Disseminated Angiolymphoid Hyperplasia With Eosinophilia: A Case Report

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A 37-year-old Cantonese man presented with pruritic erythematous papules and nodules on his face, limbs, and trunk that had been present for 10 years and aggravated for 2 years. More nodules were noticed where the skin was scratched and traumatized. The lesions were alleviated temporarily, but they did not subside entirely without therapy. The lesions responded to treatment with intravenous dexamethasone. Histopathology results indicated angiolymphoid hyperplasia with eosinophilia (ALHE), and the patient was diagnosed with disseminated ALHE (DALHE). His lesions ameliorated after treatment with prednisone.

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Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon vascular disorder of unknown pathogenesis. The lesions tend to occur locally on the head and around the ears. Generalized lesions are rare in most patients with ALHE. We describe a patient with ALHE who presented with multiple cutaneous nodules all over his body.

Case Report

A 37-year-old Cantonese man presented to our outpatient department with pruritic erythematous papules and nodules on his face, limbs, and trunk. The lesions had been present for 10 years and aggravated for the past 2 years. History revealed that 10 years prior to presentation the patient had been bitten by mosquitoes and other insects and later noticed erythema and grain-sized red papules on his face, on the back of his hands, and on his feet and limbs, with scattering distribution and mild itch. Lesions then appeared on his trunk, and nodules developed. More nodules were noticed where the skin was scratched and traumatized. The lesions were alleviated temporarily but did not entirely subside until therapy was initiated. The lesions responded to treatment with intravenous dexamethasone. In the 2 years following presentation, the lesions progressively increased in size and in number, with episodes of severe itching. The patient had no history of previous illness but reported having had multiple sexual partners.

A physical examination revealed no abnormal findings except the skin manifestations. There were innumerable 2- to 10-mm nodules on the patient's head, face, neck, trunk, and limbs that were skin-colored or dark red and hemispherical or spherical (Figure 1). The surface of most lesions was smooth with umbilication. The lesions were hard to the touch. The top of some lesions were shiny. In addition, on some lesions there were scratch marks or scabs.

Results of laboratory tests revealed a red blood cell count of 4.31×10^{12}/L, a hemoglobin level of 144 g/L, a white blood cell count of 7.7×10^{9}/L, an absolute eosinophil count of 0.858 to 1.72×10^{9}/L (reference range, 0.05–0.3×10^{9}/L), and an erythrocyte sedimentation rate of 5 mm/h. Test results for complete urine routine, stool routine, liver and kidney function, antinuclear factor, immunoglobulin, complement, human immunodeficiency virus antibody, rapid plasma regain, and *Treponema pallidum* hemagglutination were within normal limits. No growth was evident on fungal and mycobacterial cultures. Results of a chest x-ray, electrocardiogram, and ultrasound were normal. Pathologic examination of the skin lesions showed hyperkeratosis, hyperplasia of prickle cell layer, infiltration of masses of eosinophils and lymphocytes, proliferation of blood vessel endotheliocytes with partial canaliculization, and occasional lymphoid follicles in the entire dermis (Figure 2). Histopathology results revealed ALHE.

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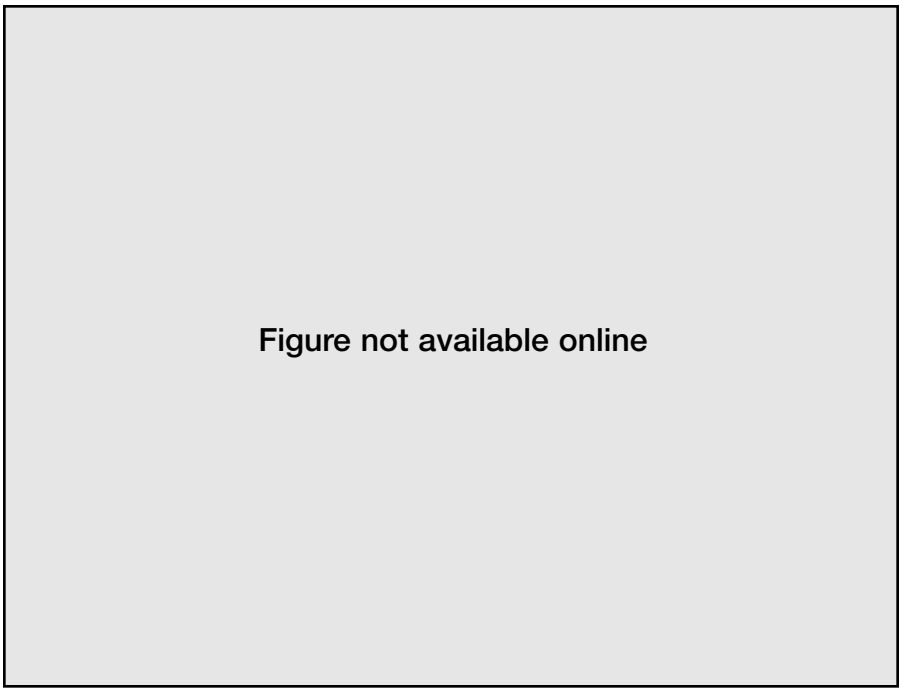

