Bilateral mandibular regional odontodysplasia with vascular nevus

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Abstract

Odontodysplasia is a rare clinicopathologic condition that has a variety of expressions, including a range of tooth coloration, formation of hypoplastic enamel affecting one or multiple teeth, bizarre radiographic appearance, and delayed eruption of teeth. An unusual case is presented of bilateral regional odontodysplasia associated with a vascular nevus of the face and neck. A review of the etiology with emphasis on the "local circulatory disease" theory is presented. (Pediatr Dent 13:303–6, 1991)

Introduction

Odontodysplasia is a clinicopathologic disorder that affects developing dental follicles (Lian et al. 1988). The reader is directed to a review by Crawford and Aldred (1989). The condition was first described as a case of unerupted primary teeth and later referred to as arrested tooth development, odontogenesis imperfecta, ghost teeth, and Rushton's shell teeth. The term odontodysplasia has been used since 1963, with regional or local as a further descriptive since 1970 (Zegarelli et al. 1963; Pindborg 1970). There are a variety of expressions. Local or regional distribution in more than one quadrant has been noted, with most affected teeth found in the maxillary anterior region (Crawford and Aldred 1989). Both primary and permanent dentitions may be involved (Crawford and Aldred 1989). The affected teeth appear malformed with hypoplastic areas, discolored from yellow to a brown opalescence. Arrested root formation is common, and many teeth fail to erupt (Gibbard et al. 1973). These underdeveloped teeth appear indistinct or ghost-like on radiographs because of the hypoplastic enamel and dentin (Crawford and Aldred 1989).

The presenting symptoms include failure or delay of eruption of teeth, premature exfoliation or extraction of teeth, abscess formation due to pulp exposures in hypoplastic surfaces, malformed teeth, and noninflammatory jaw swelling, which presents as a firm, painless, soft-tissue enlargement of the gingiva around the affected teeth (Dayal and Mani 1981; Neupert and Wright 1989).

The etiology of regional odontodysplasia (ROD) is unknown. However, numerous theories have been suggested, including local circulatory disorders, viral infections, prenatal drug exposure, neural crest cell defects, trauma, and infection (Crawford and Aldred 1989). Currently, the most commonly accepted theory is vascular etiology, since many cases have shown that a disturbance in the local circulation overlying the affected area occurred during the time of tooth development (Gibbard et al. 1973; Fearne et al. 1986). This disturbance took the form of a vascular nevus or "nevus flammeus," which is a congenital vascular lesion that disappears gradually (Walton et al. 1978). Clinically, it presents as pale pink in color and is also known as a "salmon patch" (Walton et al. 1978). The vascular theory is debatable since proof is not evident in every patient (Lustmann et al. 1975).

Odontodysplasia is considered an uncommon dental finding (Gibbard et al. 1973). Approximately 108 cases have been reported in the world literature (Crawford and Aldred 1989). This report presents an unusual case of ROD in the mandible, which crossed the midline and was accompanied by a congenital vascular nevus.

Case Report

An 11-year-old Caucasian boy was seen by an endodontist to evaluate swelling in the labial fold adjacent to the mandibular left permanent central incisor. The tooth was diagnosed as necrotic, and endodontic treatment was initiated. The patient was referred to the University of Detroit School of Dentistry with a further complaint of missing and malformed teeth.

The patient was a product of a normal pregnancy and delivery. At birth, he displayed two large, reddish, flat congenital vascular anomalies located under his chin and on the left side of his face below the ear. These lesions faded with age; however, flat, reticulated, reddish nevi were present in the aforementioned areas on examination (Fig 1, see next page). The medical history revealed he had been hospitalized at one year of age with a fever and swelling of the left side of his face; this resolved following antibiotic therapy. The etiology was never determined. At the time of examination, he was in good health. No other member of his family had any

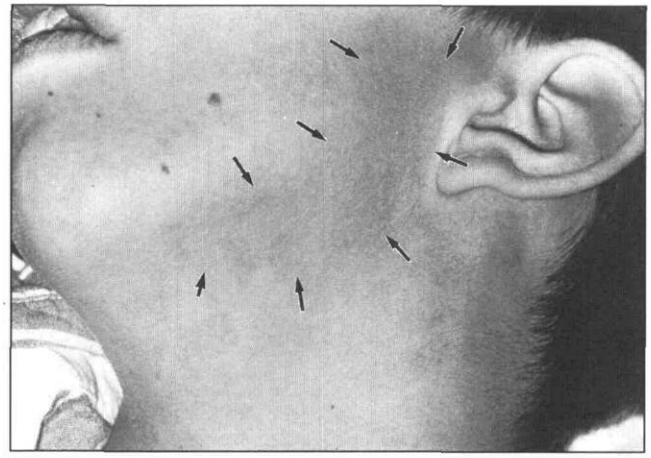

Fig 1: Vascular nevi presented in soft tissue overlying the affected area. Arrows indicate outline of lesions.

