The Effects of Ectodermal Dysplasia on Periodontal Tissues

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Abstract

Ectodermal dysplasia (ED) is a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures. ED may effect only ectodermal tissues, or may be associated with anomalies of other organs and systems. It is generally classified into three subgroups; anhidrotik (EDA), hypohidrotik (HED) and hidrotik.

All the ED appear to be genetic in etiology. However, only a small number of ED genes has been genetically mapped or cloned.

Missing teeth or delay in teething often starts to worry the parents and leads to the diagnosis of ED in the second year of life. The most important cause of periodontal problems of ED patients is the decreased level of saliva secretion. Excellent oral hygiene is crucial for the successful treatment of these patients.

By this review we aimed to determine the probable relations between periodontal diseases and ED.

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Introduction

Ectodermal dysplasia (ED) is a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures. Ectodermal dysplasia MAY Effect only ectodermal tissues, or may be associated with anomalies of other organs and systems[1].

Although ed ıs classıfıed differently, it is generally classified into three subgroups; anhidrotik (EDA), hypohidrotik (HED) and hidrotik[2].

EDA is a rare, hereditary disease that has an autosomal recessive mode of inheritance. it is characterized by partial and complete abscence of sweat and sebasseous glands, females are effected more than males[2].

HED is usually inherited as an autosomal recessive genetic trait; the disorder is fully expressed in males only. HED patients exhibit clinical signs AS; hypotrichosis, onychodysplasia, hypodontia or anodontia. Hypohidrotic ectodermal dysplasia is characterized with sparse hair, inability to sweat due to lack of sweat and sebasseous glands. The lack of sweat glands may lead to hyperthermia, followed by brain damage or death in early infancy if unrecognized[3,4].

Hidrotic ectodrrmal dysplasia is characterizeted normal sweat glands and craniofacial structures. But others findings are similar to hypohidrotic or anhidtotic type. Autosomal dominant type ectodermal dysplasia fist described by weech. This type patient have normal hair and eyebrow and missing teeth (anodontia,hypodontia) nail dystrophy[5].

About 160 clinically and genetically distinct hereditery ectodermal dysplasia types have been catalogued, only a few have been diagnosed at the molecular level[1].

Except the clasification above there are a lot of clafications about ED. Lamartine (2003) classified ED into four subgroups according to the functions of the genes discovered: cell-cell communication and signalling, cell adhesion, regulation of transcription, and development[6].

Witkop's classification is composed of[7,8]:

Type I: The most common and best characterised of these conditions is X-linked hypohidrotic ectodermal dysplasia.

Type II: Tooth and nail syndrome (Witkop sydrome) is AN autosomal dominant type of ED characterized

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by missing teeth and poorly formed nails.

Type III: X-linked autosomal dominant especially this case have normal tooth abnormal hair and nails.

Ectodermal disorders is clasificationed four major group by Freire –Maia[8,9]:

a) Hypohydrosis: Hypoplasia and aplasia of salivary and sebasseous glands
b) Hypotrichosis: Thin and sparse hair, absence of eyebrow and eyelashes.
c) Hypodontia: Anodontia or most common oligodontia
d) Onychodysplasia: Abnormalites and nail dystrophy

Several cases of EDs involving at least two of the ectodermal appendages of hair, nails and teeth have been reported in the literature.

Diagnosis Criteria

This X-linked recessive disorder affects males and is inherited through female carriers. The diagnostic tool is the typical clinical physionomy. The most characteristic findings in mEn are the reduced number and abnormal shape of teeth. The delay in teethening is often the first step in diagnosis. The men have an easily recognizable facles also referred to as an old man facies. Same infants have a premature look with scaling of the skin. This can also form a clue to the diagnosis. The number of sweat glands is reduced and both scalp and body hair are sparse, with lack of eyebrows and eyelashes. The clinical findings in carrier females are the same as those in affected males. One third of carriers appears healthy, another third of them show mild symptoms, and the last third exhibits significant symptoms, but often milder than the affected males[10].

Prenatal diagnosis of EDA has occasionally been reported. The diagnosis has been made on fetal skin biopsy, obtained by fetoscopy by 20 weeks gestation, after determination of the sex of the fetus. By histological analysis they demonstrate complete lack of or reduction in the number of polisebaceous follicles and lack of sweat glands primordia in multiple skin biopsis. The interpretation of the biopsy may be difficult. If one does not appreciate the normal regional variability of the distribution of skin appendages of fetal skin which sweat gland primordia only begin to develop at around 20 weeks of gestation. This procedure is complicated and implies a considerable risk to the pregnancy. The use of linked markers on DNA from chorionic villi has greatly improved the safety of prenatal diognosis of X-linked ED. The new method of prenatal diagnosis has major advantages as well as disadvantages. It is technically simpler and may present a lover risk to the pregnancy than fetoscopy and multiple skin biopsies[11].