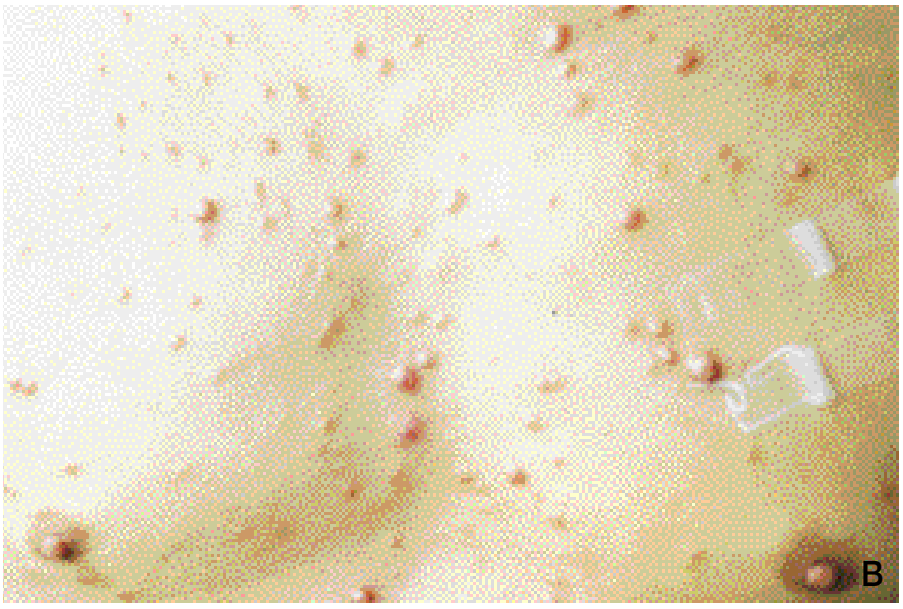

Figure 1. Innumerable 2- to 10-mm nodules on the patient's face (A) and trunk (B). The nodules are skin-colored or dark red and hemispherical or spherical.

A diagnosis of DALHE was made and the patient was placed on oral prednisone, *Tripterygium wilfordii* Hook.f. (a traditional Chinese medicine), clarityne, tranilast, and cyproheptadine. His lesions ameliorated after 3 months of treatment (Figure 3).

Comment

Although the cause of ALHE is unknown, antigenic stimulation in a patient following insect bites has been postulated. Some authors have suggested that ALHE is distinct from Kimura disease.[1,2] In these reports, the ALHE occurred in the subcutaneous tissue, skin, and maxillary antrum, whereas Kimura disease affected the subcutaneous tissue, major salivary glands, and lymph nodes. Distinctive features of ALHE are exuberant proliferation of vessels lined with cuboidal to hobnail endothelial cells with irregular nuclei and cytoplasmic vacuoles, fibromyxoid matrix, involvement of a muscular coat of blood vessels, and zonation of inflammatory infiltrate toward the peripheral portion of the lesion. Distinctive features of Kimura

