Giant Congenital Melanocytic Nevus of Face-Primary Excision with Reconstruction Using Self-Filling Osmotic Expanders

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Abstract Congenital hairy nevus or giant congenital melanocytic nevus (GCMN) especially that of the face pose a great challenge to reconstructive surgeons. These unsightly lesions can be psychologically damaging to both parent and child. A variety of treatment options exist for the management of such cases. In this review article, we present two children who had a GCMN of their face managed with self-inflatable tissue expanders (osmotic tissue expanders [OTEs]) for single stage excision, and reconstruction is presented. The uses of self-inflating tissue expanders are now a regular choice for tissue cover in wide areas and in children.

Keywords Congenital melanocytic nevus · Hairy nevus · Osmotic tissue expanders

Introduction

Congenital hairy nevus or giant congenital melanocytic nevus (GCMN) especially that of the face pose a great challenge to reconstructive surgeons. These require complex anatomical tissue cover in cosmetically and functionally significant areas. The exact risk of malignant degeneration remains unknown. These unsightly lesions can be psychologically damaging to both parent and child. A variety of treatment options exist for the management of such cases. The procedures include serial

excision, excision and skin grafting, laser therapy, chemical peel, and dermabrasion. In this retrospective review, two children who had a GCMN of their face managed with self-inflatable tissue expanders (TEs) for single stage reconstruction are presented with literature review. Only very few literature reviews are available for osmotic tissue expanders. The use of self-inflating tissue expanders is now a regularly done procedure with very good patient compliance.

Case Reports

Case 1

A 7-year-old male child was brought with congenital hairy nevus of the forehead. The lesion was extending right half and crossing the midline of forehead. The 150-cc self-inflating square expander was placed on the left side. After 6 weeks of waiting time, the nevus was excised fully deep to the muscular layer and covered with the advancement of the flap (Figs. 1 and 2).

Pathologically, it was reported as giant melanocytic nevus with no evidence of malignancy. Postoperatively, the scar was settled well and good aesthetic correction obtained. No recurrence was reported after 2 years.

Case 2

A 6-year-old female child presented with nevus on the right side of face extending over the right side of face extending to the eyebrow and eyelid area with hair growth and deep pigmentation. The marginal incision was used to place two expanders. A square osmotic expander 100 cc in the right cheek below the nevus and 50 cc expander on the frontal area above the nevus. The expanders were allowed to expand for 6 weeks

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Fig. 1 Pre operative GCMN filling right half of the forehead with long hair

with total excision of the nevi and covered the defect with advancement flaps (Figs. 3, 4 and 5).

The histopathology revealed only melanocytic nevus with no atypical cells. The case was followed up for 2 years, and there was no local recurrence.

Discussion

Giant congenital melanocytic nevi are present at birth or soon thereafter. The melanin pigment in the surface is apparent early or it may delay in appearance from age of 1 month to 2 years in the rare “tardive” type. A GCMN larger than 9.9 cm in diameter occurs in 1 per 20,000 newborns; larger than 20 cm in diameter occurs in 1 per 500,000 newborns [1–3]. An equal prevalence exists in males and females. Autosomal dominant inheritance with incomplete penetrance or multifactorial determination occurs in families with small GCMN. The

associated findings in patients with GCMN are neurofibromatosis and leptomeningeal melanocytosis [4].

GCMN have nevomelanocytes in the epidermis as well-ordered clusters and in the dermis as sheets, nests, or cords. The presence of nevomelanocytes in the lower one third of the reticular dermis is specific for GCMN. The nevomelanocytes may extend into the subcutaneous tissue. Nevomelanocytes tend to be associated with skin appendages, vessels, and nerves [2].

