Regional odontodysplasia crossing the midline: A unique case associated with bilateral impacted supernumerary teeth

L. R. Kumaraswamy Naik^{1*}, Pushparaja Shetty², K. L. Girish Babu³

¹Department of Oral Pathology, SJM Dental College and Hospital, Chitradurga, ²Department of Oral Pathology, AB Shetty Memorial Institute of Dental Sciences, Deralakatte, Mangalore, ³Department of Pedodontics, Oxford Dental College Hospital, Bengaluru, Karnataka, India

ABSTRACT

Regional odontodysplasia (RO) is an uncommon developmental anomaly affecting a localized area of dentition, with distinctive clinical, radiographic, and histological findings. It is presumed to be the result of a developmental disturbance that locally affects the odontogenic ectodermal and mesodermal tissues. In general, it is limited to only one arch and sometimes it can occasionally cross the midline. Supernumerary teeth (ST) are mammalian developmental abnormalities characterized by the presence of extra teeth in addition to the normal eruption series. Here, we report a case of RO that affected right maxillary teeth crossing the midline and showed two maxillary bilateral impacted ST.

Key words: Regional odontodysplasia, Impacted teeth, Supernumerary teeth, Bilateral



INTRODUCTION

egional odontodysplasia (RO) is a rare developmental Nanomaly involving both mesodermal and ectodermal dental components in groups of contiguous teeth first described by Hitchin in 1934.^[1] It usually affects both the primary and permanent dentition. It is more frequent in girls than in boys (1.7:1), and the maxilla is affected twice as often as the mandible.^[1,2] It is limited to only one arch, maxillary left being most common.^[3,4] It can occasionally cross the midline,^[2] and RO crossing the midline lesions are seen more frequently in mandible.^[1,4] In teeth central and lateral incisors are affected more commonly than posterior teeth. In the same quadrant degree of RO varies among affected teeth.^[3] The etiology of RO is uncertain, though many causative factors have been suggested, for example, vascular defects involving ischemia, local infections, pharmacological treatments during pregnancy, trauma, rhesus incompatibility, irradiation, neural damage, activation of latent viruses

residing in odontogenic epithelium, hyperpyrexia, metabolic and nutritional disturbances, hereditary and somatic mutation, and also neural crest migration disorders associated with hemangiomas.^[4-7] It is thought to be a noninherited condition as no cases of family history were described.^[3] Most frequent clinical symptom is missing or unerupted teeth.^[1] If teeth are erupted they usually present with gingival swelling, periapical infection and abscess formation in the absence of caries.^[8] Affected teeth in RO are colored yellowish brown, soft, hypoplastic and hypocalcified.^[9] These teeth are more susceptible for dental caries, extremely friable and can get fractured to the slightest trauma.^[3] The radiographic feature in RO is described as "ghost teeth," a term used to describe a tooth with thin and indistinct enamel and dentine, exceedingly large pulp chamber with short root and open apices.^[6] Histological examination of affected teeth reveals a fibrous enlargement of the pulp and an irregular pulp/ dentin interface, with pseudo inclusions and pulp stones. Tubular defects are found in the dentin. Follicular tissue shows a dense collagenous fibrous tissue with focal

*Address for correspondence

Dr. L. R. Kumaraswamy Naik, Department of Oral Pathology, SJM Dental College and Hospital, Chitradurga, Karnataka, India. E-mail: drkumarswamylr@rediffmail.com

aggregations of calcified bodies (enameloid) and many islands of odontogenic epithelium.^[1,10] Optimal treatment for this rare anomaly is controversial. Some authors advocate a more conservative approach, including the use of restorations to protect the permanent teeth or simply long-term follow-up whereas others argue in favor of immediate extraction of affected teeth, and then prosthetic rehabilitation.^[6]

Supernumerary teeth (ST) are mammalian developmental abnormalities characterized by the presence of extra teeth in addition to the normal eruption series. Prevalence of ST varies between 0.3% and 1.9% in the primary dentition and 1-3.6% in the permanent dentition.^[11] ST may occur singly, multiply (>5 STs), unilaterally or bilaterally, and in one or both jaws. Cases involving one or two ST most commonly involve the anterior maxilla, followed by the mandibular premolar region.^[12] It has been reported that the rate of bilateral distribution is 17-44% in cases where more than one supernumerary tooth is present.^[13] This paper reports a male case of RO affecting the permanent teeth of the right side crossing the midline affecting the left central incisor and is associated with maxillary bilateral impacted ST.

