

Cervicofacial infection in a 3-year-old child with regional odontodysplasia: A case report and literature review

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Abstract

Regional odontodysplasia (RO) also known as ghost teeth is a rare developmental anomaly affecting the mesodermal and ectodermal components of teeth with characteristic clinical and radiographic findings. The enamel and dentin are hypomineralized and hypocalcified with short roots and open apices. The affected teeth have an abnormal morphology, meaning they are fragile and thin and liable to fracture and decay. Radiographically, there is a marked reduction in radiodensity with little distinction between the enamel and dentin, hence the term "ghost teeth." RO generally affects one particular segment in one or both arches of the maxilla or mandible and can affect both the primary and permanent dentition. We report a unique case of a 3-year-old female who presented to Alder Hey Children's Hospital acutely unwell with a left-sided cervicofacial swelling from RO affecting the entire portion of the left maxilla.

Keywords: Cervicofacial infection, ghost teeth, regional odontodysplasia

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Received: 28.11.2017, **Accepted:** 29.11.2017

INTRODUCTION

Regional odontodysplasia (RO) was first published in the literature in 1947 by McCall and Wald.^[1] The word "odontodysplasia" was first used by Zegarelli *et al.* in 1963.^[2] Several other terms have been used to describe the condition including: "odontogenesis imperfecta," "localized arrested tooth development," "unilateral dental malformation" and "familial amelodentinal dysplasia." As odontodysplasia has a tendency to affect only one quadrant, "RO" is the most widely accepted term in the literature.^[3]

RO can affect both primary and permanent teeth and tends to involve several teeth within a quadrant in the jaw.^[4] It involves the maxilla twice as often as the mandible and is more likely to involve the anterior teeth.^[5] Females are

more commonly affected by the anomaly,^[5] but there is no predilection for a particular race or ethnicity.^[6]

The etiology is uncertain although a number of factors have been implicated such as local trauma, rhesus incompatibility, nutritional and metabolic deficiencies and the use of teratogenic medications.^[7-9] Currently, there have been no familial traits identified or recorded in the literature which suggest an hereditary link.

Clinically, teeth affected by RO have a distinctive appearance. The enamel and dentin are discolored with a rough and mottled surface and can be associated with pits and grooves. Affected teeth are susceptible to caries and are prone to fracture. Frequently, tooth eruption is

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10.4103/jomfp.JOMFP_146_17

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How to cite this article: Bowden E, Carroll C, Gill M, Llewelyn R. Cervicofacial infection in a 3-year-old child with regional odontodysplasia: A case report and literature review. *J Oral Maxillofac Pathol* 2018;22:S117-20.

delayed, or there is failure of eruption due to arrested root development. Teeth that do erupt are often smaller and spaced and can have gingival enlargement.^[9,10]

Radiographically, teeth have absence of roots or are short with open apices. The pulp chambers are wide in diameter often with diffuse calcification. There is marked reduction in radiodensity of enamel and dentin, and there is a little demarcation between the two which is attributable to the term “ghost teeth.”^[11]

Histologically, the tooth germ as a whole is affected. The enamel is hypocalcified and hypoplastic with an aprismatic enamel matrix. This varies in thickness which produces an irregular surface. The amelodentinal junction tends to have a scalloped appearance.^[12] The dentin is thin, and interglobular dentin has been observed interrupting dentinal tubules which are of irregular diameter. The pulp chamber is large with long pulp horns, which can contain large irregular calcifications or stones.^[13]

CASE REPORT

A 3-year-old female presented to the Accident and Emergency Department at Alder Hey Children’s Hospital in Liverpool with a 2-day history of spontaneous pain, reduced oral intake and a left-sided cervicofacial swelling. On observation, she was pyrexial at 38.4°C. All other observations were within normal limits. The patient was otherwise fit and well with no relevant medical history and no known allergies. She was born at full term with a normal delivery. Both parents reported no previous history of tooth or genetic anomalies on either side of the family.

Clinical examination revealed a diffuse swelling extending from the left buccal space to the infraorbital margin. The left eye was open and visual acuity was 6/6 on the Snellen chart. There was no evidence of cervical lymphadenopathy.

