

An alternative strategy treated giant congenital melanocytic nevi with epidermis and superficial dermis of the lesions

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Abstract

Giant congenital melanocytic nevi (GCMN) are defined as rare pigmented lesions that are believed to form between weeks 9 and 20 of gestation. It is difficult to reconstruct large defects after the removal of the lesions and it has posed a great challenge to the plastic surgeon and dermatologist.

Given all those difficulty reconstructing the defects, we try to explore an alternative way to resurfacing the defect after removal of GCMN.

Patients with GCMN received single-stage excision. Following the subcutaneous tissue and deep dermis were discarded, epidermis and superficial dermis were harvested as graft substitutes to reconstruct the defects in situ.

All of the grafted tissue survived well and skin color in the surgical area gradually became lighter. During the periodicity of follow-up, neither hypertrophic scars nor recurrence were observed. Furthermore, histopathology examination demonstrated that there are no distinct melanocytes gathered in the postoperation lesions.

For those GCMN which is difficult to reconstruct with traditional methods, resection of the lesion followed by reconstruction with epidermis skin and superficial dermis from the lesions in situ may be a feasible and alternative therapy method.

Abbreviations: GCMN = giant congenital melanocytic nevi, TBSA = total body surface area.

Keywords: epidermis skin, giant congenital melanocytic nevi, skin graft, superficial dermis

1. Introduction

Giant congenital melanocytic nevi (GCMN) developing from an embryonic neural crest abnormality are benign proliferative tumors present at birth and will reach >20 cm in adult life. A GCMN >20 cm in diameter occurs in 1 per 500,000 newborns, although smaller congenital melanocytic nevi are present in about 1% live births.^[1]

GCMNs produce significant aesthetic distortion to the affected anatomical sites and are often associated with severe psychological distress for patients and their families. Moreover, GCMN also poses the potential of malignant transformation, which might result in cutaneous or noncutaneous melanoma, characterized by high metastatic potential and a high rate of death.^[2,3] For above

reasons, it is advised that management be performed in early childhood rather than at a later age.

Having once decided to perform surgery, there are many options described: stage excision, skin grafting, tissue expansion, tangential shaving, derm-abrasion and pedicled, or free flaps.^[4,5] All above methods, however, either need multistage surgery or cause new damage to the normal skin. What's more, for those GCMN which we define as involving over 10% of the total body surface area (TBSA), it is impractical to reconstruct the defects by autologous skin graft or tissue expansion following complete resection, as it is hard for both healthcare providers and patients to accept such extensive new scar formation at the normal donor sites.^[6] As for these GCMN, we think out an alternative treatment with epidermis and superficial dermis of the lesions and we have achieved excellent results. We will present a patient with an extraordinary large GCMN of the back that was excised and where the defect was reconstructed with epidermis and superficial dermis of the lesions.

2. Patients and methods

From August 2001 to February 2012, we treated 5 patients (1 male and 4 female) presenting with giant congenital melanocytic nevus with harvested epidermis and superficial dermis from lesions. The duration of follow-up after operation was 1 to 16 years (mean of 5 years). None of the patients had complications. The affected area ranged from 11% to 19% TBSA as shown in Table 1. Written consent was obtained from all the patients and their parents involved in this study. Even as the affected lesions were very large, the treatments were performed only in a single stage. Patients received thorough resection to deep fascia under general anesthesia.

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Table 1
Details on patient characteristics.

Patient No.	Gender	1st present age, y	Main affected sites	Size % (TBSA)	Satellite nevi	Follow-up duration, y	Pathology	Complication	Hair
1	F	13	Back, buttocks	15	Yes	16	Intradermal neurofibroma	Nil	No
2	F	8	Back, left buttocks	14	Yes	1	Intradermal	Nil	No
3	F	11	Thighs	11	Yes	3	Intradermal	Nil	Yes
4	F	15	Back	13	Yes	8	Intradermal	Nil	Yes
5	M	9	Trunk	19	Yes	1	Intradermal	Nil	Yes

TBSA=total body surface area.

Epidermis skin (~200–250 μm) containing the entire epidermis and the superficial dermis were obtained from the excised tissue using a dermatome. After the ablating of the deep dermis and partial subcutaneous tissue, the harvested tissue lesion including epidermis and superficial dermis was spread onto the dermabraded area in situ and secured by interrupted sutures as shown in Fig. 1. Check whether the skin is survival and the dressing was changed every other day. Four patients had complete removal of the main affected lesions while 1 patient whose lesions involve the back, buttock, and bilateral thighs had only finished the first stage

of treatment as shown in Fig. 2. The time interval between the 2 stages is recommended to be no <3 months. The follow-up was executed regularly and different levels of pigmented skin were taken from all patients at each follow-up time point for histological examination.

