Spontaneous Regression of Generalized Angiolymphoid Hyperplasia with Eosinophilia in a 2-year-old Boy

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Sir,
Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vasoproliferative disorder characterized by solitary or multiple subcutaneous nodules, usually occurring on the head and neck and very rarely on the extremities and trunk (1, 2). It is important to be able to recognize and differentiate ALHE from other hyperproliferative diseases associated with eosinophilia, such as Kimura's disease, as well from vascular tumours, particularly from malignant angiosarcoma, which may develop in the elderly. There are numerous treatment regimes for ALHE, and complete surgical excision is recommended in persistent lesions. We report here a spontaneous regression of this unusual dermatosis, which occurred in a 2-year-old boy.

CASE REPORT

A 2-year-old Japanese boy was brought to our hospital by his parents with a tumour on his left upper arm of 1-month duration. The lesion had a tendency to bleed after minor trauma. The tumour progressively increased in size and small nodules developed around the tumour. Physical examination revealed a 2.5 × 2 cm freshly ulcerating tumour in the lateral side of the left elbow with satellite nodules (Fig. 1a). The tumour was firm on palpation. Regional or systemic lymphadenopathy was absent. Laboratory findings, including complete blood count, liver function tests and urinalysis, were all within normal limits, except for a mild elevation of soluble interleukin-2 receptor (IL-2R) (819 U/ml; normal range 145–519 U/ml). Skin biopsy revealed small blood vessels and dense inflammatory infiltrate characterized by lymphocytes intermixed with numerous eosinophils (Fig. 2a). The vascular spaces were lined by protuberant endothelial cells, some of which had cytoplasmic vacuoles (Fig. 2b). Mitosis in endothelial cells was rare. Immunohistochemistry showed a positivity of the endothelial cells for CD31 (Fig. 2c). The absence of bacteria was proven by periodic acid-Schiff (PAS), Gram and Ziel-Neelsen stains. Furthermore, a lack of cellular atypia in the infiltrating lymphocytes was consistent with immunostaining for CD3, CD4, CD8, CD20 and κ/λ light chains, which showed their polyclonal nature. In addition, we performed *in situ* hybridization for Epstein-Barr virus encoded RNA (EBER) and did not find any EBER-positive cells. Thus, the patient was diagnosed as having ALHE and treated conservatively with an ordinary dressing alone.

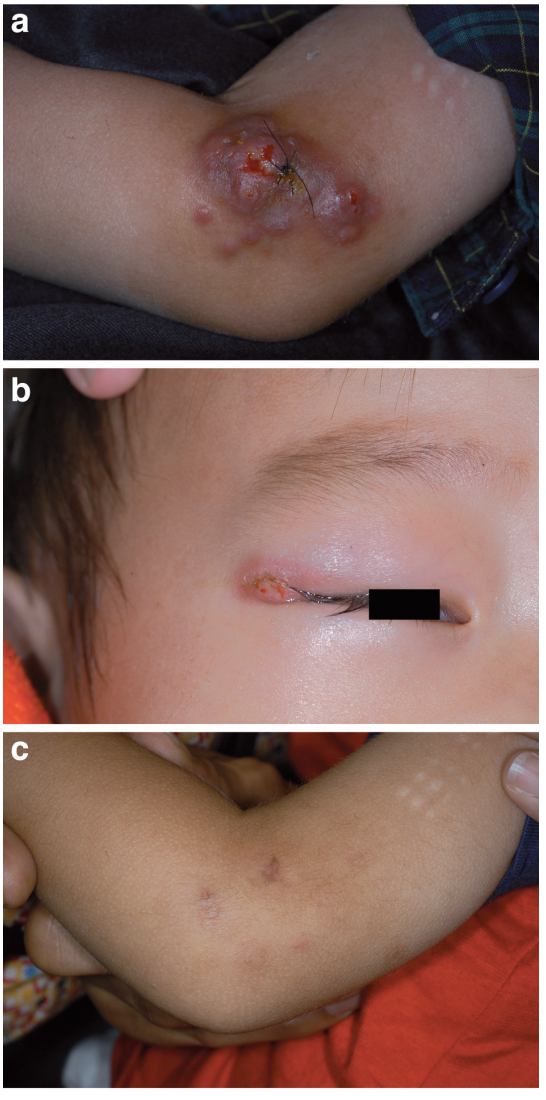

Fig. 1. Clinical images of angiolymphoid hyperplasia with eosinophilia (ALHE). (a) Dome-shaped reddish tumours on left upper extremity and (b) mildly elevated subcutaneous tumour on eyelid. (c) Disappearance of the tumours on the upper arm 6 months later.