Differential Diagnosis

The differential diagnostic problem is the distinction of autosomal recessive form of HED from EDA. Anhidrotic form is considerably less common than HED. The clinical features are quite similar in both conditions but due to the different mode of inheritance HED affects both males and females and the heterozygotes have no signs at all. For adequate genetic counseling it is thus important to recognize HED heterozygotes by dental examination and sweat tests[12].

Epidemiology

The prevalence of EDA is unknown; however the incidence in male is estimated at 1 in 100000 births although the condition is usually overlooked in infants. This X-linked recessive disorder affects males and is inherited through female carriers.This carriers–incidence is probably 17,3 in 100000 women[13].

Ethiology and Genetics

All the ectodermal dysplasias appear to be genetic in etiology. However, only a small number of ED genes has been genetically mapped or cloned. The gene associated with HED has been identified, and several splice forms of this gene exists. In situ hybridization revealed that the gene was expressed in the hair follicles and epidermis of adult skin.The discoveries of disease genes and the identification of mutations in patients represent A great progress in biomedical science[14,15].

The recent studies TNF ligand and receptor have developmental regulatory role and are tightly associated with epithelial-mesenchymal interactions and signaling pathways that regulate ectodermal appendage formation and organogenesis during the initiation of development[11].

Clinical Features

Ectodermal dysplasia is characterized by triad of signs:

1) Sparse hair (atrichosis or hypotrichosis)
2) Abnormal or missing teeth (anodontia or hypodontia)
3) Inability to sweat due to lack of sweat glands (anhidrosis or hypohidrosis)

Most patients with ectodermal dysplasia have a normal life expectancy and normal

intelligence. However, the lack of sweat glands may lead to hyperthermia, followed by brain damage or death in early infancy if unrecognized. Thus, an early diagnosis is important[11,16,17].

Craniofacial Structures

Considering how much osseous and dental tissue is missing, these patients have surprisingly normal facial structures. Clinically, this patients have skeletal abnormalities; the forehead appears square, with frontal bossing. There is a prominent supra orbital ridge. The nose has a depressed nasal bridge and is called a saddle nose. The midface is depressed and hypoplastic. The cheekbones are high and broad, although they appear flat and the lips everted and protuberant[18,19].

Hair, Nails, Skin and Skintags

Abnormalities of hair are present in all affected individuals. Most individuals have sparse, fine, slowly growing scalp hair. Sparse eyebrows and eyelashes are always found. However, bread and moustache hair are normal[2,11,14,16,17].

Nail problems occur more frequently in older individuals. About half of the affected individuals exhibit mild fingernail abnormalities and nail dystrophy. Slow nail growth and split nails are most often reported findings. Toenails were generally normal. The nail beds are more susceptible to progressive injury with age[2,11,14,16].

Most individuals repot dry skin. Affected individuals have a smooth, almost velvety skin texture. The skin of patients also seems to be thinner than expected for age[11,14,20].

Almost all affected relatives have decreased sweating, and many show heat intolerance. Some individuals only sweat in certain areas on their body. Because of the reduced number of sweat glands, there is a danger of hyperthermia. The hyperthermia may also lead to brain damage. Episodes of hyperpyrexia and severe respiratory infections are life threatening components in EDA[11,14,20,22].

Oral Structures

Missing teeth or delay in teething often starts to worry the parents and leads to the diagnosis of ED in the second year of life. A dentist should not hesitate to radiographically examine a patient whose teeth have not erupted by the appropriate age in order to exclude ED[2,14].

Mostly There is hypodontia which is known as one of the major factors of ectodermal dysplasia. In severe cases no teeth may form. More often most of the deciduous form but there are few or no permanent teeth. The teeth are usually peg shaped conical, which give an undesirable appearance. The enamel layer is thin. Enemal is rarely hypoplastic. The outer and inner enamel epithelium can not be observed in the buds from the HED fetus, but is clearly seen in the normal fetus[11,23].

Taurodontism frequently on the second deciduous molars, is a common feature. Not only the shape is abnormal, but also the number. A severe hypodontia is a universal feature among affected individuals. There are generally more teeth in the maxilla than in the mandibula. Most often the lower incisors and premolars are missing, followed by the upper premolars and incisors[11,16].

All affected individuals had sagittally underdeveloped maxillary retrusion and vertical dentoalveolar development are related to severe hypodontia.this patients had deep palatinal arch[2,21].

Also the HED oral mucosa differed from the normal specimen. The inner epithelial lining in the lower lip seemed to be intact in both the HED fetus and the specimen. In contrast, the epithelium in the palate was found to be separated from the underlying mesenchymal tissue in the HED fetus[15].

Many patients complain of dry mucous membranes in mouth and nose. Reduced salivary secretion has been spotted in some EDA patients. Autopsy has also shown absence of mucous glands in the pharynx, larynx, trachea and bronchiI. This is in agreement with the susceptibility to respiratory infections. Analysis of the saliva has revealed a reduced buffer capacity and an increased number of bacterial cultures. Most affected individuals were susceptible to dental carries[11,23-28].