Intraoral examination demonstrated a marked fluctuant buccal swelling adjacent to the upper left deciduous canine and upper left deciduous first molar and a discharging sinus with pus exudate above the upper left deciduous central incisor [Figure 1]. The left maxillary dentition was partially erupted with hypoplastic and hypocalcified teeth and associated gingival enlargement. The primary teeth had abnormal morphology and were yellowish with a mottled, irregular surface. The upper left deciduous canine had a vertical crown fracture down to the amelodentinal junction [Figure 2]. The pulp horns were exposed on the upper left deciduous first molar [Figure 3], and the upper



Figure 1: Intraoral view demonstrating a buccal sinus above upper left deciduous central incisor and characteristic hypoplastic and hypocalcified appearance of teeth

left deciduous second molar was unerupted. The remaining dentition was caries free and had developed normally.

An orthopantomogram [Figure 4] showed an abnormal morphological dentition to the upper left quadrant affecting the primary teeth from the upper left central incisor to the upper left deciduous second molar which was unerupted. It also showed that the upper left first permanent molar dental germ was affected. These teeth were radiolucent with a characteristic ghost appearance and appeared less developed in comparison to the primary teeth on the contralateral side. Normal development of the permanent successors was visible radiographically in the unaffected quadrants.

Based on the clinical and radiographic findings, a diagnosis of RO was suggested with associated cervicofacial infection. The patient was admitted and immediately commenced on empirical intravenous co-amoxiclav. She was taken to emergency theatre for incision and drainage of the left buccal swelling and extraction of the upper left deciduous central incisor, upper left deciduous lateral incisor, upper left deciduous canine and upper left deciduous first molar. The upper left deciduous second molar was left *in situ* as it was unerupted. Postprocedure, the cervicofacial swelling significantly resolved, and the patient remained in hospital for a total of 3 days.

DISCUSSION

A literature search was undertaken of PubMed, Embase and CINAHL. The terms included in the search were (Regional)*, (Odontodysplasia)* and (REGIONAL ODONTODYSPLASIA)*. Our results produced 206 papers, from which duplicates were removed, leaving 113 papers in total to review. Of the 113 papers, Embase (7),

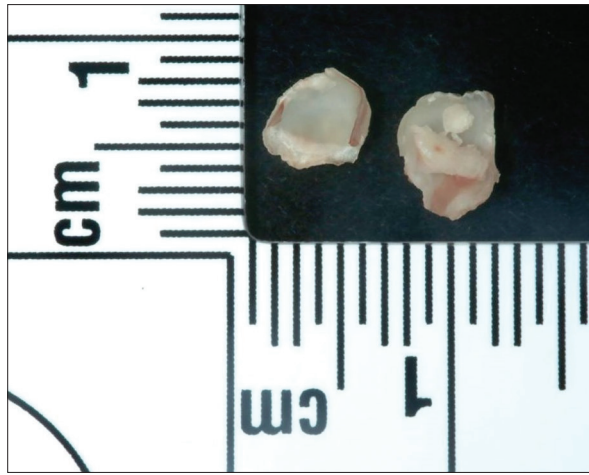


Figure 2: Extracted Upper left deciduous canine (ULC) showing vertical crown fracture

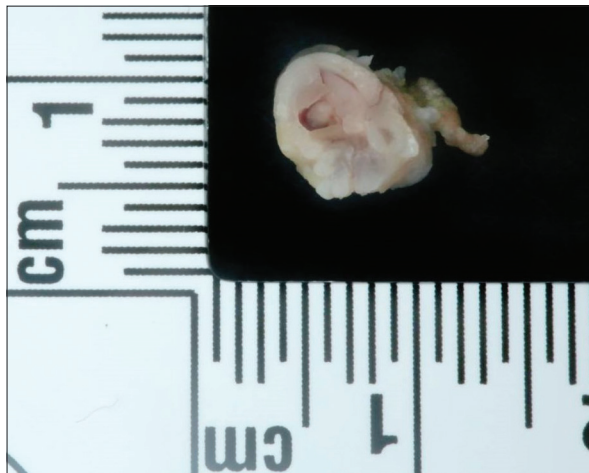


Figure 3: Hypoplastic dental tissue leading to pulp exposure and ultimately necrosis and abscess formation



Figure 4: Panoramic view showing the affected teeth in the left maxilla with a “ghost” appearance; marked reduction in radiodensity between enamel and dentin; incomplete root formation. (Note also that the upper left first permanent molar is affected)

CINAHL (4) and PubMed (102), the highest level of evidence of the papers searched were case reports and case studies. The descriptive nature of the articles examined exhibits the limited information on the topic.

