Dental Treatment for Pediatric Patients with Ectodermal Dysplasia

Dr. Claudia S. Luto
Northwell Staten Island University Hospital

Introduction

Ectodermal dysplasia (ED) is a group of disorders, an inherent condition where the ectodermal layer of the skin is abnormally developed. There are over 100 types of ectodermal dysplasia with an international prevalence of about 7 to 10,000 births. This presentation discusses a 4-year-old male patient who was referred to Staten Island University Hospital with ectodermal dysplasia.

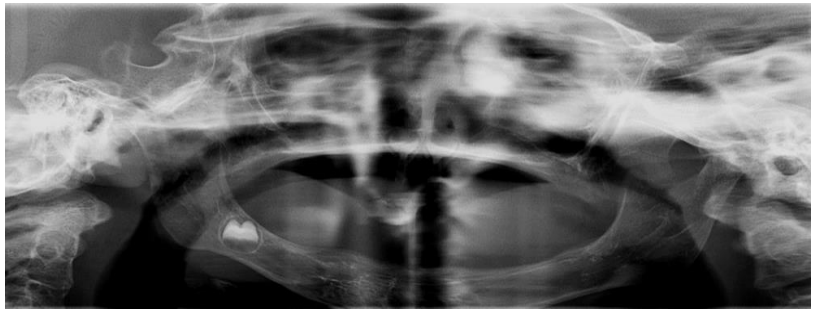

Figure 1: PAN radiograph

Literature Review

- Hereditary ectodermal dysplasia compromises of a large group of conditions in which two or more ectodermally-derived anatomic structures fail to develop. These anatomical structures can be characterized by hypoplasia or aplasia. This can be the skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear.(1)
- Dentition and hair are similarly involved in hereditary pattern.(1)
- The most common type of ectodermal dysplasia is hypohidrotic ED which is X linked recessive therefore seen mostly in males.(1)
- General appearance for ED patients includes: thin/transparent like skin, fine sparse hair, poorly developed/absence of sebaceous glands, oligodontia/hypodontia, conically/pointed shape teeth, poor enamel formation.(1)

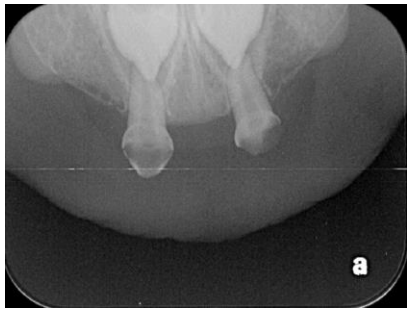

Figure 2: Occlusal radiograph of E and F

Case Description

This presentation discusses a 4 year old male patient who was referred to Staten Island University Hospital. The patient is diagnosed with ectodermal dysplasia by PCP. The chief complaint was: “ My child is missing several teeth and the ones that he does have cavities. I want to save these teeth.”. Patient reports no pain. Patient is not currently taking any medications. The patient has no reported drug and/or food allergies. Clinical examination reveals teeth E and F present. Teeth E and F with decay clinically and radiographically, decay not extending into pulp. PAN reveals tooth buds present for 8,9, and 31. Patient’s current treatment plan included teeth E and F MIDFL's with midazolam premedication and therapeutic device.

References:
1. Hekmatfar S, Jafari K, Meshki R, Badakhsh S. Dental management of ectodermal dysplasia: two clinical case reports. J Dent Res Dent Clin Dent Prospects. 2012 Summer;6(3):108-12. doi: 10.5681/joddd.2012.023. Epub 2012 Sep 1. PMID: 22991648; PMCID: PMC3442425.

Discussion

- A combination of defects to the nails, hair, skin, sweat glands, mucous membranes, and teeth may be seen. Dentally, teeth may be missing, abnormally shaped and or mal positioned.
- Cases vary with retention of primary dentition. If composite restorations can not be upheld with primary dentition, patient will need interim acrylic removable partial/complete dentures. Dentures will often be utilized in early phases as a minimally invasive option. In the future, patients will need frequent relining or remaking of a removable prosthetics to accommodate growth, fracture prostheses and/or progressing esthetic needs. As growth occurs and reaches completion, definitive treatment can be rendered which often involves dental implants

Conclusion

- There are over 100 types of ectodermal dysplasia with an international prevalence of about 7 to 10,000 births. Again, a combination of defects to the nails, hair, skin, sweat glands, mucous membranes, and teeth may be seen
- As ED is typically diagnosed early in life, a pediatric dentist is first in line for treatment. Maintaining primary dentition for a long as possible is crucial for maintaining bone density.
- Inter professional relationships for mature ED patients is vital and necessary to ensure that the patient can live with a fully functioning and restorative dentition.