Management of a Case of Ectodermal Dysplasia: A Multidisciplinary Approach

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ABSTRACT

Ectodermal dysplasia (ED) is a rare disorder involving 2 or more of the ectodermal structures, which include the skin, hair, nails, teeth, mucus glands, and sweat glands. Each person with ectodermal dysplasia may have a different combination of defects. The most common form of ED is linked to the X chromosome and usually affects men. Another form of the disease affects men and women equally. The patient's special needs are taken into consideration in a treatment plan. The purpose of this case report was to describe the dental management of a 9-year-old boy with ectodermal dysplasia whose maxilla showed the presence of mobile, conical teeth that were unrestorable and a solitary permanent left molar, while his mandible was completely edentulous. These mobile, conical teeth were extracted, and a complete denture was constructed to restore the patient's esthetics, form, and function.

(J Dent Child 2011;78(2):107-110)

Received November 16, 2009; Last Revision May 3, 2010; Revision Accepted May 4, 2010.

KEYWORDS: ECTODERMAL DYSPLASIA, ECTODERMAL STRUCTURES, X CHROMOSOME, CONICAL TEETH, EDENTULOUS JAW

Ectodermal dysplasia (**ED**) is a very rare condition occurring in an estimated 1 per 100,000 live births.[1] The disease was first described by Thurnman in 1843.[2] It embraces a long series of abnormalities whose common denominator is a morphological alteration of ectoderm-derived organs and tissues.[3] It is considered to be an X-linked or autosomal recessive condition.[4] ED is a disorder involving 2 or more of the ectodermal structures, which include the hair, nails, teeth, mucus glands, and sweat glands. There are many distinct types of ED. ED patients often have certain craniofacial features which can be distinctive: frontal bossing is common; longer or more pronounced chins are frequent; and broader noses are also very common.

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Freire-Maia and Pinherio[5] described 117 varieties of ED with multiple combinations of abnormal ectodermally derived structures. Clinically, ED may be divided into 2 broad categories: (1) the hidrotic type; and (2) the more common anhidrotic or hypohidrotic type.[6] The X-linked hypohidrotic form is also called Christ-Siemens Tourine syndrome[7,8] and is characterized by the classical triad of hypodontia, hypohidrosis, and hypotrichosis.

ED is usually a difficult condition to manage prosthodontically because of the typical oral deficiencies and afflicted individuals who are quite young to receive extensive prosthodontic treatment procedures. These treatments can restore their appearance and promote the development of positive self-image.[9,10]

The purpose of this case report was to describe the dental management of a 9-year-old boy with ectodermal dysplasia whose maxilla showed the presence of mobile, conical teeth that were unrestorable and a solitary permanent left molar, while his mandible was completely edentulous. The focus of our treatment was to improve the patient's facial esthetics and masticatory function

and allow him to integrate with his peers, thus preventing any psychological repercussions.

CASE REPORT

A 9-year-old boy reported to the department of Pediatric dentistry, DAV dental college,Yamunanagar, India with the chief complaint of absence of teeth and inability to eat properly. Physically, he exhibited several of the classical features of ED: hypohidrosis; hypotrichosis; prominent forehead; and saddle nose. His family and medical history were noncontributory. Extraorally, nevus flammeus (port wine stain) was present on the face and prognathic mandible (Figure 1).

Panoramic radiographs revealed the presence of 2 conical permanent maxillary lateral incisors with incomplete roots and a maxillary permanent left first molar (Figure 2). Intraoral examination revealed dry and sticky oral mucosa with localized mild gingivitis, conical-shaped and grossly decayed teeth, a permanent left molar in the maxillary arch, and an edentulous mandible (Figure 3). The patient's parents reported that only these conical teeth have erupted since birth, and he has not visited any dentist before. As these conical teeth were grossly decayed and mobile and could not be restored or treated endodontically, they were extracted under local anesthesia. The permanent maxillary left molar was retained (Figure 4). A removable denture was planned for the remaining edentulous area in the maxillary arch, and a complete denture was planned for the mandibular arch (Figure 5). Wax rims were prepared for the maxillary and mandibular denture base, and proper vertical height was established. A trial denture base was made from autopolymerizing acrylic resin, and the selected denture teeth were adjusted in size and set in wax. After trying, the dentures were processed in heat-polymerizing acrylic resin in the usual manner and delivered (Figure 6).

