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Age-Based Assessments of Structural Changes in the Skin of Congenital Ichthyosis Disease in Hot Climate Conditions

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Abstract The relatively high level of congenital ichthyosis in the general structure of dermatological diseases, its duration, difficulties in treatment and low efficiency are one of the current problems facing medical practice and pediatricians. Based on the data presented in the scientific literature, research on morphological and morphometric changes in skin structures in congenital ichthyosis is an urgent scientific problem.

Keywords Ichthyosis, Hot climate, Morphology of skin structure, Age dependence

1. Relevance of the Research Problem

The relatively high level of congenital ichthyosis in the general structure of dermatological diseases, its duration, difficulties in treatment and low efficiency are one of the current problems facing medical practice and pediatricians. To date, there is no single agreement on the treatment of congenital ichthyosis, and there are controversies over the use of steroid hormones [3,6,24,30,37,38,42,43]. Ichthyosis remains one of the most pressing health problems. This situation necessitates the study of ichthyosis and the assessment of its epidemiological prevalence and the appointment of long-term treatment outcomes. The prevalence of different forms of ichthyosis in the population is described as follows: vulgar ichthyosis - 1: 100 [32] X chromosomal (X-linked ichthyosis) - 1: 2000 males, (females are healthy carriers of the recessive gene); autosomal recessive ichthyosis (lamellar ichthyosis - 1: 100,000; ichthyosis, which is part of the syndrome, and congenital bullous ichthyosis-shaped erythroderma - 1: 300,000 [46].

Congenital ichthyosis is 1: 10900 among children in Minsk. The prevalence of normal ichthyosis is 1: 17500, X-linked - 1:87 600 (among boys), lamellar - 1:87 600, ichthyoform erythroderma - 1: 262 800 [27,28]. According to the literature [1,21,23], the incidence of ichthyosis among children and adults depends on the geographical area of congenital disease (for example, in the UK - 1: 230, in the US - 1: 300, in Russia - 1: 9600) and on average 1: 250 - 1: 1000, X-linked - 1: 2000 - 1: 6000, lamellar - less than 1: 300 000, ichthyoziform erythroderma - 1: 100 000 [20,10]. According to world statistics, 1 in 30,000 newborns is born with ichthyosis.

2. Objective

To reveal the age-related aspects of the morphology of structural changes in the skin in congenital ichthyosis in hot climates and to determine the ways of solving this problem.

The relatively high incidence of ichthyosiform keratoderma in the general structure of dermatological diseases, their severity, systemic nature of lesions, difficulties in treatment and low efficiency lead to the problem of timely and accurate diagnosis of these diseases and require symptomatic and systematic approach throughout life [7,11]. There are many publications in the local and foreign literature devoted to the study of morphological changes in the keratoderm, in particular ichthyosis [2,13,14,15,17,31,37,38,39,42,47]. In particular, it is noted that the morphological examination of the pathological process in the skin in ichthyosis is one of the objective methods based on evidence-based medicine [9,11].

To date, there is no complete and systematic approach to the morphological diagnosis of ichthyosis. The role of proliferative activity of epidermal cells in the development of hereditary dermatoses remains an unresolved problem and there is no specific pathoanatomical criterion for the

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diagnosis. The proliferation of keratinocytes and other cells of the epidermis, the mechanisms of interaction between cells of the immune system, and the proliferative activity of epithelial cells have not yet been adequately studied. Therefore, the study of specific pathomorphological criteria that allow to determine the state of the epidermis in one form or another of ichthyosis is one of the current problems [2,11,17].

According to the literature, the hearing of keratinized scales in which, with ichthyosis, increases with age and reaches a maximum at a late age, 20-25 years, as a result of migration of the substance develops puncture atrophy, and in adult patients it is more pronounced the back of the hands, the appearance of small wrinkles in the area of flexion, the surfaces of the limbs, cracks in the soles, and hyperkeratosis [19], however, age-related morphological changes in skin structures were not reported in these studies. The authors state that there is a tendency for periodic peeling of the skin in patients with congenital ichthyosis - once every 3-4 months, often in the summer, during bathing, and then again the formation of horn horns again. In ichthyosis, the skin usually changes depending on climatic conditions. In cold and dry weather, the disease intensifies. [29].

It should be noted that morphological changes due to climatic conditions are not sufficiently covered in the literature.

