

GIANT CONGENITAL MELANOCYTIC NEVUS OF THE FACE: A CLINICAL CASE

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This article describes a rare case of congenital anomalies involving giant melanocytic nevus of the face. Errors in the choice of treatment tactics for children with this disease and its complications can lead to poor esthetic and functional results. When selecting a method of plastic surgery to eliminate defects formed after removal of nevi, it is necessary to take into account anatomical features of the maxillofacial region. Our department has developed a complex scheme for treatment of these patients. Surgical treatment in combination with massage, myogymnastics, and regular medical observation have provided good esthetic results. An integrated treatment approach for children with giant nevi of the face allows early medico-social rehabilitation of children with this pathology.

Keywords: giant congenital melanocytic nevi, surgical tactics, complex treatment.

Introduction

Congenital melanocytic nevi are benign melanocytic tumors usually discovered immediately after birth or appear during the first weeks of life [1]. The frequency of children born with giant nevi is approximately 1 in 20,000 newborns [2].

Allen and Spitz classified nevi into the following types: epidermal, complex, intradermal, blue, and juvenile melanoma [3]. Depending on the nevus structure, melanocytes are located in the epidermal layer of the skin and/or dermis [4]. Baidurashvili et al. (2011) observed that congenital nevi are different acquired nevi—they have larger size and increased cell count [5]. Furthermore, according to the size, nevi are classified as small, large, and giant.

The problems that doctors encounter in the treatment of giant nevi of face are considerable; there is no uniform nevi classification and treatment protocols. Errors in the selection of treatment, and complications because of these errors, may result in poor esthetic and functional results.

Materials and methods

In April 2011, patient H., born in 2010, was seen for the first time in the Department of Maxillofacial Surgery of Turner Scientific and Research Institute for Children's Orthopedics. The child was born with a giant nevus of the face.

The nevus had been increasing in size at the same rate as the growth of the maxillofacial region. The defect was spread over the buccal, periorbital, temporal, and left forehead areas, as well as over the glabella, dorsal and side planes of the nose, and left upper and lower eyelids. The lesion had a uniform dark brown color (Fig. 1 A, B) and was also covered with thick hair growth. There were no other melanocytic lesions on the child's body.

After examination and consultations with the oncologist, the first step of surgery was performed—reconstructive and plastic surgery of congenital anomalies of the maxillofacial region, i.e., excision



Figure 1. A giant nevus of the face.
A — front view, B — side view

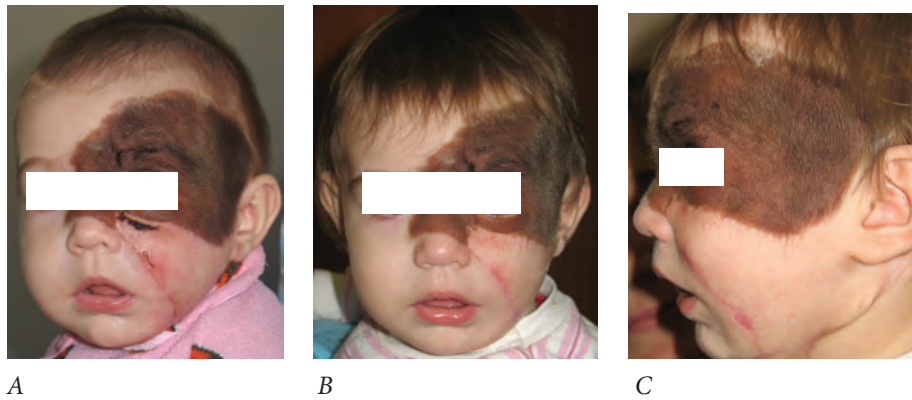


Figure 2. A — 10 days after the first stage of surgery; B, C — six months after surgery.