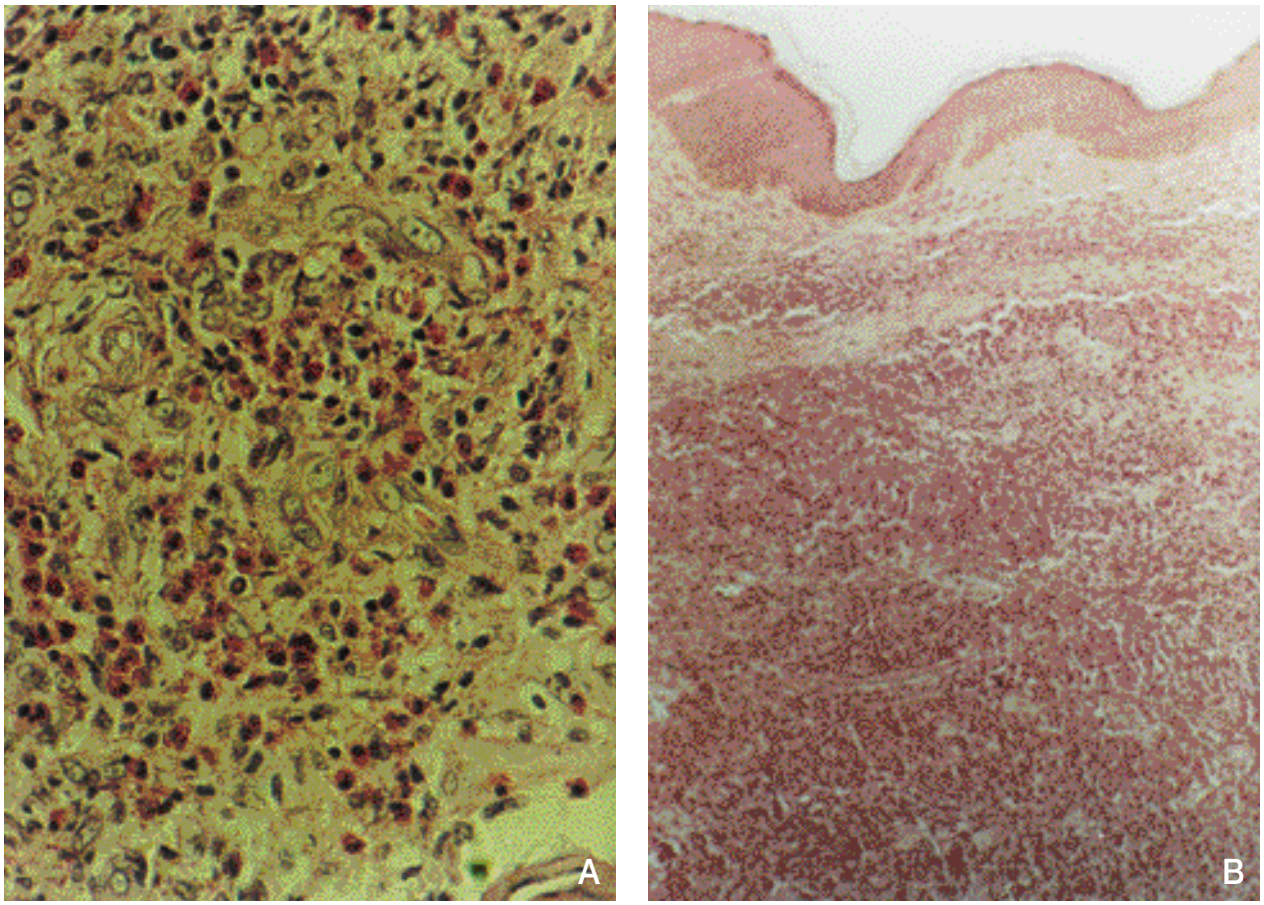

Figure 2. Hyperkeratosis and hyperplasia of the prickle cell layer, infiltration of masses of eosinophils and lymphocytes, proliferation of blood vessel endotheliocytes with partial canaliculization, and occasional lymphoid follicles in the dermis (A and B)(H&E, original magnifications ×10 and ×20).

