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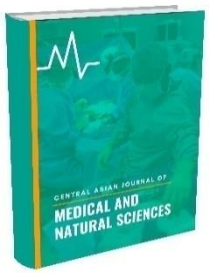

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A Clinical Observation of Epidermal Nevus

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Abstract: Some clinical cases of generalized verrucous epidermal nevus are difficult to diagnose. Hence, we demonstrate the effectiveness of using a diagnostic algorithm to identify generalized verrucous epidermal nevus for a case report from our own clinical practice. The patient had congenital skin rash that was not characterized by spreading or spontaneous involution. It bilaterally affected skin of the head, neck, body and flexor surface of extremities along the lines of Blaschko as light or dark-brown wart-like structures shaped like stripes and curls resembling 'mud splashes'. Laboratory testing revealed no significant deviation from the norm. Consultations with experts in adjacent disciplines showed no concomitant pathologies. Histological examination of a bioptate collected from the pathological focus revealed hyperkeratosis, papillomatosis, moderate acanthosis, the non-uniform granular layer, and scarce perivascular lymphohistocytic infiltrates (ie, changes typical of verrucous epidermal nevus). Differential morphological diagnosis showed that the patient did not have the Bloch — Sulzberger syndrome or other diseases manifesting as papillomatous eruptions. Thus, the proposed diagnostic algorithm included allowance for anamnestic and clinical examination data, laboratory testing, meticulous morphological analysis of the histologic patterns during examination of biopsy specimens, and differential diagnosis along with consultations with experts in the adjacent fields. The algorithm proved to be efficient for identifying the dermatologic pathology.

Keywords: nevoid neoplasms, epidermal nevi, systematized epidermal nevus, therapeutic strategy, treatment, prognosis.

Epidermal nevi are benign hamartomas that develop from the ectoderm. They can serve as one of the manifestations of epidermal nevus syndrome, i.e. be associated with cardiovascular anomalies, nervous, genitourinary system or musculoskeletal system [1-3]. Epidermal nevi are often

