Desmoplastic Hairless Hypopigmented Nevus

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PRACTICE **POINTS**

- Desmoplastic hairless hypopigmented nevus (DHHN) is a giant congenital melanocytic nevus that shows progressive loss of hair, desmoplasia, and progressive reduction in the number of nevus cells.
- Progressive reduction in the number of nevus cells in DHHN reduces the potential for malignant transformation.

To the Editor:

We report 2 cases of desmoplastic hairless hypopigmented nevi (DHHN), which are giant congenital melanocytic nevi (GCMN) that show sclerosis with progressive loss of pigment and hair. These changes in GCMN could be considered signs of regression.

A 6-year-old boy presented in the dermatology department with an asymptomatic skin lesion on the right buttock since birth. The parents claimed that the lesion was darkly pigmented at birth and gradually increased in size, with progressive reduction in color in the last 2 years. Physical examination revealed a 10×6-cm, well-defined, raised plaque on the upper medial side of the right buttock (Figure 1). The plaque was firm with a shiny smooth surface and was devoid of hair. The surface was flesh colored with scattered pigmented spots. A punch biopsy of the lesion showed increased melanin content in the basal cell layer. The upper dermis showed small nests of epithelioid nevus cells, most of them containing melanin pigment (Figure 2). In the lower two-thirds of the dermis, nevus cells were both epithelioid and spindle shaped and were arranged in between thick sclerotic collagen bundles with an increased number of fibroblasts. There was a marked reduction in the number of hair follicles. Immunohistochemical staining results were S-100 positive and CD34 negative.

A 5-year-old boy presented in the dermatology department with a large hairy GCMN covering most of the trunk since birth. In the last

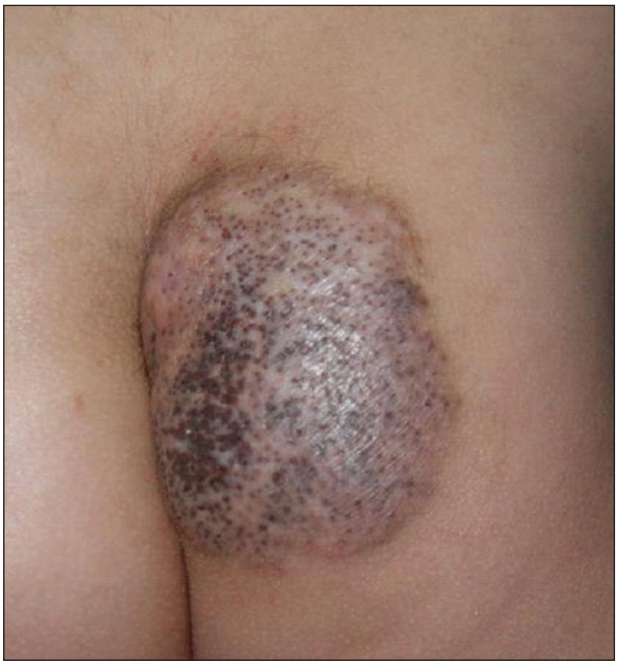

Figure 1. Giant congenital melanocytic nevus on the upper medial side of the right buttock.

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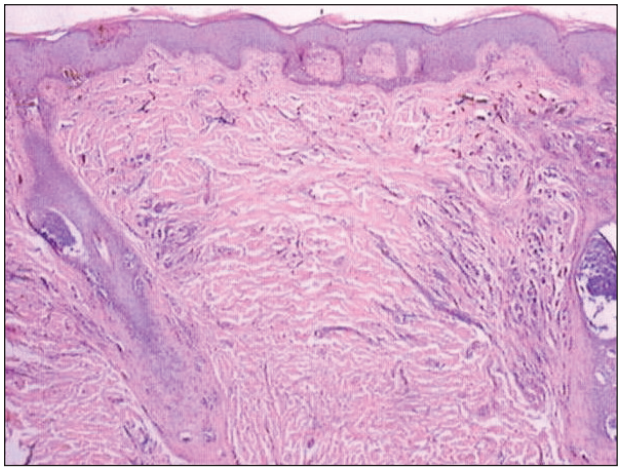

Figure 2. Histopathology showed nests of melanocytes within the upper half of the dermis. A few nests were present at the dermoepidermal junction and follicular epithelium (H&E, original magnification ×40).

