Erythema Elevatum Diutinum in a Patient With Human Immunodeficiency Virus

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Erythema elevatum diutinum (EED) is a chronic cutaneous leukocytoclastic vasculitis. This rare disease is characterized by red, brownish-purple, and yellow papules, plaques, and nodules distributed symmetrically about the extremities. There have been recent reports of the disease in association with infection with the human immunodeficiency virus (HIV). We describe the case of a 51-year-old man with HIV who presented with EED, which was successfully treated with dapsone.

Erythema elevatum diutinum (EED) is a rare chronic form of localized cutaneous vasculitis that initially presents as leukocytoclastic vasculitis of the skin and later resolves with fibrosis. It is characterized by relapse with or without persistent red, brown-purple, and yellow papules, plaques, and nodules. EED has a predilection for a symmetric distribution over elbows, buttocks, knees, shins, ankles, and interphalangeal joints. Small ulcerations, vesicles, bullae, and lesions resembling xanthomas have less frequently been described. The pathogenic mechanisms of EED are unknown, but hematologic malignancies, monoclonal gammopathies, and autoimmune and infectious diseases have been described in association with the disease.[1] The association of EED with chronic or recurrent streptococcal infections has been well documented.[2] There have been several reports of EED presenting in patients with the human immunodeficiency virus (HIV).[1-5] We present the case of a 51-year-old man infected with HIV who developed EED and discuss the presentation of EED in this population.

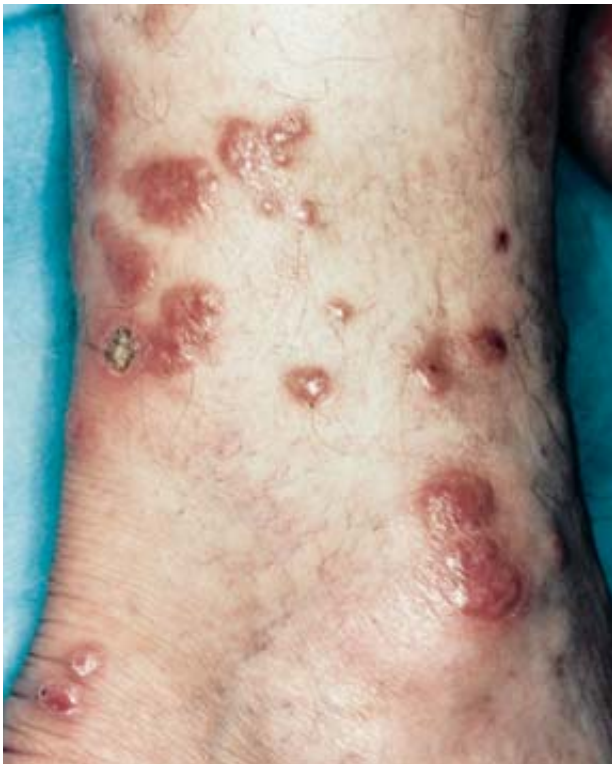

Figure 1. Yellowish-red plaques on the ankle and Achilles tendon.

Case Report

A 51-year-old Hispanic man, a long-time intravenous drug user, was referred for evaluation of multiple pruritic lesions of 4 months' duration over both lower

