5-2022

Oligodontia, Mandibular Hypoplasia and Microglossia in Pediatric Patients: Review of the Literature and Report of a Case

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Abstract

The combined findings of oligodontia, mandibular hypoplasia and congenital malformation of the tongue (hypoplasia) is relatively rare in children. This presentation discusses the case of a 6-year-old female patient who presented to the dental clinic for emergent care secondary to gross dental caries. Additional findings at the initial visit were that of oligodontia, hypoglossia, micrognathia, and mandibular hypoplasia. A review of the medical history indicated that due to the physical anomalies noted at birth, a genetic microarray was completed indicating some variants of unknow clinical significance. As these forms of pathology uncommonly present without a connection to a specific syndrome, further genetic evaluation is warranted. This presentation reviews the clinical findings noted in the case, potential treatment options for future care and the importance of appropriate collaborative care with other medical professionals.

INTRODUCTION

Oligodontia is a rare genetic disorder which represents the congenital absence of more than six teeth in primary, permanent or both dentitions. It is usually a part of a syndrome and seldom occurs as an isolated entity. Genes responsible for non syndromic oligodontia are found to be MSX1 and PAX9 genes.

A growing amount of evidence suggests that BMAL1 is a key component in mesenchymal cells development and in turn is essential for hard tissue development. In mesenchymal cells, BMAL1 defect inhibits osteoblastic and chondrocytic differentiation. Inactivation of this gene can increase bone resorption as well. Preclinical data suggests that abnormal expression of BMAL1 is associated with mandibular hypoplasia and other skeletal disorders.

Hypoglossia is a very rare condition with an unknown etiology. It has sometimes been associated with limb abnormalities and may be grouped together as hypoglossia-hypodactylia syndrome

CASE REPORT

6 year old female reported to our dental clinic regarding intraoral pain. Upon examination it was noted that she had oligodontia, mandibular hypoplasia, congenital malformation of the tongue (hypoplasia) and cognitive function was typical for her age. Guardian reported maternal drug use during pregnancy. . A review of the medical history indicated that due to the physical anomalies noted at birth, a genetic microarray was completed indicating some variants of unknow clinical significance The teeth visible intraorally appeared to be typically developed. Soft tissue was WNL. A narrow, vaulted palate was noted. #L,S were severely decayed and required extraction, due to non-restorability . During mastication, Tooth #14 was traumatically impinging upon the mandibular gingiva which was her chief complaint. A CBCT was taken confirming oligodontia and absence of cleft palate. Figures 1-4 indicate clinical and radiographic findings. Due to the patient's atypical presentation the patient was referred to a primary care provider and instructed to return for follow-up dental care. Follow-up dental was provided and unrestorable, decayed primary teeth were removed. The patient was then referred to genetics and plastic surgery for further evaluation. A CT image was obtained and evaluation was completed by a plastic surgeon who suggested that the patient may have a variant of ectodermal dysplasia. He further suggested that no intervention was needed at this time as the patient was functioning well.