3. Results

The histopathological results indicated that most of the melanocytes were located in the deeper dermis, at an approximate

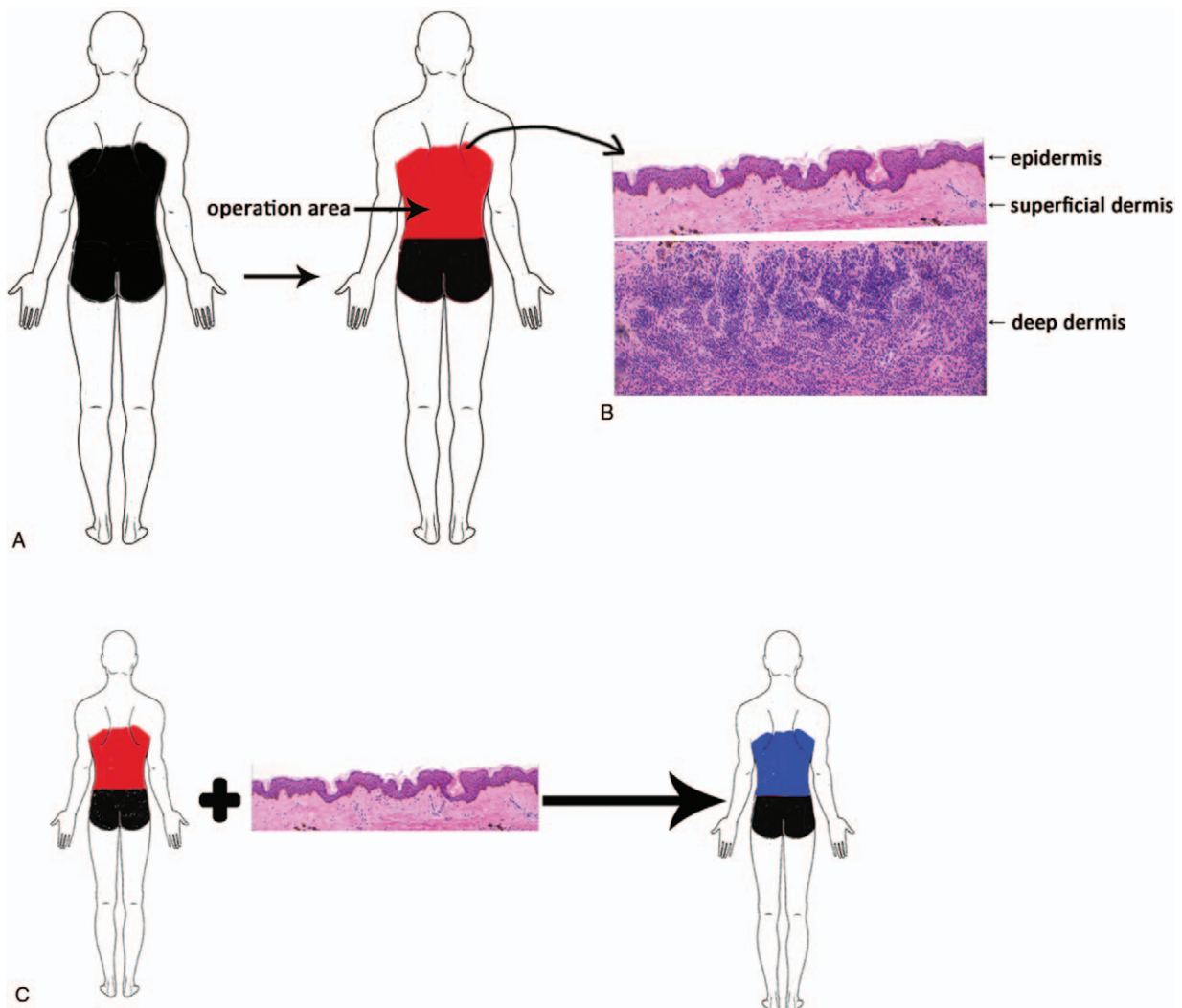


Figure 1. Pattern diagrams of surgical process.