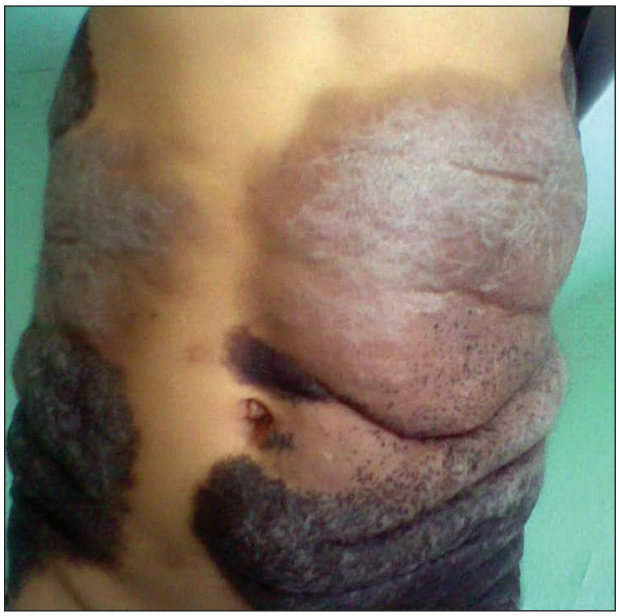

Figure 3. Giant congenital melanocytic nevus covering the anterior aspect of the trunk.

1.5 years the parents noted gradual fading of color, decreased hair density, and increased induration of the nevus. Physical examination revealed a large plaque covering the anterior aspect of the trunk (Figure 3) and the back extending down to the buttocks. The lesion formed large skin folds that were more pronounced on the back. The nevus was darkly pigmented with large areas of lighter color that were indurated, devoid of hair, and showed small spots of dark pigmentation. A punch biopsy from the lesion showed small nests of nevus cells in the upper part of the reticular dermis. In the lower part of the dermis, nevus cells were arranged in single units in between thick collagen bundles.

In 2003, Ruiz-Maldonado et al[1] described 4 cases of GCMN that showed progressive loss of pigmentation, sclerosis, and hair loss. They proposed the term *desmoplastic hairless hypopigmented nevus* for their cases and considered it as a variant of GCMN.[1] Prior to these reported cases, 2 similar cases were described. The first was a report by Hogan et al[2] in 1988 of a 7-month-old girl with a GCMN involving the occipital area and the upper back that became indurated and ulcerated with progressive involution that led to complete disappearance of the nevus. The second was a report by Pattee et al[3] in 2001 of a newborn with a GCMN located on the trunk with progressive sclerodermiform reaction. After surgical excision of the nevus, the sclerotic margin disappeared.[3]

Following the report by Ruiz-Maldonaldo et al,[1] 5 more cases of DHHN were described.[4-8] All cases of DHHN share the same clinical and histopathological features. The clinical features include a GCMN present since birth with progressive sclerosis over time and loss of both pigmentation and hair. Histologically, DHHN shows the typical changes of a congenital melanocytic nevus with decreased numbers of nevus cells, thick sclerotic collagen bundles of the reticular dermis, increased number of fibroblasts, and decreased number of hair follicles. The progressive reduction in the number of nevus cells in melanocytic nevi is considered a sign of regression. Spontaneous regression was rarely described in GCMN, and all the reported cases of regression were associated with desmoplasia.[4] Desmoplasia is thought to be induced by either melanocytes that function as adaptive fibroblasts or by fibroblasts themselves, as fibroblasts can show multifunctional differentiation capabilities.[9] The direct correlation between the increased induration of DHHN and pigment depletion supports the former hypothesis. The absence of inflammatory cells within the sections of DHHN lesions is against the possibility of an immune-mediated reaction as a cause for the clinical and histological changes seen in this rare form of GCMN. The progressive hair loss in DHHN may be explained by the progressive fibrotic changes in the reticular dermis that affect the blood supply to follicles, leading to atrophy or even absence of the follicles. The progressive reduction in the number of nevus cells in DHHN reduces the potential for malignant transformation and hence following a watchful waiting strategy is a reasonable way to manage these nevi.

We present 2 patients with DHHN, which is a rare form of GCMN that shows signs of regression. The cause of these changes is still unclear.

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