Life-threatening blood loss from scratching provoked by pruritus in the bulky perineal nevocytoma variant of giant congenital melanocytic nevus in a child

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We describe a 3-year-old girl with intractable, debilitating pruritus associated with a giant congenital melanocytic nevus, resulting in life-threatening anemia from extensive bleeding skin excoriations. Multiple conventional oral and topical antipruritic medications failed to provide relief, but the patient was successfully treated with the selective serotonin 5-hydroxytryptamine type 3 inhibitor ondansetron, suggesting a serotonin-related mechanism to her pruritus. (J Am Acad Dermatol 2005;53:S139-42.)

Giant congenital melanocytic nevi (GCMN) are pigmented skin lesions occurring in approximately 1 in 20,000 newborns.[1] Besides the obvious cosmetic disfigurement, these patients have an increased lifetime risk of malignant melanoma[2] and neural crest proliferations within the spectrum of neurocutaneous melanosis.[3] Thus far, no reports of GCMN causing pruritus have been documented to our knowledge. We describe an unusual case of a 3-year-old girl with intractable pruritus in her GCMN, who continuously bled from her excoriations.

CASE REPORT

A 3-year-old, African American girl with a history of intractable, localized pruritus in her bathing-trunk GCMN was hospitalized for acute respiratory distress and high-output heart failure secondary to severe anemia (hemoglobin, 3.1 g/dL; hematocrit, 10.7%). She had undergone 4 partial surgical reductions of the nevus in her genital/perineal area and back, all of which were successful without extensive scarring or keloid formation.

STIEFEL® Research in Dermatology Supported by Stiefel Laboratories.

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Funding sources: None.
Conflicts of interest: None identified.
Reprints not available from the authors.

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0190-9622/$30.00

doi:10.1016/j.jaad.2004.12.040

The patient's past medical history was significant for a hospitalization 7 months before caused by severe anemia (hemoglobin, 5.7 g/dL; hematocrit, 18.9%), with guaiac-positive stools and active bleeding from perianal ulcerations in the area of the nevus. She was transfused, and iron supplementation was started at that time.

Physical examination revealed a poorly nourished child with markedly increased work of breathing, bilateral pulmonary crackles, and anasarca. No hepatosplenomegaly was detected. A GCMN in a bathing-trunk distribution extended from her lower abdomen and back to her upper thighs, completely covering her perineum and buttocks, with numerous terminal hairs distributed throughout. Most notably, the labia were grossly hypertrophic and edematous. The clitoral hood measured 5 cm × 4 cm × 2 cm and had a verrucous and papillated surface (Fig 1). Erosions and shallow ulcerations oozing blood were found on her labia, perianal region, and inner buttocks. Her diaper contained bright red blood mixed with liquid stool. Although the patient had a family history of atopic dermatitis, minimal clinical features suggesting active atopic or other types of dermatitis were seen on examination.

Workup for the anemia included a peripheral blood smear showing microcytic, hypochromic red blood cells. Iron studies were consistent with iron deficiency anemia. A colonoscopy showed normal mucosa without evidence of bleeding. A coagulopathy workup was also negative.

The patient was intubated 4 hours after presentation and was repeatedly transfused. Over the next 4 days, she continued to scratch and bleed from the superficial erosions in the GCMN, saturating her diapers with blood. It became clear that the anemia

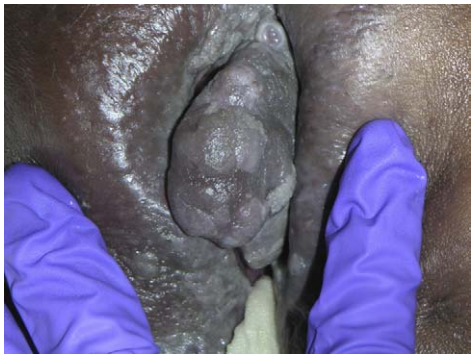

Fig 1. Clitoral hood measuring 5 cm × 4 cm × 2 cm, and showing verrucous and papillated surface with ulceration.

was secondary to blood loss from her external wounds in the nevus secondary to scratching.