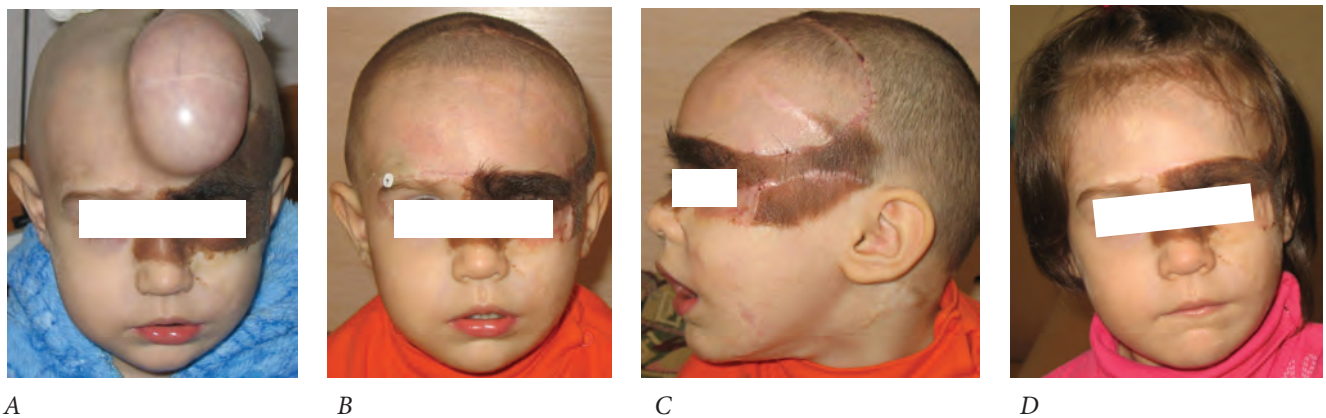


Figure 3. A — skin expansion (total fluid volume 120 ml), B, C — 10 days after the second stage surgery, D — one year after surgery.

of the nevus in the buccal area on the left side and closure of the defect with local tissues (rotation skin flap was used) (Fig. 2).

In 2012, the patient had a two-stage treatment—reconstructive and plastic surgeries. A tissue expander was inserted in the forehead, with total volume of fluid being 120 ml (Fig. 3 A); nevus was excised on the left side of the forehead, and the defect was closed by the graft, harvested from the expanded skin area. Nevus area on the left lower eyelid was excised, and combined eyelid reconstruction was performed (full thickness skin graft from behind the ear was used) (Fig. 3 B–D).

In 2013, the patient had undergone the third stage of the surgery—reconstructive and plastic surgery. A tissue expander was installed in the left temporal region with full liquid volume being 50 ml. The frontotemporal area of the nevus was excised; then the area was closed with the flap from expanded skin. Nevus was excised from the nose area, and combined skin grafting was performed

(full thickness skin graft from behind the ear was used).

In 2014, the child had the fourth stage of surgery—reconstructive and plastic surgery: excision of the nevus portion on the upper eyelid, and combined skin grafting (full thickness skin graft from behind the ear was used).

