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REGIONAL ODONTODYSPLASIA: A UNIQUE DENTAL ANOMALY WITH AN INSIGHT INTO ITS POSSIBLE ETIOLOGIC FACTORS

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ABSTRACT:
*R*egional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth. It affects the primary and permanent dentitions in the maxilla and mandible or both jaws. Generally it is localized in only one arch. Clinically, affected teeth have an abnormal morphology, are soft on probing and typically discolored, yellow or yellowish-brown. Radiographically, the affected teeth show a "ghostlike" appearance. This paper reports the case of an 11-year-old girl presenting this rare anomaly on the right side of the maxillary arch. The primary maxillary right central, the lateral and the canine were found missing. The permanent teeth had a "ghostlike" appearance radio graphically. The treatment performed was rehabilitation with temporary partial acrylic denture and periodic controls. The presentation of this case adds valuable information to dentists to review special clinical and radiographic features of RO, which will facilitate the diagnosis and treatment of patients with this condition. Since the etiology of this dental anomaly is uncertain, a review of its probable etiologic factors is summarized to get a better understanding of its cause of occurrence.
KEYWORDS: Regional odontodysplasia; Dental dysplasia; Dentition, permanent.

INTRODUCTION

Regional odontodysplasia (RO) is a rare developmental anomaly involving both mesodermal and ectodermal dental components in a group of contiguous teeth[1]. This condition was probably first described by Hitchin[2] (1934). The first report of this condition was published by McCall and Wald (1947) under the title of Arrested Tooth Development, and reported only radiographic findings[2].

The word "odontodysplasia" was coined by Zegarelli, et al[3]. (1963). Because this abnormality has a tendency to affect only one quadrant, "regional odontodysplasia" became the most accepted term to define it. The same condition has been described under other denominations, such as "odontogenic dysplasia, "localized arrested tooth development" "ghost teeth, "odontogenesis imperfecta", "unilateral dental malformation" and "familial amelodentinal dysplasia".

RO affects both primary and permanent dentitions. Generally, it is limited to only one arch and sometimes crosses the midline. The maxilla is affected twice as often as the mandible[4], the maxillary left quadrant being most commonly involved. Regarding the teeth, the central and lateral incisors are more frequently affected than the posterior teeth. It is also common that in the same quadrant the teeth are affected in the different degrees. It has been suggested that this condition is more common in girls than in boys. There has been no report of tendency towards a particular ethnic group[5].

The etiology of this dental anomaly is uncertain, although several factors, such as local trauma or infection[6], teratogenic drugs, local circulatory disorders, Rh incompatibility, irradiation, neural damage, hyperpyrexia, metabolic and nutritional disorders and vitamin deficiency, have been discussed.

The management of regional odontodysplasia is somewhat controversial and revolves around the question of whether or not to remove the affected teeth. This paper reports a rare case of regional odontodysplasia of the deciduous and permanent teeth associated with eruption disorders.

Case Report

An 11-year old female reported to the Department of Oral Pathology and Microbiology, Subharti Dental College, Meerut, with a complaint of missing upper front teeth since birth. According to her mother, the patient was born deaf and dumb and had undergone treatment for the

same. The patient became normal at 3 years of age. She had also met with an accident and had periods of memory loss which too gradually recovered.

On examination, the patient had a deficient maxilla on the right side leading to facial asymmetry. In the maxillary arch on the right side, the central incisor, lateral incisor and canine were missing or unerupted; the associated alveolar mucosa was enlarged and covered by fibrous tissue. All the teeth showed some degree of hypoplasia. The oral hygiene was poor with an active carious lesion on the mandibular right first molar**.(Fig.1)**