CASE REPORT

A 15-year-old male reported to the Department of oral medicine and radiology AB Shetty Memorial Institute of Dental Sciences, Mangalore, Karnataka, India, with a complaint of unerupted upper right front teeth. No other symptoms were present, and the patient reported no pain. The past dental/medical history was noncontributory. According to patient's parents, the milk teeth were erupted and exfoliated at right time without any complications. Both parents reported no previous history of tooth or genetic anomalies on either side of the family or in the siblings. No facial asymmetry or observable pathology was noted on extra oral examination. On Intraoral examination, there were missing right upper permanent central incisor, lateral incisor and canine [Figure 1]. On left maxillary, quadrant central and lateral incisors were partially erupting. Only central incisor was hypoplastic with yellow spots. Radiographic features revealed malformed and impacted central incisor, lateral incisor and canine on the right side. Left central incisor was also hypocalcified and malformed. Radiographic differentiation of enamel and dentin was not clear. There was pulpal chamber obliteration. Right lateral incisor showed slight follicular enlargement. The pattern was suitable for the term "ghost teeth." Two developing ST in relation to roots of right and left maxillary premolars was also noted [Figures 2 and 3]. Provisional diagnosis of RO was arrived at. Given the observed delay in dental eruption and the fragile hard tissue of the affected



Figure 1: Clinical photograph showing missing right maxillary central incisor, lateral incisor and canine. Partially erupting left central and lateral incisor can be noted



Figure 2: Occlusal view showing "ghost like" appearance of maxillary right central incisor, lateral incisor and canine. Also observe two developing supernumerary teeth in relation to roots of right and left maxillary premolars



Figure 3: Intra oral periapical radiography showing "ghost like" appearance of maxillary right central incisor, lateral incisor and canine. Left central incisor is also affected. Note developing supernumerary teeth in relation to roots of right maxillary premolar

teeth, the treatment plan was consisted of two phases, extraction of affected teeth along with bilateral ST and subsequent functional rehabilitation through prosthesis. Extracted teeth along with the dental follicle were received for histopathological examination. The teeth were of altered morphology, yellowish in color, soft to rubbery in consistency, and had very short or unformed roots with wide open apices. Histopathology sections of both decalcified and ground sections of teeth along with the soft tissue of the dental follicle was prepared. Microscopic features of follicular tissue showed collagenous stroma in whorls with many round, ovoid shaped plump cells with basophilic round, oval and irregular calcifications within the whorls. Few large areas of calcifications were formed by coalescing of smaller calcific masses [Figure 4]. There was the absence of any osteoblastic/osteoclastic cells surrounding these calcified masses. There were no odontogenic islands. Decalcified sections of the teeth revealed pulp stones and amorphous atypical dentin formation [Figure 5]. Ground section revealed large pulp stones, atypical dentine formation, and large areas of inter-globular dentine. Enamel is poorly formed and lacked the normal thickness and structure. There was the



Figure 4: Photomicrograph of follicular tissue showing collagenous stroma in whorls with many round, ovoid shaped plump cells with basophilic round, oval and irregular calcifications (H and E, stained section original magnification ×10)

absence of scalloping at dentin-enamel junction [Figures 6 and 7]. Among the extracted, ST right one was showing mild features of RO, whereas left one did not show any abnormalities. Depending upon clinical, radiographic and microscopic findings the diagnosis of RO was confirmed. Two months postoperatively, a temporary acrylic maxillary partial denture was made to preserve the alveolar ridge during the period of skeletal growth. A final rehabilitation for the loss of teeth might be accomplished after the facial bones had attained complete growth and maturity.

DISCUSSION

Regional odontodysplasia is an uncommon developmental anomaly, and it rarely crosses the midline. Though previous reports suggest involvement of upper left quadrant commonly, in the present case right upper contiguous permanent teeth was affected along with the



Figure 5: Photomicrograph of decalcified sections of the teeth revealing pulp stones and amorphous atypical dentin formation (H and E, stained section original magnification ×5)



Figure 7: Photomicrograph of ground section showing poorly formed enamel lacking the full thickness, absence of scalloping in dentin-enamel junction and large areas of pulp stones (Original magnification ×4)



Figure 6: Photomicrograph of ground section showing large areas of inter-globular dentine formation (Original magnification ×10)

left central incisor.^[2-4] Cases crossing the midline can also affect lateral incisor on the opposite side.^[4] It is more frequent in females and present case occurred in a male patient.^[2] One interesting feature, which was observed in previous literature was that, though rare in mandible, RO in these mandibular lesions tends to cross the midline frequently when compared with maxilla. Moreover, these patients are usually females (A), (B), (C). However in the present case, RO affected maxilla in a male patient. Past dental/medical history in the present case was noncontributory and according to patient's parents the primary teeth were unaffected though previous reports suggest that both deciduous and permanent teeth get affected in the same arch.^[4]

