SHORT COMMUNICATION POJ 2015:7(1) 41-44

Regional odontodysplasia: report of an unusual case

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Abstract

Regional odontodysplasia (ROD) is a rare, non-hereditary disorder affecting both primary and permanent dentitions.It is more common in maxilla and usually involves one quadrant, rarely crossing the midline. Clinically, the affected teeth have an abnormal morphology and appear discolored. Radiographically, the demarcation between enamel and dentin is not evident and the pulp chamber appears wide. Enamel and dentin are thin and hypoplastic, therefore giving the impression of "ghost teeth".

The report describes a case of regional odontodysplasia in a 6 year old boy who presented with the complaint of delayed eruption. This case study aims to report the clinical and radiological findings of the current case.

Keywords: Odontodysplasia, Ghost teeth, Developmental defects

Introduction

Regional odontodysplasia (ROD) is a rare dental anomaly affecting both primary and adult dentitions in the maxilla, mandible or both jaws, though the involvement of the maxilla is more common.1 The condition was first described by Hitchin in 1934,2 although McCall et al. [3] were the first to report this condition in 1947; in which it was named as "arrested tooth development". Its prevalence is reported to be less than 1/1000000 and only about 140 cases have been reported in the literature.[4] The etiology of regional odontodysplasia is uncertain, but several associated factors such as local trauma, infection, local circulatory disorders, neural damage, hyperpyrexia, nutritional deficiencies, teratogenic drugs, Rh incompatibility, activation of latent viruses residing inodontogenic epithelium, somatic mutations and disorders of neural crest cell migration have been advocated.[5]

One of the distinguishing characteristics of regional odontodysplasia is that the condition affects a single quadrant and rarely crosses the midline [5,6] although cases crossing the midline have been reported[7]. The involved teeth usually appear hypoplastic, with a yellowish/brownish discoloration and aberrant morphology.[8] The involved enamel is soft on probing making the teeth susceptible to caries.[3] Regional dontodysplasia has no racial predilection and affects female more than male individuals (ratio F/M = 1.4/1).[5] Radiographically, the affected teeth show abnormal morphology and hypoplastic crown and the delineation between enamel and dentin is usually not apparent. Enamel and the dentin are very thin and the reduced radiodensity imparts a "ghost-like appearance".[3] Enlarged pulp chambers, short roots and shell-like crowns are the other pathognomic radiographic characteristics[3]. The teeth may show signs of arrested development.[9] Histologically, all developing structures of the tooth are affected. A varying thickness of enamel with an irregular prism pattern is frequently seen.[10] The dentin layer is reduced with irregular dentinal tubules and presence of amorphous dentin areas and interglobular dentin and the pulp usually shows varying degrees of calcification.[11]

The treatment depends on the severity of the condition ranging from placement of protective restorations on the affected teeth to tooth extraction and prosthetic rehabilitation.

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Most authors are in favor of removing the severely affected teeth [1,12] due to structural defects and the fact that these teeth often fail to erupt.[8,9] Some prefer to retain them until skeletal growth is complete to preserve the bone, as long as they are free of infection.[1]

Case Report

A 6-year old boy was referred to Orthodontic Department, Islamic International Dental Hospital, Islamabad with the complaint of delayed eruption. The patient was a healthy six-year old boy with no relevant medical history at the time of presentation.

The prenatal, natal history was insignificant. The parents did not report any tooth abnormalities or genetic anomalies in either maternal or paternal family. The patient, however, had a chronic dental history described by the father as 'bleeding from teeth'. There was no history of any extraction and the teeth were 'lost as soon as they erupted'. He was treated by a local dentist in a rural area, of which no record was present.

Extraoral examination (Fig. 1) revealed nothing of significance. He had a normal symmetrical face and normal skin, hair and nails. The temporomandibular joints appeared normal and there was no lymphadenopathy. The boy was at the early mixed dentition stage. Maxillary arch (Fig. 3 & 4) was intact with no missing tooth, however, right primary second molar showed aberrant morphology with signs of hypoplasia. The mandibular arch (Fig. 4 & 5) was edentulous except for the permanent first molar, primary second molar and a remnant of primary first molar root on the left side. No dental abscess was seen clinically. Oral hygiene was fair.

The panoramic radiograph (Fig. 6) was taken and showed that the maxillary teeth were normal

except the right primary second molar. The tooth was hypoplastic with significant root resorption and the succedaneous second premolar was missing. All the permanent teeth in the mandible were present. The affected teeth (right permanent first molar to contralateral first premolar) displayed the typical "ghost-like" appearance; a mild radiopaque outline with no proper crown

shape or structure. The demarcation between the enamel and the dentin of the affected teeth was not clear and the enamel was hypoplastic. The left mandibular second molar was carious and periapical radiolucency was present. Ridge resorption was evident in association with the premature loss of the primary teeth.

