Pediatr Radiol (1998) 28: 802–804

Robert J. Gorlin

Otodental syndrome, oculo-facio-cardio-dental (OFCD) syndrome, and lobodontia: dental disorders of interest to the pediatric radiologist

Received/accepted: 6 May 1998

R. J. Gorlin
University of Minnesota Schools of Dentistry and Medicine,
515 Delaware St S. E., 16–127,
Minneapolis, MN 55455 USA

Abstract Three unusual dental anomalies are presented which should be of interest to the pediatric radiologist: otodental syndrome – an autosomal dominant syndrome of bizarre, greatly enlarged teeth with bulbous crowns (globodontia) that spares the incisors, in combination with sensorineural hearing loss; oculo-facio-cardio-dental syndrome – an X-linked dominant syndrome that is lethal in males, characterized by congenital cataracts, unusual facies, atrial septal defect (ASD), ventricular septal defect (VSD), and canine teeth that cease to grow only when they cut off their own blood supply by growing through the orbit and lower border of mandible; and lobodontia – a dominant disorder characterized by teeth whose crowns resemble those of a wolf.

Introduction

The study of dental dysmorphology is unfortunately becoming a lost art. Only in oral radiology courses in dental schools is time devoted to consideration of some of the fascinating genetic conditions that affect teeth. Pediatric radiologists in their formal training have almost no experience. To whet intellectual appetite and to encourage more consideration of this field, I will briefly review three conditions.

Otodental syndrome

In 1972, Levin and Jorgenson [1] described a syndrome of dental anomalies and sensorineural hearing loss. To date, several unrelated families have been described [1–10]; one case report is doubtful [11]. Inheritance is clearly autosomal dominant with variable expressivity.

The incisors of both dentitions are spared. The crowns of the canines and posterior teeth are enlarged, bulbous, and malformed with multiple prominent lobules. The deciduous dentition is more severely involved. The relation between cusps and major grooves is eliminated, hence the use of the term "globodontia". An enamel defect is frequently noted on the facial surface of the canine teeth. Premolar teeth are frequently missing or small in size. There is often delayed eruption of the deciduous malformed teeth or even of the permanent posterior teeth [2, 3, 9]. One can observe duplicated pulp chambers with denticle formation and a longitudinal dental septum and early pulpal obliteration. The molar teeth have a tendency toward conical or taurodont root form. Complex and/or compound odontomas have also been described in the posterior maxilla and mandible [2, 8, 8a, 9] (Fig. 1).

Sensorineural hearing loss to about 65 dB is found at all frequencies, but is more pronounced at about 1000 Hz. It usually plateaus by the fourth decade [4]. The age of onset of the hearing loss ranges from early childhood to middle age [4, 6, 7], which may complicate diagnosis of the disorder.

Oculo-facio-cardio-dental syndrome

Marashi and Gorlin [12, 13] described three examples of binary combination of congenital cataracts and canine radiculomegaly. They cited an earlier example of Hayward [14] in which canine radiculomegaly, delayed dentition, persistent primary teeth, oligodontia, and congenital cataracts were found in an 18-year-old fe-

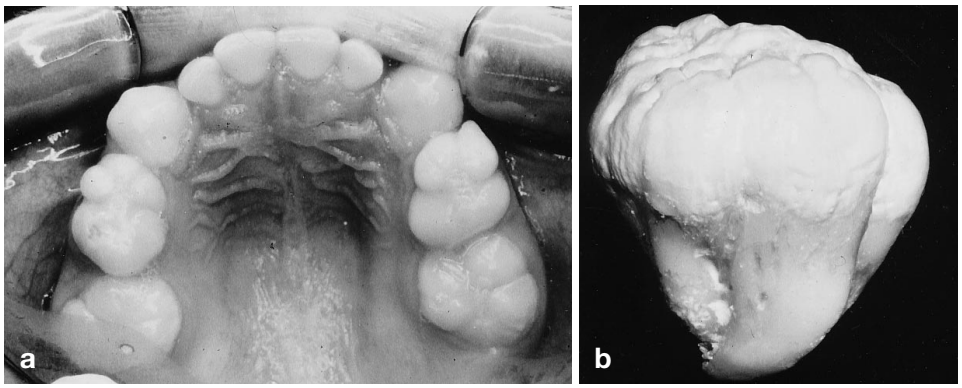

Fig. 1 Otodental syndrome **a** In occlusal view, not normal-sized incisors, markedly enlarged canine and molar teeth (globodontia). **b** Enormous crown with bizarre morphology and short fused roots

male. Marashi and Gorlin [12, 13] also noted a 20-year-old female with marked canine radiculomegaly on whom further information was not available. Gorlin et al. [15] suggested the name oculo-facio-cardio-dental syndrome.