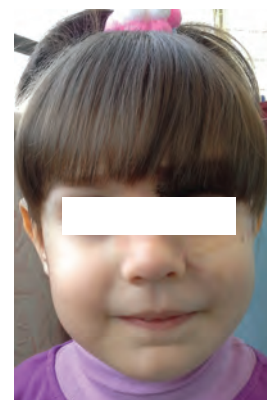


Figure 4. The results a year after the third stage of treatment

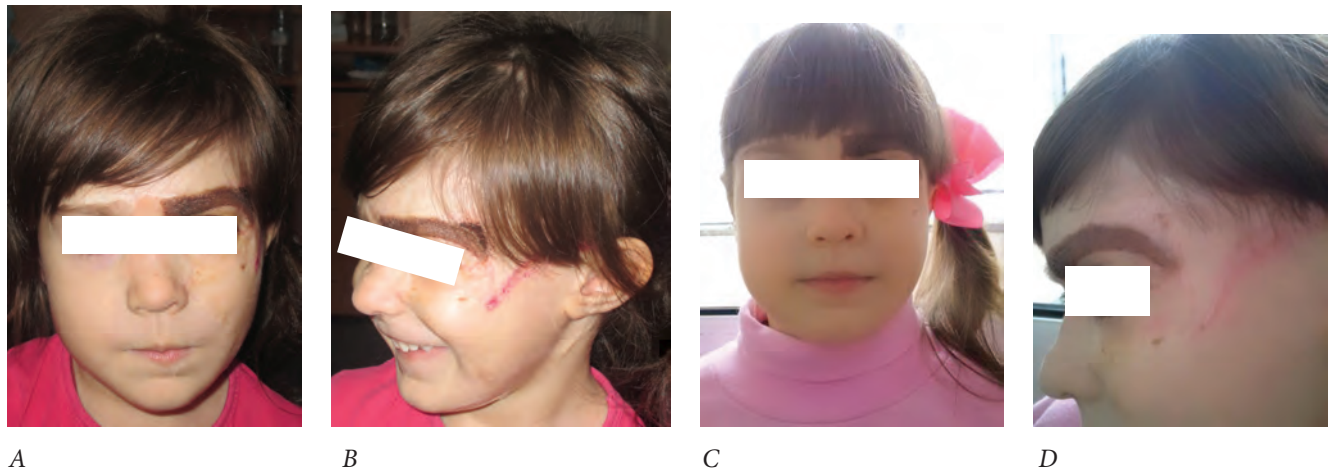


Figure 5. A, B — 10 days after the fourth stage of the treatment, C, D—a year later

To prevent formation of thick scars, after each stage of surgery, the patient received massage of the incision area and facial muscle exercises and was instructed to use scar gels.

He was under our observation for 1 year after the last surgery. Treatment results have remained stable.

Morphology

Based on a morphological study of surgical specimens, intradermal nevus was diagnosed given the abundance of small and medium-sized “nevus cells” (melanocytes), with a relatively small amount of weak oxyphilic cytoplasm, moderately basophilic nuclei, and predominantly irregular ovoid shape.

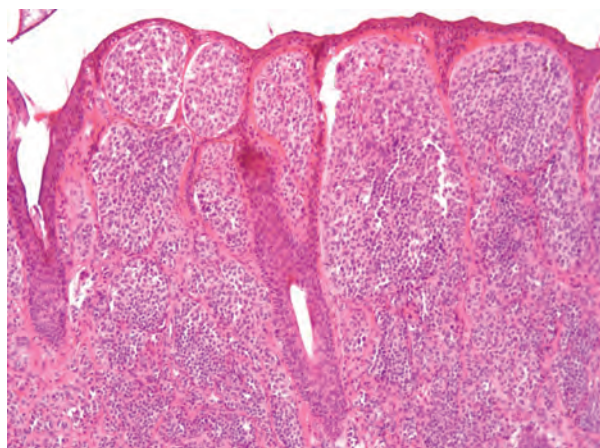
In the upper layers of the dermis (papillary layer), nevus cells that had formed were

clearly demarcated, irregularly rounded nested clusters of different sizes (Fig. 6 A). In the reticular layer, cells formed extensive “fields” (Fig. 6 B).

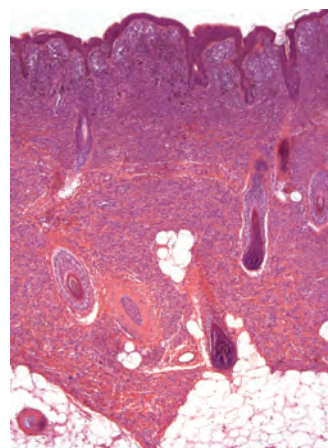
In the deeper layers of the dermis, nevus cells spread to the boundary of the hypodermis and penetrated into this along fibrous septa, which separate the cells of subcutaneous fat tissue (Fig. 7).

