Regional odontodysplasia: An unusual case report

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ABSTRACT
Regional odontodysplasia is an uncommon developmental anomaly affecting a localized area of dentition, with distinctive clinical, radiographic, and histological findings. This article reviews a case of a 14-year-old female who reported with unerupted maxillary anterior teeth. This case was rare in that it involved maxillary dentition with unerupted maxillary anterior teeth on the right side, but the left central incisor was missing. The molars on the right side were showing some amount of abnormality. Radiographically, the affected teeth had a 'ghostly' appearance, showing a marked reduction in radiodensity. Both enamel and dentin appeared to be very thin, the pulp chamber was exceedingly large, and the roots were short with wide open apices. All the characteristics were consistent with the diagnosis of regional odontodysplasia. The care and treatment of this patient required a multidisciplinary approach. The unerupted maxillary anterior teeth were surgically removed, following which temporary prosthetic restoration was provided to improve esthetics and to restore the function.

Key words: Regional odontodysplasia, odontodysplasia, multidisciplinary approach

INTRODUCTION
Regional odontodysplasia is an uncommon, nonhereditary developmental anomaly affecting dental tissues, derived from both the mesoderm and ectoderm.[1] The condition can be differentiated from other odontogenic disturbances, as all the histological elements of the dental organ are abnormal in the affected teeth, while other teeth in the same individual are normal.[2]

Crawford[3] ascribed the first report to Hitchin in 1934. McCall and Wald[3] were credited for publishing the first report of odontodysplasia in 1947, in which they termed the condition ‘arrested tooth development’. In 1954, the term ‘shell teeth’ was introduced by Rushton, which the author used to describe the radiographic findings. In 1963, Zegarelli et al. were the first to suggest the term ‘odontodysplasia’. As the condition affects a group of several adjacent teeth in a particular segment of the jaw, Pindborg added the prefix “regional” in 1970.[3]

Regional odontodysplasia exhibits no specific racial predilection. Females are more often affected than males (1.4:1).[3,4] The condition can affect both primary and permanent dentition and can occur in the maxilla, the mandible, or both. The maxilla is involved twice as much as the mandible. It is usually unilateral and rarely crosses the midline.[1]

The etiology of odontodysplasia is uncertain, though many causative factors have been suggested, for example, vascular defects involving ischemia, local infections, 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.[5-7]

The management of regional odontodysplasia is somewhat controversial and revolves around the question of whether or not to remove the affected teeth. Although many clinicians prefer to extract the anomalous teeth as soon the diagnosis is made, some would prefer to retain them as long as they are free of infection until the skeletal growth is complete.[1] This case is interesting and rare because it presents with the condition affecting the right side and the left central incisor is also missing. We present a case report of this rare anomaly with certain unique findings and a multidisciplinary treatment protocol.

CASE REPORT
A 14-year-old girl came to the Department of Pedodontics and Preventive Dentistry, Bapuji Dental College and Hospital, Davangere, Karnataka, India, with a chief complaint of missing or unerupted maxillary anterior teeth. A previous history of trauma to her maxillary anterior region about eight years back was recorded, due to which there was avulsion of few deciduous teeth and few fractured teeth, which were
subsequently removed. The patient’s medical history was noncontributory. Both parents reported no previous history of tooth or genetic anomalies on either side of the family.

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 [Figure 1]. Also there was a rotated first premolar, erupting second premolar, and unerupted first and second molar on the right side. However, on the left side of the maxillary arch only a central incisor was missing and the rest of the teeth were present. In the mandibular arch, on both sides, all the teeth were present except the third molars. All the teeth showed some degree of hypoplasia. The oral hygiene was poor with an active carious lesion on the mandibular left first molar. There was deficient maxillary growth on the right side, showing signs of arch collapse [Figure 2].

Panoramic, occlusal, and periapical radiographs were taken. The mandibular dentition was normal, as was the left maxillary dentition, except the central incisor. 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. There was complete absence of the left central incisor and the right first molar. The second molar was also malformed, but it was not as severe as the other malformed teeth. 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 representing enlarged dental follicles. Only an insignificant amount of root formation, was visible radiographically [Figures 3 and 4]. As the upper right second molar did not look as affected when compared to the other affected teeth, we could wait for its eruption. On the basis of the clinical and the radiographic findings, a provisional diagnosis of regional odontodysplasia was made. Considering the situation, the eruption of the affected teeth was not favorable except for the second molar. Therefore, it was suggested to surgically remove the affected teeth and go in for a prosthetic replacement to improve the function and for an esthetical purpose. The patient’s family was properly educated about the situation and the need for surgical removal and prosthetic rehabilitation and a complete consent and approval was obtained.

Under local anesthesia, the affected teeth were surgically removed [Figure 5]. The teeth were of altered morphology, yellowish in color, soft or rubbery in consistency, and had very short or unformed roots, and wide open apices [Figure 6]. The treatment plan included dietary counseling, instructions on oral hygiene, and prescription of mouth washes. Indirect pulp capping and subsequent restoration of the carious mandibular left first permanent molar was also performed. Surgically removed teeth were histologically examined under an optical microscope in the ground and decalcified section. Histologically, all structures of the tooth germ were affected. In the ground section, the enamel was of variable thickness, producing an irregular surface. Longitudinal ground sections revealed the presence of enamel tissue over dentin and a central enlarged pulp cavity. The dentinoenamel junction was flat and in some areas irregular. The dentin layer was abnormally thin with the mantle dentin showing a normal dentinal tubular pattern, while the deeper dentin contained a mixture of predominant interglobular dentin and poorly organized tubular dentin [Figure 7].