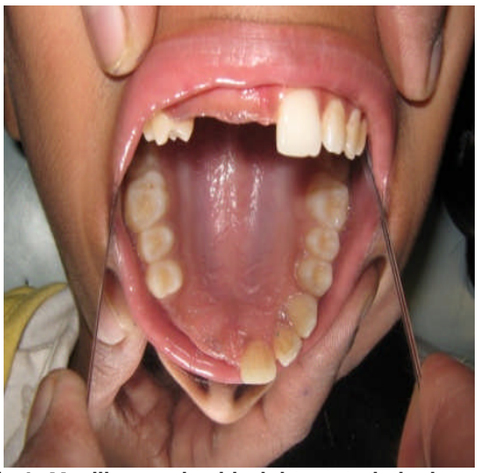

Fig.1: Maxillary arch with right central, the lateral and the canine missing

Panoramic and periapical radiograph revealed that mandibular dentition was normal as well as the left maxillary dentition. In the maxillary anterior region, the right central incisor, lateral incisor, and canine showed malformed and retarded development as compared to the age of the child. The malformed teeth had thin radio-opaque contours with no distinction between the enamel and dentin, and wide pulp chambers, giving a ghost like appearance. The crowns of the affected unerupted teeth were surrounded by radiolucent areas, probably enlarged dental follicles**.(Fig. 2A and 2B)**

Considering the situation of the affected teeth, it was suggested to surgically remove the affected teeth followed by prosthetic replacement**.(Fig.3.)** The patient's family was educated about the situation and a complete consent and approval was obtained. Surgically removed teeth were sent to the Department of Oral Pathology and macroscopic examination showed multiple discoloured teeth with abnormal morphology. Specimen marked as central incisor showed a short conical root and malformed crown. Other two specimens of lateral incisor and canine showed enamel covered malformed crown along with rudimentary roots.

Longitudinal Ground sections of received tooth specimens showed prism structure of enamel resulting in an irregular surface with prominent incremental lines, dentin which was irregular with interglobular areas and areas of haphazard deposition of dentin. **(Fig. 4A and Fig. 4B).** Small, obliterated pulp chambers with presence of pulp stones, also deficient formation of root with absence of cellular cementum was observed. H&E staining of the decalcified sections revealed the presence of teeth like structures with abnormally formed enamel and dentin with wide pulp space. Few areas of enamel spaces were seen. In most of the areas the dentin was atubular and contained clefts scattered through a mixture of globular areas, also pulp tissue containing free or attached stones with numerous areas of basophilic calcifications. **(Fig. 5 and Fig. 6).** Based on the histological features observed, the provisional diagnosis of regional odontodysplasia was confirmed.

The affected edentulous quadrant was also rehabilitated with temporary acrylic maxillary partial denture. Oral hygiene instructions were given. The patient was placed on periodic recall to review the development of maxillary arch.

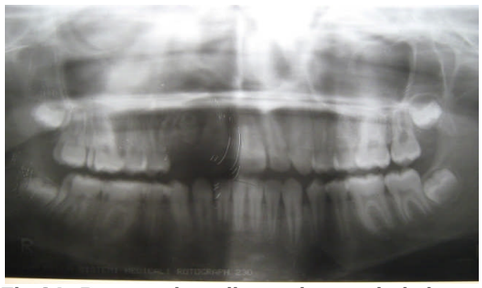

Fig.2A. Panoramic radiograph revealed shows the teeth with 'ghostlike' appearance in maxillary right quadrant. All teeth are normal except in the right maxillary quadrant

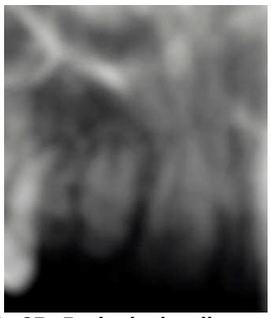

Fig.2B. Periapical radiograph showing the characteristically ghost-like appearance of regional odontodysplasia.

Discussion

RO is a relatively rare, localized developmental anomaly that affects both the dentin and enamel of a group of contiguous teeth. It occurs in both deciduous and permanent dentition, but has a marked preference for the maxilla. Although the incisors and canines are most often involved, any tooth may be affected. When primary teeth are involved, the permanent successors are also usually affected[4].

The condition tends to affect several adjacent teeth within a particular quadrant. Published cases suggest ROD occurs more frequently in females, with no racial predilection. According to a previous literature review, 138 cases of RO had been published from 1934 up to the end of the year 2004 other 15 cases were retrieved from a Pubmed database search until the reporting of the present case[7].