Recall appointments were scheduled for 24 hours, 72 hours, 2 weeks, 4 weeks, every 3 months for the first year, and every 4 months for the second year. Written oral hygiene instructions were given and explained to the patient. The patient adapted well to the dentures and has been on regular recall for three years and has shown satisfactory response to the treatment.

DISCUSSION

Hypohidrotic ED is a rare congenital disease that affects several ectodermal structures. Manifestations of the disease differ in severity and involve teeth, skin, hair, nails, and sweat and sebaceous glands. Intraoral examination in an ED patient can reveal hypodontia, with the teeth usually pegor cone-shaped. Anodontia is also possible. Dental deficiencies compromise lip support and are responsible for a decreased lower facial height. Nowak stated that "treating the pediatric patient with ED requires the clinician to be knowledgeable in growth and

Figure 1. Patient exhibiting the classical features of ectodermal dysplasia: hypohidrosis; hypotrichosis; prominent forehead; and saddle nose.

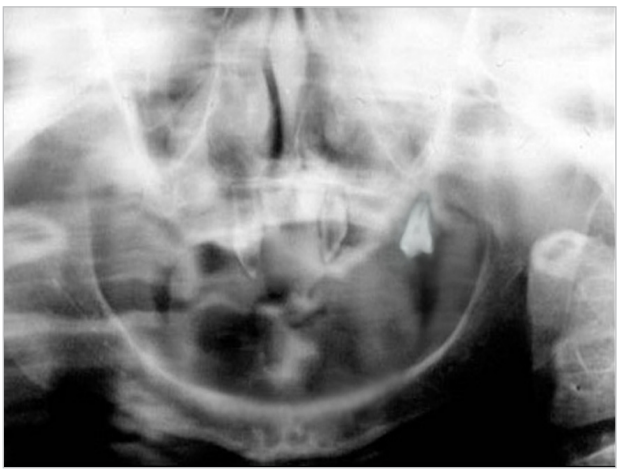

Figure 2. Panoramic radiograph revealing the presence of 2 conical permanent maxillary lateral incisors and permanent maxillary left molar.

Figure 3. Intraoral examination revealing conically shaped, grossly decayed teeth, a permanent left molar in the maxillary arch, and edentulous mandible.

development, behavioral management, techniques in the fabrication of a prosthesis, the modification of existing teeth utilizing composite resins, the ability to motivate the patient and parent in the use of the prosthesis, and the long-term follow-up for the modification and/or replacement of the prosthesis." According to Nowak, a series of introductory visits may be needed before treatment commences to attain the patient's required trust.[11]

Figure 4. Extracted conical teeth and retained permanent maxillary left molar.

Figure 5. The removable denture for the edentulous area in the maxillary arch and a complete denture for the mandibular arch.

Figure 6. Patient wearing the complete denture.

Fixed prosthodontic treatment is seldom used exclusively in the treatment of ED, primarily because many afflicted individuals have a minimal number of teeth. In addition, ED patients are often quite young when first treated, and fixed partial dentures with rigid connectors should be avoided in young, actively growing patients. This is because rigid fixed partial dentures could interfere with jaw growth, especially if the prosthesis crosses the midline.[12] Individual crown restorations have no age restriction related to jaw growth, but larger pulp sizes and shorter crown heights may cause concern. In spite of these concerns, crowns often are used to treat young ED patients.[13] Crowns and direct composite restorations often are used in combination with removable partial dentures in the prosthodontic management of these patients. They usually are necessary to provide proper contours on the hypoplastic teeth that will be used as abutments for removable partial dentures.[14,15]

Removable prosthodontics are the most frequently reported treatment modality for the dental management of ED. Because anodontia or hypodontia is typical in individuals with this condition, complete dentures, partial dentures, or overdentures are often part of the treatment provided. Although complete dentures can provide an acceptable esthetic and functional result, under development of the edentulous alveolar ridges in ED patients can compromise denture retention and stability. Periodic recalls of young ED patients also are important because prosthesis modification and replacement will be needed as a result of continuing growth and development. In addition to adjustments related to fit, occlusion of prostheses must be monitored for ages because of jaw growth. Other problems related to removable prostheses are speech difficulties, dietary limitations, and loss of the prosthesis.[11]

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