Becker Nevus of the Leg With Lipoatrophy

Becker nevus presents most commonly as a patchy hyperpigmentation with dark hairs on the upper arms or the shoulder girdle of male patients. Its prevalence has been determined at 0.52 percent in a large cohort of male French military recruits between the ages of 17 and 26 years.1 The male to female ratio of Becker nevus has been approximated to be about 4:1, although it may well be underdiagnosed in women owing to less intense pigmentation and milder or even absent hypertrichosis. An association with soft-tissue defects is common, manifesting most frequently as breast hypoplasia. Danarti et al2 defined a Becker nevus syndrome as the coincidence of Becker nevus and ipsilateral breast hypoplasia, scoliosis, spina bifida, or ipsilateral limb hypoplasia. Becker nevi of the lower extremity are exceedingly rare.

Report of a Case | We report the case of a 31-year-old woman from the United Arab Emirates who presented with a lesion that had been present since birth but darkened during adolescence. Following a cesarean section 1 year before presentation, she observed a reduction in girth of the left leg. She also complained of pain on exercise. Examination showed hyperpigmentation and lipoatrophy involving the entire left lower extremity, extending from the left lumbar region to the ankle (Figure). The lesion’s margins were irregular with satellite macules, reminiscent of an archipelago. The circumferences of the left and right thighs were 56 and 68 cm, respectively. There was only minimal hypertrichosis, and that was restricted to the ventral aspect of the left thigh. The patient underwent 7.5-MHz sonography, which revealed thinning of the subcutis of the left side (1.04 cm) compared with the right side (2.57 cm). Histopathologic analysis showed increased melanin production with pronounced pigmentation of the basal cell layer.

The patient desired to improve the cosmetic appearance of the affected leg, which required extensive plastic surgery. This, again, was declined by the patient, and she was lost to follow-up for 3 years.

Figure. Dark Pigmentation and Lipoatrophy Extending From the Hip to the Ankle

A

B

Front (A) and posterior (B) views.
Discussion | Khaitan et al,3 in 1998, were the first to describe a patient with Becker nevus of the lower limb. Subsequently, 11 more cases of Becker nevus of the leg have been reported.4 In 2 patients, the reports explicitly noted the absence of hypertrichosis. In 2 female patients, Becker nevus was associated with localized lipoatrophy: a 14-year-old girl with a soft-tissue defect on the ventral aspect of her thigh and a 45-year-old woman with lipoatrophy on the right thigh.5 The case presented herein is unusual because the Becker nevus extended below the knee and involved almost the entire leg.

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Paraneoplastic Autoimmune Multiorgan Syndrome in a Patient With Li Fraumeni Syndrome

Treatment of paraneoplastic autoimmune multiorgan syndrome (PAMS) can be challenging; a p53 tumor suppressor mutation adds a further layer of complexity.

Report of a Case | A 30-year-old African American woman with Li-Fraumeni syndrome was evaluated for a 4-month history of oral ulcerations. Her history was significant for adrenal carcinoma at age 3 years, undifferentiated leukemia at age 12 years, and breast cancer at age 18 years. Her oral ulcerations were painful causing difficulty eating and weight loss. Previous treatment with prednisone, 20 mg/d, provided limited relief.

On examination she had ulcerations affecting her labial, gingival, lingual, and buccal mucosae with no involvement of the skin or conjunctiva (Figure, A).

A biopsy from her buccal mucosa showed lichenoid mucositis (Figure, B). Direct immunofluorescence findings were negative. Immunoprecipitation assay results for bullous pemphigoid antigens 180 and 230 were negative. Enzyme-linked immnosorbent assay findings for desmoglein 3 were negative, and the desmoglein 1 level was mildly elevated at 22 U (reference range, <14 U). On indirect immunofluorescence testing, salt-split skin showed cell surface staining consistent with pemphigus, and the monkey esophagus test result was negative. Further testing on rat bladder was not done owing to timely radiologic results.

The clinical features raised concern for paraneoplastic autoimmune multiorgan syndrome (PAMS). A computed tomographic scan of her chest, abdomen, and pelvis was performed in search of an occult malignant neoplasm. The scan revealed an anterior mediastinal mass, which, on excision, was determined to be an invasive thymoma. No other organ involvement was identified.

She began treatment with prednisone, 60 mg/d, and showed improvement. Subsequently, cyclosporine, 100 mg/d (1.25 mg/kg), was added to her regimen. Excision of her thymoma did not improve control. Over the next year her prednisone dose was tapered to eventual discontinuation, and she continued taking cyclosporine as monotherapy. She improved substantially, although her response was not complete.

Figure. Photographic Images From the Subject Case

A, Severe mucosal ulceration at initial presentation. B, Lichenoid mucositis, left buccal mucosa, bandlike lymphocytic inflammation at the mucosal-submucosal junction, basal vacuolar degeneration, and pigment incontinence (hematoxylin-eosin, original magnification ×20).