Figure 2. Panel A shows the posterior view immediately after procedure, while B and C show the 6 and 18-month follow-up views, respectively. No reoccurrence and malignant transformation is observed. The color of arrowed areas is getting lighter compared with the status before. auto=autologous grafted skin, in situ=in situ grafted skin.

depth of 250 to 340 μm (Figs. 3A and 4A), except in 1 case with neurofibroma differentiation. In all cases, the survival rate of the grafted skin was $\geq 95\%$. In the early stage, the lesions site had the same color as before, and gradually, the pigments fade away, but it gradually became lighter (Fig. 5). No recurrence, diffusion, and malignant transformation have been seen during follow-up.

Biopsies at follow-up revealed that the melanocytes were eliminated and replaced by remodeled dermal tissue. The reconstructed skin resembled normal skin structure (Fig. 3B). These results were consistent across biopsies, although these samples were collected from different levels of pigmentation of the grafted skin.

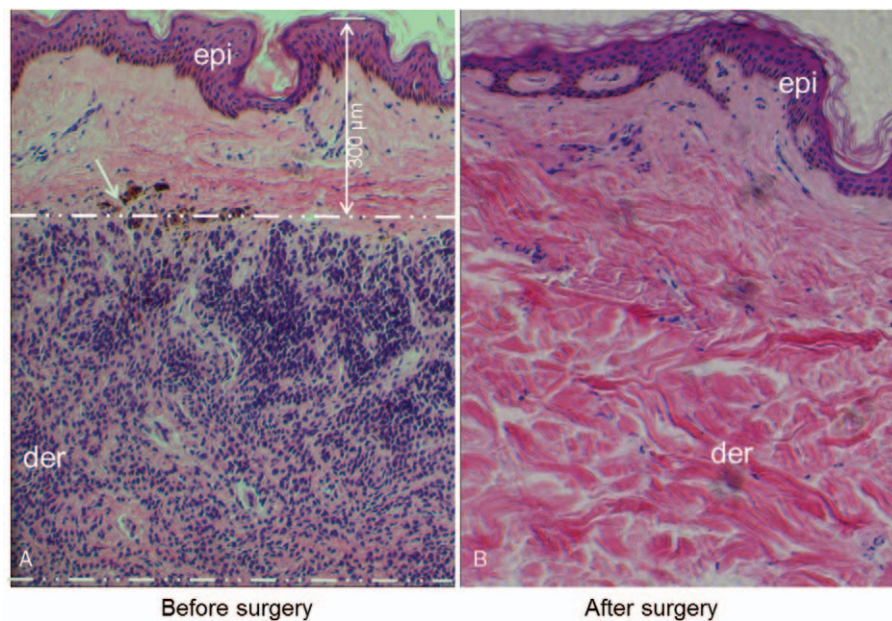


Figure 3. Histological examination of the lesion before and after the treatment by HE staining. A shows the prevalence of melanocytic cells in the dermis between the dotted red lines. B indicates the clearance of melanocytic cells with only minor melanocytic cell nests located in the basal layer of epidermis designated by white arrow. epi=epidermis; der=dermis. Magnification: 100 \times .