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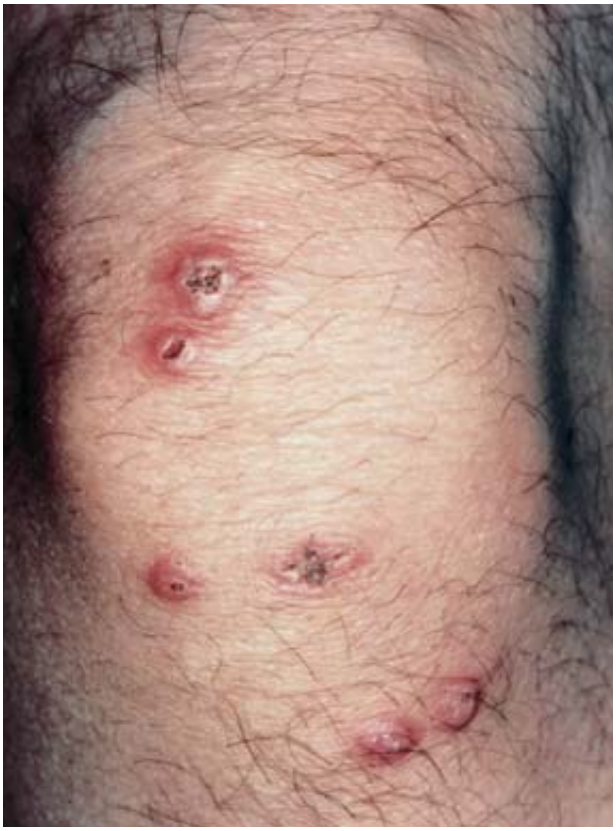

Figure 2. Crusted nodules on the knee.

Figure 3. Healed lesions of erythema elevatum diutinum with hyperpigmentation.

extremities. His medical history was significant for infection with HIV and chronic alcoholism.

Physical examination revealed a moderately nourished man with generalized lymphadenopathy. Cutaneous lesions were characterized by multiple discrete and confluent smooth, reddish-brown papules, nodules, and plaques symmetrical on the anterior aspect of the lower legs, knees, and Achilles tendons (Figures 1 and 2). In addition, a few crusted papules were present on the knees. Laboratory evaluation revealed a CD4 count of 150 and moderate elevation of liver enzymes. Antibodies for HIV, hepatitis B, and hepatitis C and a polyclonal gammopathy were present.

Histopathologic examination of multiple papules revealed a nodular inflammatory infiltrate in the dermis composed of many neutrophils, lymphocytes, and macrophages. Significant leukocytoclastic vasculitis was evident with endothelial swelling, perivascular neutrophil leukocytes, nuclear dust, and fibrinoid material within the walls of blood vessels. Focal degeneration of collagen also was noted. There was no evidence of fibrosis.

The patient responded to treatment with dapsone, 100 mg daily, with rapid clearing over several weeks (Figure 3). The patient was tapered off of dapsone and remained clear at a follow-up 3 months after discontinuation of therapy.

Comment

EED has been reported in 8 other patients with HIV,[1-5] one of whom had HIV-2.[1] There are several unique findings in patients with EED and HIV disease, although our case differs in many aspects from the majority of the others. Thus far, all patients described have been men. The age at onset of EED has tended to be earlier than usual, with a mean age of 35 years (range, 22–58 years). The mean CD4 lymphocyte count in the patients reported is 181×10^6/L (range, $112–314\times10^6$/L). One patient was reported to have a normal CD4 count.[2]

Of the 8 previous cases, 6 had homosexuality as their HIV risk factor; one patient was an intravenous drug user; and one had received an infected blood transfusion. Our patient was, as noted, a longtime intravenous drug user. Many of the patients described have tended to present with late nodular lesions of EED.[3-5] Our patient had early lesions, based histologically on the lack of fibrosis and presence of neutrophils.

Treatment options for EED include dapsone,

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tetracycline, niacinamide, colchicine, antimalarials, and steroids. Surgical excision is another therapeutic option.[1,5] Dapsone is the treatment of choice. Our patient responded rapidly to dapsone, 100 mg daily, with tapering. In contrast, 3 of the patients described by LeBoit and Cockerell[5] were treated with dapsone; there was no response in 2 of the patients; and therapy was discontinued in the third secondary to hepatitis. The authors theorized that the lack of response to dapsone in these patients might reflect the preponderance of fibrosis rather than neutrophils in advanced lesions.

In conclusion, HIV appears to be a potential etiologic factor in the development of EED. LeBoit and Cockerell[5] speculated that either opportunistic infection or some aspect of HIV infection provided the antigenic stimulus for EED in their patients. Physicians should be aware of this condition in patients infected with HIV because it may be the first sign of infection,[1] or may be mistaken for Kaposi sarcoma or bacillary angiomatosis.

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