congenital, but can occur throughout life (acquired). After emergence, they usually do not show a tendency to spread and rarely undergo spontaneous involution [4, 5]. At the site of the nevus, the development of benign and malignant tumors of the epidermis and its appendages is possible [6]. According to epidemiological data, epidermal nevi equally often affect both sexes with a frequency of 1: 1000. Usually, cases of this pathology are sporadic and rarely occur in members of the same family [6]. Most often, epidermal nevi are localized on the flexion surfaces of the arms and legs, where they are located longitudinally (usually linearly) along the course of nerves, large vessels, and embryonic lines of skin closure [6, 7]. Sometimes they occupy an extensive surface of the body, and on the body they have a transverse direction. Scalp nevi are usually smooth and hairless. There are nevi on the mucous membranes of the oropharynx, genitals, etc. [2, 6]. The surface of a warty nevus largely depends on its histological type and localization. So, the epidermolytic type of nevus resembles a vulgar wart with an erythematous base. Nevi on the palms and soles also have a warty surface, on the bends of the limbs - velvety and macerated. The elements are usually asymptomatic. Only when localized in the periungual region, they can cause recurrent paronychia and splitting of the nail plate. The warty surface of many epidermal nevi is caused by the presence of hyperkeratosis and papillomatosis. Initially, they are colorless, slightly scaly spots that thicken with age, become warty and hyperpigmented. Clinically, these are dense, clearly demarcated warty growths of a dirty gray or slightly brownish color (hard epidermal nevus) [5]. Histologically, the verrucous nevus is manifested by orthohyperkeratosis, papillomatosis, acanthosis with lengthening of the epidermal processes, consisting of highly differentiated spinous cells with a pronounced basal layer and uneven expression of the granular layer, sometimes parakeratosis. In some cases, against this background, the structures of the skin appendages are determined: sebaceous, sweat glands, hair follicles and others [6, 8]. There are two main types of features of the histological structure of epidermal nevi. The first type includes solitary and widespread nevi, characterized by a noticeable compact hyperkeratosis, thickening of the granular layer, moderate or severe acanthosis with lengthening of the papillae of the dermis, as well as papillomatosis of varying severity; histologically, many of these lesions resemble seborrheic keratosis. The second type includes systemic epidermal nevi with the presence of "granular degeneration" of cells in the upper and middle sections of the Malpighian layer of the epidermis and thickening of the granular layer. Similar changes are found in extrabullous areas with congenital bullous ichthyosiform erythroderma. It should also be borne in mind that the histological picture of individual epidermal nevi, as well as their clinical manifestations, may change as the elements mature [6]. Spontaneous involution is rare. The appearance of nodules or ulcers on the surface of a nevus usually indicates the development of tumors, such as basal cell carcinoma, Bowen's disease, or squamous cell carcinoma. At the same time, malignant transformation of a warty nevus occurs less frequently than a sebaceous or apocrine nevus. A verrucous epidermal nevus can be associated with a melanocytic nevus, a sebaceous gland nevus, as well as diseases of the central nervous system, epilepsy. Some clinical cases of widespread, and especially systemic epidermal nevus present diagnostic difficulties. In this regard, we present a description of our own observation. Patient I., 22 years old, turned to a dermatovenerologic dispensary with complaints of rashes, located almost throughout the skin. The patient noted that he had rashes from birth and they periodically undergo inflammation, sometimes with the presence of mucus-like secretions on the surface. The pathological process on the skin was widespread. The rashes were located on both sides along the Blaschko lines on the scalp, neck, trunk, flexor surfaces of the arms and legs in the form of light and dark brown warty formations in the form of stripes, curls, in places resembling "splashes of mud". On the body, they were in the form of transverse stripes that did not cross the midline of the body; in the lateral part of the body, the rash was S-shaped; on the limbs - linear and vertical. Based on the history and clinical data, the patient was preliminarily diagnosed with a verrucous epidermal nevus, but due to the presence of hyperpigmented papillomatous rashes on the trunk in the form of "swirls" and "mud splashes" papuloverrucous stage of Bloch-Sulzberger syndrome.

As you know, Bloch-Sulzberger syndrome (pigment incontinence) is a systemic ectomesodermal disease, inherited dominantly, X-linked with a lethal effect for male embryos. In 90-95% women are sick, and the disease of men is regarded as a result of spontaneous mutation. The clinical picture of Bloch-Sulzberger syndrome depends on the stage of the process. From birth or in the first days / weeks of life (less often), erythematous vesicular, papulovesicular rashes occur, located mainly on the lateral surfaces of the trunk and proximal extremities with a tendency to a strip-like arrangement (stage I). Eosinophilia is detected in the blood. Then some of the elements acquire a verrucous character (stage II). After regression of rashes (stage III), pigmentation remains in the form of characteristic "splashes", "swirls" and stripes. Gradually, after 15-20 years, hyperpigmentation is replaced by mild atrophy, sclerosis and depigmentation (stage IV). The staging of the disease is sometimes poorly expressed, at the same time there may be bullous, papular and pigmented foci. Often stage III appears without prior symptoms. This may be in cases where stages I and II have passed in the prenatal period or, due to the worn out symptoms, have gone unnoticed. In addition to skin changes, most patients have various ecto- and mesodermal defects: dental anomalies, hypotrichosis, nail dystrophy, changes in the eyes, skeleton, and central nervous system [9]. For differential diagnosis, the patient underwent clinical and laboratory examination. Dysmorphic changes, as well as asymmetry of the face, trunk and extremities, disturbances in the development of hair, nails, skull bones were not revealed during examination. Family history is not burdened. The patient was followed up by a neurologist for a long time with a diagnosis of post-traumatic encephalopathy. Somatogenically caused depressive disorder. " General blood and urine tests, blood biochemical parameters were within normal limits. The patient was consulted by related specialists (therapist, ophthalmologist, surgeon, orthopedist, neurologist, dentist), as a result of which the pathology of the internal organs and the skeleton was not identified. With preliminary diagnoses “Verrucous nevus? Bloch-Sulzberger Syndrome? Acanthosis nigricans? " the patient was referred for biopsy. The results of a histological examination of a biopsy specimen from a pathological focus located on the skin of the abdomen: “Hyperkeratosis. Follicular hyperkeratosis. Papillomatosis. Moderately pronounced acanthosis. The granular layer is uneven. The content of melanin in the basal layer of the epidermis is increased. In the upper dermis there are scanty perivascular lymphohistiocytic infiltrates. Nevus cells are missing. " Conclusion The morphological picture corresponds to the warty epidermal nevus. On the basis of a clinical examination and data from a histological examination of the skin, the patient was diagnosed with “Bilateral verrucous epidermal nevus, non-epidermolytic subtype”.

