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Case Report

Removable partial dentures for a patient with ectodermal dysplasia: A clinical report

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ABSTRACT

The orofacial characteristics of ectodermal dysplasia include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, frontal bossing, a depressed nasal bridge, protruberant lips, and hypotrichosis. Patients with this disease often need complex prosthodontic treatment. The options for a definitive treatment plan may include fixed, removable, or implant-supported prostheses, singly or in combination. However, financial constraints and other priorities can prevent the patient from choosing the most desirable treatment. This clinical report describes the diagnosis and treatment of ectodermal dysplasia in a 33-year-old man. The treatment included endodontic, restorative and definitive removable partial dentures fabricated to establish an acceptable therapeutic occlusal vertical dimension.

Ectodermal dysplasia syndromes have been described as a group of disorders of morphogenesis displaying 2 or more of the following signs and symptoms: (1) trichodysplasia, (2) dental anomalies, (3) onychodysplasia, and (4) dyshidrosis. These malformations result from developmental defects in tissues in which progenitor cells were originally derived from the ectoderm of the embryo. Congenital malformation of teeth, hair, nails, or sweat glands may occur either as single isolated malformations or as part of an ectodermal dysplasia syndrome.[1] Freire-Maia and Pinheiro classified ectodermal dysplasias into 11 possible groups, based on all possible combinations of 2 or more defects.

Ectodermal dysplasias appear to be inherited as an X-linked recessive trait, with the most common condition among the ectodermal dysplasias being hypohidrotic (anhidrotic) ectodermal dysplasia, also known as Christ-Siemens-Touraine syndrome.[2] Clarke suggested that the X-link trait is transmitted with the gene being carried by the female partner and manifested in the male partner. Anhidrotic ectodermal dysplasia is considered to be triad of hypodontia or anodontia, hypotrichosis, and a hypohidrosis, and associated with other components that result from defective development of structures of ectodermal origin (Fig. 1). It is 1 of 132 possible types of clinical ectodermal dysplasia syndromes. Anhidrotic ectodermal dysplasia is found in all races, with an incidence of 1–7 per 100,000 live births. Affected males usually have extensive dental involvement, missing most of their deciduous and permanent dentition. Hypotrichosis occurs with the scalp hair being thin in childhood. Body hair and eyebrows are also sparse (Fig. 2).

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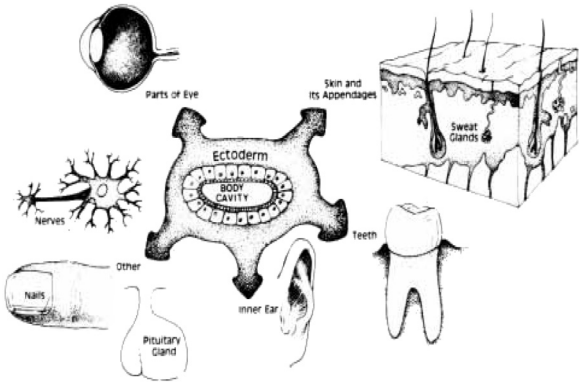

Fig. 1 – Schematic drawings of affected structures derived from ectodermal embryonic layer.

The most common oral characteristic is hypodontia or anodontia in many cases, reflecting the complete suppression of dental ectoderm. A few teeth may be present but with retarded eruption. Because of the lack of teeth and resultant loss of vertical dimension, the lips are protuberant, the vermilion border is indistinct, and the alveolar process does not develop in the absence of teeth and, hence, is missing incisors, canines, and premolars, when present, often have conical absence of teeth and the oral mucosa often appears dry.[1] Histopathologic evaluations show aplasia of the labial, buccal and lower respiratory glands. The pharyngeal and laryngeal mucosa may be atrophic, resulting in dysphonia.

A multidisciplinary team approach to treatment is recommended. Removable partial dentures are most often the treatment of choice; Periodic recall is also necessary in the patients. In addition, osseointegrated implants are an alternative treatment in older persons with anhidrotic ectodermal dysplasia. This clinical report describes the characteristics and prosthodontic treatment of a patient with anhidrotic ectodermal dysplasia.