Two factors influence the treatment of congenital nevomelanocytic nevi: the potential for malignant change and the cosmetic appearance. Each case requires tailoring of the operation(s) to fit the anatomic defect. The presence of an enlarging nodular mass indicates malignant change and requires immediate treatment. This mass may represent a rare neuroectodermal sarcoma. The incidence of malignant melanoma appears higher in the scalp, back, and buttocks and requires early and wide removal. The prospective study of GCMN turning malignant is found to have 8.5 % [5]. Excision of the nevi begins in 6–9 months. Surgical excision with reconstruction is the mainstay of treatment [6]. Chemical peels, dermabrasion, and laser treatments are adjunctive treatment choices [7, 8]. All of the adjunctive treatment methods have been associated with scarring. Furthermore, adjunctive treatment measures have not been demonstrated to decrease the malignant potential. If surgical excision is not feasible, management consists of examination and high-quality photographic documentation for life [8].

The goals of treatment are to remove as much as feasible the GCMN and reconstruct the defect, preserving function and maintaining the aesthetic appearance [7, 9]. Early removal of the GCMN is advisable. If direct closure after complete excision is not possible, reconstruction may include serial excision, excision with skin grafts and skin flaps, tissue expansion with subsequent flap rotation or full thickness skin grafting, autologous cultured human epithelium, artificial skin replacement, and free tissue transfer about tissue expansion.

Fig. 2 Postoperative after 3 weeks

Fig. 3 Preoperative large nevus extending to the eyelid and eyebrow

Cultured epidermal auto grafts [CEA] have been used successfully to obtain surface coverage after excision of giant hairy nevi. Dermal regenerate templates prior to skin grafting have been reported in the literature as a substitute for tissue expansion and rotation flaps. Evaluation of all small and medium GCMN for prophylactic excision should take place before the age of 12 years, since the potential for malignant change rises sharply after this age [8, 10, 11].

These GCNM of face and head region always carry deep pigmentation with hair growth. Excision of the lesions in these areas is difficult due to the anatomical peculiarities of the eyebrow, eyelid, forehead, and facial skin features. The aims of surgical excision are to remove the nevi to maximum and extend and reconstruct with tissues of the same color, texture, and thickness. The various methods used are the split skin graft, full thickness graft, and flaps. The split skin graft will “take” well, but the color, appearance of surface, and contracture are not good for face. Full-thickness graft can be used only for limited areas, and its availability and “take” are limited. Even though flaps with the same skin color and thickness

is ideal, but it is difficult to get such light colored and thin flaps [2]. Free flaps are like forearm flap; anterolateral thigh flap in a child is difficult technically due to long procedures and microvascular anastomotic difficulties.

Alternate to solve all these problems are the tissue expanders [12, 13]. Classical expanders and self-inflating expanders [osmotic tissue expanders] are available. In the classical expander’s regular injections of the expanders is a difficult part for the children. The self-inflating expanders are a good choice for the problems [14, 15]. After planning for the local tissue transfer, the expanders are inserted by a small incision at the margin of the nevi into the expanding area and allowed for the wound healing in its normal way. By absorbing fluid from the body, the expanders attain its maximum size by 4–6 weeks. In our cases the first procedures were done under General anesthesia and expanders [150 cc in 1st case and 2 expanders of 100 cc and 50 cc in 2nd case] are placed. After 6 weeks, the lesions were excised completely and the expanded tissue is transferred as advancement flaps. Only two procedures were required for the whole surgical treatment. The expansion

Fig. 4 Intraoperative and postoperative picture showing the advancement flap

Fig. 5 Postoperative after 21 days of surgery profile view

procedure requires only one procedure of implantation and a second surgery for its removal and reconstruction. The two procedures are well tolerated by the children in our series.

Postoperative care of the flap with topical smoothening agent will give a good scar. The cases are presented for the message that such self-inflating expanders are the best choice in children for procedures, which require tissue transfer as in the case of GCNM. In a normal case, no complications are seen with these expanders. In our experience of 20 cases of self-inflating expanders, wound necrosis occurred in two cases but early transfer of expanded tissue could save the flap. The cost of the expanders is the only disadvantage in these cases [16]. The follow-up of our cases were done for a period of 1 year with no recurrence.

Conclusion

Management of giant melanocytic nevi is difficult, and early excision and reconstruction give good cosmetic and anatomical correction. Multiple tissue expanders, preferably osmotic expanders, are very useful in reconstruction in children because it is a two-stage surgery with no interim or post-excision procedures.

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