The patient was referred to Paediatric Medicine department to rule out any systemic cause. The laboratory examinations revealed that serum calcium, phosphorus and blood CP were normal. Patient's serum alkaline phosphatase was raised and Vitamin-D deficiency was found. On the basis of our clinical and radiological findings, a diagnosis of regional odontodysplasia was proposed. Treatment plan was discussed with the parents. Conservative management was planned involving the restoration of the maxillary right second molar with stainless steel (SS) crown. The mandibular left second molar was to be retained to prevent further bone loss so pulpectomy with SS crown was advised. Removable partial denture was to be given to restore function and programmed follow-up visits were planned.

Figure 1 Extra-oral picture of the patient

Figure 2 & 3 Intra-oral pictures showing intact maxillary arch with a malformed primary second molar on right side.

Figure 4 & 5 Intra-oral pictures showing premature loss of the involved teeth in mandibular arch and carious primary left second molar.

Figure 6 OPG showing 'ghost-like' teeth in the mandible

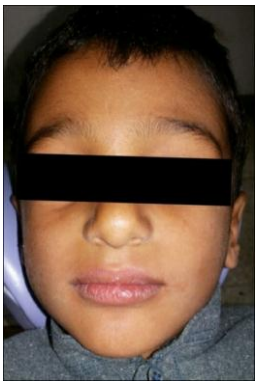

Figure 1: Extra-oral picture of the patient

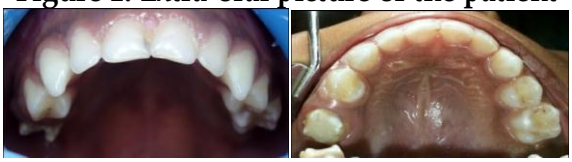

Figure 2 & 3 Intra-oral pictures showing intact maxillary arch with a malformed primary second molar on right side.

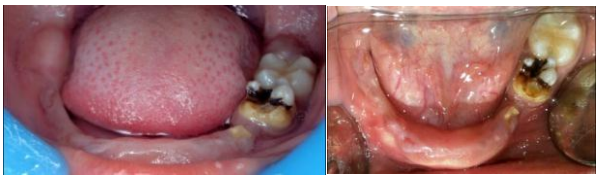

Figure 4 & 5 Intra-oral pictures showing premature loss of the involved teeth in mandibular arch and carious primary left second molar.

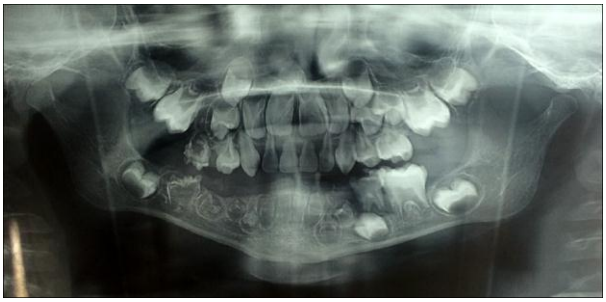

Figure 6 OPG showing 'ghost-like' teeth in the mandible

Discussion

Regional odontodysplasia is a rare developmental anomaly1 therefore the report of this case seems to be imperative.

The patient in this report had the classic radiographic presentation of "ghost teeth" in the mandible extending from right permanent first molar crossing the midline to opposite first premolar, a total of ten teeth being involved. According to the Lustmann et al.13 the condition is more prevalent in maxilla and this anomaly rarely crosses the midline. In this case, mandible was involved and crossing of midline was observed. Based on the literature, regional odontodysplasia is more prevalent in females, and our case was a boy.

The affected teeth are prone to caries which may be a possible explanation for the premature loss of the involved deciduous teeth. Vitamin D affects bone and mineral metabolism in the body,[14] and its deficiency adversely affects bone health. Low levels of Vitamin D could possibly account for the bone loss associated with early exfoliation of the involved primary teeth in the patient.

Vitamin deficiency has been advocated to be a possible etiological factor for regional odontodysplasia [2,5,15,16] although the exact etiology is still uncertain. Vitamin D deficiency could be a potential etiological factor for ROD in the reported case.

A differential diagnosis of ROD includes dentine dysplasia, amelogenesis imperfecta, dentinogenesis imperfecta and hypophosphatasia. The developmental anomalies, however, usually affect the entire dentition rather than a localized segmental involvement.[17]

Treatment of ROD is challenging and requires a multidisciplinary approach. Consultations between pediatric, prosthodontic, orthodontic, and surgical specialties are often necessary. Factors such as the patient's age, medical history, extent of the lesion, eruption of the teeth and esthetics need to be carefully considered.[8,9]

In the reported case, the long-term prognosis is poor because of the amorphous morphology and meager development of the associated teeth. Extraction of these teeth will be necessary once the patient's skeletal growth is complete. Dental implants may be a feasible long-term treatment option for a stable fixed or removable prosthesis in future.

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