Malignant Melanoma Arising at the Site of a Previously Excised Giant Congenital Melanocytic Nevus

Patients with giant congenital nevi are at increased risk for malignant melanoma. Most reported cases involve development of melanoma during childhood, usually before age 6 years.1-3 Herein, we report a case of a man with a giant congenital nevus and satellite nevi who underwent multiple excisions and grafting procedures as a child and developed malignant melanoma more than 30 years later.

Report of a Case | A man in his 40s with a history of a giant truncal congenital nevus and many satellite nevi presented with concern for a tender nodule on his right posterior thigh (Figure 1). Notably, over a 4-year period in childhood, he underwent 10 to 11 serial surgical excisions with accompanying split-thickness graft reconstruction to attempt nevus removal. Histologic analysis showed the presence of nevus cells at the deep margin of the final excision. He had no family history of skin cancer.

At presentation, the nodule had been palpable for 2 months and had not improved with oral antibiotic treatment prescribed by his primary care physician. On physical examination, he had a 2.0 × 1.5-cm subcutaneous, tender nodule with an overlying well-healed skin graft of the upper right posterior thigh. There was no palpable lymphadenopathy. Biopsy showed a dermal-based atypical melanocytic proliferation with epithelioid nests of pleomorphic melanocytes with prominent nucleoli and infiltration into the subcutis. Multiple mitotic figures, focal lymphovascular space invasion, and necrosis were seen (Figure 2). These findings were consistent with metastatic melanoma; no epidermis was present in the specimen. The patient underwent positron emission tomography/computed tomography, and the findings were negative for metastatic disease.

The patient underwent wide local excision and sentinel lymph node biopsy. Histologic examination revealed a nevoid melanoma, lymphovascular space invasion, and foci of residual benign congenital melanocytic nevus. Microscopic metastatic disease was present in 2 of 4 right inguinal sentinel lymph nodes. Completion lymph node dissection revealed no additional disease. His tumor was negative for a BRAF V600E mutation. The patient was referred to medical oncology for evaluation of his stage IIC melanoma and underwent treatment with pegylated interferon.

Discussion | The patient underwent several staged excisions and grafting procedures as a child to address his giant congenital melanocytic nevus (GCMN) and numerous congenital satellite nevi. His melanoma was diagnosed 34 years after the final stage of his GCMN excision. To our knowledge, melanoma in a site of excision and grafting has been reported only 4 times.2,4-6 It has also been reported after dermabrasion for congenital nevi.7 The risk for malignant melanoma in patients with GCMN has been variably reported but is agreed to be greater than that of patients without GCMN.1

As in this patient, it has been postulated that melanoma arises more deeply in patients with GCMN.1 Our patient’s age at presentation is older than average. In 205 patients in the New York University Large Congenital Melanocytic Nevus registry,7 70% of melanomas (7 of 10) developed in patients 3 years or younger, and 30% (3 of 10) developed in patients 35 years or older. Similarly, in a cohort of 1008 patients with large or multiple congenital melanocytic nevi, only 27% of melanomas (4 of 15) were diagnosed in patients older than 30 years.2 Though most melanomas arising in patients with GCMN have been reported in young patients,1 the risk for malignancy persists with age.
Congenital satellite nevi are known risk factors for neurocutaneous melanosis. Their impact on the risk for malignant melanoma is not as well established, but some studies point to a possible increased risk in patients with GCMN and satellite nevi. In summary, our patient is an example of the need for continued dermatologic follow-up and high index of suspicion for new nodules appearing in patients with a history of GCMN.

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**Perineal Groove: A Report of 2 Cases**

A perineal groove is a rare and usually uncomplicated congenital malformation of the perineum characterized by a wet sulcus that extends from the posterior fourchette to the anus. Of 12 previously described cases, only 1 has been reported in the dermatology literature.

**Report of Cases**  
*Case 1.* A 4-month-old term white girl was referred to pediatric dermatology by her pediatrician and a pediatric surgeon for evaluation of a congenital perineal lesion. The mother's pregnancy was unremarkable, and the infant was healthy. The lesion had been stable and uncomplicated since birth. Barrier creams were attempted for presumed diaper dermatitis without response. Physical examination showed a well-demarcated, erythematous, superficial ulcer anterior to the anus (Figure, A). A fungal culture was negative. At the 1-month