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საქართველოს სამედიცინო სიახლენი

NEW APPROACHES IN THE DIAGNOSTIC OF ICHTHYOSIS DISORDERS

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The lack of a holistic conception of the pathogenesis of ichthyosis complicates the development of effective and reliable treatment methods, making it impossible to predict the clinical course of this dermatosis. For the diagnosis of ichthyosis, it is necessary to use a complex of morphological and laboratory methods of research, which differ in accuracy of results and complexity of implementation. Today known methods of diagnosing macroscopic skin changes in dermatosis have a number of shortcomings [2,3,5-7]. Firstly, the offered dermatological criteria are meant mainly for diagnosis of melanoma. Secondly, they do not evaluate the severity of the disease. Thirdly, offered dermatological criteria for non-tumor skin diseases remain at the level of treatment concepts and are not standardized.

The generally accepted clinical examination of the patient is based on the subjective perception of the visible signs of skin lesions by a doctor, that can lead to diagnostic errors. Therefore, the development of new methods for ichthyosis diagnosis is urgent, which will facilitate the verification of the diagnosis and accordingly improve the quality of treatment. One of the modern and available methods of diagnosis in dermatology is dermatoscopy, which is essentially epiluminescent microscopy with the study of skin microdermabrases (Ø 25 mm) to study skin dyschemia. As our experience shows, the use of dermatoscopy does not require special preparation on the part of the patient, has no contraindications. One of the advantages of the dermatoscopy method in the diagnosis of ichthyosis is the ability to clearly distinguish the lesion type, with its subsequent fixation on digital carriers. It allows to analyze the received images, to get consultation from other specialists, and also provides possibility of objective dynamic observation in the background of treatment.

Despite the existing clinical and laboratory data regarding the etiology and pathogenesis of ichthyosis, the achievement of modern therapies, some aspects of the development of this pathology remain poorly understood and the research data are controversial, which inhibits the development of promising methods for diagnosis and especially treatment of this serious disease. That is why the purpose of our research was to study the macroscopic changes in the skin of patients with various forms of ichthyosis in order to improve and optimize its treatment.

Material and methods. A comprehensive examination of 12 patients with ichthyosis under the supervision of a dermatovenereologist was conducted during the research. The study included cases that were verified in Vinnytsya Regional Dermatovenerologic Dispensary. Among them there were 8 men and 4 women. The patient's age ranged from 11 to 55 years. All patients received treatment in accordance with the clinical protocols for the provision of medical care approved by the Ministry of Health of Ukraine.

For the diagnosis of macroscopic skin changes, the dermatoscopy was performed with the device HEINE DELTA 20®, (Germany) the results were recorded with a Canon 550D camera (Japan) at 1:35 or 1:30, at a sensitivity of IZO 200 using an adapter attached to the dermatoscope. Apart from the dermatoscopy all patients have undergone standard clinical and laboratory examinations. All affected skin areas were examined and compared with unharmed ones. The condition of the skin vessels was separately determined. General-clinical examination and a pathomorphological study were conducted as well.

Results and their discussion. The task was performed with the help of our algorithm for dermatoscopic evaluation of the severity of ichthyosis (Table) [1]. As a result, during dermatological examination in mild form of ichthyosis, dry skin, uneven or follicular hyperkeratosis, fine-plate peeling with the presence of dirty-gray scales, atrophy of hair follicles and hair loss, and also characteristic changes in smoothness were observed, there was a pseudo-pixel mesh formed by keratin in the form of scales, in the form of stripes, mesh and loop structures resembling cerebrium-shaped structures. The vascular pattern was presented by the same type of elongated or short capillaries of narrow diameter without branches, and a small amount of helical capillaries was noted. Few spotted capillaries were in the form of dots (Fig. 1), ("sparkling champagne").

