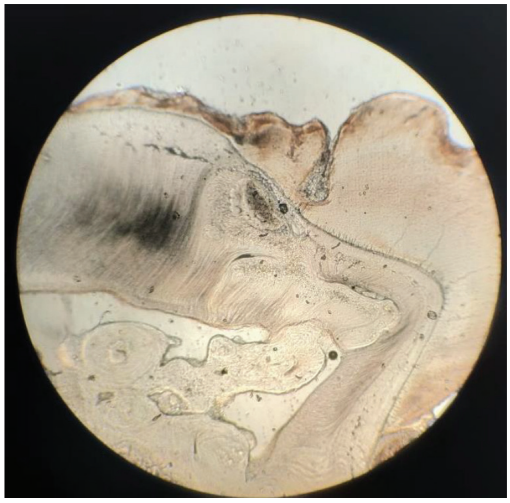


Figure 4. Case 1: Histopathological ground section analysis of tooth shows irregular enamel deposition and large interglobular dentin with irregular dentin tubules arrangement.

to be more prevalent in females as found in the first case[3,5-7,9].

Clinically, affected teeth appeared to be Chalky white, discolored, and irregular surface with pitting involving the tooth surface and partially erupted or unerupted into the oral cavity. The thin enamel is soft on probing and teeth are typically discoloured, yellow or yellowish-brown. It is possible to find some teeth without any alterations in the affected quadrant[6]. Similar findings were noted in case 1 and case 2. In the present study, case 1 shows missing teeth #12, 13, and 16 and partially erupted #15, and 17 with a white chalky appearance of teeth and case 2 shows teeth having irregular surfaces with pitting involving teeth #11, 12, 13, 21 with altered size and shape[3,6,7]. Common clinical features of RO are gingival enlargement, periapical infection and abscess formation in the absence of caries. Alotaibi *et al.* concluded that failure of tooth eruption was followed by swelling of the affected area. The affected

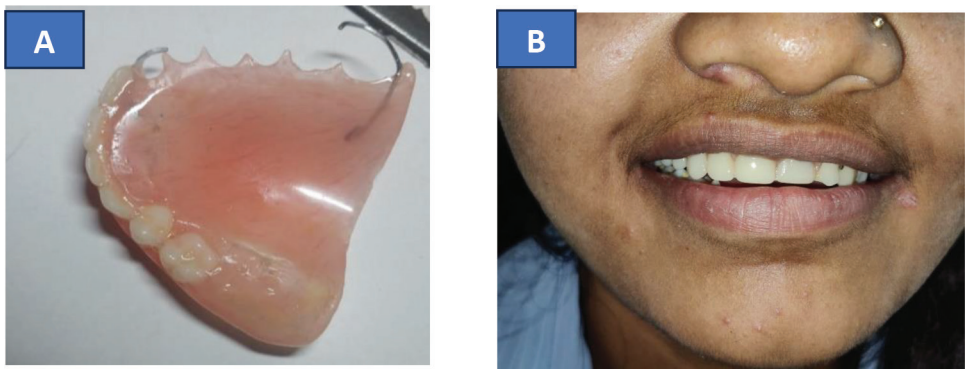


Figure 5. Case 1: **(A)** Removable partial denture for restoration of missing teeth in patient with Regional Odontodysplasia; **(B)** smile view of patient with RPD. rehabilitation with temporary maxillary partial acrylic denture.

3. Discussion

Regional odontogenic dysplasia is a rare developmental anomaly[3,5]. This condition affects the dentin and enamel of a group of adjacent teeth. Its exact aetiology is uncertain, although several factors are involved. In many cases, RO is probably misdiagnosed as malformed teeth or odontomas. It occurs in both primary and permanent dentitions, with a marked preference for the maxilla. According to Lustmann *et al.*, the maxillary arch is affected more frequently and rarely crosses the midline[3-6]. In this present report, RO affected only one quadrant in the maxilla, but it crossed the midline and involved the opposite quadrant incisor, as previously reported by Magalhães *et al.* in both cases[6]. RO seems teeth are irregular with malformed roots which prevent tooth eruption[9].

Radiographically, Koskinen *et al.* and Fatemeh *et al.* said that there is reduced radiodensity of enamel and dentin, both of which are less radiopaque than unaffected teeth and give a Ghost-like appearance[3,5-8]. In this report, all affected teeth had reduced radiodensity of enamel and dentin in comparison to unaffected teeth and unerupted teeth 11, 14, 21 with altered size and shape seen in case 1 and case 2 shows crown and root structures of teeth were malformed teeth #11, 12, 13 and 21, and there was low contrast between enamel and dentin indicating hypomineralization. An ill-defined radiolucency was seen in the periapical region of teeth # 11 and 21 with loss of lamina dura suggestive of apical abscess with widened pulp chamber giving Ghost teeth appearance.

According to Zegaralli *et al.*, Rashidian *et al.* and Jahanimoghadam *et al.*, the histological findings of RO show a marked reduction in the amount of dentin and enamel, a widening of the pre-dentin layer and the presence of large areas of interglobular dentin and an irregular tubular pattern of dentin. Sometimes attached denticles are seen in the pulp cavity[2,3,5,7,10]. In a present case report, case 1 shows there is irregular deposition of enamel and enamel spindle. Dentin shows large areas of interglobular dentin with irregular arrangements of dentinal tubules. The attached denticles are seen in the pulpal cavity supporting our diagnosis.

The treatment plan for RO patients must consider several factors, including the patient's age, medical history, previous dental history, and the attitudes of the patient and their parents towards dental treatment, as well as the number of affected teeth. There has been considerable debate regarding whether to extract or preserve affected teeth. Ziegler *et al.*[10] suggested that a combined surgical, orthodontic, and prosthetic approach is necessary for treating RO. In this case, the proposed treatment for case 1 involved the extraction of impacted and affected teeth, followed by the provision of a removable appliance with occlusal adjustment to increase the vertical dimension for 12 months, and subsequent rehabilitation using implant prostheses. For case 2, the extraction of 11 and 13, root canal treatment for 12 and 21 and prosthetic rehabilitation were advised.

4. Conclusion

In concluding this case report on regional dysplasia, the diagnosis was substantiated through a comprehensive analysis integrating clinical observation, and radiological and histological examination. This triad of diagnosis approach was instrumental in establishing a definitive diagnosis of RO. Integration of these findings not only underscores the complexity of this dental anomaly but also highlights the importance of a multidisciplinary approach in the accurate diagnosis of such rare conditions.

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