In recent years, the incidence of fetal abnormalities has increased, including fetal developmental delays, abnormal structures of fetal organs, and births with anomalies. In the structure of perinatal pathology, the birth of a fetus with congenital malformations occupies one of the leading positions every year. Statistics show that in Russia, 35% of children are born with congenital pathologies. One of the many congenital defects is ichthyosis. The origin of this disease depends on many factors and occurs in newborns in a ratio of 1: 250 - 1: 1000 [10,20]. Ichthyosis is a pathological process with synonyms such as diffuse keratoma, hereditary dermatosis, diffuse disorder of the "fish coin" type of frostbite, which is an inherited disease that is passed from generation to generation by autosomal recessive and autosomal dominant [18,25,45]. The etiology and pathogenesis of ichthyosis have not been adequately studied. However, there are opinions that a decrease in the concentration of vitamins A and E plays an important role in the development of this disease. A number of authors [20,25] have suggested that as a result of impaired protein metabolism, the accumulation of amino acids in the skin and an increase in cholesterol levels lead to an increase in the total weight of the fetus. Immune syndrome in ichthyosis is a form of ichthyosis [40,41]. This syndrome is characterized by premature birth, neonatal asphyxia, and ichthyosis. After the birth of ichthyosis, the disease progresses for several weeks and persists for a lifetime, the disease often takes the form of atopic dermatitis and recurrent infection. The patient's epidermis thickens, hyperkeratosis and droplets are detected in the stratum corneum.

In addition, the corneal and granular layers of the patients will have multilayered membranes in a curved position [33,34,35,41].

Diagnosis of ichthyosis is somewhat difficult during the antenatal period. During amniocentesis for analysis of amniotic fluid during pregnancy, as well as material for biopsy examination from the chorionic villus sampling can be obtained. However, such invasive methods increase the risk of early miscarriage of the fetus. Therefore, these methods are rarely used in practice. Perinatal biopsy results indicate that the fetus has ichthyosis before birth. UTT can detect fetal ichthyosis in the third trimester of pregnancy. The location of the fetal arms of ichthyosis, the openness of its mouth, can be obtained by some information through hypoplasia of the nasal ridges. Pregnancy can be detected by increased fertility and exogenous amniotic fluid. However, none of these signs are specific. The presence of ichthyosis can only be suspected [5,26,27,28,30]. Diagnosis of ichthyosis is not difficult, as all manifestations of the disease are visible after the birth of the baby. This is a special sign of desquamation of the skin, the covering of the skin with gray-looking coins. The oral cavity is not opened, the lips are wrinkled, sclerotic changes in the arms and legs are detected [4,5,18]. Changes in the analysis conducted by some experts nonspecific changes increase in cholesterol, decrease in esterol are important in the early diagnosis of ichthyosis [12].

The diagnosis of ichthyosis is based on clinical signs. In suspicious cases, it is based on histological examination. Diagnosis in newborns is difficult because it must be distinguished from Leiner-Mussu's descriptive erythroderma and Ritter's exfoliative dermatitis. In recent years, molecular-genetic methods to identify disease form, medical-genetic prognosis and gene defects have been widely used [16,44]. To date, more than 50 forms and types of hereditary ichthyosis have been identified through the use of genetic research methods [21].

There are many publications in the local and foreign literature devoted to the study of morphological changes in keratoderm, in particular ichthyosis [2,13,14,15,17,31,39, 43,47]. The researchers performed a pathomorphological analysis of patients with vulgar ichthyosis and ichthyoziform erythroderma after skin biopsy after treatment with retinoids, and recommended that it be divided into two types according to pathomorphological changes: mild and severe. Mild form of ichthyosis is characterized by the following pathological signs in vulgar and ichthyosiform erythroderma: a decrease in the number of granular keratinocytes or the absence of a granular layer, moderate hyperkeratosis, intact corneal structure, focal acanthosis. The following pathohistological signs are characteristic of a severe form of ichthyosis: hyperkeratosis combined with acantholysis, granular degeneration of epidermal cells (acanthokeratolysis), formation of bosaphyl and eosinophil bodies in the granular layer, development of vacuolar degeneration in all layers of epidermis, spongiosis, parakeratosis, parakeratosis. Histochemical examination revealed focal alcyanophilia in the collagen fibers of the stratum corneum and dermis [11]. A characteristic feature of all types of ichthyosis is the

persistence of lifelong skin problems, suffering in severe cases, as well as the influence of family members and relatives on the patient [36].

3. Conclusions

Based on the above data, research on morphological and morphometric changes in skin structures in congenital ichthyosis is an urgent scientific problem.

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