Giant congenital melanocytic nevi (GCMN) are benign proliferative tumors present at birth consisting of melanocytic cells (nevus cells) that are found in the epidermis, dermis, or other tissues and will reach >20 cm in adult life. The incidence of GCMN is described to be <1:20,000 newborns, although smaller congenital melanocytic nevi are present in about 1% live births. The larger garment-like GCMN, as presented in this case report, has an even rarer incidence of approximately 1:500,000.

GCMN are associated with the risk of developing malignant melanoma, which is recently reported to vary from 0% to 3.8%. Other authors have reported higher incidence although these studies are relatively old with a small number of patients. Still there is a controversy of the real incidence of malignancy leading to more conservative management in many units compared to a few decades ago. Regardless of the malignant potential, these patients also have to be carefully evaluated due to the association of central nervous system involvement and often huge psychosocial impact.

Having once decided to perform surgery, there are many options described: staged excision, skin grafting, tissue expansion, tangential shaving, dermabrasion, curettage in the first week of life, and pedicled or free flaps.

An additional treatment option that has been established during the last 2 decades for the treatment following skin defects after primarily large burns is the use of Integra dermal regeneration template (Integra Life Sciences, Plainsboro, N.J.). However, the use on patients with GCMN is limited. We will present a patient with an extraordinary large GCMN of the back that was excised and where the defect was reconstructed with Integra dermal regeneration template and split-thickness skin transplant.

CASE REPORT

The patient was born abroad after a normal pregnancy. Immediately after delivery, a very large voluminous GCMN of the back covering about 16% of the total body surface area was found. The nevus was excised and a large skin defect was reconstructed with Integra dermal regeneration template and split-thickness skin transplant.

Summary: Garment-like giant congenital melanocytic nevi are very rare, and those being treated are most often offered excision and split-thickness skin transplantation. Due to the risk of restricted mobility secondary to shrinkage and hypertrophic scarring of the transplant, we treated to date the largest reported giant congenital melanocytic nevus with Integra dermal regeneration template (Integra Life Sciences, Plainsboro, N.J.), giving a more functional skin reconstruction. In addition, the dermal regeneration template had to be covered with split-thickness skin transplant including multiple smaller nevi due to lack of larger area with normal skin.

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the total body surface area and additional multiple smaller nevi on the body were demonstrated. The patient and his family immigrated to Norway, and he was referred to our hospital when he was 1.5 years old (Fig. 1). His main complaints were severe itching and difficulty in normal everyday activities and sleeping due to the large tumor.

Based on the patient’s complaints, we decided to perform a two-stage operation with (1) excision of the large tumor (Fig. 2) and coverage with Integra (Fig. 3) and (2) 3 weeks later split-thickness skin transplantation of the 100% vascularized Integra. The split-thickness skin transplantation was harvested with 0.008-inch thickness, meshed 1:1.5, and fixed with vacuum-assisted closure technique at 80 mm Hg (Smith & Nephew, Hull, United Kingdom). Due to multiple smaller nevi on the rest of the body, the split-thickness skin transplant was harvested including these lesions. After additional 5 days, the bandage was removed and 90% of the split-thickness skin transplant had healed. A secondary split-thickness skin transplantation procedure was performed, and 2 months after the primary operation the defect was completely healed (Fig. 4).

**DISCUSSION**

Reconstruction of very large GCMN offers substantial surgical challenges. After excision, reconstruction of the full-thickness defect can be performed with different techniques, although as in this case, with a very large voluminous tumor, many of the conventional procedures would have been inappropriate. The tumor was too large for staged excision, tissue expansion, and pedicled or free flaps. Complete excision with skin transplantation could have been performed, but due to the risk of transplant shrinkage and the potential restricted movement of the back, especially in relation with increased growth, in addition to the risk of hypertrophic scarring, we wanted to perform a two-stage operation with Integra with a thin split-thickness skin transplant.

Integra dermal regeneration template consists of a two-layer skin regeneration system. The outer layer is a thin silicone film that acts as the patient’s epidermis. The inner layer is made of a complex matrix of cross-linked fibers that acts as a scaffold for regeneration of dermal skin cells. About 3 weeks after application, the silicone layer is replaced with a
Tønseth et al. • Treatment of Melanocytic Nevus with Integra

thin (0.004–0.008 inch) epidermal skin graft. This procedure gives a more functional and very pliable skin reconstruction compared with conventional skin transplantation, which is desired on skin defects in areas which are exposed to much movement and load. In addition, the donor-site morbidity is reduced because one can harvest and use thinner split-thickness skin grafts. Although Integra has its advantages, the main concerns have been the risk of infection, the need for two-stage procedure, and its costs.

Schiestl et al. presented the use of Integra in 12 children with GCMN covering 1–12% of the total body surface area (mean, 4.2%). The age of the patient when treated ranged from 0.6 to 11 years (mean, 5.8 years). In 8 children, Integra implantation was primarily successful, and in 4 patients, a partial or complete removal and reimplantation were necessary due to complications. The functional and cosmetic outcome was rated excellent in 58%, good in 25%, and fair in 17%. Unlike the patients described in the article by Schiestl et al., our patient had a larger GCMN and multiple smaller nevi, which made coverage of the Integra with split-thickness skin transplant without including these lesions almost impossible. The postoperative result therefore showed a reconstructed skin defect with these multiple smaller nevi, although with a very good functional condition.

The recent research on CMN has led to a more conservative approach when it comes to the role of surgery. In an article from Great Ormond Street Hospital, the following conclusions are given: (1) untreated CMNs can lighten spontaneously, (2) routine surgery has not been demonstrated to reduce the risk of malignancy and is, therefore, for cosmetic reasons only, (3) early surgery has not been shown to be advantageous and carries increased anesthetic risk, and (4) there is some evidence that surgical intervention may adversely affect the behavior of the CMN cells. Our department agrees with these statements. However, as described in this article, some patients with GCMN can in addition to cosmetic concerns have significant functional limitations, which should also be evaluated as an indication for surgery.

CONCLUSION

Treatment of extraordinarily large GCMN with Integra dermal regeneration template can give very satisfactory results and should be considered in areas that are exposed to much movement and load.

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PATIENT CONSENT

Parents provided written consent for the use of the patient’s image.

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