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 immunosorbent 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).
Discussion | Paraneoplastic pemphigus was coined over 20 years ago by Anhalt et al1 to describe an autoimmune blistering disorder in the setting of a malignant or benign neoplasm. More recently the term PAMS has come into use, in recognition of the polymorphous nature of this multiple-organ condition.2 Diagnosis rests on clinical, histopathologic, and immunologic criteria, and these have been suggested by Czernik et al.2 PAMS is a heterogeneous condition mediated by either a predominantly autoantibody or cell-mediated cytotoxic (CMC) effect. The predominance of a CMC mechanism is associated with the lichenoid variant.2,3 The best outcomes are seen in cases where the underlying malignant neoplasm is fully resectable, with resection leading to improvement in 15 of 20 patients in one series.4 Unfortunately, our patient did not demonstrate this.

Glucocorticoids form the backbone of therapy, with additional immunosuppressive agents being required in most cases. Options for additional immunosuppressive therapy include cyclosporine, mycophenolate mofetil, cyclophosphamide, intravenous immunoglobulin, plasmapheresis, and rituximab.2,3,5 It has been proposed that many of these adjuvant immunosuppressants increase the risk of developing a malignant neoplasm. While this potential increase in risk may be acceptable in a patient at average risk, in the present case, it added a layer of complexity to our patient with a p53 mutation. We opted to use cyclosporine as a steroid-sparing agent, and our patient subsequently continued taking cyclosporine as monotherapy with a good response.

To our knowledge, this is the first reported case in English of treatment for PAMS in the setting of Li-Fraumeni syndrome. We recommend that an oncologist be involved in the choice of therapeutic agents for the treatment of PAMS in the setting of a p53 mutation.

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COMMENT & RESPONSE

A Variant of AESOP Syndrome (Adenopathy and Extensive Skin Patch Overlying a Plasmacytoma)

To the Editor | We read the letter by Foo and collaborators1 with great interest. What surprised us is not only that their enumeration of AESOP cases is wrong, but also that they failed to underline the profound differences in the histopathologic characteristics with respect to the original case of Lipsker et al.2 In fact, the number of reports of AESOP cases that have been published so far is 11 and not 9. Foo and collaborators may find the list in an article by our research group.3 The histopathologic features of their patient differ because of the absence of dermal mucin deposition, which Dr Foo and collaborators do not even mention and which is present in all other reported cases, and because of the presence of pleomorphic and multinucleated histiocytes (CD68+) and myofibroblasts, which have never been reported before. We believe that the histopathologic features of their case are not “comparable to the cases described by Lipsker et al.4,5,10,11,12” as they assert, but are distinctive. The reasons for such differences are difficult to understand. The possibility of an atypical type of multinucleated angiohistiocytoma cannot be ruled out because it is suggested by the vascular proliferation and the pleomorphic and multinucleated CD68+ histiocytes.4 Alternatively, if we consider the lesion as an unusual histologic variant of skin patch in the setting of AESOP, it may be that the different nature of the underlying tumor is related to the production of a different panel of cytokines diffusing from the tumor into the tissues of the chest wall and stimulating a different cellular component, according to the concept of “contiguous inflammation of the skin.”5(p49)

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