CLINICAL VIGNETTE

Necrotic Papules and Plaques in a 76-Year-Old Man

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Case

A 76-year-old man presented to dermatology with 6-month history of enlarging bumps on extremities and buttocks. Lesions were asymptomatic without any systemic symptoms. On exam, multiple 0.3 centimeter to 1.4 centimeter erythematous to violaceous papules and plaques were scattered on extremities and buttocks. Several larger lesions showed central necrosis and crusting.

Two 4-millimeter punch biopsies were performed, one from a larger necrotic lesion, and one from a newer smaller lesion. Both biopsies demonstrated epidermal acanthosis, spongiosis with overlying parakeratosis, focal neutrophilic spongiosis/ microvesicles, superficial perivascular lymphocyte predominant infiltrate, and focal exocytosis/epidermotropism of epidermal lymphocytes. One specimen showed extravasated erythrocytes. Histopathology and clinical presentation point to a diagnosis of pityriasis lichenoides et varioliformis acuta (PLEVA)

Discussion

Pityriasis lichenoides is an uncommon inflammatory skin disorder. The acute form is pityriasis lichenoides et varioliformis acuta (PLEVA), also known as Mucha-Haberman disease. The chronic form is pityriasis lichenoides chronica. While some experts believe the two forms to be separate entities, many believe they exist on the spectrum of the same condition.[1]

While PLEVA can occur at any age, it is most commonly seen in the second or third decade of life.[2] There is a slight male predominance. No geographic or racial predispositions have been reported.

The etiology of PLEVA is poorly understood. One theory is that it is a lymphoproliferative condition. This has been supported by studies demonstrating a dominant T-cell clone.[3,4] Other support include rare reports of development of lymphoma following PLEVA. A second theory is that PLEVA is an inflammatory skin condition triggered by infections. Pathogens that have been reported to trigger PLEVA include Epstein-Barr virus, adenoviruses, human immunodeficiency virus, human herpes virus 8, varicella-zoster virus, parvovirus B19, *Toxoplasma gondii*, *Staphylococcus aureus*, and *streptococcus pyogenes*.[5] Reports of PLEAVA occurring after vaccines further support this theory.[6-8]

PLEVA starts with acute crops of erythematous macules and papules that rapidly evolve into papulovesicles and papulopustules, often with necrotic or hemorrhagic centers.[5] The lesions favor trunk and flexural areas of extremities, but can occur anywhere on the body. There is usually no mucosal involvement. As some lesions heal, new ones appear, resulting in lesions at various stages of development. Lesions tend to be asymptomatic and usually heal without scarring, though often resulting in post-inflammatory hyper/hypopigmentation. While most cases of PLEVA are self-limited and do not have systemic symptoms, febrile ulcerative PLEVA is a severe and potentially lethal variant. In febrile ulcerative PLEVA, patients develop large necrotic plaques and ulcers that can reach several centimeters in diameter.[9] Systemic involvement include high fever, fatigue, interstitial pneumonitis, abdominal pain, arthritis, cardiomyopathy, and central nervous system vasculitis.

Diagnosis of PLEVA is based on clinical presentation and histology.[1] Skin biopsy classically demonstrate parakeratosis, spongiosis, epidermal acanthosis, and infiltration of lymphocytes and histiocytes that lead to vacuolar alteration of basement membrane. Other common features are exocytosis of lymphocytes, erythrocyte extravasation, and epidermal necrosis.

The differential diagnosis for PLEVA is broad and should include lymphomatoid papulosis, pityriasis lichenoides chronica, arthropod bite reactions, varicella, disseminated herpes simplex virus, Gianotti-Crosti syndrome, pityriasis rosea, guttate psoriasis, secondary syphilis, and Langerhans cell histiocytosis.[5] The disease that most closely mimic PLEVA is lymphomatoid papulosis. In lymphomatoid papulosis, the skin lesions tend to last much longer than PLEVA and may develop into nodules, plaques, or tumors. Histologic and immunohistochemical features are important in distinguishing between the two entities. Biopsies of lymphomatoid papulosis show dermal infiltrate of large atypical lymphocytes that are CD30-positive. Unlike PLEVA, histology of lymphomatoid papulosis show little or no vacuolar degeneration of the basal layer and few or no necrotic keratinocytes.

First-line therapy for PLEVA usually consists of either oral antibiotics or phototherapy.[1] For adults, doxycycline or minocycline at 100mg once or twice daily is commonly used. In pediatric population, erythromycin 30mg to 50mg/kg can be effective.[10] Azithromycin has also been reported to be helpful

in both children and adults, though data is more limited.[11] Phototherapy with ultraviolet (UV) is another treatment option. Both narrowband UVB and UVA have been reported to be helpful.[12,13] Of the two modalities, narrowband UVB has a better safety profile regarding risk of skin cancer. An additional benefit of UV phototherapy is that it can be continued at reduced frequency to prevent relapses of PLEVA. For recalcitrant disease, other treatment options include methotrexate, acitretin, dapsone, or cyclosporine.[1] Urgent treatment of febrile ulcerative PLEVA is crucial since it carries mortality rate upwards of 25%. Methotrexate and systemic corticosteroids have been reported to have variable efficacy.[9]

Conclusion

In summary, PLEVA is a poorly understood inflammatory skin condition. Although oftentimes self-limited, a potentially life-threatening variant exists. It is crucial for internists to recognize this condition and urgent referral should be placed to dermatology for evaluation and treatment.

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