Wilkie et al. [16] reported a mother and daughter with congenital cataracts, microphthalmia, and septal heart defect, suggesting that this combination represented a new syndrome. The face was long and narrow, the philtrum long, and the nose had a high bridge. In the lateral view, the nose appeared pointed. The tip was broad with well-delineated cartilages. Eye findings included congenital cataracts, secondary glaucoma, microphthalmia, and horizontal nystagmus. In the daughter, posterior embryotoxon was noted and there was oligodontia with persistent primary teeth. The columella was broad. In the mother, the teeth had variable root length and canine radiculomegaly. The mother had an ASD; the daughter, both ASD/VSD. Cutaneous syndactyly of toes 2–3 was observed in the daughter.

Aalfs et al. [17] described two unrelated female patients with a long narrow face, microphthalmia, microcornea, congenital cataracts, high nasal bridge, short nose with broad tip, long philtrum, sensorineural hearing loss, persistent primary teeth, oligodontia, ASD, VSD and cutaneous syndactyly of toes 2–3. One of the two had cleft palate, the other had a septate vagina. Additional cases were added by Gorlin et al. [15] and Obwegeser and Gorlin [18, 19].

The occurrence of this syndrome in two generations and in seven females but in no males suggests that we may be dealing with a syndrome that is X-linked dominant and lethal in the male.

The patient appears to have a long narrow facies which is characterized by a sharp nose with a clearly defined bifid tip. A high nasal bridge was noted in at least five patients. Eye changes evident at birth consist of congenital cataracts and microphthalmia or microcornea with resultant or secondary glaucoma.

Surely the most unmistakable finding is canine radiculomegaly. This is not manifest orally, but its unique nature is seen on a panoramic radiograph of the jaws (Fig. 2). The root ends of the canine teeth do not close until adulthood, the roots continuing to grow until the orbit and lower border of the mandible are reached by the maxillary and mandibular canines, respectively.

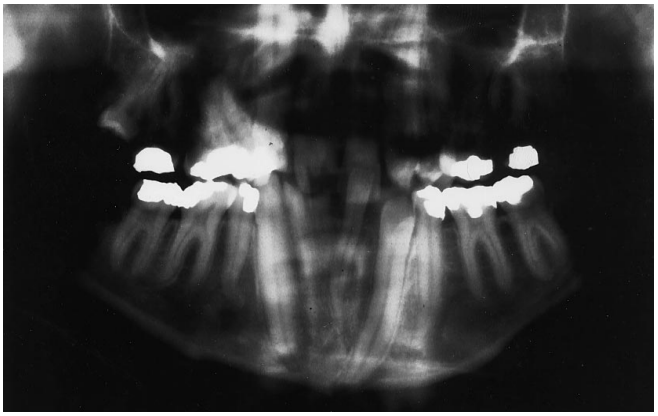

Fig. 2 Oculo-facio-cardio-dental syndrome. Canine teeth grow until the apex pierces the lower border of the mandible and cuts off its own blood supply

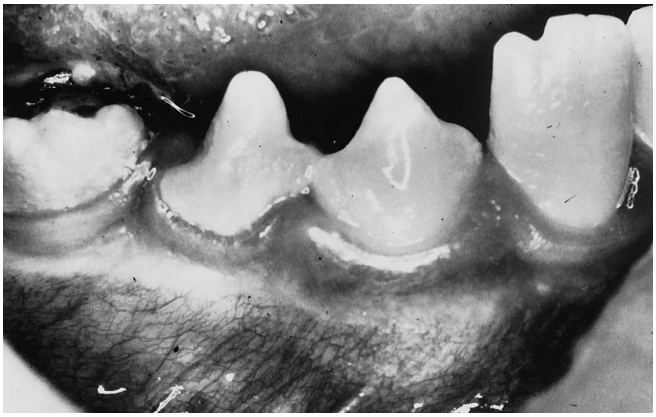

Fig. 3 Lobodontia. Teeth resemble those of a wolf

This dental anomaly is accompanied by oligodontia and persistence of primary teeth.

