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Case Report

Prosthetic Rehabilitation of a Pediatric Patient With Ectodermal Dysplasia: A Case Report

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Abstract

Ectodermal dysplasias represent a heterogeneous group of hereditary disorders characterized by abnormal development of tissues derived from the ectoderm, including teeth, hair, nails and sweat glands. In pediatric dentistry, these cases are uncommon but clinically significant due to the functional and psychosocial implications associated with oral manifestations.

Objective: To describe the clinical management and prosthetic rehabilitation of a pediatric patient diagnosed with Hypohidrotic Ectodermal Dysplasia (HED).

Materials and Methods: A 6-year-old female patient attended the pediatric dentistry clinic accompanied by her mother, who reported delayed dental eruption. Clinical and radiographic examinations confirmed partial anodontia consistent with hypohidrotic ectodermal dysplasia. A removable functional-esthetic prosthesis was fabricated to restore oral function and improve facial appearance.

Results: The installation of the prosthetic device significantly improved the patient's esthetics, phonation and masticatory function, contributing to proper maxillomandibular development and psychosocial well-being.

Conclusion: Early diagnosis and prosthetic intervention in pediatric patients with ectodermal dysplasia are essential to promote normal oral and facial growth, as well as to improve quality of life.

Keywords: Hypohidrotic Ectodermal Dysplasia; Pediatric Dentistry; Oral Rehabilitation; Anodontia; Oligodontia; Maxillofacial Development; Removable Prosthesis

Introduction

Dysplasia is a cellular alteration characterized by variations in volume, shape and organization. There are several degrees of dysplasia, depending on severity: mild, with minimal changes; moderate, as an intermediate state and severe. Regarding malignancy potential, dysplasia can be classified into three groups: non-malignant, premalignant and malignant. Dysplasias can be pure, occurring as isolated forms or complex or syndromic, when they form part of a clinical syndrome.

Ectodermal dysplasias represent a heterogeneous group caused by elements of different types or nature, presenting alterations characterized by abnormal development of structures derived from the ectoderm. These congenital syndromes affect one or more ectodermal components without a progressive cellular course. Approximately 150 types of ectodermal dysplasia have been described; however, Hypohidrotic Ectodermal Dysplasia (HED) is the most common form.

HED, first described in 1838 by Wedderburn and known as Christ-Siemens-Touraine syndrome, is a rare genetic disease typically inherited as an X-linked recessive disorder, with affected male progenitors and carrier females, although autosomal mutations are possible. This condition causes abnormalities in the development of ectoderm-derived structures, including nails, teeth, hair, sweat glands and sebaceous glands.

Therefore, HED is clinically characterized by the triad of hypohidrosis, hypotrichosis and hypodontia. Patients may present, among other alterations, fine and smooth skin, hypohidrosis, which can trigger fevers of unknown origin or hyperpyrexia, low set pointed ears, prominent lower lip, periorbital pigmentation, dystrophic nails, altered mammary glands, short stature and intellectual delay.

In the comprehensive management of HED, dentistry plays a special role due to numerous oral manifestations, such as hypodontia, anodontia or oligodontia; lack of alveolar ridge development; loss of vertical dimension; narrow and short jaws; deep palate; dry mucosa; prominent lips; and cleft lip or palate. Existing teeth may present anomalies including malposition, rotation, retrusion, conical crowns, spacing, malformations and short roots. These conditions make the dentist an indispensable professional to improve the quality of life of patients with this disorder. The objective of this study is to analyze the case of a pediatric dental patient with ectodermal dysplasia and her prosthetic rehabilitation.

Materials and Methods

Methodologically, this is an observational descriptive study presenting the clinical experience of an unusual case or unexpected evolution, providing descriptive information for medical and community education based on CARE guidelines by Gagnier, et al. A 6-year-old female school-aged patient presented to the pediatric dental clinic asymptomatically, accompanied by her mother, who reported absence of tooth eruption. Both patient and guardian were cooperative. Extraoral and intraoral examinations were performed following the ALOP Pediatric Dentistry Clinical Procedure Reference Manual. Panoramic radiographs and intraoral photographs were used for evaluation and analysis.

Case Report

The patient exhibited abnormal shape and size of primary teeth (A and O), edentulous spaces, loss of the mucocutaneous line at the lips, altered scalp growth and hair emergence, brittle nails and dry skin. Complementary exams included a panoramic radiograph to confirm the presence of permanent tooth germs. The mother reported no family history of agenesis (Fig. 1). Radiographic analysis confirmed the absence of permanent teeth #8, #7, #5, #9, #10, #11, #12, #20, #28 and #29. Considering the altered shape and size of teeth A and O, along with oligodontia and clinical features, ectodermal dysplasia was diagnosed. Prosthetic rehabilitation was planned to improve oral quality of life (Fig. 2).

