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LARGE AND GIANT MELANOCYTIC NEVI OF THE MAXILLOFACIAL AREA IN CHILDREN: FEATURES OF THE MORPHOLOGICAL STRUCTURE AND SURGICAL TREATMENT

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Background. Congenital melanocytic nevus is a benign pigmented neoplasm composed of nevus cells that clinically manifest at birth. When choosing a treatment for nevi, the possibility of recurrence as well as the threat of tumor malignancy should be considered.

The aim of this work is to justify the surgical removal of a large and giant nevi of the face as a method of treatment justified by the pathomorphological structure.

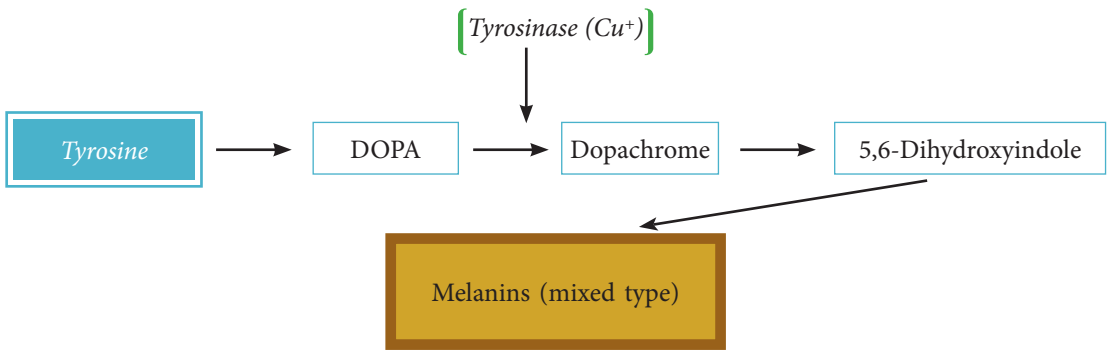
Materials and Methods. In 40 children of different ages born with large and giant nevi of the face, we used various types of plastic surgery to eliminate any defects formed after the excised nevi. We accounted for the features of the maxillofacial area: local plasty, expander dermotension, and transplantation of free skin grafts. We performed a total of 68 surgical interventions. Sixteen patients underwent the surgery once and 24 patients underwent secondary surgery (from 2 to 4). We also developed a scheme of the staged surgical treatments and conservative methods.

Results. All patients had stable positive results that were studied by comparing the outcomes of different surgical treatment options and accounting for various morphological characteristics of the removed nevi.

Keywords: congenital giant melanocytic nevi, congenital large melanocytic nevi, morphological structure, surgical treatment.

BACKGROUND

Congenital melanocytic nevi are non-malignant melanocytic tumors whose presence may be determined in utero, and they are revealed immediately following childbirth [1]. Giant nevi occur in 1 in 20 000 newborns [2]. Most researchers believe that the occurrence of nevi is associated with the migration of pigment cell predecessors (melanoblasts) from the neuroectodermal tube to the basal layer of the epidermis during fetal development [3]. Melanocytic nevi comprise melanocytes—cells that produce melanin, the pigment synthesized from the amino acid tyrosine under the influence of tyrosinase (Scheme 1) [4]. The presence of dark brown melanin in nevus cells determines the color of the neoplasm in macroscopic studies [5].



Scheme 1. Melanin synthesis

Most authors determine the following intraclass morphological variants for the classification of melanocytic nevi: boundary, complicated, intradermal, blue, and juvenile melanomas [6]. Depending on their structure, melanocytes are located in the epidermal layer of the skin and/or in the derma [7, 8]. A.G. Baindurashvili et al. (2011) stress that congenital nevi differ from acquired nevi by their larger dimensions and higher cellularity [9]. In children, congenital giant nevi are characterized by numerous features: large skin segments are damaged in varying localizations; dimensions of the giant nevus proportionally increase with the child's growth; and morphological characteristics may conform to a combination of complicated, epidermal, and dermal variants of nevi [10]. In 6%–10% of cases, a giant congenital melanocytic nevus may be a predecessor to a malignant melanoma [6].

