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Prosthodontic rehabilitation of a 6-year-old patient with ectodermal dysplasia: A case report

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Abstract

Ectodermal dysplasia (ED) is an inherited disorder that affects the ectodermal-derived structures of developing embryos, such as skin, teeth, hair, nails, sweat glands, and sebaceous glands. It typically presents as missing teeth (anodontia/hypodontia), sparse hair (hypotrichosis), lack of sweat glands (hypohidrosis), abnormal nails [onychodysplasia], and palmoplantar hyperkeratosis. There are two major types of this condition: X-linked anhidrotic or hypohidrotic and hidrotic (autosomal type). Oral manifestations of ectodermal dysplasia include missing teeth, with or without cleft lip and palate, conical or peg-shaped teeth, delayed eruption of permanent teeth, thin knife-edge-shaped alveolar ridges, etc. This case report details the prosthodontic rehabilitation of a 6-year-old patient with hypohidrotic ectodermal dysplasia and typical dental manifestations of oligodontia.

Keywords: Ectodermal dysplasia, oligodontia, hypohidrosis, prosthodontic rehabilitation

Introduction

Ectodermal dysplasia (ED) is a heterogeneous group of diseases that manifest in ectodermal tissues, including hair, teeth, nails, sweat glands, and sebaceous glands. It is characterized by trichodysplasia (abnormal hair), anodontia or oligodontia (abnormal or missing teeth), onychodysplasia (abnormal nails), and dyshidrosis (abnormal or missing sweat glands). There are 170 distinct ectodermal dysplasia subtypes known to exist. Ectodermal dysplasia (ED) is present in about 7 births out of 10,000 worldwide. Clinically, there are two primary categories based on the involvement of the sweat glands: (1) hypohidrotic or anhydrotic (Christ-Siemens-Touraine syndrome), in which sweat glands are either absent or significantly reduced in number; (2) hydrotic (Clouston's syndrome), in which sweat glands are normal. Defective dental and hair involvement is quite similar in both types. Hypohidrotic ectodermal dysplasia is the most common type, showing an X-linked inheritance pattern; males are more susceptible than females. The inheritance pattern for the hydrotic type is autosomal dominant. Frontal bossing, sunken cheeks, a saddle nose, thick and everted lips, wrinkled and hyperpigmented skin around the eyes, and wide, low-set ears are characteristics of typical faces. The majority of patients have brittle, thin, sparse, and lustreless scalp hair. Conical or peg-shaped teeth, hypodontia or total anodontia, delayed eruption of permanent teeth, a decrease in the alveolar ridge height, and a reduction in the lower third of the face's vertical dimension are among the dental manifestations. This case report details the prosthetic rehabilitation of a 6-year-old patient with hypohidrotic ectodermal dysplasia having a typical dental manifestation of oligodontia using upper and lower removable partial dentures.

Case Report

A 6-year-old patient was referred from NRS Medical College to the Department of Pediatric and Preventive Dentistry, Dr. R. Ahmed Dental College and Hospital, complaining of the absence of multiple teeth in relation to the upper and lower jaws. Family history reveals, patient's sister also has thin scalp hair without any other manifestations. His mother gave a history of thin and fine hair growth, dry skin, and decreased sweating. Extraoral examination reveals frontal bossing, a prominent supraorbital ridge, a depressed nasal bridge, retarded malar prominence, thick everted lips, sparse hair, scanty eyebrows, dry, scaly skin, and reduced lower facial height.

Intraoral examination revealed absence of all the teeth except four primary second molars in each quadrant and thin alveolar ridges with reduced vertical bone height. Panoramic radiographs revealed unerupted first permanent molar tooth germs and developing tooth germs of the second permanent molar and second premolar in each quadrant. Other noticeable features were reduced sweating and dry, scaly skin. The patient was diagnosed with hypohidrotic ectodermal dysplasia based on clinical and radiographic findings.

Fig 1: Frontal view showing typical clinical manifestations of ED

Fig 2: Sparse scalp hair

Fig 3: Side view

Fig 4: Intraoral view

Fig 5: Maxillary Occlusal View

Fig 6: Mandibular occlusal View

Fig 7: Panoramic radiograph

By considering the patient's age, number of teeth present, economic background, and preservation of the alveolar bone integrity present at the time, removable maxillary and mandibular partial dentures were decided in this case. The primary impression was taken with irreversible hydrocolloid [alginate] and cast made with dental stone. Custom trays fabricated with cold-cured acrylic resin for the final impression. The final impression was taken with condensation silicone using the functional impression technique. Bite registration was done in the patient's mouth. Registered bite transferred to the articulator, and teeth setting done. Trial denture adjustments are done by checking phonetics. Denture fabrication is done by using heat-cured acrylic. Denture insertion was done. The patient was satisfied both functionally and aesthetically. The patient was given instructions for the proper maintenance of the oral and prosthetic hygiene.