The RO etiology is uncertain; numerous factors have been suggested and considered as local trauma, irradiation, hypophosphatasia, hypocalcemia, and hyperpyrexia[3,8]. RO has been in association with, epidermal nevus syndrome, orbital coloboma, hypoplasia of the affected side of the face, metabolic disturbance, rhesus incompatibility, medications during pregnancy, and ectodermal dysplasia.

The RO has also been related to the activation of latent viruses in the odontogenic epithelium, to the presence of nevus, hemangiomas and hydrocephaly, dolicocephaly and clinodactyly[9], but most of the cases present no relevant information. Walton and his coworkers observed that in three cases of regional odontodysplasia that they reported, all three patients had vascular nevi of the overlying facial skin as infants[10]. Heredity did not appear to be a factor as no family members were identified with dental anomalies. However, it is noteworthy that the patient described here did not present with any of these conditions. No association could be found with the history of cured deaf and dumb condition of the patient.In conclusion, the cause of the present case seems to be unknown.

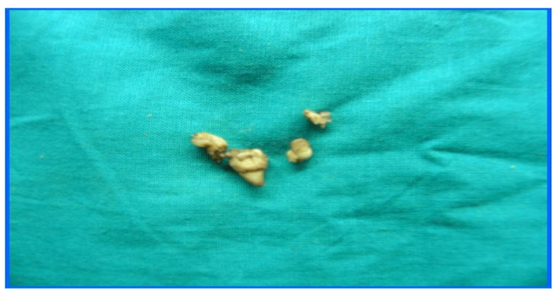

Fig.3.Surgically removed teeth with altered morphology,yellowish in colour, very short & unformed roots

Conditions which show some similarities to RO include amelogenesis imperfecta, dentinogenesis imperfecta, and dentinal dysplasia types I and II, shell teeth, and hypophosphatasia. All these anomalies tend to affect the entire dentition instead of showing segmental distribution[11]. Clinically, affected teeth have an abnormal morphology and an irregular surface contour, with pitting and grooves on the surface. The teeth appear to be discolored, hypoplastic and hypocalcified. The thin enamel is soft on probing and teeth are typically discolored, yellow or yellowish-brown[12]. It is possible to find some teeth without any alterations in the affected quadrant. Affected teeth are more susceptible to caries and are extremely friable.

Radiographically, there is a lack of contrast between the enamel and dentin, both of which are less radiopaque than unaffected counterparts. Additionally, enamel and dentin layers are thin, giving the teeth a 'ghost-like' appearance[6]. The pulp chambers are noticeably enlarged with open apices and enlarged follicles. The unerupted teeth affected by RO may be incorrectly diagnosed as undergoing root resorption or being an odontoma[13].

Histologically, all structures of the tooth germ are affected. Areas of hypocalcified enamel are visible and enamel prisms appear irregular in direction. Coronal dentin is fibrous, consisting of clefts and a reduced number of dentinal tubules; radicular dentin is generally more normal in structure and calcification. Pulpal calcification of various degrees is also commonly seen.

The mineral content of the affected enamel has been found to be higher than that of dentin in microradiographic studies[9,14]. The greater density of the enamel is not evident in conventional radiographs, probably because of the thinness of the enamel layer in affected teeth. It has been proposed that an imbalance of necessary proteins might lead to the structural disorganization seen in this anomaly, such as the metalloproteinases (MMPs) which are enzymes that play a key role in dental development[15].

The patient in this report exhibits several aspects of the common clinical, radiographic and histopathologic features related to regional odontodysplasia. According to the literature, abscess formation is the main reason for extraction of affected teeth, but absence of eruption of permanent teeth was the chief complaint of our patient. There was no sign of gingival swelling or abscess formation.

Several factors must be considered to determine the best treatment option for a child with RO, such as age of the patient, any relevant medical history, previous dental experience, child's and parental attitude regarding dental treatment and number of affected teeth. There has been much debate as to whether affected teeth (with or without abscesses) should be extracted or saved[16].

