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CASE REPORT

Prosthodontic treatment of patients afflicted with hypohidrotic ectodermal dysplasia: Report of two cases

Tratamiento prostodóntico de pacientes con displasia ectodérmica hipohidrótica: reporte de dos casos

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ABSTRACT

Ectodermal dysplasia is a hereditary disorder characterized by the abnormal development of certain ectodermal-origin tissues and structures. Hypohidrotic type is the most commonly observed form of ectodermal dysplasia. Diagnosis is based on the absence or decrease of sweat glands. Dental treatment of oral deficiencies characteristic of this syndrome is commonly quite complex. Physiological and psychosocial reasons dictate the importance for these patients to receive dental treatment at early ages. The present clinical report describes characteristics and prosthodontics treatment of two siblings afflicted with hypohidrotic ectodermal dysplasia.

RESUMEN

La displasia ectodérmica es un desorden hereditario caracterizado por un desarrollo anormal de ciertos tejidos y estructuras de origen ectodérmico. La forma más comúnmente observada de displasia ectodérmica es del tipo hipohidrótica. El diagnóstico se basa en la ausencia o disminución de glándulas sudoríparas. Comúnmente es una condición complicada en cuanto al tratamiento odontológico de las deficiencias orales características de este síndrome. Es importante que estas personas reciban atención dental a temprana edad por razones fisiológicas y psicosociales. Este reporte clínico describe las características y el tratamiento prostodóntico de dos hermanos con displasia ectodérmica hipohidrótica.

Key words: Ectodermal dysplasia, hypohidrotic, full dentures, lingualized occlusion.
Palabras clave: Displasia ectodérmica, hipohidrótica, dentaduras totales, oclusión lingualizada.

INTRODUCTION

Ectodermal dysplasia is a hereditary disorder characterized by the abnormal development of certain ectodermal-origin structures.[1-7] Among the structures at risk we can count hair, sweat and sebaceous glands, nails, teeth, the eye's conjunctive, the crystalline, anterior pituitary glands and ears. Central nervous system defects can also be found.[1-3] Hypohidrotic ectodermal dysplasia can exhibit either a recessive autosomal pattern or a pattern linked to chromosome X. Nevertheless, the most common type is the one linked to chromosome X, exhibited in males.[2] Moreover, due to mutation of gene Xq12q13,[3,4] anhidrotic or hypohidrotic ectodermal dysplasia can appear in a family lacking any history of this condition.

Female carriers of this disease might be afflicted with a variable degree of clinical implications, which might vary from undetectable signs to the manifestation of considerable signs of hypodontia, hypotrichosis and unilateral chest hypoplasia.[5] This is only one of the 192 types of described dysplasia cases. Ectodermal dysplasia is present in all ethnicities, it is estimated that 7 out of every 10,000 births present some type of ectodermal dysplasia, and one out of 100,000 male births presents the anhidrotic variance.[1-9] Affected males exhibit extensive dental absence, among other characteristics such as prominent forehead, depressed nasal bridge, protuberant lips and undefined vermillion border.[1-3,5-9]

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During childhood, hypohidrosis can be found in variable degrees. Frequently with severe cases of hyperthermia.[10]

Hypodontia or anodontia are the most common oral manifestation; they reflect complete suppression of the dental ectoderm. Some teeth can be present with delayed eruption; they are usually malformed or conical teeth. It is common to find dry oral mucosa due to the decrease or absence of salivary glands. Likewise, due to absence of teeth in the residual ridge, it fails to develop suitably, in some cases it even appears missing; in consequence, there might be decrease in facial vertical dimension.

In these patients, the appearance of their teeth is extremely important since it can affect their self-esteem. Psychosocial and physiological reasons mandate providing dental care for patients at an early age. Periodic revisions are also needed in order to preserve and follow up the patients' oral health status.

Most frequent prosthetic treatment is the manufacture of full prostheses, although fixed prostheses or prostheses over implants can also be used. Prosthesis manufacturing enhances sagittal and vertical skeletal relationship during growth and development, since they provide improvement in esthetics, phonetics and masticatory efficiency.

The present clinical report describes characteristics and treatment of two siblings afflicted with hypohidrotic ectodermal dysplasia.