Cardiac anomalies of various types (ASD, VSD, mitral valve prolapse) have been documented in at least five of the patients. Cleft palate or submucous cleft palate was documented in three of the seven patients. Cutaneous soft-tissue syndactyly of toes 2–3 was found in four of seven patients. Various miscellaneous findings included a septate vagina in one patient and sensorineural hearing loss in another.

Lobodontia

There is general reduction in crown size and there may be hypodontia. The incisors tend to be invaginated or to be shovel-shaped. The cingulum of incisors and premolars is accentuated (20–23). The canines and premolars have pointed cusps with crowns resembling those of a wolf, causing Robbins and Keene [21] to label the condition lobodontia (Fig. 3). The molars are multituberculate with single conical roots. Inheritance is autosomal dominant.

References

1. Levin LS, Jorgenson RJ (1972) Familial otodental dysplasia: a "new" syndrome. Am J Hum Genet 24: 61a
2. Beck-Mannagetta J, et al (1984) Odontome and pantonale Hörstörung bei otodentalem Syndrom. Dtsch Zahnärztl Z 39: 232–241
3. Chen RJ, Chen HS, Lin LM et al (1988) "Otodental" dysplasia. Oral Surg Oral Med Oral Pathol 66: 353–358
4. Cook RA, Cox JR, Jorgenson RJ (1981) Otodental dysplasia: a five year study. Ear Hear 2: 90–94
5. Gundlach KKH, Witkop CJ Jr (1977) Globodontie – eine neue erbliche Zahnformanomalie. Dtsch Zahnärztl Z 32: 194–196
6. Jorgenson RJ, Marsh SJ, Farrington FH (1975) Otodental dysplasia. Birth Defects Orig Artic Ser 11: 115–119
7. Levin LS, Jorgenson RS, Cook RA (1975) Otodental syndrome. A new ectodermal dysplasia. Clin Genet 8: 136–144
8. Salmerón JI, et al (1996) Odontogenic tumours and dental anomalies in children. Cranio-Maxillofac Surg [Suppl 1]: 151

8a. Santos-Pinto L, et al (1998) Otodental syndrome: Three familial case reporto. Pediatr Dent 20: 208–211

9. Winter GB (1983) The association of ocular defects with the otodental syndrome. J Int Assoc Dent Child 14: 83–87
10. Witkop CJ Jr (1976) Globodontia in the otodental syndrome. Oral Surg 41: 472–483
11. Stewart DJ, Kinirons MJ (1982) Globodontia. Br Dent J 152: 287–288
12. Marashi AH, Gorlin RJ (1990) Cataracts and radiculomegaly of canines. Oral Surg Oral Med Oral Pathol 70: 802–803
13. Marashi AH, Gorlin RJ (1992) Radiculomegaly of canine teeth and congenital cataracts: confirmation of a syndrome. Am J Med Genet 42: 143
14. Hayward JR (1980) Cuspid gigantism. Oral Surg Oral Med Oral Pathol 49: 500–501
15. Gorlin RJ, Marashi AH, Obwegeser HL (1996) Oculo-facio-cardio-dental syndrome. Am J Med Genet 63: 290–292
16. Wilkie AO, Taylor D, Scambler PJ, Baraitser M (1993) Congenital cataract, microphthalmia and septal heart defect in two generations: a new syndrome? Clin Dysmorphol 2: 114–119
17. Aalfs CM, Oosterwijk JC, van Schoeneveld MJ, et al (1995) Cataracts, radiculomegaly, septal heart defects, and hearing loss in two unrelated adult females with normal intelligence and similar facial appearance: Confirmation of a syndrome. Clin Dysmorphol 6: 93–103
18. Obwegeser H, Gorlin RJ (1997) Oculofacio-cardio-dental syndrome. Clin Dysmorphol 6: 1–3
19. Obwegeser HL, Gorlin RJ (1997) Oculo-facio-cardio-dental (OFCD) syndrome. Clin Dysmorphol 6: 281–283
20. Brook AH, Winder M (1979) Lobodontia – a rare inherited dental anomaly. Report of an affected family. Br Dent J 147: 213–215
21. Robbins I, Keene HF (1964) Multiple morphologic dental anomalies. Oral Surg 16: 683–690
22. Schulze C (1976) Erbbedingte Formanomalien sämtlicher Zähne – ein neues Syndrom? Zahnärztliche Welt Rundschau 85: 80–84, 178–182
23. Shuff RY (1972) A patient with multiple conical teeth. Dent Prosth 22: 414–417