

Surgical Management of Giant Congenital Hairy Nevi Without Skin Graft or Other Methods of Closure

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Background: Congenital hairy nevus refers to pigmented skin lesion presents at birth. Giant hairy nevus (GHN) is disfiguring and carries a great emotional distress on the patient and family. It also imposes a higher risk for developing malignant melanomas and other tumors. The management of GHN depends on the lesions' size, location and its propensity to become malignant. When surgery is indicated, the simplest approach is a staged serial excisions. The longstanding challenge is in reconstructing the raw surface defect, especially if large.

Method : A case of congenital GHN on the trunk, abdomen and bilateral upper thighs was presented and the data was taken from the medical record from the Plastic and Reconstructive Surgery Division of Cipto Mangunkusumo Hospital.

Results : Patient was managed surgically with a technique similar to dermabrasion, by scalpel instead of dermabrator, to peel the skin in partial-thickness. In each surgery, not more than 10% of the total body surface area was excised and left to heal secondarily. Compression was used to reduce risk of hypertrophic scar formation. Subsequent operation was ideally 3 weeks apart, to allow for the excised surface to epithelialize and the body to return to homeostasis.

Summary : Although visible, the patient's family prefer the resulting scar than the initial lesion, because the skin is lighter, hairless, and less frightening. At two-years follow up, the patient still refuse further operation due to economical and social issues.

Keywords : congenital nevi, giant nevi, melanocytic nevi, hairy nevi

Pendahuluan: Nevus kongenital berambut (GHN) adalah lesi kulit terpigmentasi yang dibawa sejak lahir. Bila ukurannya besar, lesi ini sangat mengganggu penampilan dan dapat mengakibatkan gangguan emosional pada pasien dan keluarganya. GHN juga berpotensi menjadi melanoma maligna maupun tumor lain. Tata laksana GHN bergantung pada ukuran lesi, letak, dan tendensinya untuk menjadi lesi ganas. Bila diputuskan melakukan intervensi bedah, yang umum dilakukan adalah eksisi serial bertahap. Namun yang selalu menjadi tantangan adalah bagaimana menutup defek yang ditinggalkan, terutama bila luas.

Metodologi: Melaporkan satu kasus GHN pada batang tubuh, abdomen, dan paha atas bilateral yang dilakukan operasi rekonstruksi di Divisi Bedah Plastik dan Rekonstruksi RS Cipto Mangunkusumo. Data diambil dari rekam medis dan dokumentasi saat pembedahan.

Hasil : Pasien ini dilakukan intervensi bedah menggunakan teknik seperti dermabrasi, namun menggunakan pisau bukan dermabrator, untuk mengeksisi sebagian ketebalan kulit. Pada tiap operasi, tidak lebih dari 10% area luas tubuh dieksisi. Luka kemudian dibiarkan sembuh sekunder. Untuk mencegah terbentuknya skar hipertropik, dilakukan kompresi pada daerah luka. Idealnya, operasi berikutnya berjarak 3 minggu kemudian, agar kulit sudah reepitelisasi dan fisiologis tubuh kembali seimbang.

Ringkasan: Meski menyisakan bekas, keluarga pasien lebih memilih skar pasca operasi dibanding lesi awal karena warna kulit tidak segelap awal, dan rambut-rambut hilang. Pada follow up 2 tahun setelah operasi, pasien masih belum menjalankan operasi keduanya atas alasan sosial dan ekonomi.

Kata Kunci: congenital nevi, giant nevi, melanocytic nevi, hairy nevi

Melanocytic nevi are pathologic accumulation of melanocytes of neuroectodermal origin¹. Congenital melanocytic nevi are lesions appearing at birth, and acquired nevi are those appearing postnatally. Congenital melanocytic nevus is often accom-

panied by abnormal hair growth and also known as congenital hairy nevi. By definition in several literatures, the lesion is said to be giant when: it affects a major part of an area such as face or hand, the greatest diameter of the lesion measures more than 20 cm, by surface it covers

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>1% total body surface area in head and neck region or >2% elsewhere, the lesion surface encompasses area greater than 100 cm² or when the nevi cannot be excised in a single operation²⁻⁶.

Giant hairy nevus is a rare phenomena, presents at about one in every 20,000 births⁷. The clinical presentation varies in size, location, and macroscopy; with most lesions occurring in the trunk, followed by the extremities then head and neck⁸. The lesion may change with age. Hairless, pale brown, flat lesion evolves over months and years to be hyperpigmented, hypertrichosis, eroded or ulcerated, with verrucous texture and nodularity⁹. Eighty percent patients has satellite nevi elsewhere¹⁰. The different presentation and progression of congenital nevi than acquired nevi can be explained histologically^{1,9}. Congenital nevi display larger cell size with greater cellularity in varying architecture and morphology. The cells spread to deeper dermis, the subcutaneous, reaching as deep as the glands, follicles, vessels, nerves, up to fascia or muscle. This explains why some lesions has pruritus, tenderness, xerosis, and anhydrosis.

Giant congenital hairy nevus (GCN) are disfiguring and will invoke psychosocial problems on the patient and family. Many feel ostracized, embarrassed, and has low self-esteem. Study shows 30% of patients have social problems and 26% suffer from emotional distress^{11,12}. Aside of the aesthetic and psychosocial reasons, GCN also imposes a higher risk for developing malignant melanomas and other tumors such as liposarcoma or rhabdomyosarcoma when the pathologic melanocytes infiltrate the fat or muscles. The risk congenital nevi transforming into melanoma is 8.2%, a 52% higher risk than the general population¹³.

