Ectodermal Dysplasia: Case Report

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Abstract

Ectodermal dysplasia is a hereditary disorder that occurs as a consequence of disturbances in the ectoderm of the developing embryo. Primarily affected tissues are the skin, hair, nails, eccrine glands, and teeth. The disorders are congenital, diffuse, and nonprogressive. The dental management of patients with such dysplastic conditions necessitates a multidisciplinary approach. This case report details the dental rehabilitation of a 16-year-old female to maximize opportunities for the best possible oral function and esthetics.

Introduction

- The term ectodermal dysplasia (ED) is used to describe a disorder when two or more of the structures of the ectoderm have not properly formed.
- Inheritance pattern: X-linked recessive (most common), autosomal dominant and autosomal recessive.
- Males are most often affected, and incidence is estimated to be 1 case per 100,000 individuals.
- Classic EDs are caused by the mutation or deletion of certain genes.
- Hypohidrotic ectodermal dysplasia (HED) is the most common form, and it is usually transmitted as an X-linked recessive trait in which the gene is carried by the female and manifested in the male.

Clinical Features:

Individuals present with varying degrees of abnormalities depending on the subtype of ED, which can include:

Extraoral manifestations:

- Sparse, slow-growing, light- colored hair. Risk of elevated body temperature (hyperthermia). Absent, thin, thick, grooved nails. Cleft lip and/or palate is common. Syndactyly, digital duplication. Severe skin erosions. Dry eye, corneal erosions/opacification and hearing impairment.

Oral manifestations:

- Hypodontia or anodontia, conical teeth, late eruption, alveolar ridges and bone are often under-developed , decreased saliva production, enamel problem, consequently generalized spacing which impairs both aesthetic as well as the masticatory function.

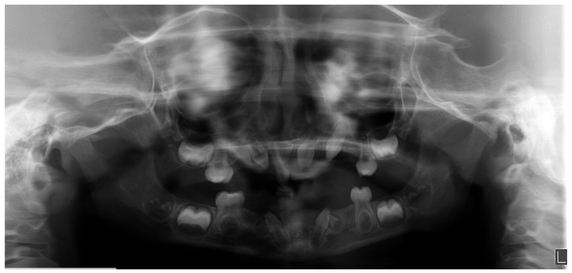

Figure 1. Panoramic radiograph at age of 3-year-old

Case Report

16-year-old female presented to our clinic with CC “I do not like my teeth, and I wish to have a nice smile”. Patient has Ectodermal Dysplasia with a family history of Ectodermal Dysplasia. She is not taking any medications and is allergic to shellfish.

Patient’s first dental visit was at the age of 3 with the parents’ CC “she has only 5 teeth in her mouth”.

Clinical and radiographic examination showed:

- Sparce/thin hair, eyebrows, and eyelashes.
- Posterior crossbite on the left side
- Conical shaped anterior teeth
- Missing teeth

Figure 2. Extraoral photograph

Figure 3. Panoramic radiograph at age of 16-year-old

Figure 4. Intraoral right side

Figure 5. Intraoral left side

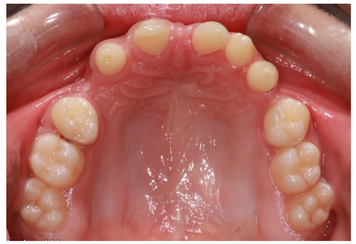

Figure 6. Intraoral maxillary view

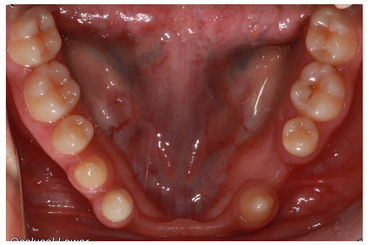

Figure 7. Intraoral mandibular view

Management

In our pediatric department, the required restorative treatments and caries control were done, and patient was referred to periodontic, orthodontic, oral surgery and prosthetic departments, a multidisciplinary team worked on the treatment plan as follows:

- Equally spread the upper anterior teeth in order to have space for crowns.
  - Extrude impacted canine #6
  - Intrude #10-11-22-26-27 to achieve an even plane of occlusion
  - Correct the tooth's rotation of #29
  - Keep the space for #21 and between #23-25 for future implant placement.
  - Extract A, C. H, J, R
  - Place implant for #4.5.7.12,13, 20.21, and #27, Implant bridge on 23xx 26
  - Crown #6,8,9.11, 22.27

The oral surgery will evaluate for possible jaw surgery in the future to prevent lower facial collapse.

Figure 8. intraoral frontal view

Figure 9. Intraoral frontal view with active ortho treatment and temporary anterior lower prothesis

Discussion

Patients with ectodermal dysplasia require lifetime dental care. It will be necessary to create new prosthesis as the person grows in order to accommodate their evolving oral structures and enable chewing. The greatest practical and aesthetically pleasing outcomes will eventually come from orthodontic therapy and the prosthetic insertion of implant-supported dentures. Patients with ED have particular dental needs, and a multidisciplinary approach is the most effective way to treat them. Patient compliance is essential to the outcome of treatment, which may involve a prolonged procedure.

References

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