Thus, clinically in favor of a verrucous nevus was evidenced by the absence of staging of the process, since the rashes from the very beginning were papillomatous in nature with no transition to the hyperpigmented and atrophic stages, which is characteristic of Bloch-Sulzberger sidrome. Eosinophilia was not observed in the blood. It was important that our patient was male, and with pigment incontinence, almost all boys are not viable. Clinical examination and consultations of related specialists made it possible to exclude the association of skin rashes with anomalies in the development of the cardiovascular, nervous, genitourinary system or the musculoskeletal system, i.e. manifestations of epidermal nevus syndrome. The histological picture of verrucous epidermal nevus and Bloch-Sulzberger syndrome also has a number of differences. Since the patient had hyperpigmented rashes of a verrucous nature on the skin, it made sense to carry out morphological differential diagnosis with stage II of pigment incontinence, which is characterized by acanthosis, irregularly - papillomatosis and hyperkeratosis, the presence of numerous dyskeratotic cells, and in the basal layer of the epidermis - vacuolization of cells and a decrease the content of melanin in them. In the dermis, a moderately pronounced chronic inflammatory infiltrate with a small number of melanophages, penetrating into the epidermis, is determined [10]. Due to the fact that our patient was morphologically absent in the epidermis phenomena of dyskeratosis, pigment incontinence and vacuolization of basal cells, and in the papillary layer of the dermis there was practically no

inflammatory infiltrate with an admixture of melanophages, we excluded the presence of Bloch-Sulzberger syndrome. In differential diagnostic terms, it was important to exclude diseases also characterized by papillomatosis, acanthosis and hyperkeratosis. In this regard, we were guided by the indications from the fundamental works [6, 8, 10] that suprabasal acantholysis, atypia and discomplexation of epidermal cells and uneven proliferation of the epidermis downward (unlike actinic keratosis) are morphologically uncharacteristic for the analyzed pathology; the presence of increased proliferation of basal cells, as well as horny pseudocysts (unlike basal cell papilloma) is uncharacteristic; the presence of large vacuolated cells in the granular layer of the epidermis (unlike the common wart) is uncharacteristic, as well as the predominance of atrophy of the epidermal processes over their elongation (unlike Acanthosis nigricans). The differential morphological diagnostics carried out by us made it possible to exclude diseases with the named triad of symptoms that characterize "papillomas", and to come to the conclusion that the patient has a verrucous epidermal nevus. The presented clinical case deserves attention not only because of the rarity of this pathology, but also due to the fact that the pathological process in the observed patient was widespread. In addition, recurrent exacerbations in the patient in the area of verrucous eruptions increase the risk of malignancy of the nevoid process. In this regard, the patient was given recommendations for dispensary observation by a dermatologist at the place of residence. Thus, the diagnostic algorithm built by us, including taking into account anamnestic data, clinical examination results, laboratory data, conclusions of related specialists, as well as a differential diagnostic search, along with a thorough morphological analysis of histological pictures in the study of biopsy material, was the key to success in recognizing the widespread verrucous epidermal nevus.

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