Between the 2 hospitalizations, the patient had been seen in the emergency department numerous times with uncontrolled bleeding from excoriations of her perineal and perianal region. These wounds had been attributed to constant scratching of her GCMN because of intractable pruritus, which became progressively worse, impairing the patient's ability to walk. During this period, the following treatments were tried, all without beneficial effect. Oral medications included doxepin, hydroxyzine, and diphenhydramine. Topical treatments included doxepin cream, calamine, camphor, menthol, mupirocin, ketoconazole, iLEX skin barrier, zinc oxide, hydrogel, hydrocortisone, anesthetics, and emollients. The pruritus worsened and the patient's labia enlarged and became hypertrophic (Fig 2).

During the second hospitalization, the patient's perineal wounds were managed with hydrocolloid dressings, barrier cream, iLEX, and Vaseline. Oral doxepin, hydroxyzine, diphenhydramine, and analgesics plus topical antipruritics, antifungals, and anesthetics failed to relieve the pruritus. Despite hand restraints, the patient awoke at night, using her feet and inner thighs to scratch.

At this time, a trial of ondansetron 0.6 mg orally twice daily was started on hospital day 7, to which the patient responded with mild improvement of her pruritus. She was discharged on ondansetron; upon 6-week follow-up the patient's pruritus was significantly improved, her wounds were healing, and further reduction surgeries were scheduled.

ANATOMIC PATHOLOGY

Specimens derived from the surgical reductions of this lesion featured the characteristic histologic appearance of a GCMN. In the initially resected skin, the epidermis showed mild hyperkeratosis and

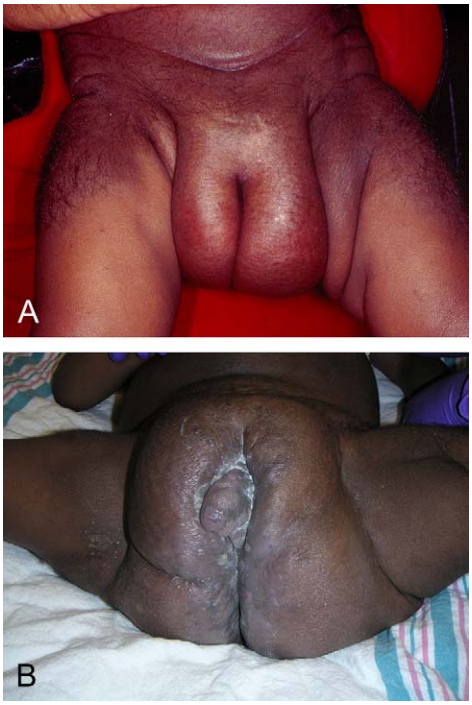

Fig 2. Comparison of patient's perineal region (**A**) 2 years before and (**B**) during hospitalization, showing remarkable hypertrophy and growth of the clitoral hood.

moderate hyperpigmentation of the basal layer. The dermis revealed a diffuse proliferation of nevus elements arranged in nests with prominent melanin deposits and extensive formation of Wegner-Meissner–like bodies (*lames foliacées*), recapitulating the neural crest derivation and ability to undergo neuroid differentiation of these nevus cells. This proliferation extended very deeply into the dermis and subcutaneous tissues, where the melanocytes exhibited the characteristic spindle morphology (Fig 3, *A*). These features have been previously recognized as prominent in massive nevomelanocytic lesions involving the perineal area, representing the nevus variant named *bulky perineal nevocytoma*.[4]

In view of the intractable pruritus, immunohistochemical labeling of mast cells was carried out with the use of antibodies to detect CD117 (c-kit) and showed a moderately increased number of CD117-positive cells uniformly distributed at all levels of the nevus involvement (Fig 3, *B*). Biopsies from the vulva and clitoral hood showed an intradermal nevus with hyperkeratosis, papillomatosis and acanthosis, and ruled out the presence of mastocytoma and human papillomavirus infection.

DISCUSSION

Pruritus is thought to be mediated peripherally by unmyelinated C fibers (distinct from those