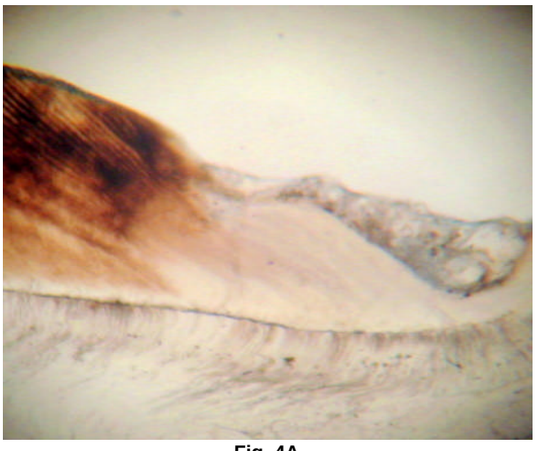

Fig. 4A

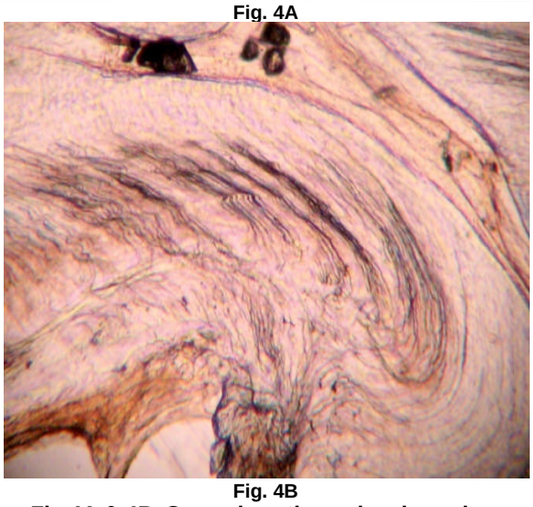

Fig. 4B

Fig.4A & 4B. Ground sections showing prism structure of enamel resulting in an irregular surface with prominent incremental lines, dentin which was irregular with interglobular areas and areas of haphazard deposition of dentin.

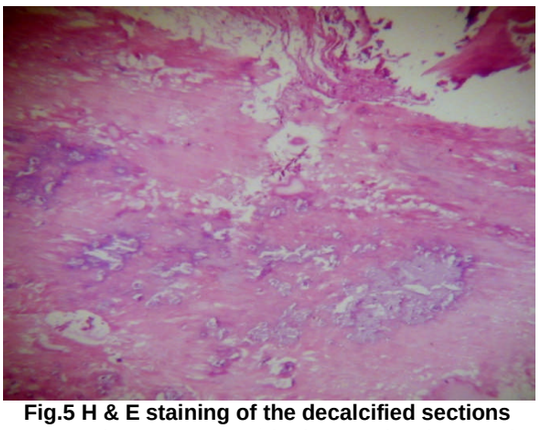

Fig.5 H & E staining of the decalcified sections revealed the presence of teeth like structures with abnormally formed enamel and dentin with wide pulp space.

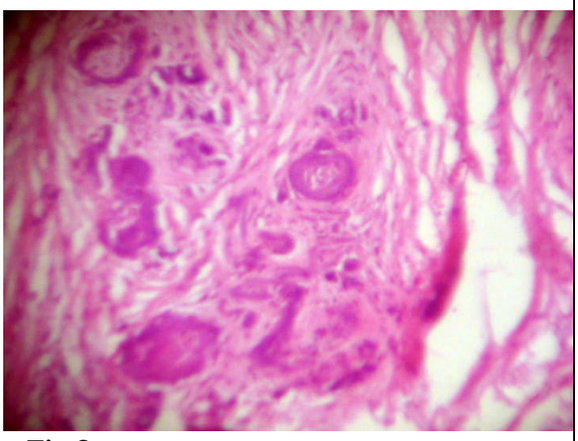

Fig.6. Decalcified sections showing pulp tissue containing free or attached stones with numerous areas of basophilic calcifications. (10 X)

Treatment

The care and treatment of a child with RO requires a multidisciplinary approach. Treatment planning should be designed for each individual case of RO, taking into account factors such as the age of the patient, the medical history, the extent of involvement, the eruption of the teeth, aesthetics, the development of pathology and the wishes of the patient and parents[14].