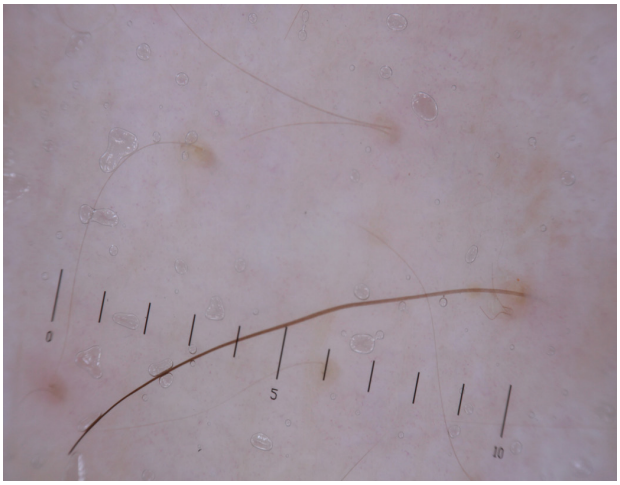

Fig. 1. Pseudo-pixel mesh, nets and loop-shaped structures, dotted capillaries in the form of dots ("sparkling champagne"). Patient P. Mild form of ichthyosis. Dermatoscopy

Detected dermatological signs are clearly defined during examination and are reproduced when fixing on digital carriers. We offer specific dermatoscopic criteria for ichthyosis: this is a "splash of champagne", a rather stable pathognomonic sign of the capillaries lesions on the background of hyperkeratosis, characteristic of ichthyosis. We can explain

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Table. Algorithm for estimating basic dermatological criteria of ichthyosis skin lesions

The severity of the disease	Dermatoscopy criteria
Mild form	Skin pattern is smoothed **- 1 point.** Pseudo-pixel mesh formed by keratin of scales, in the form of bands, mesh and looped structures resembling cerebroform structures - **1 point.** The vascular pattern is represented by the same type of elongated or short capillaries of narrow diameter without branches - **1 point.** A small number of helical capillaries is **1 point.** Rarely spotted capillaries in the form of dots ("splashes of champagne") - **1 point.** **Score from 1 to 5 points.**
Severe form	Skin pattern is of atypical form - in the form of massive cubic structures - **2 points.** Expressed hyperkeratosis in the form of massive multilayer crust and conglomerates, separated by cracks in the stratum corneum of the epidermis - a symptom of "melting ice" - **3 points.** Massive horny crust - **2 points.** Atrophy of salivary hair follicles - **3 points.** Hair on smooth skin is much weakened and deformed or absent - **2 points.** The vascular pattern is depleted, predominantly represented by the same type of "splash of champagne" - **3 points.** **Score from 6 points and up.**

its appearance by the presence of lesions in all epidermis layers, which visually manifests itself at a multiple magnification possible only with a dermatoscopia examination.

In contrast to the mild form in patients with severe forms of ichthyosis, the macroscopic skin pattern was characterized by manifestations of marked hyperkeratosis, follicular areas with massive dirty-gray scales and cracks, sometimes even deep ones. For the severe form of ichthyosis, specific dermatoscopic signs were also characteristic: skin pattern is atypical in the form of massive cubic structures, expressed hyperkeratosis in the form of massive multilayer crust and conglomerates, separated by cracks in the stratum corneum of the epidermis - a symptom of "melting ice" (Fig. 2), presence of massive cornea and atrophy of the salivary hair follicles.

Fig. 2. Symptom of "melting ice". Severe form in the course of ichthyosis. Patient D. Dermatoscopy

The symptom of "melting ice" offered by us to describe dermatoscopic signs of severe forms of ichthyosis, points to deep disorders of keratinization, which are characteristic of this disease and are associated with severe course of illness. In our opinion this symptom suggests the need for more active therapeutic intervention, it is possible to use systemic keratinides to improve the functioning of keratinocytes and to normalize keratinization processes.

Thus, with the application of the above mentioned method a dermatopoietic evaluation can be clearly defined at the initial phase of ichthyosis as well as the degree of severity.

Clinical Example 1. Patient P, born in 1997, residing in Gorodokivka, Kryzhopilsky district, addressed Vinnytsya Regional Dermatovenerologic Dispensary with the following complaints: on the background of severe dry skin, a large number of dirty-gray colored scales, crins, and cracks were noted. After clinical examination the given diagnosis was - Ichthyosis.

After a dermatoscopic examination with a dermatoscope with fixation of detected macroscopic changes in the skin (Fig. 1) and the use of the algorithm for assessing the basic dermatoscopic criteria for skin lesions with ichthyosis (Table), the symptoms namely are:

1. Skin pattern is smoothed - 1 point.
2. Pseudo-pixel mesh formed by keratinum scales in the form of bands, mesh and loop structures resembling cerebroform structures - 1 point.
3. The vascular pattern is represented by the same type of elongated or short capillaries of narrow diameter without branches - 1 point.
4. A small number of helical capillaries - 1 point.

Total number of points - 4.