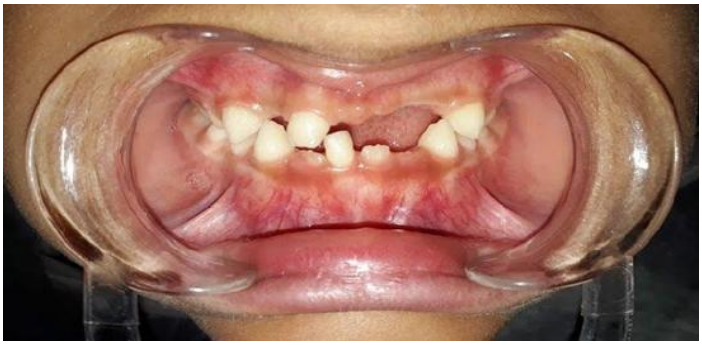

Figure 1: Initial Intraoral Photograph. Edentulous spaces in the anterior maxilla.

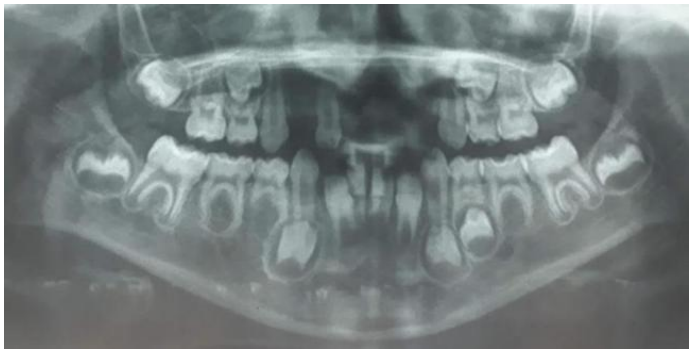

Figure 2: Initial Panoramic Radiograph. Oligodontia and absence of permanent tooth germs #8, #7, #5, #9, #10, #11, #12, #20, #28 and #29. Altered shape and size of teeth A and O.

Treatment Objectives

To clinically manage a pediatric patient with ectodermal dysplasia and provide prosthetic rehabilitation. A functional and aesthetic prosthetic device was selected to promote functional orthopedics, stimulate transverse maxillary development, improve mastication, psychological development and social integration through aesthetics and functionality. Fixed prosthetic devices were preferred due to patient comfort, tissue safety and suitability for long-term use. The guardian provided informed consent. Prior to prosthesis fabrication, the patient received preventive treatment to reduce caries risk and improve plaque control. Functional impressions of the upper arch were taken using alginate and the device was designed with orthodontic bands, 0.8 mm wire for the body and 0.9 mm wire for the tube and double-bar system, tailored to the patient's maxilla to avoid occlusal or soft tissue trauma (Fig. 3).

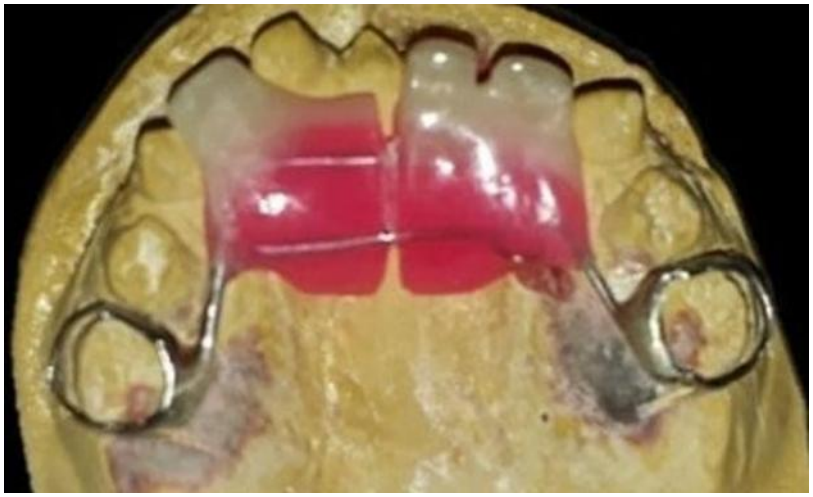

Figure 3: Functional aesthetic prosthetic device on the model.

The prosthesis was cemented with dual cement on teeth 55 and 65. The patient tolerated the device well, reporting no discomfort or functional difficulty.

Clinical Evolution

The guardian and patient were instructed that excessive salivation, speech and swallowing difficulties could occur during adaptation. Instructions on device use, nutrition and hygiene were provided. Follow-up occurred at 4 weeks to assess adaptation, soft tissue health and prosthesis maintenance, with subsequent check-ups every 6 weeks. Interdisciplinary consultations were planned due to rapid maxillary growth at this age, which may affect prosthesis fit (Fig. 4).