The problems encountered by clinicians during the treatment of giant nevi remain unresolved. There is no unified scheme for treatment. The anatomic and physiological characteristics of the maxillofacial zone create numerous problems in determining the surgical strategy. The results depend on many factors such as the localization, extent of the pathological process, and method of treatment. The skill of the surgeon is also of great importance. In the selection of the treatment strategy, one should keep in mind the continued growth of incompletely resected nevi, the possibility of recurrence in completely resected nevi, and the risk of neoplasm malignization before conducting therapeutic intervention in one operation or staged operations.

MATERIALS AND METHODS

Between 2010 and 2014, in the Department of Maxillofacial Surgery at the Scientific Research Children's Orthopedic Institute (SRCOI) of G.I. Turner, 40 children between the age of 1 and 18 years and with congenital large and giant nevi in the maxillofacial zone underwent surgery. Preoperative examinations revealed no concomitant abnormalities in the patients. Before the operations, all children were examined by an oncologist.

The most frequent localizations of large and giant facial nevi were the cheek and the periorbital zones.

In total, 60 surgical operations were performed in 40 patients. Of these, 16 underwent one operation and 24 underwent 2–4 operations.

Within the indicated period, samples (skin grafts with nevi of different size) from 40 pediatric patients (n = 68; 100% of the cases) were sent to the morphological laboratory of the SRCOI of G.I. Turner after each surgical session. After the standard pretreatment, histological preparations stained with hematoxylin and eosin were examined using an optical microscope (Axio Scope.A1; Carl Zeiss, Germany). Morphological (histological) studies of samples (in some cases, more than once) from the 40 patients revealed the presence of intradermal nevi in 32 patients (80%; 54 histological preparations) and mixed nevi in eight patients (20%; 14 histological preparations; Diagram 1). No boundary nevi were identified in any samples. There were no signs of tumor tissue malignization in any observation.

RESULTS AND DISCUSSION

Intradermal nevi

On visual inspection, the neoplasms were characterized by large dimensions, different shapes, and quite sharp boundaries; prevalence of dark brown coloring of the outer surfaces of the nevi; and frequent presence of numerous dark hairs in the nevus "spot" (Figs. 1A and 2A).

Intradermal nevi were characterized morphologically by an abundance of nevus cells (melanocytes), which were diffusely located throughout the thickness of the derma; the concentration of melanocytes in the papillary layer of the derma and in the upper half of its cellular layer was significantly higher than in the deep layers of the cellular layer near the hypoderm (Figs. 1B and 2B). Melanocytes were not observed in the fatty tissue of the hypoderm. The coloring of the hematoxylin and eosin preparations conclusively revealed a presence of a small number of nevus cells in the upper (extending off of the derma) areas of the fibrous septi of the hypoderm.

Mixed nevi

From a macroscopic point of view, mixed nevi were also of varied size and irregular shape with rather sharp boundaries, but had less intensive, uneven, light brown (sometimes brownish) coloring of the external derma than intradermal nevi. Fine, not very long, unevenly spread dark brown and fair

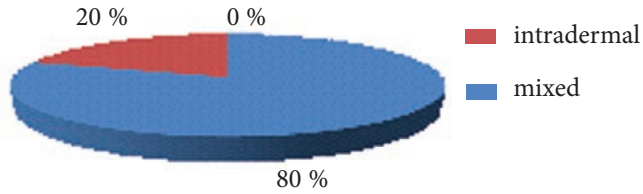
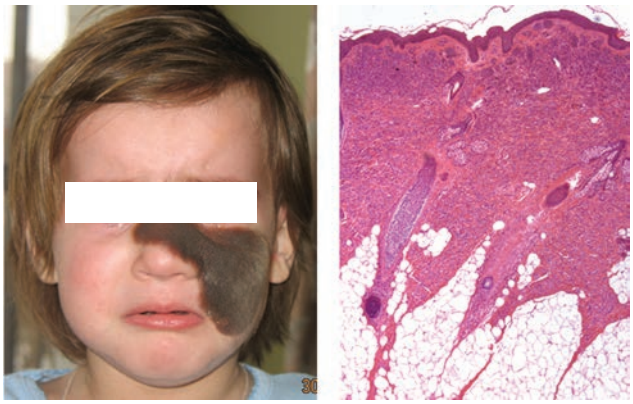