Similar tumours had also developed on his legs, abdomen, eyelid and scalp within 2 months (Fig. 1b). Although multiple treatment modalities, including intralesional corticosteroid injection and cryotherapy, were proposed, the patient's parents were reluctant to allow them. However, an improvement was noted, with a decrease in size and a slightly grey appearance on the surface of some tumours, and complete resolu-

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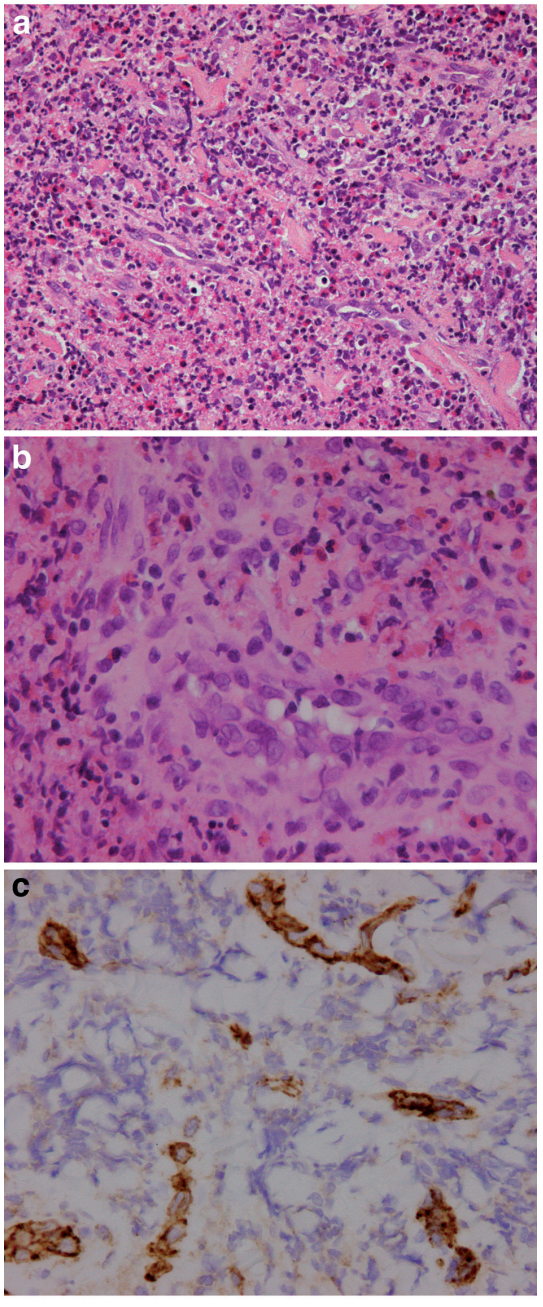

Fig. 2. Histopathology of the tumour on the upper arm. (a) Vascular proliferation with a rich lymphoeosinophilic infiltration is seen. (b) The vascular spaces were lined by protruding, partly vacuolated endothelial cells. (a, b; H&E stain). (c) Immunohistochemical staining shows the positivity of the endothelial cells for the marker CD31. Original magnification: (a) ×200; (b) ×400; (c) ×400.

tion was noted 6 months later (Fig. 1c). No recurrence has been reported after 2 years.

DISCUSSION

Originally described by Wells & Whimster in 1969 (2), ALHE is a relatively rare condition of unknown frequency and uncertain pathogenesis, and is considered to be a reactive lesion rather than a tumour. It affects women more commonly than men, during young adulthood to middle age, with few cases reported in children and the elderly. The case described here is unusual, in that it occurred in a 2-year-old boy in a generalized pattern. While most patients have only a single lesion, approximately 20% of affected patients have multiple lesions (1). In the case of multiple lesions, they usually occur in contiguous areas, and disseminated lesions, as in our case, are exceptional (3, 4). Even in such cases, the diagnosis of ALHE can be made on the basis of the unique histopathological features, consisting of the proliferation of large endothelial cells lining vascular spaces, and lymphocytic and eosinophilic infiltrate. In our case, the increase in small blood vessels lined by protuberant, vacuolated endothelial cells, together with eosinophilic infiltration, closely matched the histopathological features of ALHE.

ALHE usually presents as a persistent nodule or tumour. Numerous therapeutic approaches have been tested, including destructive techniques such as surgery, electrodessication and radiotherapy. Progressive enlargement of ALHE may require total excision. Topical imiquimod has been reported to be effective for ALHE (5). On the other hand, the occurrence of spontaneous regression has also been reported (3, 6), indicating the presence of a clinical spectrum among ALHE lesions. In some cases the lesions are persistent for a long time; in others they regress spontaneously. In our patient, rapid enlargement with the appearance of multiple lesions was followed by spontaneous regression, thus avoiding the need for aggressive treatments. This supports the view that ALHE may be observed for several months, awaiting spontaneous regression prior to surgical and other therapies.

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