Pityriasis Lichenoides Et Varioliformis Acuta (Mucha-Habermann Disease)

January 01, 2008
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Figure
A 10-year-old boy presented with a persistent rash that began several months earlier as recurrent crops of papules and a few vesicles with crusting. Varicella was initially diagnosed, and the patient was treated unsuccessfully with over-the-counter drying lotions. The patient denied systemic symptoms and pruritus and was in his usual state of health otherwise. Review of systems, family history, and social history were unremarkable. There was no history of travel or new exposures during this period.

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The patient was referred to a dermatologist who confirmed the diagnosis of pityriasis lichenoides et varioliformis acuta (PLEVA).

PLEVA (also known as Mucha-Habermann disease) is a rare skin condition of unknown origin. Although most common in young adults, PLEVA can affect any age group, including children. PLEVA and pityriasis lichenoides chronica are at opposite ends of the same spectrum; the terms "acute" and "chronic" refer to the timeline of the individual lesions rather than the clinical course. The disease is characterized by insidious onset and a lack of symptoms other than mild pruritus and low-grade fever. Males are more commonly affected than females. There is no geographical or racial predisposition.

Pityriasis lichenoides is often described as a papulosquamous disorder based on its appearance; however, the predominance of CD8+ T cells in biopsy specimens suggests that it may be a lymphoproliferative disorder. Although commonly considered benign, malignant transformation with progression to cutaneous T-cell lymphoma has been reported. "Epidemic" outbreaks during the fall and winter and onset or flaring of the disease after infections provide further support of an immune-mediated hypersensitivity against an infectious agent. PLEVA is associated with infection with HIV, Epstein-Barr virus, Cytomegalovirus, Parvovirus, group A beta-hemolytic Streptococcus, and Staphylococcus aureus.

The rash consists of reddish brown, round to oval papules of 2 to 10 mm that appear in crops either singly or in clusters. A violaceous center may develop with micaceous scale. The lesions can become vesicular or pustular, may undergo hemorrhagic necrosis, and frequently result in postinflammatory hyperpigmentation (or less commonly varioloid scarring). The rash most commonly involves the trunk, thighs, and upper arms; the face, scalp, and palms/soles are involved in about 10% of cases.

Febrile ulceronecrotic Mucha-Habermann disease (a severe subtype of PLEVA, characterized by a more destructive rash with fever and systemic involvement) is associated with up to 25% mortality. The differential diagnosis includes insect bites, varicella, viral exanthema, Gianotti-Crosti syndrome, psoriasis guttata, lymphomatoid papulosis, and secondary syphilis. This variant is a medical emergency. Systemic corticosteroid therapy is the treatment of choice.

In many patients with PLEVA, first-line therapy with erythromycin estolate or ethylsuccinate (30 to 50 mg/kg divided in 3 equal doses daily) for up to 8 weeks can result in remission; tapering over several months is necessary to prevent recurrence. Topical corticosteroids and oral antihistamines may provide symptomatic relief but have no real effect on the disease course. Other variably effective therapies include psoralen plus UV-A, UV-B phototherapy, tetracycline, gold, methotrexate, oral corticosteroids, and dapsone. When first-line therapies fail, UV light therapy can be used. Systemic immunosuppressant therapy is reserved for more severe cases. With any of these treatments, PLEVA can persist for months, even years, and relapse is common. Therapy with erythromycin initially controlled this patient's rash; however, his symptoms eventually recurred despite an extended taper. He is currently receiving UV-B therapy and has had no further recurrence.

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