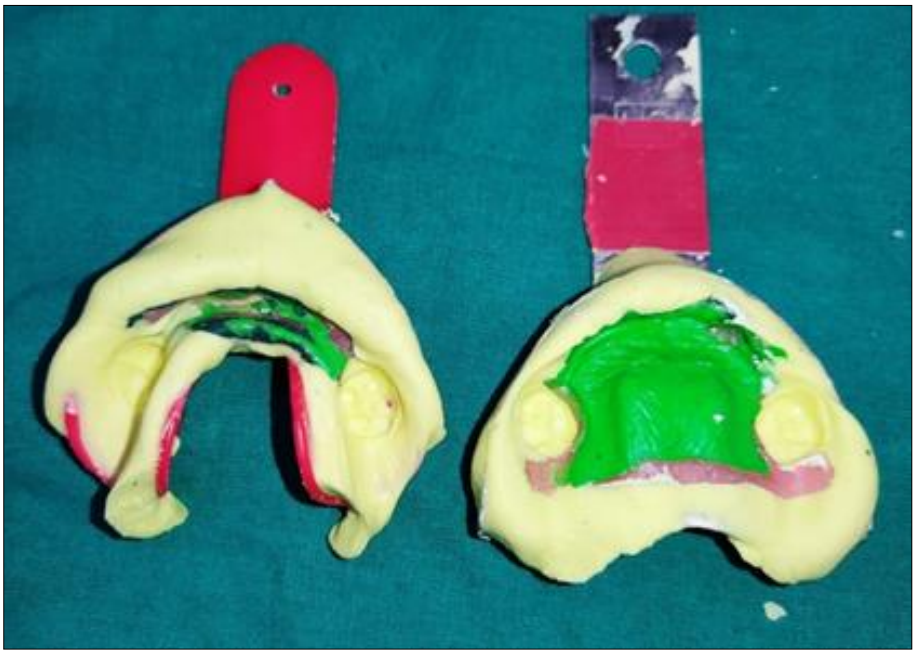

Fig 8: Secondary Impression

Fig 9: Teeth Setting

Fig 10: Denture Insertion

Fig 11: Final Acrylic Denture Insertion

Discussion

The primary dental need for ectodermal patients is prosthetic rehabilitation to restore chewing function, aesthetics, and social confidence. The National Foundation for Ectodermal Dysplasia states that the objective of dental care for people with ectodermal dysplasia is to create a dentition that is age-appropriate and enhances speaking, swallowing, mastication, oral and facial development, and aesthetics.

A removable or fixed partial denture, an overdenture, a complete denture prosthesis, or an implant-supported prosthesis are some of the available treatment choices. Age, the psychosocial environment, the presence of teeth, oral hygiene, the occlusal vertical dimension, bone volume, and the growth and development of the jaw are all factors that must be taken into consideration when planning dental treatments. Early prosthodontic intervention minimizes the related emotional and physiological issues in these patients by assisting the child in adjusting to the prosthesis and developing normal appearance, speech, mastication, swallowing, and temporomandibular joint function.

Prosthodontic rehabilitation is a challenging task because of reduced alveolar bone height with "knife-edge" morphology and limited remaining tooth structure. It is often advisable to wait until adolescence to replace teeth with implants, as these replacements are typically limited to those with finished craniofacial growth.

For young patients, the use of removable partial denture (RPD) is a reversible treatment that can significantly improve functions and aesthetics without jeopardizing compromised dentitions.[10] Relining, rebasing, or remaking of the removable prosthesis is necessary as the child grows to accommodate changes in growth and preserve normal oral function and aesthetics. Vergo advised creating a new prosthesis every 4-6 years and relining/rebasing an intraoral prosthesis in a growing patient every 2-4 years. Patients with ED should have periodic dental recalls at regular intervals so that prostheses can be adjusted or replaced according to the patient's growth and development.

Conclusion

For this patient, rehabilitation with a removable partial denture was a feasible, affordable, and acceptable technique that enhanced speech, function, appearance, and psychosocial state. Both the patient and parent were well satisfied with the treatment. For the treatment to be successful over the long run, recall appointments and careful maintenance of dental and prosthetic hygiene are required. Patient was advised for dental implants in the future, after the completion of dentofacial growth.

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