OTODENTAL SYNDROME

Vidhi Shah[1]

[1]Department of Pediatric and Preventive Dentistry, College of Dental Sciences and Research Centre, Ahmedabad, India

NARRATIVE REVIEW

ABSTRACT

Otodental syndrome is an autosomal dominant disorder that is characterized by bulbous formations of canine and molar teeth, sensorineural hearing loss and eye coloboma. Management of this syndrome is interdisciplinary, with primary focus on eyes, teeth and ears.

How to cite: Otodental Syndrome. The Quadrant. 2023;1(3):14-15. https://doi.org/10.5281/zenodo.11114831

HISTORY & ETYMOLOGY

The otodental syndrome was recognized in 9 families by 2006. The syndrome is also known as otodental dysplasia, familial odentodysplasia, globodontia and oculo-oto-dental syndrome.[1]

ETIOLOGY

This condition is inherited on an autosomal dominant basis. The locus for this condition is mapped on 20q13.1, in a 12-cM chromosomal region.[2]

FACIAL MANIFESTATIONS

Patients present with the notable features of otodental syndrome in early childhood to middle age. The most striking clinical facial features includes sensorineural high frequency progressive bilateral hearing loss of 65 dB at 1000 Hz.

A range of ocular defects have been observed. These include iris pigment epithelium defects, severe chorioretinal coloboma, microcornea, microphthalmos, lens coloboma and asymmetry

How to cite: Otodental Syndrome. The Quadrant. 2023;1(3):14-15.https://doi.org/10.5281/zenodo.11114831

of eyes.[2]

ORAL MANIFESTATIONS

Oral manifestations include gingival hyperplasia around erupting teeth, delayed eruption, premolar microdontia and absence of premolars. The most characteristic oral manifestation is globodontia, where large bulbous canines and molars can be seen without any prominent cusps or globules (Figure 1). Fused tooth germs, duplicated and calcified pulp chambers with a thistle-tube configuration have been reported. Odontomas are underreported and no consistent observations have been made pertaining to malocclusions.[3]

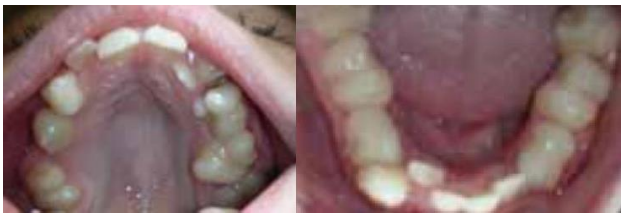

Figure 1 Dental Features of Otodental Syndrome

MANAGEMENT

Management can include extraction of teeth to orthodontic correction and is usually based on correction of individual symptoms. This includes ad hoc treatment of eyes and hearing aids if necessary.[2,3]

REFERENCES

1. Bloch-Zupan A, Goodman JR. Otodental syndrome. Orphanet Journal of Rare Diseases. 2006;1(1). doi:10.1186/1750-1172-1-5

2. 1. Enright S, Humphrys AK, Rea G, James JA. Globodontia in the Otodental Syndrome: A rare defect of tooth morphology occurring with hearing loss in an eight-year-old. Dental Update. 2015;42(10):927–32. doi:10.12968/denu.2015.42.10.927

3. Coltzer J. Otodental Syndrome: A Case Report. Pediatric Dentistry. 2005;27(6)482–5.

How to cite: Otodental Syndrome. The Quadrant. 2023;1(3):14-15.https://doi.org/10.5281/zenodo.11114831