GENETICS AND GENODERMATOSES

CLINICAL CASE OF ECTODERMAL GENODERMATOSIS - THE BLOCH-SULZBERGER SYNDROME

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Background: Bloch-Sulzberger Syndrome (pigment incontinence syndrome) is a genetically determined disease. It is characterized by the staged course of skin manifestations, as well as the damage to organs and tissues that are of ectodermal origin (central nervous system, the visual analyzer, appendages of the skin).

Observation: A female child was under our supervision, who immediately after birth was marked by numerous vesicle-pustular elements of the rash on the erythematous background with a strained tire. The efflorescences were located linearly along the lines of Blaschko on the skin of the lower, upper limbs and trunk. On the 4th month, the appearance of papular elements with a verruxial surface was noted on the site of the former eruptions.
From an anamnesis - the child is from the second pregnancy with gestosis in the late term, family history is without features. The Bloch-Sulzberger syndrome was diagnosed based on the dynamics of the clinical picture. A diagnosis of cystic degeneration of the frontal lobes, dysgenesis of the corpus callosum was established according to the magnetic resonance imaging of the brain.
Currently, the girl is 1.5 years old. At the moment, on the skin of the lateral surfaces of the trunk, upper and lower extremities, more in the region of the thighs and lower legs, there are hyperpigmented spots in the form of bands, curls, "mud splatters". Neurological diagnosis at present is spastic tetraparesis with a rough delay of psychomotor and speech development. At examination at the ophthalmologist is diagnosed a complex hypermetropic astigmatism of both eyes. She receives intermittent therapy courses from a neurologist, rehabilitation therapy.

Key message: Timely detection of clinical manifestations of the Bloch-Sulzberger syndrome and differential diagnosis are an important stage for conducting additional methods of investigating a possible multi-organ pathology.