Diagram 1. Percentage of intradermal and mixed nevi according to histological analysis

hairs were revealed in relatively small proportion (Figs. 3A and 4A).

In histological examinations of mixed nevi, diffuse localization of melanocytes was revealed both in the epidermal layer and almost throughout the thickness of the derma (Fig. 3B). In some observations, the density of nevus cell distribution in the deep layers of cellular dermal layer was significantly less pronounced than in the upper skin layers (Figs. 3B and 4B). During optical microscopy of the hematoxylin and eosin-stained preparations, pathologically changed skin nevus cells were not revealed in the regularly formed fatty tissue of the hypoderm. In some samples, rather deep penetration of nevus cells into the upper areas of the fibrous septi of the hypoderm was observed.

Pseudopapillomatosis of the epidermal layer combined with significant hyperkeratosis was observed in three patients with mixed nevi (37.5%) during histological analysis (Fig. 4B).



A *B*

Figure 1. A. Macroscopic view of an intradermal nevus: a dark brown formation of irregular shape, with rather distinct boundaries and significant number of dark hairs. B. Histological image of an intradermal nevus (hematoxylin and eosin stain; uv. ×130)

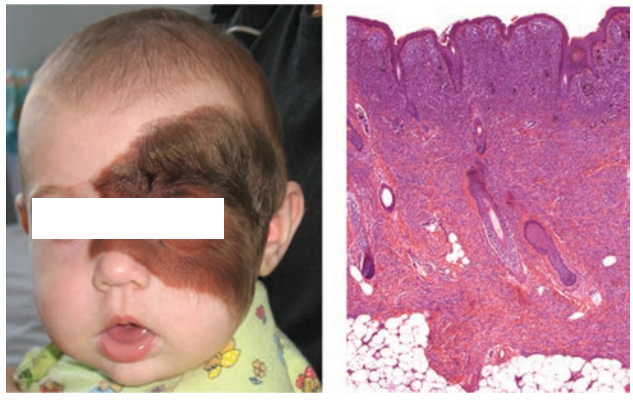


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Figure 2. A. Macroscopic image of an intradermal nevus: a dark brown formation of irregular shape, with rather distinct boundaries and significant number of irregularly distributed dark hairs. B. Histological image of an intradermal nevus (hematoxylin and eosin stain; uv. ×130)