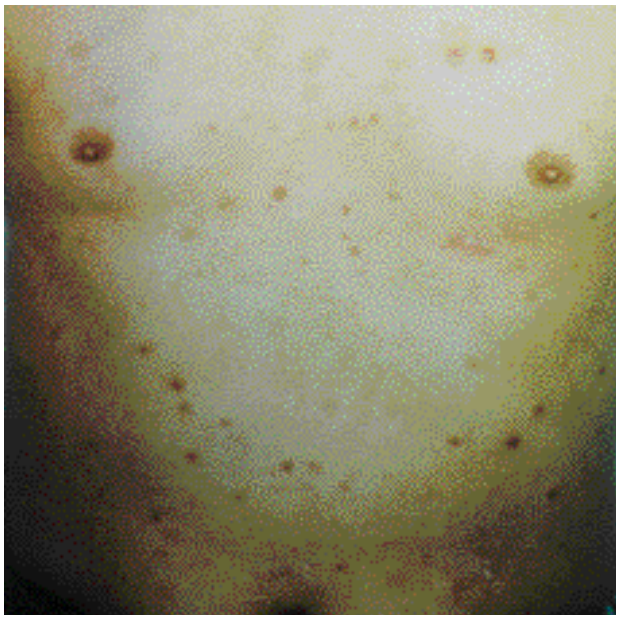

Figure 3. The patient's lesions ameliorated after 3 months of treatment.

disease are a florid lymphoid infiltrate with prominent lymphoid follicles, vascularization of germinal centers, germinal center necrosis, marked eosinophilia with or without eosinophil abscess formation, proliferation of high endothelial venules, and sclerosis.[2]

ALHE has a slight predilection for middle-aged to elderly women. It usually presents as persistent, recurrent, reddish brown dermal papules or nodules that appear primarily on the head and neck, most commonly the ears and preauricular area. Although most patients have only a single lesion, approximately 20% of affected patients have multiple lesions.[3] Our patient is a middle-aged man with a 10-year history of innumerable pruritic 2- to 10-mm nodules on his face, limbs, and trunk that were skin-colored or dark red and hemispherical or spherical in shape. Peripheral blood eosinophilia is common. Histology indicates proliferation of vascular channels with a surrounding infiltrate of lymphocytes and eosinophils.

We believe that the features were consistent with the diagnosis of ALHE. Because our patient has extensive lesions over his entire body, the diagnosis of DALHE was made. The differential diagnosis of DALHE includes other skin diseases with extensive papules and/or nodules, such as leprosy, leishmaniasis cutis, syphilis, sarcoidosis, neurofibromatosis, tuberous sclerosis, eruptive xanthoma, xanthoma disseminatum, eruptive keratoacanthoma, Cowden disease, multiple trichoepithelioma, multiple fibrofolliculoma, multiple trichodiscomas, progressive nodular histiocytoma, and generalized eruptive histiocytoma.

Treatment of ALHE involves excision, electrodesiccation and curettage, radiotherapy, carbon dioxide laser therapy, pulsed dye laser therapy, intralesional and systemic corticosteroids, cryosurgery, pentoxifylline, cessation of estrogen therapy, systemic vinblastine sulfate, oral retinoid, indomethacin, and intralesional interferon alfa-2a.[3] Our patient responded to treatment with oral corticosteroids and the traditional Chinese medicine *Tripterygium wilfordii* Hook.f. (the therapeutic mechanism of this drug may be due to its anti-inflammatory and immunosuppressive effects).[4] Follow-up is still in progress.

REFERENCES

1. Kuo T-T, Shih L-Y, Chan H-L. Kimura's disease: involvement of regional lymph nodes and distinction from angiolymphoid hyperplasia with eosinophilia. *Am J Surg Pathol*. 1988;12:843-854.
2. Chan JK, Hui PK, Ng CS, et al. Epithelioid haemangioma (angiolymphoid hyperplasia with eosinophilia) and Kimura's disease in Chinese. *Histopathology*. 1989;15:557-574.
3. Shenefelt PD, Rinker M, Caradonna S. A case of angiolymphoid hyperplasia with eosinophilia treated with intralesional interferon alfa-2a. *Arch Dermatol*. 2000;136:837-839.
4. Xu Wenyan, Zheng Jiarun, Lu Xueyu. *Tripterygium* in dermatologic therapy. *Intern J Dermatol*. 1985;24:152-157.