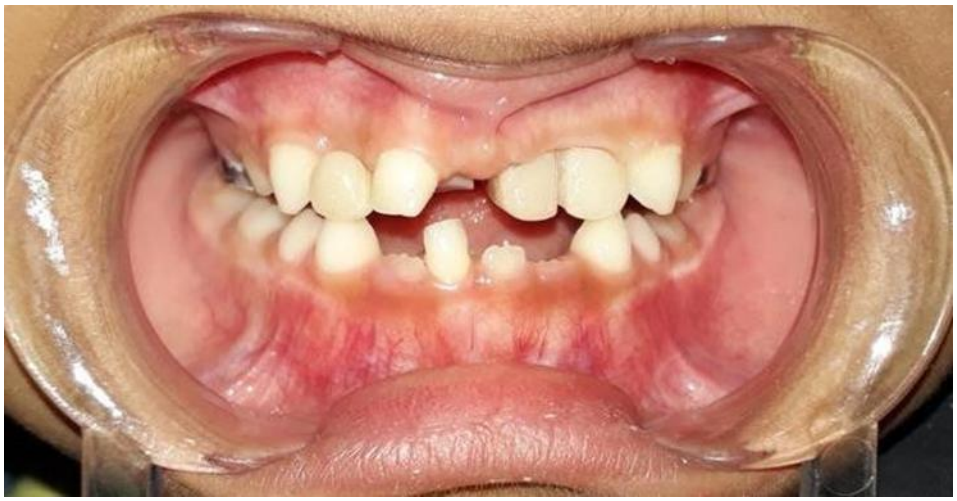

Figure 4: Prosthesis *in-situ* during follow-up.

Discussion

Ectodermal dysplasia is a hereditary disorder affecting ectodermal structures, including central and peripheral nervous system, sensory epithelium, epidermis, glands, pituitary and dental enamel. Tooth abnormalities, including hypodontia, anodontia or oligodontia, are common and may limit function, reduce arch length, induce improper habits and negatively impact self-esteem. Comprehensive evaluation considering all patient characteristics is essential for accurate medical-dental diagnosis and effective management. Alarcón, et al., recommend fixed prostheses with orthodontic bands for children who are less cooperative with removable partial dentures, to preserve supporting tissues, ensure functional mastication, swallowing and phonation, provide harmonious dentofacial aesthetics and prevent antagonist tooth extrusion. In this case, a double-bar tube system was added to facilitate maxillary development, improving upon previously described designs.

Conclusion

Early diagnosis and intervention can significantly improve aesthetics, speech and maxillary development in pediatric patients with ectodermal dysplasia, enhancing clinical outcomes with functional aesthetic prosthetic devices. These devices offer a treatment option for children with anterior permanent tooth loss, whether congenital or due to trauma or early childhood caries, emphasizing the innovative role of pediatric dentists in solving such cases. Successful use requires fully erupted primary second molars and careful clinical supervision.

Conflict of Interests

The authors declare that they have no conflicts of interest.

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References

1. Alarcón C, Perona M, Mock A. Aesthetic and functional alternative of the anterior sector in deciduous dentition. Rev Cient Odontol Univ Sur. 2017;5(2).
2. Alves N, De Oliveira R, Figueredo N. Hypohidrotic ectodermal dysplasia: A syndrome of interest for dentistry. Int J Odontostomatol. 2012.
3. Bassil J, Antunes A, Tedesco T, Reyes A, Rodrigues G. Oral rehabilitation in patients with ectodermal dysplasia: One-year follow-up. Case report. Odontología. 2016;19(2).
4. Campuzano S, Santos-Juanes J, Medina A, Sánchez J. Hypohidrotic ectodermal dysplasia: Case report. Departments of Pediatrics and Dermatology, Central University Hospital of Asturias, Oviedo, Spain. 2004.
5. Gagnier JJ, Kienle G, Altman DG, Moher D, Sox H, Riley D, et al. The CARE guidelines: Consensus-based clinical case report guideline development. J Clin Epidemiol. 2013.
6. Huamán-Guerrero M, Pichardo-Rodríguez R, De La Cruz-Vargas J. How to write a case report: Methodological principles. Rev Fac Med Hum. 2016.
7. Orozco J, Puello C, Hernández D, Roviro C. Dental management of patients with Chris-Siemens-Touraine syndrome: Case report. Rev Odontol Mex. 2018;22(1).
8. Universidad Central de Venezuela. Practice: General aspects of tumors. Faculty of Dentistry, Department of Pathological Anatomy. Caracas. 2002
9. Zaror C, Hope B, Diaz J, Jans A. Fixed prosthesis with tube-bar system in pediatric dentistry: A 12-month clinical follow-up case report. Rev Clin Periodoncia Implantol Rehabil Oral. 2015.