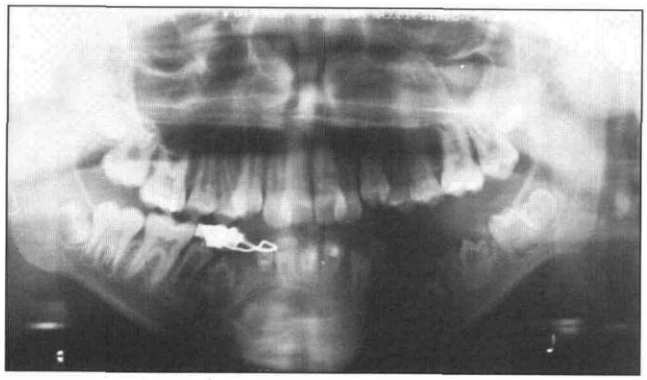

Fig 2: Bilateral manifestation of ROD.

dental abnormalities.

The intraoral clinical and radiographic examinations revealed a structurally normal, age-appropriate maxillary permanent dentition with severe crowding of the incisors. In the mandibular arch, the right posterior segment and the four permanent incisors were visible clinically, while the dentition in the entire permanent left posterior quadrant was unerupted. The dysplastic permanent left first molar and the primary left first and second molars had been extracted in response to episodes of noninflammatory swelling of the gingivae around the affected teeth. An orthodontic appliance was present in the mandibular right quadrant, and a temporary restoration was noted in the access opening of the mandibular left central incisor (Fig 2).

Clinically, ROD affected both the primary and permanent dentitions. The permanent and primary anteriors were yellow with hypoplastic enamel (Fig 3). Radiographically, the unerupted teeth appeared underdeveloped with large pulp chambers and little demarcation between the enamel and dentin. The radiodensity of these teeth was reduced in comparison to the unaffected mandibular teeth. The dental follicles

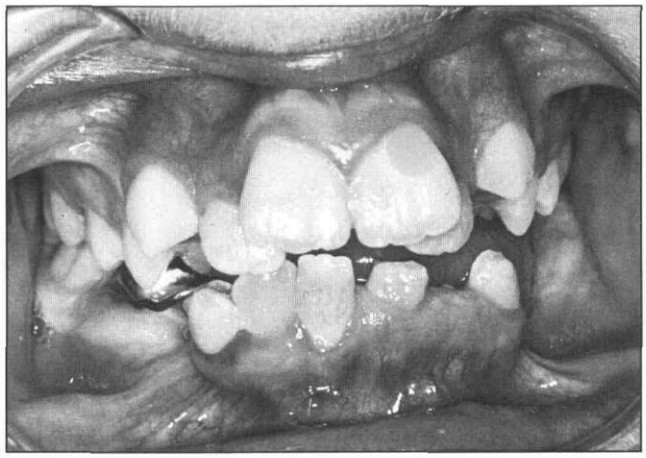

Fig 3: Hypoplastic and discolored permanent and primary teeth.

surrounding the unerupted canines were enlarged, and small radiopaque foci representing dystrophic calcification were identified in the follicles surrounding the partially erupted lateral incisor (Fig 4).

Discussion

Epidemiologic data derived from more than 100 reported cases indicate that regional odontodysplasia is 2.5 times more common in the maxilla (Crawford and Aldred 1989). However, our patient presented with a mandibular lesion. When more than one quadrant is involved, ROD can be seen on the same side of both jaws or bilaterally in either the maxilla or mandible, as in this patient (Crawford and Aldred 1989). Our case is somewhat unusual, however, in its pattern of bilateral distribution (Lustmann 1975 found that affected teeth were located on both sides of the

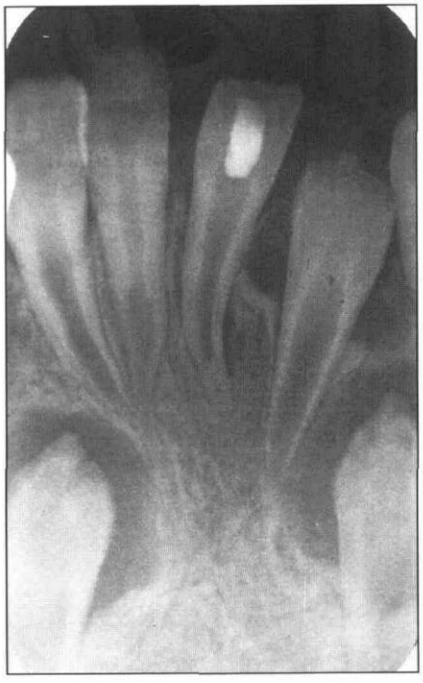

Fig 4: Radiodensity of affected teeth was reduced as compared to normal teeth; note large pulp chambers, large follicles around the canines, and small radiopaque foci in the follicles of the partially erupted lateral incisor.

midline in only 11 of the 51 cases reported to that time).

