Missing Teeth Mystery: Case Report of the WNT10A Gene Mutation

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Introduction

Oligodontia is defined as congenitally missing 6 or more permanent teeth not including third molars. The prevalence of oligodontia is 3.9% in the North American population. Identification of such cases is important because a gene mutation causing oligodontia may also be part of a syndrome affecting skin, nails, eyes, ears, and skeleton. Specifically, the WNT10A gene may be associated with odontoonychodermal dysplasia and schopf-schulz-passarge syndrome which may present with thin hair, smooth tongue surface, and/or microdontia.

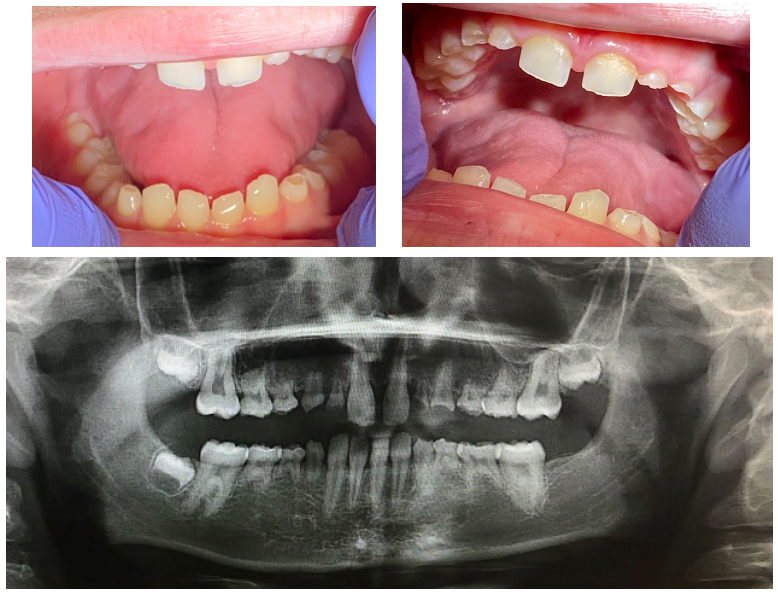

Figure 1. Photos and orthopantomogram of the patient's dentition at the time of consult appointment

References:

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2.Zhou M, Zhang H, Camhi H, Seymen F, Koruyucu M, Kasimoglu Y, Kim JW, Kim-Berman H, Yuson NMR, Benke PJ, Wu Y, Wang F, Zhu Y, Simmer JP, Hu JC. Analyses of oligodontia phenotypes and genetic etiologies. Int J Oral Sci. 2021 Sep 30;13(1):32. doi: 10.1038/s41368-021-00135-3. Erratum in: Int J Oral Sci. 2021 Nov 12;13(1):35. PMID: 34593752; PMCID: PMC8484616.

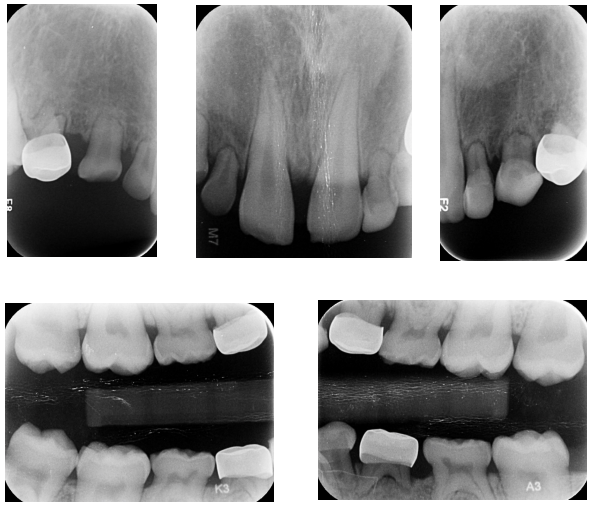

Figure 2. Post restorative intraoral bitewings and periapical radiographs

Clinical Presentation

This presentation discusses a 14-year-old boy who was sent for a consult at University Hospital's Rainbow Babies and Children's Hospital for the management of 19 congenitally missing permanent teeth diagnosed on an orthopantomogram. The patient is ASA-1, taking no medications, and has no known drug allergies. The patient did not report any dental sensitivity or pain either. The patient's physical exam consisted of normal hair distribution, symmetric face, and normal philtrum. Both patient's mother and maternal uncle have a history of congenitally missing 2 maxillary lateral incisors. Upon clinical examination, the patient was noted to have 26 erupted teeth of which 13 are permanent and remaining are primary. Severe attrition was also present on the incisal/occlusal edges on the primary anterior, canines, and molars which was attributed to the patient's habit of chewing ice.

Genetic Testing Results

The patient had his genetic testing completed at University Hospitals on April 10, 2022. The sample was collected via a buccal swab and then sent for a sequence analysis for deletion/duplication testing of the 73 genes related to ectodermal dysplasia and related disorders. The genetic results tested positive to be heterozygous with low penetrance for a pathogenic variant of the gene WNT10A. The pathogenic variant causes a premature translational stop signal resulting in an absent protein product. The patient is heterozygous for the WNT10A mutation, putting him at an increased risk of tooth agenesis which explains our clinical findings. Those who are homozygous for the WNT10A pathogenic variant are also at a 15% increased risk of developing additional signs and symptoms of ectodermal dysplasia.

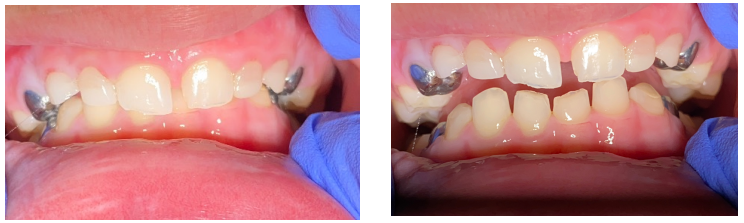

Figure 3. Post restorative photographs of patient's dentition

Treatment

The patient was scheduled for treatment at University Hospital's Rainbow Babies and Children's Hospital for restorative on November 30, 2021, and December 20, 2021. The main goal for the patient's treatment was to make sure the primary dentition could be maintained for as long as possible to preserve alveolar bone. This would set the patient to be a good candidate for implants once he is eligible. Due to the extensive attrition, anterior resin crowns were placed on teeth C D G H M R, and stainless-steel crowns were placed on teeth B I L S. An indirect pulp cap using Biodentine was placed on teeth C D G H I and R because the wear was close to the pulp. A preventive resin restoration was placed on the occlusal surface of tooth 30 and sealants were placed on teeth 3 14 15 19.