

A Surprising Case of the Combination of the Congenital Malformation of the Arm and Skin. What do we do?

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Abstract

The paper presents a very rare combination of childhood congenital malformations in children in the form of aplasia of the hand and nevus of the skin. The location of a benign skin tumor in the upper third of the forearm of an underdeveloped limb significantly limited the fit and use of the prosthesis, the only hope for improving the function and cosmetics of the child's hand. Parts of the prosthesis in contact with the nevus in the elbow bend area could cause her mechanical irritation during use with negative consequences. A decision was required to optimize the nature of prosthetics and prevent severe complications. In this case, the option of rehabilitation was rational methods of reconstruction of the prosthesis or adequate surgical treatment of the tumor.

Keywords: Congenital Malformations; Nevus; Aplasia; Child; Prosthetics

Introduction

The term "nevus" (lat. Naevus) was first used by R. Virchow (1863) to refer to birthmarks of the skin, and J. Jadasson (1914) gave a broader interpretation, including malformations and some benign skin tumors in this group [1-3]. Congenital pigmented nevi are benign tumors and are common in children; they are formed from special cells containing the pigment melanin [4,5], are associated with the vicious development of neuroectoderm and are divided into 3 main groups according to histological structure: junctional or borderline nevus, intradermal and mixed [6]. The localization of nevi is very diverse, although most often they are located on the face and neck, less often on the limbs and trunk of the child. In accordance with the growth of the child, congenital nevi increase, but their course is usually benign. Until puberty, malignancy (transition to melanoma) is not observed, and for the growing child and his relatives, the main concern is a cosmetic defect in the area of the nevus, especially if the formation is located on open areas of the body, face or hands [6].

In the literature we studied, there were no clinical observations describing the combination of a congenital malformation of the upper limb and a congenital malformation of the skin-nevus. In this regard, it seemed to us advisable to present options for solving the problem of prosthetics of an underdeveloped hand.

Purpose of the Study

To show possible ways to rehabilitate a child with a combination of congenital malformations of the skin and upper limb.

Materials and Methods

A surprising case of a combination of a congenital malformation of the left upper limb (congenital aplasia of the hand) and a nevus in the forearm of the same hand in a 9-year-old girl is considered. The child's upper limb would be represented by a soft-tissue rudimentary formation in the form of a small palmar part and a general shortening of the arm up to 7 cm in comparison with a healthy limb. The rudiment of the hand was turned to the palmar side at an angle of up to 90 degrees, and the range of motion in the wrist joint did not exceed 15 - 20 degrees (Figure 1).



Figure 1: Congenital transverse defect (congenital aplasia of the left hand), shortening of forearm till 7 cm.

The nevus along the outer surface of the upper third of the forearm, at an angle to the axis of the forearm and measured $3.5 \times 4.5 \text{ cm}$. Its proximal part was localized at the level of the elbow bend (Figure 2).



Figure 2: Skin nevus of upper third of forearm has light brown color and hair covered.

Partial restoration of the gripping function of the fingerless hand and improvement of the cosmetic condition of the upper limb could only be achieved with the help of an active upper limb prosthesis. However, there was a high risk of mechanical trauma to the nevus at all mandatory technological stages of the prosthesis manufacturing (removal of a plaster cast, fitting the socket, fitting the prosthesis, and most importantly, its subsequent regular use). This was especially true of the skin of the upper third of the forearm in the area of the upper part of the socket and the elbow hinge, in which flexion and extension ensure the grip and opening of the artificial fingers of the prosthesis hand (Figure 3).



Figure 3: The main parts of below elbow prosthesis (cuff on the shoulder, elbow hinge, socket for the arm), requiring reconstruction for skin nevus.

Results and Discussion

Usually, the indication for removal of the tumor is aesthetic considerations, from the point of view of correcting a cosmetic defect. In our situation, the indication for surgery was the possible mechanical trauma of the nevus with the details of the prosthesis: its receiving sleeve and the elbow hinge.

One of the possible options for preventing pressure on the skin with the details of the prosthesis was the decision to make a hole in the socket or the so-called "window" in the prosthetics in the projection of the nevus. However, when bending and unbending the arm at

the elbow, there is always the effect of piston-like displacements of the skin relative to the walls of the receiving sleeve. It would not have been possible to avoid the risk of trauma to the nevus even if there was a "window".

Our patient was offered a scheme for cutting soft tissues according to Limberg, including excision of the pathological focus, taking into account the location of the lines of force of the human body [7] in the upper third of the forearm and providing for the correct location of the lines of skin incisions (Figure 4).

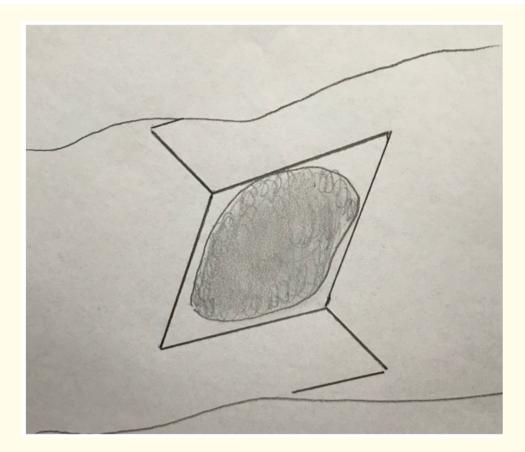


Figure 4: The optimal skin cuts pattern for Limberg, when skin nevus is cut.

Unfortunately, the girl's relatives refused the proposed operation and preferred prosthetics of the underdeveloped limb in the usual way.

Conclusion

In the presence of a combination of congenital malformations of the limb and skin, the optimal solution is the surgical correction of the latter. The choice of the method of treatment is individual, depending on the size of the tumor, its localization and the state of the surrounding tissues.

In the conditions of modern medicine, in addition to surgical correction of a skin defect, a nevus is removed using a laser, cryodestruction, electrocoagulation, and a radio knife.

After removal of the tumor, it is possible to achieve the required functional and aesthetic result with the help of prosthetics and eliminate the risk of tumor malignancy in the child in the future.

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