

Congenital Giant Pigmented Nevi

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Short Commentary

Congenital melanocytic nevus – represents a benign proliferation of dermal melanocytes, which clinically is observed at birth or appears during the first weeks of life. Congenital nevi vary in size, macroscopic manifestations and histological characteristics [1], they appearing in about 1% of newborns. Most nevi are characterized by small size, but there are large (LCN) and giant congenital nevi (GCN) as well, which have a large area, and sometimes extend to the entire segment. GCN are relatively infrequent congenital malformation which under macroscopic study typically characterized by intense pigmentation and often can have a differently expressed hair cover [2].

Histological studies show that the GCN are characterized by an abundance of not very large and medium-sized "nevus cells" (melanocytes), with a relatively small amount of weakly oxyphilic cytoplasm, moderately basophilic nuclei, mostly having incorrect ovoid shape. In the upper layers of the dermis (papillary layer region) nevus cells often form clearly delimited, not quite rounded nested clusters of different sizes in the cytoplasm and contain brown pigment (melanin) in the form of small lumps.

In the deep dermis GCN nevus cells extend up to the boundary with the epidermis and can penetrate into it, by fibrous septa, that divide cells of subcutaneous fat, and appear directly in its tissue. There are data concerning an increase in the number of macrophages in the nevus tissue and their increased activity, expressed in particular in the strong absorption of the melanin pigment by macrophages [2].

Congenital giant pigmented nevi are often significant cosmetic problem and a significant psychotraumatic factor for the child. It is believed that malignant melanoma, which develops from the other options of nevi, usually occurs in areas of the dermal epidermal connection (in the surface layers of the skin), whereas melanomas of the GCN originates mainly in the deep layers of the dermis [2-4], which obviously is the condition of preserving the possibility for malignancy of GCN residues at its incomplete removal.

The reasons of occurrence and development of the GCN are discussed in literature, although there is a fairly well-established opinion that the GCN develop from belated during migration and then proliferating cells of primitive neuroectodermal origin (from the neural crest cells) [5]. There are data that these tumors have diverse morphological manifestations and clinical behavior [6]. Due to the fact that the majority of congenital giant pigmented nevi undergo early surgical and medical treatment for cosmetic reasons and as a prophylactic measure, the true incidence of their malignancy is difficult to determine [2].

There is also no consensus on the need to remove nevi and a clear view about the choice of treatment. Currently in the literature are described and in practice are applied a few basic methods for elimination of GCN - landmark excision with plastic, free cleavage or

full layered autotransplant, the use of flaps obtained by tissue dermatension, dermabrasion, and laser curettage [7]. Several authors have reported about surgical treatment of GCN using microsurgical techniques [8-10].

Below there is a case of a giant congenital pigmented nevus from Georgia, which since child's birth was observed by us in collaboration with colleagues - oncologists. The diagnosis of congenital giant pigmented nevus was confirmed by the results of histological examination

Histological preparation from 16.09.08 (left groin area): the stratum corneum is loosened and is composed of 4-10 corneocytes rows. In the granular layer, forming 2 rows, content of keratohyalin granules is uneven, sometimes meager. Prickly layer has 3 to 4 rows of keratinocytes, but one of the sites has 15 rows.

In the basal layer are relatively few apoptotic cells. Papillary layer is edematous, nipples are not expressed, the boundary of the epidermis to the dermis is almost flat. In the papillary layer nested cluster of nevus cells have the form of spheroids (Figure 1).



Figure 1: Congenital melanocytic nevus

Between the spherical formations individual nevocytes were marked. Close to the cell nests and inside of nevus cells the blood capillaries have been found. Nevus cells differ in the content of melanin, in one nest, they are overloaded by melanin, in other – do

not contain it. In the deeper parts of the dermis nevus cells are fragmented, diffuse and do not contain melanin. Diagnosis: congenital dermal nevocytic nevus.

Histological preparation from 16.09.08 (the front surface of the right thigh): structure of the skin is the same as in the previous preparation. The difference is that in the nests of melanocytes, cells have distinctly fibroblast-like cell structure and only individual of them contain melanin in the cytoplasm. The lower parts of the dermis filled by nevus cells lying more densely than in the previous preparation and having the morphology of fibroblasts. Diagnosis: dermal nevocytic nevus.

Mother and father of the child Georgians. The baby was born at term, the growth of 50 cm, weight 3450 g, mother - primipara. Pregnancy and childbirth were without complications. At one of the regular examination of the child, the skin lesions in the form of dark and light brown pigment spots were found, which captures part of the back, buttocks, abdomen, groin and upper thighs (Figure 1).

On the background of pigmented lesions of the skin, as well as on the periphery of the lesions, single pustular eruptions, as well as serous-purulent crustose elements were observed. For the sanitation of secondary pyococcus infection, an adequate course of treatment, including antihistamines, broad-spectrum antibiotics and topical antibacterial agents were prescribed.

In other parts of the body (lower limbs, genitals) multiple pigmented spots with a diameter of 1 to 4.5 cm were observed. The color of the rash was vary from light brown to gray-brown, dark and sometimes black tint. In the back and buttocks nevi are raised above the surface of the skin, creating an uneven rough terrain. A large portion of the surface of the nevus (on the back and hips) was covered with thick, rigid, vellus, mostly blond hair.

Patients with congenital giant pigmented nevus lifelong must be controlled by a dermatologist, and depending on the indications – by oncologist. Due to the high risk of malignant degeneration of

pigmented nevi in melanoma, risk factors: skin type, age, family history and ultraviolet irradiation should taken into account. Parents need to protect children from prolonged sun exposure and, especially, from sunburn. It is recommended to wear closed clothes, use sunscreen means, abstaining from exposure to the sun during the hours of the most intense radiation. The regular (at least 2 times a year) visits to dermatologist is needed.

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