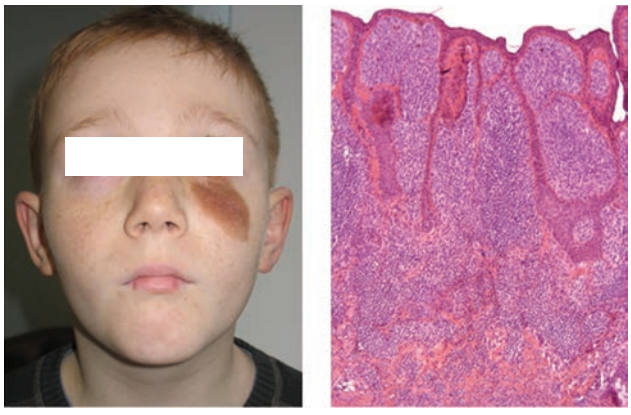

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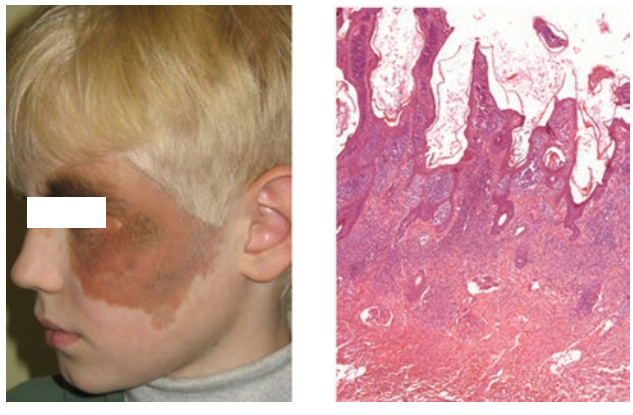
Figure 3. A. Macroscopic image of a mixed nevus: a dark brown formation of irregular shape, with rather sharp boundaries and moderate amount of unevenly spread fine hair. B. Histological image of a mixed nevus (hematoxylin and eosin stain; mp. ×130)



A *B*

Figure 4. A. Macroscopic image of a mixed nevus: a large, light brown formation of uneven coloring and irregular shape, with rather sharp boundaries and a moderate amount of unevenly spread fine hair. B. Histological image of a mixed nevus with pseudopapillomatosis and hyperkeratosis (hematoxylin and eosin stain; mp ×130)

Deep penetration of melanocytes into the hypoderm through its fibrous septi necessitates the removal of not only the pathologically changed skin areas but also, at least, the upper hypoderm layer.

Reconstructive and plastic surgeries

The method of choice is a resection of the nevus with the upper layer of subcutaneous fat, deviating 0.2 cm from the boundary, with subsequent grafting of the defect. If it is impossible to remove it in a single stage, staged resection of nevus tissue is performed.

We used the following methods to repair the defects formed after the nevi resection in the maxillofacial zone: local tissues, free autografts, or grafts with skin obtained as a result of dermotension. It is necessary to combine several surgical treatment options for the treatment of giant nevi.

A choice of plastic surgery method is individualized and depends on the size and localization of the defect. One of the peculiarities of facial skin defect closure is the increase of their dimensions due to traction of mimic muscles, which are closely related to the facial skin. This significantly complicates precise planning of the operations.

A defect formed with cheek nevus excision was closed with local tissues. Large rotation flaps obtained from the lower zone of the face and neck were used (Fig. 5B). Skin in this area meets the esthetic criteria. Complete resection of the formation was conducted in eight patients diagnosed with large cheek nevi. This was one of the treatment stages for 23 patients with the diagnosis of giant nevus. No complications were observed.

Tissue dermotension is one of the methods of choice for nevi localized in the zone of the forehead and the hairy part of the head. In treatment of large and giant nevi, this technique gives good esthetic and functional results, which are particularly