Results and discussion

We strongly believe that surgical removal of the giant nevus of the face, confirmed by histological examination (spread of melanocytes into deep tissues), is the treatment of choice. If one-step removal of nevus is impossible, multiple-step procedures should be used. We used the following methods to correct the defects formed after the



A



B

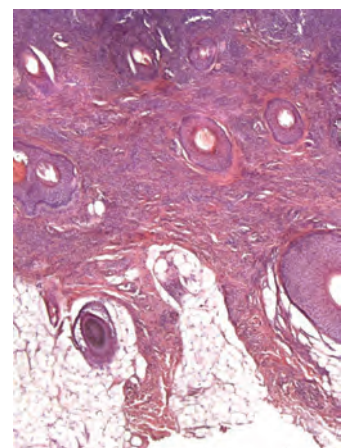


Figure 7. Dermis–hypodermis border (H&E stain)

Figure 6. Intradermal nevus histology (H&E stain; A — $\times 260$, B — $\times 130$)

removal of nevi: local skin grafts, free skin autografts, and skin grafts harvested after tissue expansion. In the treatment of giant nevi of the face, all these techniques can be used in combination.

The choice of method of plastic surgery is specific for each patient, and depends on the size and location of the defect. One of the features of nevi of the maxillofacial region is that it is localized in close proximity to vital organs. If treatment strategy is improperly selected, severe scarring may form, resulting in ectropion, lip eversion, nasal atresia, microstomia, and/or limitation of mouth opening. Another feature is the deficit of adjacent tissue, which limits the use of local tissue to close the defect. Furthermore, the use of skin for grafts from the surrounding areas (e.g., from behind the ear, submandibular region, or neck) is very limited, for a satisfactory esthetic result of similar color and thickness. Use of free skin grafts from distant parts of the body that are very different from the face skin in texture, thickness and color, often leads to re-pigmentation and the need for repeat corrective operations.

While choosing a method of correction of the facial skin defect, it is necessary to take into account the anatomical features of the face. Difficult anatomical relief dictates the conditions: the face should be separated into zones and these zones restored consecutively, separating non-moving areas of the face from moving areas.

In our department, the protocol of treatment for children with this pathology is developed taking into account the features of the maxillofacial region. When a nevus is excised in the buccal region, the defect is closed with local tissues. We use large rotation flaps derived from the lower zone of the face and neck. The skin in the area is best suited for esthetic performance. Tissue extension is a method of choice if a nevus is located on the forehead and scalp. In the treatment of giant nevi of the face, this technique gives good esthetic and functional results, which is especially important on exposed skin areas. This method allows for removal of large areas of giant nevi using excess skin harvested in the area adjoined to the nevus. Free skin autografts are used in areas where it is impossible to use local or expansion tissues. This is particularly true for the area of the nose and orbit, where the donor site is behind the ear. The

skin in this area is similar to facial skin in color and texture so that re-pigmentation was not observed.

Perhaps the most significant role in getting good and lasting esthetic results is that of facial muscle massage and exercises. Long-term outpatient follow-up visits and active parental supervision is needed until the full formation of the maxillofacial region. Strict adherence to the principles of integrated treatment not only prevents the development of scar deformities but also significantly improves the quality of life of children with this defect.

Conclusion

In cases of giant nevi of the face, severe deficiency of skin for grafts results in the need for a large number of surgical interventions. When treating children with giant nevi of the face, the best tactic is to use an integrated approach. It is necessary to combine different methods of plastic surgery to eliminate defects, while paying attention to the particular features of the maxillofacial area. Massage and facial muscles exercises play a key role in achieving good, lasting results of surgical treatment. Constant long-term monitoring and active participation of parents are necessary. Abiding by all of the principles of integrated treatment allows early medical and social rehabilitation of children with this defect.

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