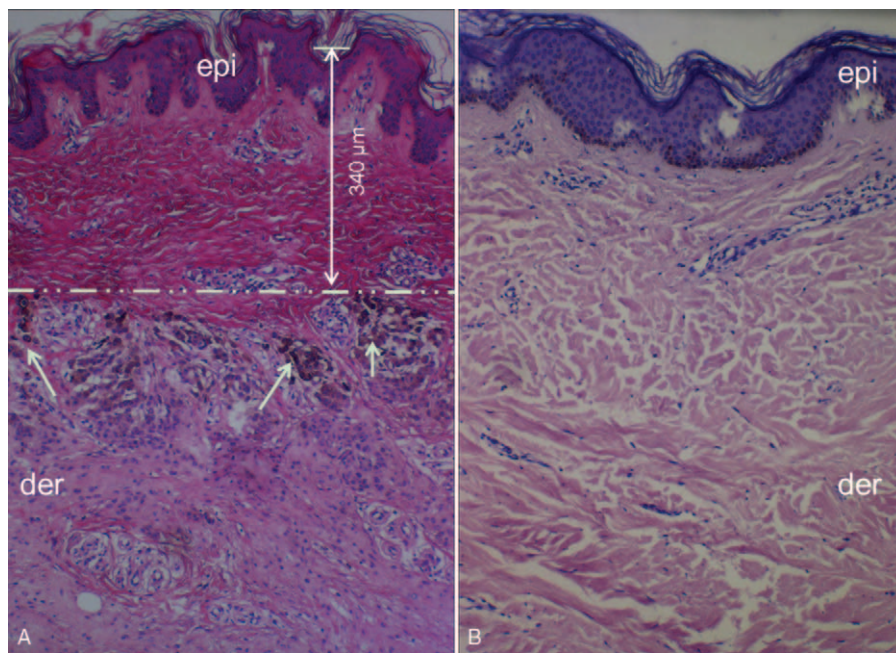


Figure 4. Histological examination of the affected skin is performed before and after the treatment. Panel A shows the prevalence of melanocytic cells indicated by white arrows in the dermis between the dotted white lines of the original lesion. Panel B demonstrates the clearance of melanocytic cells after treatment of 12 year's follow-up. epi=epidermis; der=dermis. Magnification: 100 \times .

3.1. Representative case 1

This 8-year-old girl presented with GCMN affecting the whole back. Furthermore, numerous satellite nevi were distributed all over her body as shown in Fig. 2A. The histopathological results showed that most of the melanocytes were located in the deeper dermis as shown in Fig. 4A. The main lesion was removed completely in one stage, and the wound was reconstructed with epidermis and superficial dermis harvested from the lesion itself. Initially, the grafted skin was as dark as before surgery (Fig. 2B), however, it progressively became lighter and brown without obvious hypertrophic scar formation (Fig. 2C). Neither malignancy transformation nor recurrence was observed during a 12-year follow-up. Recent biopsies showed that the dermis was free of melanocytic cells as shown in Fig. 4B. The patient is now

married and receiving multiple resections of the remaining small lesions at our clinic.

4. Discussion

The definition of giant congenital melanocytic nevi is a lesion affecting 2% of TBSA in newborns and toddlers, or a diameter >20 cm in older children and teenagers.^[1] Although the GCMN may invade the subcutaneous tissue or deeper structures, they main concentrated in dermis especially the deep dermis considering their histologic features (Figs. 3A and 4A).

Malignant transformation should be the primary consideration when we try to treat GCMN. The risk of malignant transformation in GCMN has been reported to range between 0% and 42%.^[7,8] As above mentioned the GCMN can also bring



Figure 5. Panel A illustrates a preoperative view, while Panel B 2-week postoperative and Panel C 12-year follow-up. The lesion was flat and the color is obviously lighter with only mild hypertrophic scars formed between skin grafts indicated by white arrows.

great psychological and physic damage. All the 5 cases documented here have numerous satellites nevi all over the surface of the body. Thus, it is challenging to remove all the affected lesions toward entirely eliminating the potential for malignancy and surgical excision should be performed in early childhood.

Traditional surgery techniques including tissue expansion are mature, but its limits have inhibited its applying in GCMN treatment. Morimoto et al^[9] had treated the GCMN combining inactivated autologous nevus tissue by high hydrostatic pressure and a cultured epidermal auto graft and achieved great results. And this method not only diminishes the damage of the normal skin, but also acquires good appearance. Its drawbacks of multistage surgery and asymmetry of epidermis, however, is obviously.

In our study, we ablated the lesions completely. And then, the subcutaneous tissue and deep dermis were removed, it is better to leave the epidermis as thin as possible so as to eliminate most of the nevus cells. Following the epidermis and superficial dermis were harvested from the lesions as graft substitute, it was grafted onto the lesions defects in situ. All of our grafts survive well and the pigments fade away gradually following the nevus cells that produce the pigment will not be present. As the follow-up results, we can see rare melanocytes in dermis as shown in Figs. 3B and 4B. That is probably why the in situ grafted skin becomes less pigmented after surgery. The remaining melanocytes and melanin at the base of the epidermis may contribute to the maintenance of dark color. It is an interesting hypothesis that could lead to better treatments that combine this method with laser abrasion to remove the superficial remaining melanocytes and melanin. For the first patient, we have followed up 16 years, and there are no complications including infections, malignant transformation.

Although malignant transformation was not seen in all cases in our study, periodical follow-up, usually every 6 months, was

strictly required. Also, both patients and their parents were asked to observe the lesions closely, including lymph node palpation and any changes to the color, texture, or size of the lesions. In this study, we only report on 5 cases, some of which have insufficiently long follow-up durations and, therefore, need to be tracked longer term. Broader basic and clinical research into this new technique is also required.

In conclusion, for patients older than 8 suffering from “super” GCMNs, grafting with in situ split thickness skin harvested from the lesion provides a new modality, eliminating the sacrifice of normal skin and reducing the risk of malignant transformation.

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