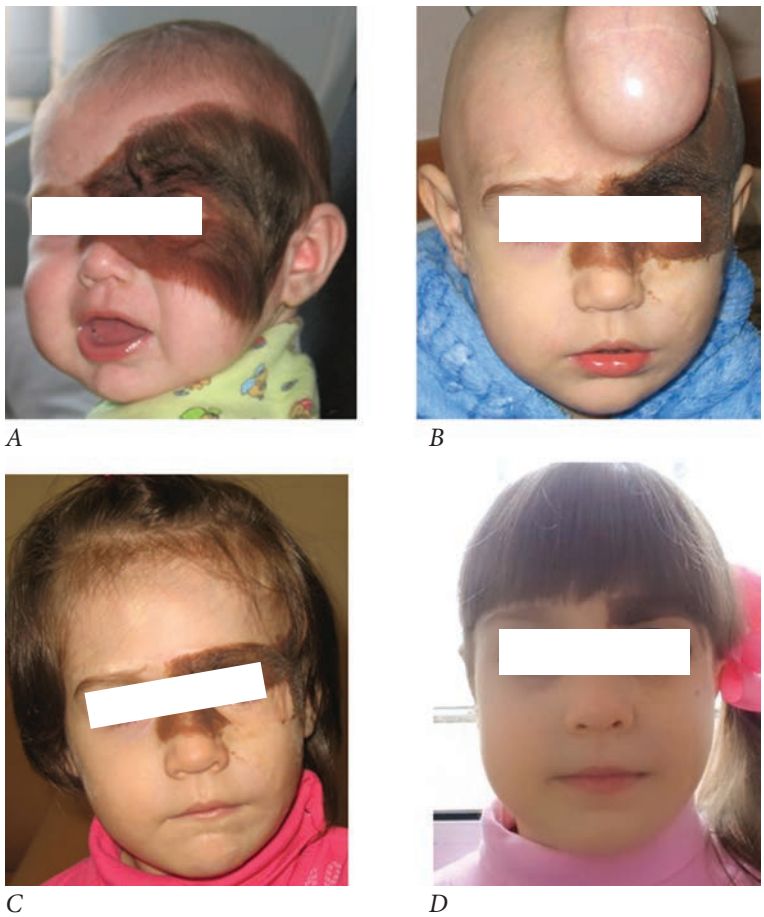


Figure 5. Diagnosis: giant facial nevus. A. Before the treatment; B. One year after the first stage of surgical treatment (resection of cheek nevus area and closure of the defect with local tissues). Tissue dermotension with expander was conducted as the second stage (120 ml). C. One year after surgical treatment (expander removal, resection of nevus site in the forehead area on the left, closure of the defect with the obtained flap and resection of nevus area of lower eyelid on the left, closure of the defect with free skin grafting). D. The result 1 year after the third and fourth stages of surgical treatment (resection of nevus in nasal and upper eyelid areas and closure of the defects with free skin grafting)

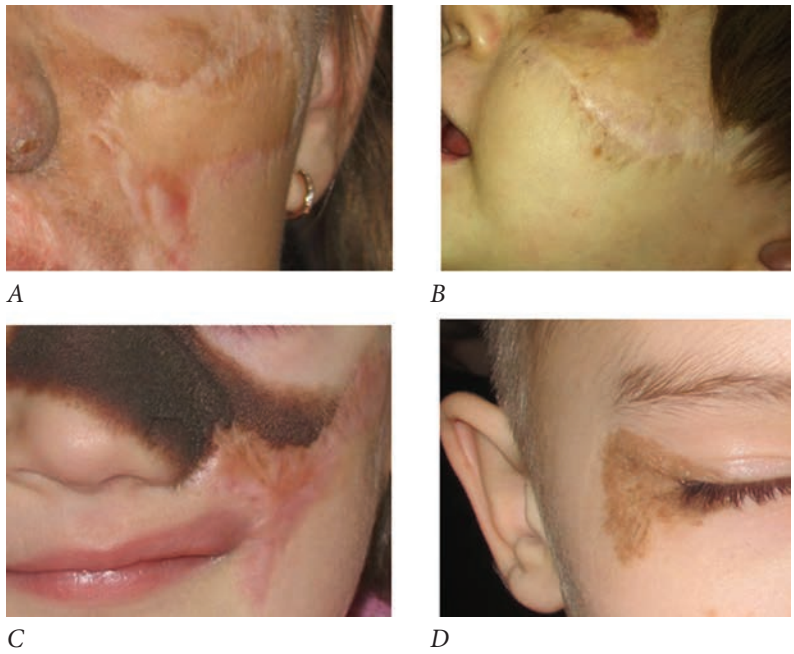


Figure 6. A. Color of skin autografting differs from the color of surrounding skin and is close to the color of nevus. B. Hair can be observed in the zone of autografting. C. Coarse postoperative scar that deforms the corner of the mouth and red border. D. Recurrence of pathological process after resection of nevus with carbon laser.

important on areas of open skin. This method allows the resection of giant nevi with a large volume using the surplus of the skin obtained in the area bordering the nevus (Fig. 5B and C). This method of treatment was used in 18 patients (as a radical operation in six patients and as one of the treatment stages in the remainder).

