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Surgical management of congenital hairy nevus of face with Limberg flap, a case report

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Abstract

Introduction: Cosmetic deformity of the face is always a subject of concern for the patient. Besides the aesthetic considerations, the possibility for large congenital nevi to become malignant is significant and emerges as an important factor in the treatment and management of this entity.

The case report is about a four years old female child who reported with a large black hairy pigmented lesion on the left side of the mid-face region. The child often faced discrimination at school. The child's parents brought her to our institute for the correction of this major cosmetic defect.

Procedure: Surgical management involved excision of the lesion and reconstruction of the defect using a Limberg flap. In the immediate post operative phase, regular topical antiseptic ointment application was done for 2 weeks. Suture removal was done after 2 weeks and regular application and massage with an Allantoin based cream was done for the next 6 months.

Summary: Healing for the patient was uneventful and scars gradually decreased with massaging. There was no recurrence of the CNN. At the 2 year follow-up, the scar marks had become very faint. According to the testimony of the patient, the patient feels increasingly confident and more socially accepted as a result of the procedure.

Conclusion: We would like to propose that with proper planning and analysis, Limberg flap can produce satisfactory functional and aesthetic reconstructive results for many full thickness defects of the facial region.

Keywords: Congenital hairy nevus, Limberg flap, Rhomboid flap, Congenital melanocytic nevus.

Introduction

The congenital nevomelanocytic nevus (CNN), commonly known as the congenital hairy nevus, refers to a pigmented surface lesion present at birth.[1] Melanocytic nevus of the face is a type of congenital nevus, which is rare, hair bearing and have high risk of becoming malignant. Besides the aesthetic considerations, the possibility for large congenital nevi to become malignant is significant and emerges as an important factor in the treatment and management of this entity.

Management and treatment options for patients with CNN varies with the lesion's size, location, and propensity for malignant transformation. Usually surgical management of large CNN lesions are addressed at the age 6 months[5,8,9] for better cosmetic prognosis. Serial excision[10] and reconstruction with skin grafting, tissue expansion, local rotation flaps, and free tissue transfer[11] are few of the accepted surgical modalities used in surgical management of CNN.

Case Report

The case report is about a four years old female child who reported with a large black hairy pigmented lesion on the left side of the mid-face region. The child was subjected to ridicule at school and was brought for the correction of this major cosmetic defect by her parents. A provisional diagnosis of black hairy nevus was reached. Surgical management involved excision of the lesion and reconstruction of the defect using a Limberg flap.

The lesion involved the left side of the cheek over the zygomatic arch region. The lesion extends 4cm behind the nasiolabial fold, 5cm anterior to the tragus of the ear, superiorly 3cm below the lateral canthus of the eye and inferiorly extending to the line joining the angle of the mouth and external pinnae. The lesion measured 2cm X 2.5cms in size. (Fig. 1)

While treating a CNN of the face, aesthetic considerations are very important. The incision was designed in the form of a parallelogram with two angles of 120° and two of 60°. These angles, however could vary depending on the shape of the lesion or defect. All sides of the rhomboid and all sides of the flap were measured to be equal. The flap designed for closure was extended off the 120-degree corner with an additional limb which was drawn parallel to the closest edge of the rhombus (STEP 1). After the rhombus defect was excised (Fig. 3) (T), the transposition flap (X) was elevated (STEP 2). The two edges of the transposition flap (P', Q') was juxtaposed to approximate the edges of the rhomboid defect (P, Q) (STEP 3).The flap was transposed and the standing cone deformity (Z) was corrected with a Burow's triangle excision (STEP 3). Closure of the defect was done in two layers (STEP 4). (Fig. 2)