In the present case, parents were also unaware/could not recollect any birth injury or medication or any other disease related to etiology of tooth abnormality. However in previous reports there were some relevant history, which can be related for etiological factors.^[3,4] Though numerous systemic etiological factors were suggested for RO,^[4-7] we believe systemic factors are less responsible as they usually produce generalized defects, and these defects are represented in each teeth depending upon a time of insult. Since the parents and siblings of RO patients be clinically normal, a noninherited local ischemia which can be a result of somatic mutation could be an etiological factor as suggested by many authors.^[3,9,14] However in the present case, as seen in previous reports no such clinically observable vascular ischemic lesions were seen near the affected area.^[1] According to Barbería et al. it was suggested in one of their cases that, etiology could be a history of extensive infection and contralateral vascular and nerve supply could be the cause for RO to cross the midline. However, it only explains etiology of RO affecting permanent teeth and not for deciduous dentition.^[4] However in RO cases of maxilla crossing the midline, factors, which occur locally can affect through vascular/ neural agents due to contralateral supply. However, it is not true in every case that involve maxilla and nor all cases involves both guadrants.^[1] Hence, studies at molecular level are to be taken up to evaluate the etiology of RO.

Though ST are common in occurrence, the etiology still remains unclear. Many theories are proposed to explain the cause for developmental anomaly.^[12,13,15] Among them localized and independent hyperactivity of the dental lamina is the most accepted cause for the development of ST.^[12] Furthermore as ST are mostly seen in individuals with some other dental anomalies and developmental disorders, it is thought that their development may be influenced by a combination of hereditary and environmental factors.^[13] Hence, Hattab *et al.* describe hyperdontia as "a multifactorial inheritance disorder which originates from hyperactivity of the dental lamina."^[16] It has been

reported that other dental anomalies, such as hypodontia, taurodontism, germination and macrodontia are likely to be observed with ST.^[13] In only one case it is reported until now that RO is associated with multiple ST.^[9] And we believe that this is the first case to report in which we saw simultaneous occurrence of bilateral ST and RO crossing midline. ST may also cause diastemata, root resorption of adjacent teeth, malformation of adjacent teeth such as dilaceration, and loss of vitality of adjacent teeth. Moreover, these are considered to be the effect of local pressure and not a coincidental finding.^[12] It has also been reported that no relationship between ST and other anomalies could be found and that ST should be considered a distinct entity.[13] Similarly in the present case, it is difficult to relate any common etiological factor for RO and ST as one is considered to be nonhereditary disorder whereas other is a multifactorial hereditary defect.^[3,9,16] Though they were considered to be distinct entities we believe any teeth in the region of RO if susceptible will be affected and will show the features of RO as seen in the present case, wherein only right ST was affected. Degree of involvement in RO is different in each tooth and in the present case left central was less affected than right central and lateral incisor. Right canine and developing STs were very small and showed delay in development. Development of ST depends upon a time of induction of dental lamina for abnormal proliferation, and three types of ST were described in the literature.^[15] They are predeciduous type, prepermanent type and postpermanent type. Moreover, we believe in the present case it is postpermanent type, as unaffected quadrants showed full set of teeth accept all third molars. Moreover, when bilateral ST occur they usually occur at same time.[15]

Radiographic evaluation in delayed eruption of missing teeth is very important for diagnosis and in the present case features of lack of radiographic distinction between enamel and dentine, large pulp stones, open apex and malformed teeth all together suiting the term ghost teeth as reported previously.^[1,3,4] Histopathologically features in the present case accentuates the morphological and radiographic alterations of the affected teeth in the form of alteration in enamel thickness and structure, atypical dentine formation with large areas of interglobular dentine, presence of large pulp stones. One more important feature seen in the present case was the lack of scalloping in dentinoenamel junction that is rarely reported in literature.^[17] Few features like absence of osteoblastic/osteoclastic rimming surrounding the calcified masses could suggest failed efforts to form normal enamel as reported previously.^[1] ROs are probably misdiagnosed as malformed teeth or odontomes. Other conditions, such as dentinal dysplasia, shell teeth, hypophosphotasia, dentinogenesis imperfecta or amelogenesis imperfecta can mimic some features of RO. However, these disorders tend to affect the entire dentition.^[8]

Treatment of RO is controversial and should be directed toward severity of tooth involved, its susceptibility for subsequent infection, development of alveolar bone, and patient treatment needs.^[1,3,4] Few authors suggest long term followup and other insists on immediate extraction and further prosthetic rehabilitation. Moreover, in most of the cases later treatment was adapted in literature.^[1] In the present case observing the arrest/delay in development of teeth, patient age, and considering the susceptibility of RO affected teeth for infection it was decided to extract the severely affected teeth (sparing partially erupted left central incisor) along with bilateral impacted ST. Prosthetic rehabilitation with implants will be planned in the future after complete development of the alveolar ridge.

