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ECTODERMAL DYSPLASIA IN PEDIATRIC DENTISTRY: A CASE REPORT

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Summary: Providing of dental care to patients with appeared and hidden congenital malformations both single organs and body systems is presented as difficult question of practical public health. Ectodermal dysplasia - is a group of diseases characterized by the presence of abnormalities in the development of ectoderm-derived tissues (skin, hair, nails, teeth, fat and sweat glands). Clinical case of 6-years boy with congenital hypohydrotic dysplasia in pediatric dentistry practice was presented.

Keywords: ectodermal dysplasia, pediatric dentistry

Introduction. Providing of dental care to patients with appeared and hidden congenital malformations both single organs and body systems is presented as difficult question of practical public health. According to complexity and multiplicity of negative clinical situation for these type of patients they need a highly qualified dental care and co-working with related medical professionals for full treatment and rehabilitation [1].

Ectodermal dysplasia is a group of diseases characterized by the presence of abnormalities in the development of ectoderm-derived tissues (skin, hair, nails, teeth, fat and sweat glands). Usually specialists use the digital classification of ectodermal dysplasia: 1 - hair dysplasia 2 - teeth dysplasia, 3 - nail dysplasia, 4 - sweat glands dysplasia. Ectodermal dysplasias are birth defects of ectodermal origin structures (including the skin and its appendages). Ectodermal dysplasia appears as several independent forms and as a number of diseases with different clinical picture.

Ethyological factors of ectodermal dysplasia are considered to overheat during pregnancy and to dehydration of pregnant woman's body. Ectodermal dysplasia - a congenital abnormality and can be transmitted genetically. However, the disease is not progressing throughout life, affects certain areas of the epidermis and does not spread to other. In some cases ectodermal dysplasia may develop after ultraviolet light and X-rays radiation, chemical irritation. Epidemiology of ectodermal dysplasia is 1: 100 000 newborns [2].

Common clinical symptoms of ectodermal dysplasias. Usually patients with ectodermal dysplasia may not sweat or may have decreased sweating because of a lack of sweat glands or their disfunctions. Children with ectodermal dysplasia may have problem with fever control. Mild desease or illness can cause extremely high fevers, as the skin

can't sweat and body temperature control is poor. Affected adult patients have low tolerance to warm environment and need special measures to keep a normal body temperature.

Other symptoms include:

1. Abnormality of nails
2. Abnormality or missing of teeth
3. Absent or decreased tears
4. Decreased skin color (lower pigmentation)
5. Heat intolerance
6. Inability to sweat
7. Large forehead
8. Low nasal bridge
9. Hearing loss
10. Non-adequate temperature regulation
11. Poor vision
12. Thin, sparse hair
13. Thin, dry skin [3].

Most common clinical forms.

Today the next most common clinical forms of ectodermal dysplasia are known:

• Christ-Siemens-Touraine Syndrome (EDA-gene defect, K recessive): a combination of innate lack of sweat glands, partial or complete absence of teeth, hypotrichosis, bone deformities of the nose, cheilitis, and bluish skin pigmentation. Women-carriers of hidden gene during an iodine test show a special sweat glands distribution in the skin - in form of spirals or V-shaped often more pronounced on one side of the body. Synonyms: ectodermal hypohydrotic dysplasia, Christ-Siemens Syndrome, anhydrotic ectodermal dysplasia, Siemens syndrome, idiopathic multiform keratosis.

• Ectodermal Hydrotic Dysplasia (HED gene defect, R): abnormal development of the ectoderm manifested by dysplasia of epidermis and skin appendages - teeth dysplasia, rickets, cheilitis, conjunctivitis, congenital hair and nails dysplasia (nail thickening, curtailment or lack of hair on the head), often accompanied by palmo-plantar keratoderma, skin hyperpigmentation, strabismus and mental retardation; sweating is not interrupted.

Synonyms: Clouston syndrome, Naegeli-Franceschetti-Jadassohn ectodermal dysplasia, Clouston hydrotic ectodermal dysplasia.

Clouston syndrome may associate with deafness due to an extended deletions, exciting site of gene and connexin-gene HED (13ql2).

As usual ectodermal dysplasias occurs in two types:

1) above mentioned hypohydrotic dysplasia or Christ-Siemens-Touraine syndrome. Rapp-Hodgkin's syndrome with cleft lip, alveolar process, hard and soft palate is less common syndrome; hypohydrotic form may be inherited by X-linked recessive and rarely in an autosomal recessive manner. You can find in literature a description of familial cases of this disease and the development of ectodermal dysplasia occurrence for children of consanguineous marriages.

2) hydrotic dysplasia or Clouston syndrome; may be inherited in an autosomal dominant manner

Also about 20 rare form of ectodermal dysplasia (with impaired or normal sweating) are distinguised:

• Christianson-Fourie syndrome, differs from Clouston syndrome by lack of skin lesions;

• Bazan syndrome - ectodermal dysplasia without dermatoglyphic patterns of palms with nails dysplasia and four-fingered palmar crease.

• Ectodermal hypohydrotic Dysplasia combined with hypothyreoidism and corpus callosum agenesis. The main clinical symptoms: severe mental retardation, agenesis of the corpus callosum, hypohydrotic ectodermal dysplasia, primary hypothyreoidism, thyroid gland ectopia. Gormone tests show low concentrations of

T3 and T4 - gormones, increased level of thyreotropic gormone [4, 5, 6, 7, 8].

Diagnosing. For the final verification of the diagnosis for children with suspections of ectodermal dysplasia in dental practice the following clinical studies are recommended:

- an X-ray study of the jaws (defined by the presence or absence of teeth folicules);
- perspiration test (to define sweat gland presence and function; these test are not allowed for children under one year old);
- microscopy of hair (in cases of ectodermal dysplasia only pivotal part of the hair without cortex may be observed).