CLINICAL REPORT

A 19 year old male and his 12 year old sister attended the Implantology and Oral Prosthesis Department of the Graduate and Research School, National School of Dentistry, National University of Mexico (UNAM), for examination, assessment and prosthetic treatment.

Both patients exhibited characteristic traits of hypohidrotic ectodermal dysplasia, which included the following: protuberant lips, depressed nasal bridge, mild alopecia well as sparse eyebrows and eyelashes *(Figures 1A and 1B)*. The male exhibited syndactyly in both hands *(Figure 1C)*.

Both patients wore full dentures and exhibited decrease in vertical dimension. Intra-oral exploration revealed dry mucosa as well as small, thin and undeveloped residual ridges *(Figures 1D and 1E)*. Patients had been wearing full dentures for two years. In the boy's denture, a fissure was found at the level of the midline. The girl's denture was maladjusted due to the growth of both upper and lower jaws.

During interview they mentioned that their father had been diagnosed with anhidrotic ectodermal dysplasia, and was wearing a full denture.

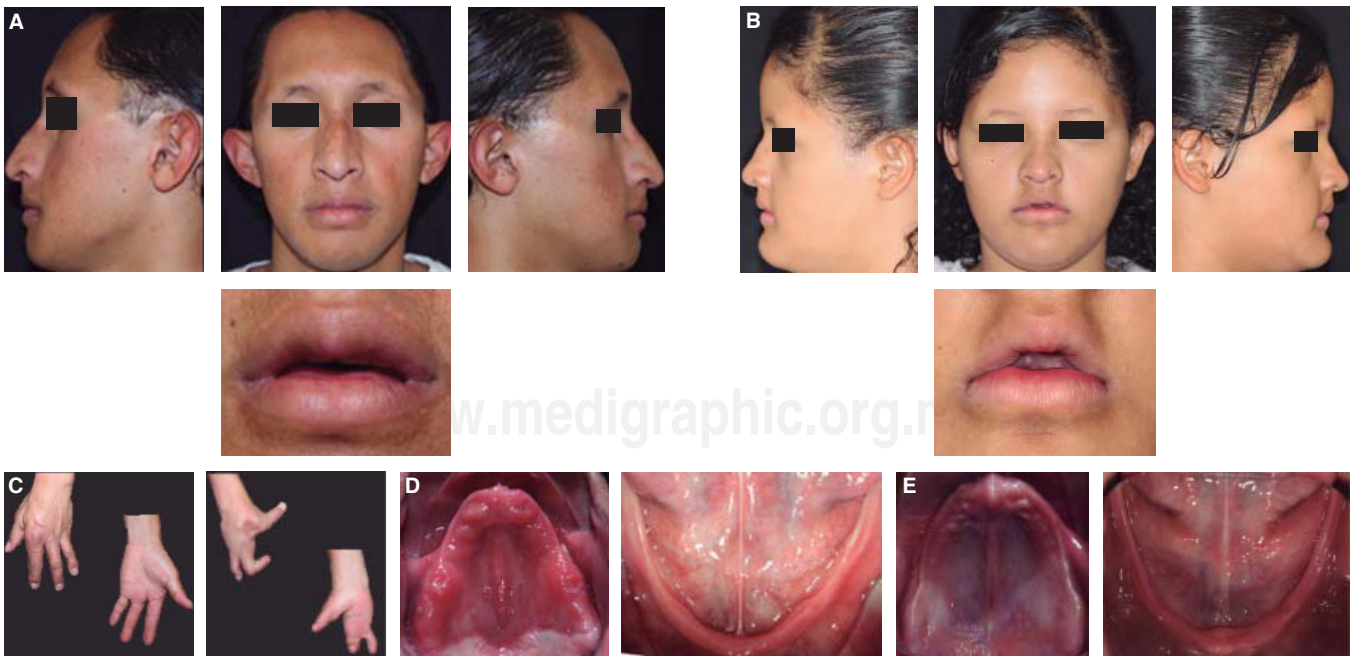

Figure 1. Patients' clinical characteristics. **(A and B)** Characteristic facial traits of dysplasia patients. **(C)** Syndactyly exhibited by the male patient. **(D and E)** Intraoral photographs showing total de-centration state.