CONCLUSION

The presentation of this case would help dentists to review characteristic clinical, radiographic and histological features of RO. The therapeutic considerations of RO should be based on the degree of the anomaly, the functional and esthetical needs of each case.

References

1. Gardner DG, Sapp JP. Regional Odontodysplasia. Oral Surg. 1973; 35:351-65.
2. Hitchin AD. Unerupted deciduos teeth in a youth aged 15 1/2. Br Dent J. 1934; 56:631-3.
3. Zegarelli EV, Kutscher AH, Applebaum E, Archard HO. Odontodysplasia. Oral Surg. 1963; 16:187-93. http://dx.doi.org/10.1016/0030-4220(63)90031-8
4. Crawford PJM, Aldred MJ. Regional odontodysplasia: a bibliography. J Oral Pathol Med. 1989; 18:251-63. PMid:2549236 http://dx.doi.org/10.1111/j.1600-0714.1989.tb00394.x
5. Gerlach RF, Jorge J Jr, de Almeida OP, Coletta RD, Zaia A. Regional Odontodysplasia. Report of two cases. Oral Surg Oral Med Oral Pathol. 1998;85:308-13

6. Rushton MA. Odontodysplasia: ghost teeth. Br Dent J1965; 119:109-13. PMid:14332067
7. Tervoten SA, Stratmann U, Mokrys K, Reichart PA. Regional Odontodysplasia: a review of the literature and report of four cases. Clin Oral Investig. 2004; 8:45-51. PMid:14714196
8. Gardner DG, Sapp JP. Ultrastructural, electron-probe, and microhardness studies of the controversial amorphous areas in the dentin of regional odontodysplasia. Oral Surg Oral Med Oral Pathol. 1977 Oct;44(4):549-59. http://dx.doi.org/10.1016/0030-4220(77)90298-5
9. Kahn MA, Hinson RL. Regional odontodysplasia. Case report with etiologic and treatment considerations. Oral Surg Oral Med Oral Pathol. 1991 Oct; 72(4):462-7. http://dx.doi.org/10.1016/0030-4220(91)90561-P
10. Walton JL,Witkop CJ, Jr Walker PO. Odontodysplasia. Oral Surg, 1978; 46:676. http://dx.doi.org/10.1016/0030-4220(78)90464-4
11. SÃ¡nchez-Quevedo C, Ceballos G, RodrÃguez IA, GarcÃa JM, Alaminos M. Acid-etching effects in hypomineralized amelogenesis imperfecta. A microscopic and microanalytical study. Med Oral Patol Oral Cir Bucal. 2006 Jan 1;11(1):E40-3. PMid:16388292
12. Fearne J, Williams DM, Brook AH. Regional odontodysplasia: a clinical and histological evaluation. J Int Assoc Dent Child. 1986; 17:21-5. PMid:2822816
13. Amado Cuesta S, Gargallo Albiol J, Berini AytÃ©s L, Gay Escoda C. Review of 61 cases of odontoma. Presentation of an erupted complex odontoma. Med Oral. 2003 Nov-Dec; 8(5):366-73. PMid:14595262
14. Hamdan MA,Sawair FA,Rajab LD, Hamdan AM. Regional Odontodysplasia: A review of the literature and report of a case.Int J Paed Dent2004; 14:363-70. PMid:15331002 http://dx.doi.org/10.1111/j.1365-263X.2004.00548.x
15. Spini TH, Sargenti-Neto S, Cardoso SV, Souza KC, de Souza SO, de Faria PR et al. Progressive dental development in regional odontodysplasia. Oral Surg Oral Med Oral Pathol Oral Rad Endo. 2007; 104:40-5. PMid:17613259 http://dx.doi.org/10.1016/j.tripleo.2007.02.027
16. Cahuana A, Gonzalez Y, Palma C. Clinical management of regional odontodysplasia. Pediatr Dent. 2005; 27:34-9.

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