Treatment and care. Specific treatment for patients with ectodermal dysplasia does not currently exist. General recommendations are: optimization of microclimate, eliminating of overheating, ussig of moisturizing skin creams and immunotherapy courses for respiratory diseases prevention.

In the cases of teeth form and eruption violations dentist consultation is recommended to provide a ptothetic treatment. Different kinds of syndactyly and cleft lip and palate surgical are indications for general, plastic and maxillo-facial surgeons consultations to correct the birth defect. Patients should avoid overheating and prolonged sun exposure [1, 2, 8].

Case report. A male child 6 years age came to dental clinic for pediatric dentist with a primary diagnosis — hypohydrotic ectodermal dysplasia, primary adentia (Fig.1-3).

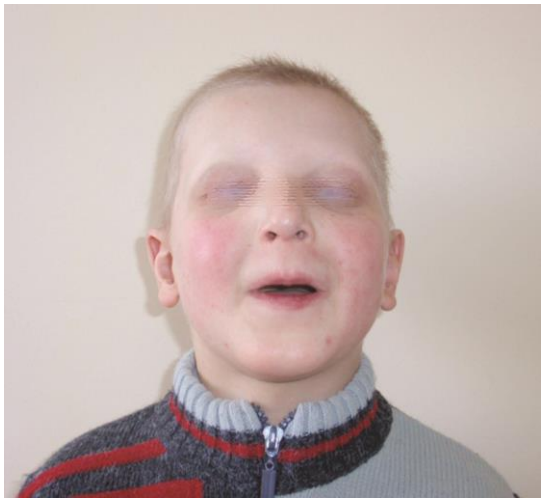

Figure 1. 6-years boy with hypohydrotic ectodermal dysplasia: face appearence

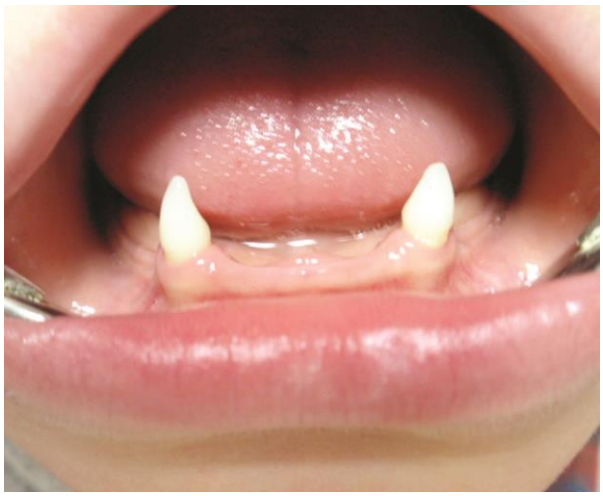

Figure 2. Primary adentia on lower jaw.

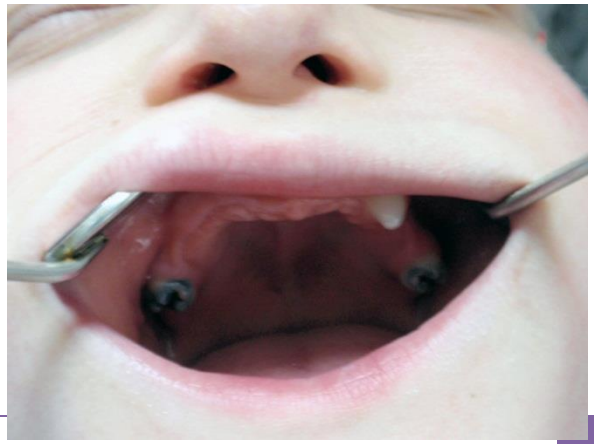

Figure 3. Primary adentia on upper jaw.

Other clinical symptoms were identified in above-mentioned patient:

1) hypoplasia of the sweat glands, which manifested by decreased sweating and the development of signs of overheating of the body during exercise and at higher ambient temperature;
2) hypotrichosis (hair covering disoder) - eyebrows and eyelashes were short, sparse and bright and were absent in some areas altogether; fur-like hair, with a weak pigmentation and slow growth;
3) partial alopecia;
4) late teeth eruption - out of sequence and timing, conical shape with caniniform tops of crowns of erupted teeth;
5) the total number of teeth was reduced to 5: 2 canine on lower jaw and 2 temporary molars and right upper canine were on upper jaw;
6) hypoplasia of the alveolar process of maxilla and alveolar part of mandible;
7) hypoplasia of the front and brain parts of the skull with the features of the "old man's face": a large forehead with a clearly protruding brow ridges and frontal mounds, small saddle nose with hypoplastic wings, sunken cheeks, blurred the boundaries of the upper and lower lips, lips were quite large and twisted;
8) ears deformity - pointed and stretched upward;
9) impaired skin structure - a thin, wrinkled and dry;
10) hypoplastic lacrimal glands - history was marked by dry eyes and conjunctivitis;
11) boy was followed up by a dermatologist about eczema;
12) patient had frequent rhinitis and acute respiratory infections (more than 6 cases per year) which may indicate a low immunity as well as the defects of the mucous glands in respiratory tract;
13) nasal passages had abundant crusting of nasal secretions;
14) blood analisys showed a decrease in hemoglobin levels, the color index of blood and signs of mild dysproteinemia;
15) patients had some mental retardation, and decreased mnemonic abilities
16) he had closed character and was and socially disoriented;
17) speech and pronunciation were disodered by primary adentia and and constant nasal congestion;
18) mild hearing loss caused by the periodic formation of cerumen in the ear canals;
19) myopia.

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