1. Clinical report

The patient was a 33-year-old man. Clinical examination revealed permanent teeth in the maxilla (first, second molars and canines of both the quadrants). In the mandible, all of the anterior permanent teeth were missing; only the mandibular first and second permanent molars of both the quadrants were present. In addition, the patient had an undeveloped mandibular alveolar ridge The vertical dimension of occlusion (VDO) was maintained due to the retained posterior molars. The patient exhibited typical characteristics of ectodermal dysplasia, including saddle nose, fine, sparse hair in eyebrows and in the eyelid with partial anodontia. His medical history was unremarkable. A diagnosis of partial anodontia was made from the clinical and radiographic features. The patient had not, however, received comprehensive dental care because of financial constraints. Ironically his wife was also suffering from the same syndrome and consequently their two offsprings (daughters) inherited the condition. Recent studies of fine genetic mapping have shown that other inheritance modalities may also be involved. Mutations in the ectodysplasin-A (EDA) and ectodysplasin-A receptor (EDAR) genes are responsible for X-linked and autosomal HED (Figs. 3–6).

The patient exhibited mild localized gingivitis and was referred for a comprehensive periodontal evaluation, including oral hygiene instructions.[3] Diagnostic casts were prepared, duplicated, and mounted on a semi adjustable articulator (Hanau H2 series). Complete rehabilitation of the dentition was planned, and the patient was informed of the

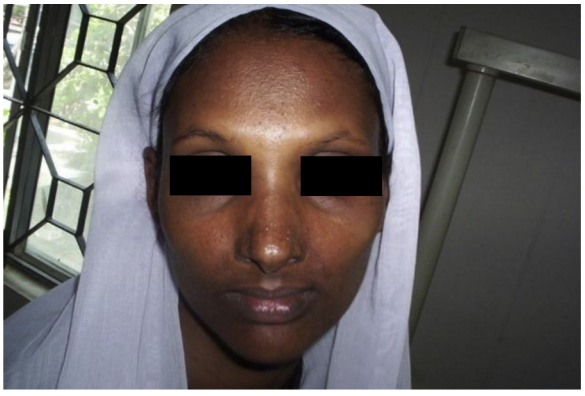

Fig. 3 – Patient's wife.

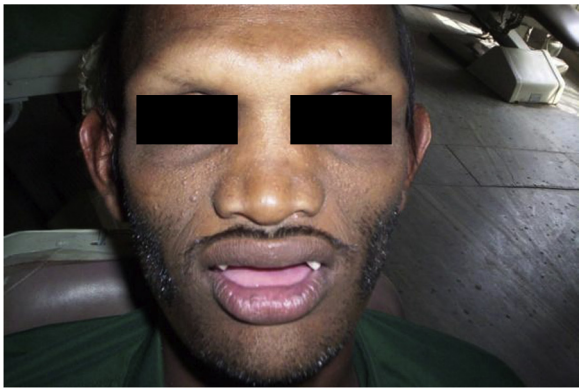

Fig. 2 – Patient extra oral profile.

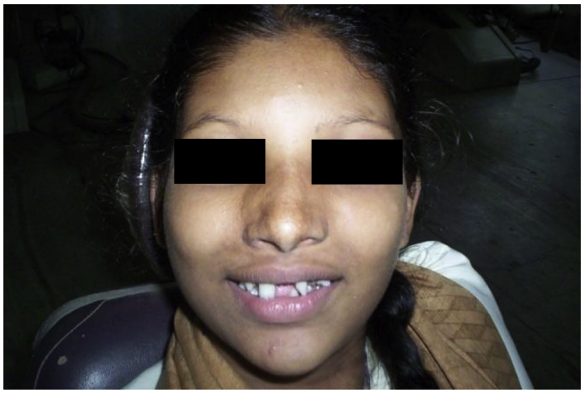

Fig. 4 – Patient's elder daughter.