Saliva, consisting of 99% water and 1% organic and inorganic components, has several important functions in the oral cavity. One of them is to modulate the oral microflora by favoring the attachment and proliferation of certain microorganisms and promoting the clearance of others. Saliva also protects the oral tissues from desiccation and exogenous insult from acids and degradative enzymes. Lactoferrin binds iron, which is an important nutrient factor for many microbial species. It also displays bacteriostatic and/or bactericidal activity towards several microbial species. Lactoferrin has been found in increased concentrations in hyposalivation. A high lactoferrin concentration in saliva can result from damage to the salivary glands, gingival inflammation or leakage of serum through inflamed mucosal membranes[29].

ED patients has reduced salivary secretion. Decreased salivary flow and alterations in salivary composition cause a clinically oral imbalance

manifested by increased caries incidence, susceptibility to oral candidiasis, xerostomia, difficulties with speech, mastication and swallowing, altered taste perception and halitosis. It has been mentioned that dentures may not be suitable for patients with hyposalivation; however, dentures could be the only restorative choice. The tongue adheres to and dislodges the denture, causing decreased retention of partial and totally removable prosthesis and resulting in abrasions, sore spots, ulceration and irritation, all unpleasant and painful experiences for the patient. A constant feeling of dry mouth is very uncomfortable and annoying. There are no universally accepted methods for the treatment of dry mouth. The aim is to relieve oral discomfort and to keep the mouth moist. A simple method is the use of water followed by saline solutions, tea and water with sodium bicarbonate. Medication capable of stimulating the salivary glands may be prescribed for certain individuals.

With the diminished salivary output xenogenic patients are more prone to caries, and therefore diligent oral hygiene and regular dental care are essential. Antibacterial mouthwashes such as 0.12% chlorhexidine are useful to inhibit the development of dental plaque and gingivitis since patients with xerostomia tend to have a greater susceptibility. The use of topical fluorides should be based on the severity of the patient's condition as well as individual caries risk. Biotene is available as a sugar-free chewing gum,an alcohol-free mouthwash and a toothpaste, while Oralbalance is available as a moisturizing gel. Multiple co-factors such as hydrogen peroxide and halides, enable the efficiency of the anti-microbial activity. The patients dont use alcohol caffein, cigarette and should use vitamin C[23-28].

The Effects of Ectodermal Dysplasia on Periodontal Tissues

The most important cause of periodontal problems of ED patients is the decreased level of saliva secretion. Saliva is an important defence factor for oral cavity. If the saliva secretion decreases, bacterial plaque and food accumulation occurs easily. Also the oxygen contain of saliva protects oral cavity against anaerob bacterial proliferation, caries and halithosis. In addition, saliva proteins and electrolytes inhibit bacterial proliferation and buffer oral acid formation. Because of all these factors, individuals become prone to periodontal diseases. Also these individuals, mobile prosthethic restoration usement become harder. Because saliva lubricates and protects oral mucosa, also increases holding capacity[23-29].

Treatment

Children with ED present many and different clinical problems from early childhood though adolescence. Clinical manifestations of HED also cause considerable social problems in affected patients. Dental treatments of the clinical traits of HED can have a profound impact on these patients. The ability to look and feel like their pers is imperative for the psychological development of these patients[2,14].

All patients should be referred for management of the oral manifestations of their HED. The aim of treatment is to determine the amount of support to patients affected by HED such as restoring missing teeth, establishing normal vertical dimensions and providing support for the facial soft tissues. Conventional prosthodontic treatment (complete dentures, overdentures or a combination of fixed and removable partial dentures) is hard because of the anatomic abnormalities of existing teeth and alveolar ridges. The conically shaped teeth and 'knife edge' alveolar ridges result in poor retention and instability of prostheses. There is usually a need to remake dentures in young patients as they grow[16,22].

Conclusions

In conclusion, when confronted with multiple dental agenesys, the clinician should look for an association of ED signs, because ED may also be detected. The major goal of dental management is to provide the patient with optimal aesthetics and function so that the patient could develop physically, emotionally, and socially like other healthy individuals. Dental management allows these patients preventive and supportive aesthetic activity, and as a result avoids social problems associated with partial or full dentures, particularly in young people.

Excellent oral hygiene is crucial for the successful treatment of these patients. Patients should use daily topical fluoride for prophylaxis against new caries attacks early placement of partial or full dentures is commonly recommended from the age of two or three years onward. The denture must be periodically modified as alveolar growth, erupting teeth and rotational jaw growth change both the alveolar,occlusal and basal dimensions. They have also a limited retention and stability. For this reason, in young children we prefer a treatment with crowns and bridge.

It is commonly agreed that osseo-integrated implants should be not placed before cessation of growth. Even in young adults, alveolar growth can

be remarkable. Impaired salivary secretion rates constitute an increased risk for dental diseases, namely caries. The fragile oral mucosa may affect the clinical situation as well as possibilities to wear removable prostheses.

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