Normal teeth generally are not found mesial to the affected dentition (Gibbard et al. 1973). In addition, ROD is usually seen in consecutive teeth, with no normal teeth interposed (Crawford and Aldred 1989). The present case is consistent in these respects. It also has been reported that the permanent first molar is dysplastic only if the adjacent primary molars exhibit ROD (Gibbard et al. 1973). The mandibular right permanent first molar and primary second molar were both normal in this patient. On the left side, however, the mandibular left permanent first molar and primary second molar had been extracted, possibly because they were both affected with ROD.

The patient's presenting complaints of missing and abnormal teeth are typical of this condition. Delayed eruption of the permanent dentition is common, as is extraction of dysplastic teeth as a result of abscesses in the absence of caries (Pruhs et al. 1975; Anneroth and Ramstrom 1980). The unusual clinical and radiographic appearance of the teeth can be related to defective production of enamel and dentin. The enamel is hypoplastic, and the presence of residual enamel matrix indicates incomplete mineralization of the matrix (Crawford and Aldred 1989). A differential diagnosis may include amelogenesis imperfecta, dentinogenesis imperfecta, dentinal dysplasia I and II, shell teeth, hypophosphatasia, rickets, Turner's tooth, nursing caries, and calcifying odontogenic fibroma (Crawford and Aldred 1989).

A microradiographic examination of ROD by Kerebel et al. (1989) demonstrated hypomineralized incremental strands of tissue interspersed throughout normal layers of enamel. This indicates that amelogenesis is not only defective, but actually is interrupted as the tooth forms. Some layers of enamel are actually well mineralized (Kerebel and Kerebel 1982; Fearne et al. 1986), but this does not affect the radiographic image; the severe hypoplasia imparts a subnormal radiopacity to the enamel. The dentinal layer is similarly hypoplastic, and dentinal tubules are reduced in number and irregular in arrangement, although the mantle dentin usually is normal (Kinirons et al. 1988). Interglobular dentin and poor calcospherite formation are detected in varying degrees (Crawford and Aldred 1989). Histologic examination of dentin has revealed the presence of "clefts" or widened tubules extending through the mantle dentin to the pulp (Abrams and Groper 1966). It is the hypoplasia and hypocalcification of hard tissue, combined with the large pulp chambers (Lian et al. 1988), that accounts for the "ghost-like" radiographic appearance of ROD. Clefts in enamel and dentin may allow entry of microorganisms into the pulp, causing pulpitis and periapical inflammation without caries.

Numerous theories on the etiology of ROD have been proposed, but compelling evidence is lacking for most of them. Rushton (1965) investigated viral infections as the possible cause of ROD. However, other authors have been unable to detect them, nor do the enamel defects in ROD resemble defects caused by known viruses such as rubella or cytomegalovirus (Crawford and Aldred 1989). Likewise, no specific group of prenatal drug exposures can be correlated to the defect (Crawford and Aldred 1989). Neural crest cells that fail to migrate are being investigated as a possible cause (Crawford and Aldred 1989). Trauma to the dentition has not been well supported as a possible cause; however, several investigators have reported it (Anneroth and Ramstrom 1980). Walton and associates (1978) suggested that local vascular disturbances may be involved in ROD. They reported three ROD patients with congenital cutaneous angiomas, and drew attention to three previously reported cases with similar vascular lesions. Our patient exhibited remnants of vascular birthmarks on the skin overlying the left side of the mandible. Although the cutaneous lesions themselves are not likely to cause alterations in tooth formation, they may be a marker of an underlying vascular disturbance that could affect odontogenesis, as discussed by Fearne et al. (1986). The fact that both primary and permanent teeth are affected in ROD leads to the conclusion that the etiologic agent must be present from the second trimester of gestation, when calcification begins in the primary dentition, through the early teenage years during calcification of the permanent teeth. Therefore, if a cutaneous angioma is related to alteration in regional blood supply to the teeth over a sufficient period of time, this theory could be plausible. Unfortunately, vascular lesions are not identified in all patients.

Another proposed factor in the pathogenesis of ROD is local or systemic infection (Zegarelli et al. 1963; Rushton 1965; Fearne et al. 1986). Our patient's history of fever and swelling of the left side of the face in infancy is intriguing since this was the side with the affected teeth. However, if infection were to result in damage to primary teeth, it would have to affect the dental lamina in utero. Furthermore, attempts to identify viral particles or antibodies in ROD patients have been fruitless (Gibbard et al. 1973).

Summary

ROD with an unusual presentation of bilateral mandibular distribution accompanied by a cutaneous vascular lesion is presented. Although this is another case of ROD coexisting with vascular lesions, the causal relationship remains uncertain.

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