In summary, an interesting case of simultaneous occurrence of RO and bilateral impacted ST is reported. Better understanding of any etiological association if present, when dental anomalies occur simultaneously will be only possible when similar cases are reported and studied in large scale. Etiology of RO remains obscure, and there is a need for more studies based on molecular aspects.

ACKNOWLEDGMENTS

Dr. G.S. Kumar, Professor and Head, Department of Oral Pathology, S.D.M. College of Dental Sciences, Dharwad, Karnataka, India.

REFERENCES

- Al-Tuwirqi A, Lambie D, Seow WK. Regional odontodysplasia: Literature review and report of an unusual case located in the mandible. Pediatr Dent 2014;36:62-7.
- Scariot R, Morosini I, Pereira CT, Amenabar JM, Rebellato N, Gugisch R. Regional odontodysplasia: A case report. Rev Odonto Cien 2012;27:246-50.

- Magalhães AC, Pessan JP, Cunha RF, Delbem AC. Regional odontodysplasia: Case report. J Appl Oral Sci 2007;15:465-9.
- Barbería E, Sanz Coarasa A, Hernández A, Cardoso-Silva C. Regional odontodysplasia. A literature review and three case reports. Eur J Paediatr Dent 2012;13:161-6.
- Ozer L, Cetiner S, Ersoy E. Regional odontodysplasia: Report of a case. J Clin Pediatr Dent 2004;29:45-8.
- Cahuana A, González Y, Palma C. Clinical management of regional odontodysplasia. Pediatr Dent 2005;27:34-9.
- Gomes MP, Modesto A, Cardoso AS, Hespanhol W. Regional odontodysplasia: Report of a case involving two separate affected areas. ASDC J Dent Child 1999;66:203-7.
- Upadhyay V, Chaturvedi TP, Pandey RK, Chaurasia A, Singh P. Regional odontodysplasia crossing midline: A rare case report. Int J Clin Pediar Dent 2011;4:159-61.
- Hema HM, Preetha A. Regional odontodysplasia An unusual case occurring along with supernumerary teeth. Indian J Dent 2002;3:29-32.
- Carlos R, Contreras-Vidaurre E, Almeida OP, Silva KR, Abrahão PG, Miranda AM, et al. Regional odontodysplasia: Morphological, ultrastructural, and immunohistochemical features of the affected teeth, connective tissue, and odontogenic remnants. J Dent Child (Chic) 2008;75:144-50.
- Naik KL, Shetty P, Shaila M, Sreelatha SV, Chowdappa HT. Unicystic ameloblastoma associated with impacted supernumerary premolar: An unusual case report. Arch Int Surg 2013;3:198-200.
- Scheiner MA, Sampson WJ. Supernumerary teeth: A review of the literature and four case reports. Aust Dent J 1997;42:160-5.
- Açikgöz A, Açikgöz G, Tunga U, Otan F. Characteristics and prevalence of non-syndrome multiple supernumerary teeth: A retrospective study. Dentomaxillofac Radiol 2006;35:185-90.
- Fearne J, Williams DM, Brook AH. Regional odontodysplasia: A clinical and histological evaluation. J Int Assoc Dent Child 1986;17:21-5.
- Kasat VO, Saluja H, Kalburge JV, Kini Y, Nikam A, Laddha R. Multiple bilateral supernumerary mandibular premolars in a non-syndromic patient with associated orthokeratised odontogenic cyst: A case report and review of literature. Contemp Clin Dent 2012;3:S248-52.
- Hattab FN, Yassin OM, Rawashdeh MA. Supernumerary teeth: Report of three cases and review of the literature. ASDC J Dent Child 1994;61:382-93.
- 17. Kappadi D, Ramasetty PA, Rai KK, Rahim AM. Regional odontodysplasia: An unusual case report. J Oral Maxillofac Pathol 2009;13:62-6.

How to cite this article: Naik LK, Shetty P, Girish Babu KL. Regional odontodysplasia crossing the midline: A unique case associated with bilateral impacted supernumerary teeth. J Pediatr Dent 2014;2:96-100.

Source of Support: Nil. Conflict of Interest: None declared.