RO may be misdiagnosed as another dental anomaly such as amelogenesis imperfecta, dentinogenesis imperfecta, hypophosphatasia and odontomas as they may display similar features; however, RO tends to be segmental in nature. In this particular case, as the upper left quadrant teeth were affected, this supports the diagnosis and subsequent management plan. Unilateral involvement of the maxilla is more likely over involvement of the mandible.^[5]

The enamel defects seen were severe, and the quality of the tooth structure was suboptimal. This leads to the longitudinal fracture of the upper left deciduous canine with ingress of microorganisms into the pulpal and periradicular tissues. This resulted in a periapical abscess and subsequent fascial space infection. In the adjacent teeth, the pulp horns were visible through the thin dentinal walls and this could have led to pulpal irritation followed by pulpal necrosis and abscess formation.

According to the literature, the most common reason to extract the teeth is due to abscess formation.^[3] Treatment of RO, however, is a controversial subject, and in our opinion, it needs to follow a “case-specific” approach. This is dependent upon a number of factors including patient age and cooperation, severity of odontodysplasia, associated signs and symptoms and medical comorbidities. In the pediatric patient, options for treatment should be explored with the parent or guardian. A number of papers suggest early extraction with prosthetic rehabilitation to prevent the development of dental pathology in the absence of caries.^[12] Other authors have adopted a preventive approach with restoration of affected teeth with composite resin material.^[14] Preservation of these teeth can provide space maintenance and thus allow for later prosthetic rehabilitation if required. Retention of noninfected teeth would be advantageous as it would preserve the alveolar bone, promote normal facial growth and remove the psychological impact of early tooth loss.^[7,15,16]

The management plan of this case involved dental extractions of affected teeth as the patient was systemically unwell with a spreading cervicofacial infection. There was a periapical abscess on the upper left central incisor with a discharging sinus. The teeth were also not amenable to restorative treatment because of hypocalcified tooth tissue, wide pulp chambers and lack of micromechanical retention.

We appreciate that this case will ultimately involve a multidisciplinary team approach, and the treatment plan will be a prolonged and complex process. Management will depend on the presence and severity of odontodysplasia in the permanent dentition.

Short-term planning may include preservation of the upper left first permanent molar with a stainless steel crown to act as a distal abutment for a temporary prosthesis. This would aim to restore esthetics, optimize masticatory function, prevent over-eruption of the opposing dentition and achieve space maintenance. We recognize that the patient who is currently 3 years of age is too young to tolerate fabrication of a partial denture. Therefore, she is on a regular long-term follow-up plan to monitor the developing permanent tooth germs and assess the severity of odontodysplasia in the permanent dentition and dentoalveolar growth in the maxilla.

Long-term options may comprise elective extraction of affected teeth and placement of implant-retained restorations which may require an alveolar bone graft. Implant placement would not be considered a viable option until craniofacial growth is complete.

A further treatment option to explore is autotransplantation of lower premolars to the affected quadrant. Autotransplantation, however, is technique sensitive and dependent on the surgical experience of the clinician.^[17] Previously reported complications of this technique include pulpal necrosis, ankylosis and transplant failure.^[7]

CONCLUSION

This case illustrates the unique clinical and radiographic features of RO. The patient could initially present to their general dental practitioner for assessment, and therefore, dental professionals must be familiar with dental anomalies to allow for prompt diagnosis, monitoring and appropriate intervention to minimize the clinical implications for the patient's dentition and quality of life.

Although numerous cases have been reported, the literature is limited to that of individual case reports. As a result, there is no accepted consensus on the best management strategy. Further research including epidemiological and experimental studies is required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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