Free skin autografts were used in areas where it was not possible to close the defects with local tissues or apply tissue dermotension. These were the nasal and orbital areas (Fig. 5C and D). The postotic area was a donor site. In this area, the skin matches facial skin by color and texture. Free skin grafting was applied in 19 patients (as radical operations in two patients and as one of the treatment stages in the remainder). Repigmentation was not observed.

The treatment outcomes were evaluated by a three-score scale: good, satisfactory, and unsatisfactory. The evaluation criteria were graft color, absence of repigmentation, estheticity of the scarring, and presence or absence of deformations. All patients showed good, stable results.

All patients were followed up at least for 1 year after the operation, and the results remained stable.

Except for primary patients, in our hospital, there were 11 patients who had undergone previous operations in other hospitals. On admission to the hospital, the patients complained of cosmetic defects: the color of grafting significantly differed from the color of the facial skin in three patients (we believe that these unsatisfactory results were related to the use of free skin grafting taken from the skin of distant sites of the body, as it differs by texture, thickness, and color from the facial skin); two patients had "hair remainders" (skin grafts contained hair follicles); four patients had straining scars near the crucial organs; and two patients had a relapse of pathological process. Patients in Figure 6A–C underwent combined skin grafting, without taking into account peculiarities of maxillofacial zone. In the patient in Figure 6D, the nevus tissue was removed with a carbon laser. The risk of recurrence is high following treatment with resection of only the upper dermal layer of derma (Fig. 6D) because the nevus cells penetrate deep in the tissues; this is confirmed by morphological studies. All patients required repeated reconstructive operations.

CONCLUSION

The treatment of large and giant nevi in the maxillofacial zone remains a complicated problem. Treatment methods involving the resection of only the upper derma layer lead to recurrence. In the treatment of giant nevi affecting several facial zones, staged surgical removal and a combination

of different methods of reconstructive and plastic elimination of defects that take into account the peculiarities of maxillofacial zone are indicated. Mistakes in the selection of the therapeutic strategy and ensuing complications may lead to the deterioration of esthetic and functional results.

A thorough morphological study of surgical samples obtained during the resection of melanocytic nevi is necessary to determine the nature of the pathological process. It also confirms the necessity of the surgical resection of congenital large and giant melanocytic nevi.

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БОЛЬШИЕ И ГИГАНТСКИЕ МЕЛАНОЦИТАРНЫЕ НЕВУСЫ ЧЕЛЮСТНО-ЛИЦЕВОЙ ОБЛАСТИ У ДЕТЕЙ. ОСОБЕННОСТИ МОРФОЛОГИЧЕСКОГО СТРОЕНИЯ И ХИРУРГИЧЕСКОГО ЛЕЧЕНИЯ

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Введение. Врожденный меланоцитарный невус — это доброкачественное пигментное новообразование, состоящее из невусных клеток, которое клинически проявляется при рождении. При выборе тактики лечения невусов нельзя забывать о возможности возникновения рецидива, а также об угрозе малигнизации новообразования, поэтому встает вопрос о радикальности выбранного метода.

Целью данной работы является обоснование хирургического удаления больших и гигантских невусов лица как метода выбора, обусловленное патоморфологическим строением.

Материалы и методы. У 40 детей разных возрастов, родившихся с большими и гигантскими невусами лица, были использованы разные варианты пластического устранения изъянов, сформировавшихся после иссечения невусов, с учетом особенностей челюстно-лицевой области: местная пластика, экспандерная дермотензия и пересадка свободных кожных трансплантатов. Проведено 68 хирургических вмешательств: 16 пациентов были прооперированы однократно, 24 пациента подвергались повторным (от 2 до 4) оперативным вмешательствам. Разработана схема этапного хирургического лечения и консервативных мероприятий.

Результаты. У всех пациентов получены стойкие положительные результаты, изучение которых проводилось как в сопоставлении исходов различных вариантов хирургического лечения, так и с учетом особенностей морфологических характеристик удаленных невусов.

Ключевые слова: врожденные гигантские меланоцитарные невусы, врожденные большие меланоцитарные невусы, морфологическое строение, хирургическое лечение.

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