Hematoxylin and eosin staining of the decalcified sections revealed the presence of teeth-like structures, with abnormally formed slight enamel and dentin with wide pulp space [Figure 8]. 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, with numerous areas of amorphous (basophilic) material [Figure 9]. Based on the histological features, the provisional diagnosis of regional odontodysplasia was confirmed.

Two months postoperatively, a temporary acrylic maxillary partial denture, with a bite plane, was made, to preserve the alveolar ridge during the period of skeletal growth [Figure 10]. Oral hygiene instructions, and dietary analysis and advice were given. The patient was placed on periodic recall to review and monitor the development of the maxillary arch.

The patient will be kept under review until she reaches adulthood, when a final rehabilitation for the loss of teeth might be accomplished after the facial bones had attained complete growth.

**DISCUSSION**

Regional odontodysplasia is a relatively rare, nonhereditary, localized developmental anomaly of the dental hard tissues of a group of contiguous teeth. Many cases are probably misdiagnosed as malformed teeth or odontomas. It occurs in both deciduous and permanent dentitions, and it has a marked preference for the maxilla. Other conditions, such as, dentinal dysplasia, shell teeth, hypophosphatasia, dentinogenesis imperfecta, or amelogenesis imperfecta can mimic some features of regional odontodysplasia. However, these disorders tend to affect the entire dentition.

Although many theories have been proposed, in this particular case it has not been possible to determine a precise etiological factor. The relationship of trauma to the upper front teeth region about eight years back could not be directly co-related to odontodysplasia. It was clear that the undeveloped...
maxilla on the right side was due to the teeth being affected by odontodysplasia. In the present case, along with the affected right teeth, the left central incisor and the right first permanent molar were missing. Also the second molar was affected to a lesser extent, although one could wait for its eruption. On account of these factors, the present report was considered interesting and rare. [8]

The case showed features that were in accordance with the literature, in that the condition seemed to affect females more...
than males, and most patients came to the dentist complaining of delayed eruption or gingival swelling. The clinical presentation of in this case was failure in eruption of teeth, which could be related to odontodysplasia. In this case, radiographs showed that the unerupted teeth had not achieved complete rhizogenesis. The affected teeth had an abnormal morphology with an irregular surface contour, with pitting and grooves, and a rough surface with defective mineralization. The teeth appear to be hypoplastic, hypocalcified, and show yellowish or brownish discoloration.

Radiographically, the affected teeth have been described as having a “ghostly” appearance, showing a marked reduction in radiodensity. Both the enamel and dentin appear very thin, and the pulp chamber is exceedingly large; the roots are short with open apices, and pulp stones or denticles are present in the affected as well as the normal adjacent teeth.

Histologically, all structures of the tooth germ are affected. In the ground section, the enamel is of variable thickness, producing an irregular surface. The enamel prisms are irregular and show hypoplastic and hypocalcified areas, whereas, the enamel closer to the dentinoenamel junction appears more normal. Invagination extending from the enamel surface into the dentin have been reported. The dentinoenamel junction is irregular and scalloped. The dentin is thin, and the tubules are reduced in number and tortuous in shape. The most significant feature is the presence of large areas of amorphous dentin containing irregularly shaped foci of a poorly formed dentinal matrix, within which, capillaries are sometimes present. Interglobular dentin and globular masses interrupting the dentinal tubules are frequently seen. Cellular dentin and amorphous areas within the coronal dentin are usually evident. Prominent interglobular dentin is also seen in radicular dentin.

Since the clinical, radiographic, and histological features of regional odontodysplasia are so characteristic, dentists should face no difficulty in the diagnosis of this abnormality. The greater difficulty arises in treatment planning, as no consensus as to the best option has yet been reached. The main question is whether to remove the affected teeth...
or not, but we believe that this decision should be taken after the assessment of each individual case of regional odontodysplasia. Some authors prefer extraction and replacement with removable, and later, fixed prostheses, while others have suggested keeping the unerupted teeth, and waiting for their calcification and thus hope for its eruption. From the evaluation it was decided that the eruption of the affected teeth was questionable, so it would be better to surgically remove the affected teeth and provide prosthetic rehabilitation for the patient.

With the consent of the patient and parents, the unerupted teeth were removed and temporary rehabilitation was made with a temporary acrylic prosthesis. The patient will be kept under review until she reaches adulthood, when a final rehabilitation for the loss of teeth may be accomplished, after the facial bones had attained complete growth.

The care and treatment of a child with regional odontodysplasia requires a multidisciplinary approach. Consultations between pediatric, prosthodontic, and orthodontic specialties are necessary in each case of odontodysplasia. Treatment planning should be designed for each individual case, taking into account factors such as the age of the patient, the medical history, the extent of involvement, the eruption of the teeth, esthetics, the development of pathology, and the wishes of the patient and parents.

CONCLUSION

The treatment of regional odontodysplasia requires a multidisciplinary approach and an organized team effort. In the present case an intermediary prosthesis was delivered. However, further treatment in the form of orthognathic surgery, bone grafting, and implant therapy needs to be anticipated in the future.

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