Spontaneously Regressive Angiolymphoid Hyperplasia with Eosinophilia: A Case Report with Evidence of Dendritic Cells Proliferation

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To the Editor: A 79-year-old male presented with multiple asymptomatic, erythema, and nodules on the right-front scalp for about three weeks. The number of the nodules gradually increased. Physical examination revealed multiple violaceous, reddish nodules measuring 0.5–1.0 cm and infiltrative plaques located on the right frontal scalp, without evidence of lymphadenopathy [Figure 1a]. Serological examination revealed the elevated level of leukocytes. Peripheral eosinophil and serum IgE were normal. Histopathology revealed vascular hyperplasia in the dermis. The larger vessels were lined by characteristic "hobnail" endothelial cells, which protruded into the lumen and have ovoid nuclei and intracytoplasmic vacuoles. There was mixed inflammatory infiltration of prominently eosinophils, histiocytes, lymphocytes, and neutrophils [Figure 1c]. Immunohistochemical examination revealed dendritic cells of epidermis and dermis increased, with positive CD1a staining, the linear density of CD1a positive dendritic cells was 70–80/mm (normal range 42.7 ± 17.9/mm) [Figure 1d]. Based on the history, clinical examination, and histopathology, the patient was diagnosed with angiolymphoid hyperplasia with eosinophilia (ALHE). The patient did not use any topical drug. The lesions regressed spontaneously after one month [Figure 1b].

ALHE is an uncommon, benign disorder that presents as solitary or multiple red-brown dome-shaped papules or nodules occurring most frequently on the head and neck. The disease is idiopathic. Trauma, hyperestrogenemia, infectious agents, atopy, reactive hyperplasia, and benign neoplasia have been implicated in the minority of cases. The pathogenesis of ALHE is still under controversial. Various hypotheses have been put forth, including a reactive process, a neoplastic process, and infectious mechanisms. Kempf *et al*.[1] postulated that ALHE might present CD4+ T-cell lymph-proliferative disorder, rather than a true vascular lesion.

Central to the histology of ALHE is the proliferation of blood vessels of varying sizes lined by plump endothelial cells. Inflammation is the second defining characteristic. Lymphocytes and varying amounts of eosinophils diffusely surround and may infiltrate the blood vessels. Depending on the stage of the lesion, the vascular or inflammatory component may predominate. In the active growing ALHE, the vascular component predominates, whereas in the late stages of the disease lymphocytes become more prominent.[2] ALHE is different from Kimura's disease clinically and histopathologically. The typical presentation of ALHE is papules or nodules, while the Kimura's disease is subcutaneous mass. Histologically, the proliferation of blood vessels was superficial in ALHE, while the Kimura's disease is deeper, florid lymphoid follicles with germinal center formation were usually seen. Moreover, Kimura's disease is a systemic immune-mediated

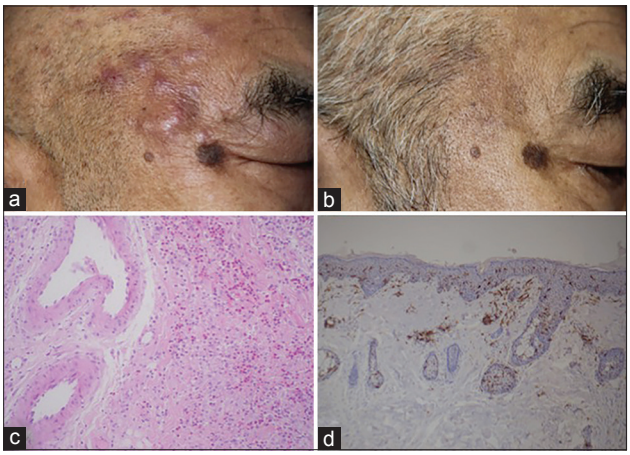

Figure 1: The patient presented with multiple violaceous, reddish nodules located on right frontal scalp (a); the lesions spontaneously regressed after one month (b); larger vessels were lined by characteristic "hobnail" endothelial cells, which protruded into the lumen and have ovoid nuclei and intracytoplasmic vacuoles (c, hematoxylin-eosin staining, original magnification ×200); dendritic cells of epidermis and dermis increased, with positive CD1a staining (d, immunohistochemical staining, original magnification ×100).

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process that commonly presented with eosinophilia, high level of IgE, and may be associated with renal disease.

In our patient, the histopathology supports the diagnosis of ALHE. However, the infiltration of inflammatory cells was more prominent, suggesting the disease was in the late stage. A rare character should be pointed out is the infiltration of dendritic cells in the epidermis and dermis, which has been confirmed by immunohistochemistry. This change has not been reported before. Dendritic cells are considered to be one of the major antigen-presenting cells in the skin. The macrophages of dermis have scavenging and phagocytic activities, as well as anti-inflammatory properties that contribute to microbial clearance, skin homeostasis, and wound repair. The predominantly increasing dendritic cells in the epidermis, which stained CD1a, accompanied with multiple histiocytes in deep dermis probably indicated the underlying immunological mechanism. Treatment of ALHE is often pursued to provide symptomatic relief and address cosmetic concerns. Surgical excision is commonly used. Other alternative treatments have been reported with variable levels of success. These treatments include laser therapy, systemic or intralesional corticosteroid injection, cryotherapy, imiquimod, tacrolimus, isotretinoin, radiotherapy, interferon-alpha 2a, anti-interleukin-5 antibody, photodynamic therapy, and methotrexate. Spontaneous resolution has also been reported. Adler *et al*.[3] conducted a systematic review of the literature, within the 593 cases, spontaneous resolution, occurring alone or after attempted treatment, was reported in only 17 cases (2.9%).

In our case, we provide support for a reactive process for ALHE, according to the prominently increasing dendritic cells and histiocytes. Moreover, the short course of the disease and spontaneous regression also reflected this point from the other aspect. We hypothesize that the dendritic cells may be involved in the pathogenesis of ALHE, especially in very early stage. While it is yet to be confirmed by large-scale research, and further work should be done to explain the pathogenesis of the findings.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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