Another associated rare but deadly condition, neurocutaneous melanosis, occurs when melanocytic cells infiltrate into the central nervous system, found in up to 11.4% of GCN cases¹⁴. A patient with GCN has the highest risk for malignant transformation before the age of 3-years¹⁵. The risk of melanoma decreases as the patient gets older, but increases with larger nevus size¹³.

The management and treatment of GCN has been a longstanding debate, whether or not to obliterate asymptomatic lesion, and by what means if treatment is warranted. Factors to be considered are the size of the lesion, its location, and the propensity to become malignant. The three indications of GCN treatment are curative, preventive, and aesthetic¹⁶. When malignant transformation is suspected, removal of lesion is curative. If the risk of a lesion developing into melanoma is considered high, treatment is for preventive purposes. When the lesion disturbs one's view of themselves, treatment is warranted to improve psychological and social aspects.

Removal of GCN can be attempted with or without surgery. Non-surgical means include the use of laser, chemical peels, dermabrasion, or curettage¹⁷. These modalities are limited to removing melanocytes up to a certain dermal depth and often do not totally remove lesions. A more popular approach among the surgeons are surgical removal of visible lesions, with the challenge in reconstructing presenting defect after nevi excision. Up to a certain size, a staged serial surgical excisions is the most straight forward approach. In very large lesions however, serial excisions may require too many stages that it is more rational to excise a larger area of lesion then utilize one of the reconstructive options to cover for the defect. The choices of reconstruction include closure of defect by direct cutaneous advancements, split or full thickness skin grafts, tissue expansion followed by local flaps, use of skin substitutes, or performing free tissue transfers. We report an alternative surgical method of treatment in a case of diffuse giant congenital hairy nevus.

METHODS

A 6-years-old girl came to our clinic with a complain of hairy dark-grey lesions presented since birth covering half of her anterior and posterior trunk, the abdomen to the pubic and inguinal region, the lower back and gluteal area, bilateral anterior and posterior upper thighs, and multiple satellite nevi across other body parts (Figure 1). This pattern of lesion is also known as the Bathing Trunk nevus. The parents wished for the lesions to be removed.



Figure 1. A 6-year-old girl with a diffuse giant congenital hairy nevus. Left: anterior view. Middle: posterior view. Right: Closer view of the lesion displaying hirsutism of the area with nevi involvement.



Figure 2. First stage split-thickness nevi excision not exceeding a 'superficial second degree burn' on the anterior trunk and abdomen.



Figure 3. Four-weeks post split-thickness skin excision with some deeper excised part showing hypertrophic scarring. Compression dressing was then worn.



Figure 4. Scar appearance one-year post split-thickness skin excision.

Due to depth of the nevi, chemical peels, dermabrasion, and laser surgeries are not liable. The extensive surface involvement made serial excision with direct closure impossible to attain within a reasonable amount of surgeries and time. Skin grafts, tissue expansion, and free tissue transfer are also ineffective owing to the minimum availability of normal skin. The family also has a limited financial resource hence skin substitutes are not opted for this case. Parents were advised on the limited reconstructive options applicable to the patient, and explanation was provided regarding the calculated risk of such lesion developing into malignancy at the time of presentation—in which judging by the

age of patient and the smooth even surface of lesions, the risk of melanoma was not imminent. Even so, the parents wished for the lesion to be removed.

Serial split-thickness dermal excisions were done in multiple stages, with a technique similar to dermabrasion but in this case a scalpel was used to peel partial-thickness of the dermis, not exceeding a superficial second-degree burn depth. In each surgery, lesion of approximately 9% (<10%) total body surface area is excised then left to heal secondarily. Figure 2 shows the first stage excision of lesion on the anterior trunk and part of the abdomen. Non-adhesive moist dressing principles were applied and

dressing changed every 2-3 days under tolerable level of pain. Epithelialization was attained within two-weeks post surgery. Figure 3 depicts the state of lesion four-weeks post split thickness skin excision, with slight hypertrophy of some wound on the abdomen because of the deeper skin excision done. Compression dressing was then worn to reduce the risk of hypertrophic scar formation. Figure 4 displays the resultant scar appearance one-year post excision.

RESULTS

In the management of this diffuse GCN with limited reconstructive options, although the split-thickness skin excision without skin grafting—or other method of defect closure—results in scarring, the scar quality was acceptable to the family. A study showed that although aesthetically displeasing, most family and patients with congenital hairy nevus prefer scars rather than the original lesion most likely because scars are more socially acceptable¹¹. Other disadvantage of this technique is the unexcised deeper lesion, leaving some melanocytes behind. These cells still hold the propensity of becoming malignant some day. The advantages gained include fulfilling parental preference, reduced risk of melanoma because a great part of the melanocytes are excised, lighter and hairless skin color.

DISCUSSION

The staged operation should be timed at least 3 weeks apart, allowing for the raw surface to epithelialize and homeostatic balance to return to baseline. One year after the first excision, the second operation was not yet done because the patient had been out of reach. Recently the patient and her parents returned to our clinic expressing their plan of having the next stage of nevi excision, but it is not yet performed to date due to administrative delay from the foundation who was going to facilitate her further treatment.

CONCLUSION

Although visible, the patient's family prefer the resulting scar than the initial lesion,

because the skin is lighter, hairless, and less frightening. At two-years follow up, the patient still refuse further operation due to economical and social issues.

Hence, we conclude that even though this technique is not ideal aesthetically, it serves the treatment goals in this case, or perhaps other cases alike.

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