The diagnosis is: Vulgar Ichthyosis of mild severity, appropriate treatment prescribed after which the patient's condition improved.

Clinical Example 2: Patient D, born in 1976, residing in Bar, Vinnytsya Oblast, addressed Vinnytsya Regional Dermatovenerologic Dispensary with complaints

of severe inflammation, especially in the folds of the skin, blisters in the large folds, surface erosion on areas of skin peeling, bad breath in skin rejection, thinning of hair on the scalp, runny nose, skin lesions in the palms and feet in the form of peeling stratum corneum with the formation of cracks located along the flexural folds. After the clinical examination, the diagnosis is established: Ichthyosis.

After dermatoscopic examination of the skin using the algorithm of dermatoscopic evaluation of the severity of ichthyosis using a dermatoscope (Fig. 2) and the use of the algorithm (Table), the symptoms namely are:
1. Skin pattern of atypical form - in the form of massive cubic structures - 2 points.
2. Expressed hyperkeratosis in the form of massive multilayer crust and conglomerates, separated by cracks in the stratum corneum of the epidermis - a symptom of "melting ice" - 3 points.
3. Massive horny crust - 2 points.
4. Atrophy of salivary hair follicles - 3 points.
5. Hair on smooth skin is much weakened and deformed or absent - 2 points.
6. The vascular pattern depleted, predominantly represented by the same type of "splash of champagne" - 3 points.
The total score is 15.

Final diagnosis: Bullous congenital ichthyozyme erythroderma, severe form. The patient is prescribed an appropriate treatment with repeated dermatoscopic monitoring throughout the year.

The revealed criteria for the severity of the course of ichthyosis in our opinion will allow standardization of groups of patients who have the same diagnosis but different course and require a specific approach to treatment, as the therapy of this disease also remains not enough researched and developed, that significantly affects the presence of complications, deterioration of life quality. For ichthyosis, as for chronic dermatosis, an extremely important task of treatment is the control of the disease, minimizing complications [8]. By reviewing current approaches to therapy, researchers point out [4] that existing local treatment methods for ichthyosis can only control and somewhat reduce some of the local skin manifestations. Consequently, there is a need to strike a balance between the effectiveness of treatment and the patient's commitment to the chosen therapy program. In our study, specific dermatoscopic signs are established on the background of different forms of ichthyosis, allowing to separate the mild and severe forms of ichthyosis, which greatly improves and optimizes treatment.

Conclusions. The offered method in comparison with other known methods of diagnosis of various ichthyosis types is precise, easy to use, allows to determine the state of the epithelium namely skin and vascular patterns and timely to diagnose ichthyosis in the early stages, to evaluate the degree of its severity, to shorten the time of examination of patients and to prescribe the appropriate treatment.

Dermatoscopy is a promising method for diagnosis of ichthyosis and control of the course of the disease.

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SUMMARY

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The aim of this study was to study the macroscopic changes in the skin of patients with various forms of ichthyosis to improve and optimize treatment.

A comprehensive examination of 12 patients with ichthyosis under the supervision of a dermatovenereologist was conducted at VOKSHVD. For the diagnosis of macroscopic skin changes, the dermatoscopy was performed

by the HEINE DELTA 20 ® (Germany) apparatus, the results were recorded with a Canon 550D camera (Japan) at 1:35 or 1:30, with a sensitivity of IZO 200 using an adapter included with the dermatoscope. All affected skin were examined, which were compared with unharmed ones.

The task was performed using the algorithm developed by us for dermatoscopic assessment of the severity of ichthyosis. The presence of typical dermatoscopic signs in examined patients with ichthyosis, which is manifested by the smoothness of the skin pattern, the presence of a pseudo-pigmental mesh, formed by keratin of scales in the form of lines, mesh and loop structures resembling cerebriform components, are established. The algorithm of estimation of expressiveness of dermatoscopic features for patients with light and severe clinical course of ichthyosis is proposed. Also, specific dermatological signs were observed in the mild clinical course of ichthyosis, in particular the phenomenon of «sparkling champagne», and in severe clinical course, the «melting ice» phenomal.

The use of the dermatoscopic method makes it possible to objectively evaluate the severity of the ichthyosis and apply a differentiated approach to choosing the most effective treatment schemes.

Keywords: ichthyosis, macroscopic changes, dermatoscopy.

