Ectodermal Dysplasia In A Completely Edentulous, Rare Clinical Scenario: A Case Report

Dr. Deepika Bhayana, Dr. Rahul Bhayana, Dr. Swati Goley, Dr. Hemendra Pratap, Dr. Shabista Jabi, Dr Rohit Bahuguna

1Reader, Department of Oral medicine and Radiology, Narayan swami dental hospital and college, Dehradun. 
2Principal & Head, Department of prosthodontics, Narayan swami dental hospital and college, Dehradun. 
3Senior lecturer, Department of Oral medicine and Radiology, Narayan swami dental hospital and college, Dehradun. 
4Senior lecturer, Department of Prosthodontics, Narayan swami dental hospital and college, Dehradun. 
5Senior lecturer, Department of Pediatric dentistry Narayan swami dental hospital and college, Dehradun. 
6Reader, Department of prosthodontics, subharti dental college, Meerut.

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ABSTRACT

Ectodermal dysplasia (ED) is a rare genetic disease caused by developmental disturbances of embryonic ectoderm derived tissues, organs, and other accessory appendages. The congenital missing of teeth is usually bilateral. Anodontia or hypodontia may be associated with other ectodermal disturbances, such as anhidrosis, asteatosis, hypotrichosis, and salivary glands defects. One such case report of ED is demonstrated here.
INTRODUCTION:
Ectodermal dysplasia (ED) is a rare genetic condition in which two or more tissues originating from the ectoderm show primary developmental abnormalities. The skin, hair, nails, exocrine glands, and teeth are the most commonly afflicted tissues. Congenital, diffuse, and non-progressive diseases are seen. Hypohydrotic (anhidrotic) ED and hidrotic ED are the two most frequent disorders in this category. The most frequent manifestation, hypohydrotic ED (also known as Christ-Siemens-Touraine syndrome), is inherited as an X-linked recessive condition. It is marked by a number of defects (e.g., hypohidrosis, anomalous dentition, onychodysplasia, and hypotrichosis). Frontal bossing, sunken cheeks, a saddle nose, thick and everted lips, wrinkled and hyperpigmented skin around the eyes, and wide, low-set ears are all common traits. Conical or pegged teeth, hypodontia or anodontia, and delayed eruption of permanent teeth are all dental symptoms. In people with hypohydrotic ED, exocrine sweat glands may be missing or limited and rudimentary. Mucous glands in the upper respiratory tract, as well as the bronchi, esophagus, and duodenum, are sometimes lacking. Short stature, ocular abnormalities, reduced tears, and photophobia are all prevalent symptoms. Therefore, the aim of this report is to treat the completely edentulous maxillary and mandibular arch with conventional complete denture.

CASE REPORT
A 19-year-old male patient presented to the OMDR outpatient department at Subharti Dental College and Hospital in Dehradun with the primary complaint of entirely missing teeth in both the upper and lower arch, as well as difficulties in eating as a result of the missing teeth. According to the report, all of the teeth were never erupted. He also detailed a history of hair loss (alopecia), dryness, and reduced sweating. The physical characteristics of the patient confirmed the diagnosis. It was also confirmed that there were no mental, nervous, or maxillofacial abnormalities, as well as irregular sweating. Extraoral examination revealed sparse fine hair, high-set orbits, pronounced chin, prominent and backward lips, sunken cheeks, broad and low-set ears, dryness (absence of sweat gland), and brittle nails (Picture 1, 2, 3, 4 & 5). There was no other case of ectodermal dysplasia in the family, according to the history. Intraoral examination revealed the entire lack of teeth, thin alveolar ridges, reduced vertical bone height, and loss of sulcus depth in the posterior regions of the maxillary and mandibular jaws; full anodontia was also confirmed by panoramic radiography. Orthopantomogram (OPG) was taken which revealed completely edentulous maxillary and mandibular arch. (Picture 7) Patient was sensitized about this condition, the maxillofacial and nutritional consequences, and the prosthetics management according to age; therefore, a conventional complete denture was elected as treatment. (Picture 8) On the basis of clinical and radiographic features, Ectodermal dysplasia was diagnosed.
DISCUSSION
The EDs are a diverse set of genetic illnesses characterized by primary developmental abnormalities of two or more embryonic ectoderm derived tissues that affect about one in every 100,000 births. The hypohidrotic type (ChristTouraine Syndrome) is X-linked and is defined by the typical triad of hypodontia, hypotrichosis, and hypohydrosis, while the hydroptic form (Clouston syndrome) damages the teeth, hair, and nails while sparing the
The most common oral symptom is hypodontia or anodontia of the deciduous and permanent dentition, which is associated with conical-shaped teeth. Anhidrosis is characterized by extraorally fine, sparse, lusterless fair hair over the scalp, severe scaling of the skin, and unexplained pyrexia and heat sensitivity. There is evidence of normal intelligence. Frontal bossing, sunken cheeks, depressed nasal bridge, thick everted protuberant lips, wrinkled hyperpigmented periorbital skin, and a large low set of ears are the other extra oral traits. The maxillary central incisors and canines, which have a conical crown morphology, are the most usually seen intraorally missing permanent teeth. In rare cases, one or both jaws may be missing teeth, preventing the development of the alveolar processes.

The diagnosis is made based on the patient's medical history, as well as a complete clinical and radiographic examination. 1 through 7 Radiographs, such as OPG radiographs, were taken in our case to confirm the diagnosis.

Anodontia (totally edentulous maxillary and mandibular teeth) was present in this case report since childhood, with extra oral characteristics such as hair loss, dryness due to involvement of the sweat gland, and sunken cheeks due to skeletal development. These clinical characteristics supported in the diagnosis of ED. Radiographs, such as OPG, were taken in our case to confirm the diagnosis.

Dental prostheses can also help to increase the tone of masticatory muscles and compensate for the loss of vertical dimension. Mastication problems have been identified as a prominent issue resulting from tooth loss. 8 The facial profile and expression improved dramatically with complete dentures, as seen in this case study; mastication and eating patterns also improved.

For tooth replacement and masticatory function rehabilitation, implant-supported or retained dentures are the treatment of choice. This option has been confirmed in healthy young individuals as well as those with ED, according to the current case study. However, implant therapy may fail due to a lack of alveolar bone, implant embedding, relocation, or displacement by facial growth, eventual trauma to tooth germs, ankylosed teeth, and multidimensional craniofacial growth constraints. 9 To avoid these concerns, the conventional complete denture used in this report was an attempt to adequately rehabilitate this patient and at the very least promote oral function and esthetics while not impairing the mandibular and maxillary bones' natural growth.

When implant therapy is recommended, the fundamental issue is a lack of bone. The prosthetics' retention and stability are extremely tough to achieve.

Dryness of the oral mucosa, as well as underdeveloped maxillary tuberosities and alveolar ridges, are problematic variables for denture resistance and stability in patients with ectodermal dysplasia. 10 These case studies emphasize the need of precise treatment planning as well as the influence of anodontia on ectodermal dysplasia diagnosis.

CONCLUSION

EDs are rare genetic illnesses with many overlapping features that make classification challenging. Individuals with ED experience substantial social challenges as a result of their clinical presentations. It affects the patients' oral functions as well as their normal body functioning. Quick diagnosis and prosthetic rehabilitation using a multidisciplinary approach are the keys to success in ED management. The use of a traditional complete denture allowed for the preservation of masticatory and phonetic functions as well as improved facial attractiveness. Improved self-esteem and social reintegration were indicators of patient satisfaction.

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