Fig. 1: Pre-op

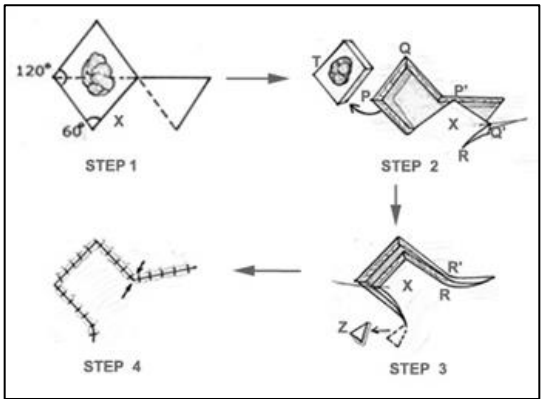

Fig. 2: Design of the Limberg flap

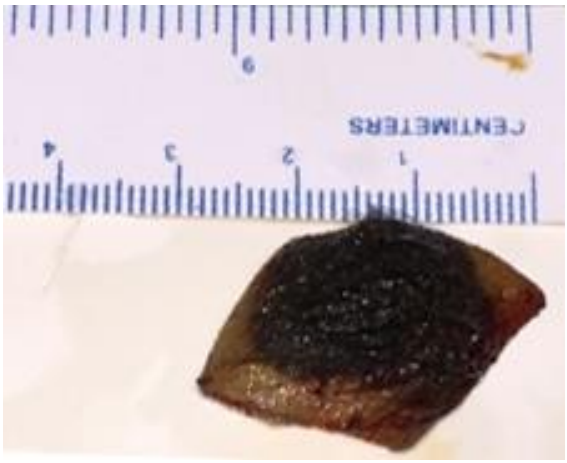

Fig. 3: Excised nevi with the surrounding skin in a rhomboid shape

Post-op care & follow-up: The patient was advised regular application of topical antiseptic ointment for 2 weeks (Fig. 4). After suture removal patient was advised regular application and massage with a Allantoin based cream for 6 months. During the follow-up period the patient was regularly photographed and evaluated for the aesthetic outcome. Healing for the patient was uneventful and scars gradually decreased with massaging. There was no recurrence of the CNN. At the 2 year follow-up, the scar marks had become very faint (Fig 5). According to the testimony of the patient, the patient feels increasingly confident and more socially accepted as a result of the procedure.

Fig. 4: 1 week post-op

Fig. 5: 6 months post-op

Discussion

CNN originates from neuroectodermal cells between the 8th to 24th weeks of pregnancy. Body protein HGE/SF (hepatocyte growth factors/scatters factors) is thought to be responsible for triggering these neuroectodermal cells to develop, migrate and scatter. They are often round or oval, clearly demarcated and sometimes slightly intact.

The cumulative risk of developing cutaneous melanoma in patients with CNN has been the topic for research in many scientific studies. Ulceration, bleeding, focal growth, pain, pruritus or significant pigmentation change are a few possible indications of malignant transformation of the CNN. According to a study performed at the University of Pennsylvania Medical Centre, patients with large or giant CNN had a 5.7% cumulative 5-year risk of developing a cutaneous melanoma.[6] Dutch nationwide pathology database

suggests that a standardized incidence rate of 12.2% of developing melanoma is associated with CNN. Moreover, patients with giant CNN are at an increased risk of 51.6% compared to general population rates.[7] For small CNN, risk rates have been reported between 0.8% and 4.9%.[7]

Chemical peels, dermabrasion and laser surgeries are the prominent adjunctive treatment modalities. Patients with small lesions are put up on regular follow-up for observation and the variations in the size of the lesion are monitored with regular photographic documentation. In few particular cases, cultured epidermal autographs are used to treat the defect.[11] As these adjunctive treatment options are incapable to eradicate the nevus cells completely, surgical excision still remains the main treatment modality.

The amount of pliability of the adjacent skin often determines the success of transposition flaps such as the rhomboid (Limberg) flap. The pliability of the adjacent skin can be noted by pinching various areas between the thumb and forefinger.[2] The elevation of the Limberg flap comprises a simple surgical technique, which requires sufficient subcutaneous fat. During the flap elevation procedure, the dissection must be carried past its base to prevent an elevated bump when it is transposed. Limberg flap are widely used in skin cancer, lupus, cystic acne, spina bifida, etc.

Summary

The success of facial reconstructive surgeries depends on the aesthetic sense of surgeons and though knowledge in the properties of local flaps. Thorough analysis of defect is necessary for choosing correct procedure. We would like to propose that with proper planning and analysis, Limberg flap can produce satisfactory functional and aesthetic reconstructive results for many full thickness defects of the facial region. Aesthetic reconstructive surgical procedures such as this can play a great role in increasing the social acceptability of a child suffering from isolation due to a congenital facial cosmetic defect.

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