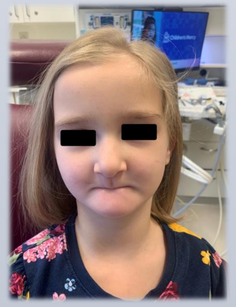

Figure 1

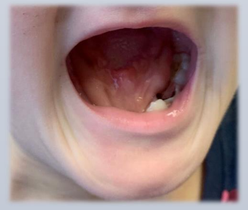

Figure 2

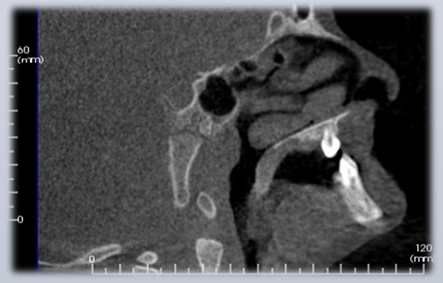

Figure 3

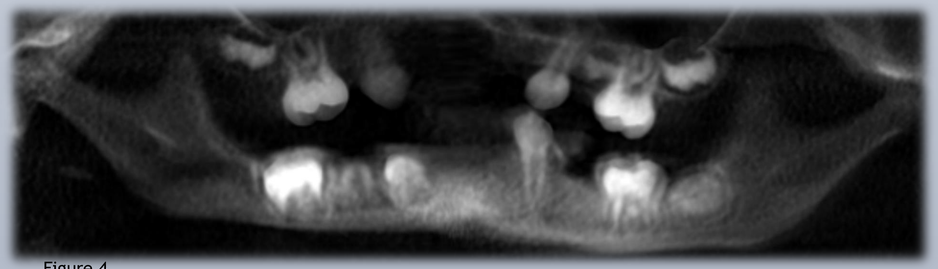

Figure 4

DISCUSSION/CONCLUSION

Individually, oligodontia, mandibular hypoplasia and hypoglossia are often associated with some type of syndrome. However, this patient demonstrated all three clinical findings, and per the current literature reviewed, the findings were not consistent with clinical attributes associated with a single syndrome. Due to the medical history of the patient, it was postulated that the findings may have been due to a combination of genetic aberration and teratogenic exposure. The plastic surgeon who performed a clinical evaluation suggested a diagnosis of an atypical variant of ectodermal dysplasia (ED). While ED may explain the oligodontia, it fails to explain the presence of the hypoglossia and mandibular hypoplasia. Future dental rehabilitation will likely include: placement of bone graft and dental implants Additional genetic counseling was also recommended.

References

1. Tangade P, Batra M. Non syndromic oligodontia: case report. Ethiop J Health Sci. 2012;22(3):219-221.
2. Aljerian A, Gilardino MS. Treacher Collins Syndrome. Clin Plast Surg. 2019 Apr;46(2):197-205. doi: 10.1016/j.cps.2018.11.005. Epub 2019 Jan 30. PMID: 30851751.
3. Voigt S, Park A, Scott A, Vecchiotti MA. Microglossia in a Newborn: A Case Report and Review of the Literature. Arch Otolaryngol Head Neck Surg. 2012;138(8):759–761. doi:10.1001/archoto.2012.1324
4. NCBI Full Report: Aglossia-adactyly syndrome. https://www.ncbi.nlm.nih.gov/medgen/354928
5. Thorp MA, de Waal PJ, Prescott CA. Extreme microglossia. Int J Pediatr Otorhinolaryngol. 2003 May;67(5):473-7. doi: 10.1016/s0165-5876(03)00003-x. PMID: 12697349.
6. Al-Ani AH, Antoun JS, Thomson WM, Merriman TR, Farella M. Hypodontia: An Update on Its Etiology, Classification, and Clinical Management. Biomed Res Int. 2017;2017:9378325. doi: 10.1155/2017/9378325. Epub 2017 Mar 19. PMID: 28401166; PMCID: PMC5376450.
7. Ruiz-Heiland G, Lenz S, Bock N, Ruf S. Prevalence of WNT10A gene mutations in non-syndromic oligodontia. Clin Oral Investig. 2019 Jul;23(7):3103-3113. doi: 10.1007/s00784-018-2731-4. Epub 2018 Nov 14. PMID: 30426266.
8. Chen G, Tang Q, Yu S, Xie Y, Sun J, Li S, Chen L. The biological function of BMAL1 in skeleton development and disorders. Life Sci. 2020 Jul 15;253:117636. doi: 10.1016/j.lfs.2020.117636. Epub 2020 Apr 3. PMID: 32251631.
9. Sharma G, Vengal M, Pai KM, Nagpal A. Hypoglossia Type 1A: report of a case and review of literature with focus on clinical investigations. Indian J Dent Res. 2012 Mar-Apr;23(2):264-70. doi: 10.4103/0970-9290.100438. PMID: 22945721.