РЕЗЮМЕ

НОВЫЕ ПОДХОДЫ К ДИАГНОСТИКЕ СТЕПЕНИ ТЯЖЕСТИ ИХТИОЗА

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Целью данного исследования явилась оптимизация лечения больных различными формами ихтиоза на основании изучения макроскопических изменений кожи.

Комплексно обследованы 12 больных ихтиозом. Для диагностики макроскопических изменений кожи проводилась дерматоскопия аппаратом HEINE DELTA 20® (Германия), результаты фиксировались фотоаппаратом Canon 550D (Япония) при выдержке 1:35 или 1:30 и чувствительности IZO 200 с помощью адаптера, входящего в комплект дерматоскопа. Исследовали все пораженные участки кожи, которые сравнивали с неповрежденными.

Оценка тяжести ихтиоза осуществлялась с использованием разработанного авторами алгоритма. У обследованных больных установлены типичные дерматоскопические признаки ихтиоза, проявляющиеся сглаженностью кожного рисунка, наличием псевдо-пигментной сетки, сформировавшейся кератином чешуек в виде линий, сетки и петлистых структур, напоминающих церебриформные компоненты. Для больных с легким и тяжелым клиническим течением ихтиоза предложен алгоритм оценки выраженности дерматоскопических признаков. Выявлены специфические дерматоскопические признаки легкого клинического течения ихтиоза, в частности, феномен «брызги шампанского», а для тяжелого клинического течения - «тающего льда».

Предложенный метод позволяет объективно оценить тяжесть ихтиоза и применить дифференцированный подход к выбору наиболее эффективных схем лечения.

რეზიუმე

იხტიოზის სიმძიმის ხარისხის დადგენისადმი ახალი მიდგომები

[1]ს. დმიტრიენკო, [2]მ. შინკარუკი-დიკოვიცკაია, [3]ს. ვერნიგოროდსკი, [4]ტ. ტეპლა

მ. პიროგოვის სახ. ვინიცის ეროვნული სამედიცინო უნივერსიტეტი, [1]კანისა და ვენდაავადებათა კათედრა; [2]თერაპიული სტომატოლოგიის კათედრა; პათოლოგიური ანატომიის, სასამართლო მედიცინის და სამართალის კათედრა, უკრაინა

კვლევის მიზანს წარმოადგენდა მკურნალობის ოპტიმიზაციისათვის იხტიოზის სხვადასხვა ფორმით დაავადებული პირების კანის მაკროსკოპული ცვლილებების დადგენა.

კომპლექსურად შესწავლილია იხტიოზით დაავადებული 12 პირი. კანის მაკროსკოპული დიაგნოსტიკა ჩაატარდა HEINE DELTA 20® (გერმანია) აპარატით,შედეგები დაფიქსირდა ფოტოაპარატით Canon 550D (იაპონია) 1:35 ან 1:30 გამძლეობით და IZO 200 მგრძნობელობის ადაპტერით. გამოკვლეულია კანის ყველა დაზიანებული ნაწილები, შემდგომში მოხდა მათი შედარება ჯანმრთელ კანთან. იხტიოზის სიმძიმის შეფასება ხდებოდა ავტორების მიერ მოწოდებული ალგორითმის მეშვეობით. გამოკვლეულ ავადმყოფებში გამოვლინდა იხტიოზის ტიპიური დერმატოლოგიური ნიშნები, რომლებიც გამოხატული იყო კანის ზედაპირის სიგლუვით, ცერებროფორმული კომპონენტის მაგვარი კერატინული ქერცლის ხაზებისაგან და მარყუჟებისაგან შემდგარი ფსევდო-პიგმენტური ბადით. მსუბუქი და მძიმე კლინიკური მდგომარეობის იხტიოზის დერმატოლოგიური ნიშნების გამოხატვის შეფასებისთვის შემოთავაზებული ალგორითმის მეშვეობით გამოვლენილია მსუბუქი კლინიკური მიმდინარეობის სპეციფიური დერმატოლოგიური ნიშნები,კერძოდ,ე.წ. "შამპანიურის შხეფები", ხოლო მძიმე კლინიკური მიმდინარეობისთვის - "ნადი ყინულის ფენომენი".

შემოთავაზებული მეთოდი საშუალებას იძლევა ობიექტურად შეფასდეს იხტიოზის სიმძიმე და გამოყენებული იქნას დიფერენცირებული მიდგომა მკურანლობის ეფექტური სქემების შერჩევისათვის.