

Unilateral Angiolymphoid Hyperplasia with Eosinophilia of the Right Upper Extremity: a reported case

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ABSTRACT :

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Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vascular proliferative disease with unknown etiology. The predilection sites are head and neck but uncommon on extremities.

We report a 36-year-old Thai female with multiple lesions of ALHE on her right upper extremity. She had high serum IgE level. The histopathology revealed typical findings of ALHE. The immunohistochemical staining was positive for capillary but negative for lymphatic markers and HHV 8. No arteriovenous shunting was found.

Key words: Angiolymphoid hyperplasia with eosinophilia (ALHE), vascular proliferative disease

บทคัดย่อ :

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สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Angiolymphoid Hyperplasia with Eosinophilia (ALHE) เป็นเนื้องอกของเส้นเลือดชนิดหนึ่ง ที่พบได้ไม่บ่อย และยังไม่ทราบสาเหตุที่แน่ชัด โดยที่รอยโรคพบบ่อยที่บริเวณศีรษะ ใบหน้า และลำคอ มักไม่พบบริเวณแขนหรือขา

รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงไทย อายุ 36 ปี ที่มาด้วยรอยโรคของ ALHE ที่บริเวณแขนขวา ตรวจพบระดับ IgE ในเลือดสูง ลักษณะทางจุลพยาธิวิทยาเข้าได้กับ ALHE ชัดเจน การย้อมพิเศษขึ้นเนื้อพบว่าให้ผลบวกต่อ marker ของ capillary แต่ให้ผลลบต่อ lymphatic และ HHV8 และไม่พบ arteriovenous shunt

คำสำคัญ: โรค Angiolymphoid Hyperplasia with Eosinophilia (ALHE), เนื้องอกของเส้นเลือด

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) was first described in 1969 by Wells & Whimster.¹ ALHE is now clearly known as the separate clinicopathologic entity from Kimura's disease.² ALHE occurs in young to middle-aged adults especially in woman. Multiple tan, brown or dull red superficial lesions of ALHE favored head and neck especially around ears. The pathogenesis is still unclear. History of trauma, arteriovenous shunting³⁻⁴ and viral infections were all postulated.⁴⁻⁵

The histopathology characterized by proliferation of lobulated blood vessels lined by epithelioid endothelial cells with sparse to heavy perivascular infiltration composed of lymphocytes and eosinophils without germinal center. The lesions may be asymptomatic or can be painful, pruritic and bleeding.⁴

The authors report a patient with unilateral multiple lesions of angiolymphoid hyperplasia with eosinophilia that involved the right upper extremity.

Case Report

A 36 year-old healthy Thai female patient presented with multiple papules varying in size on right upper extremity for 3 years. She noticed that the first lesion arised on her distal right forearm. Later, new multiple lesions developed and extended proximally. There were occasionally spontaneous bleeding and itching. She received topical and systemic steroid from a primary hospital but was not responded to the treatment. Multiple excisions were done but new lesions gradually developed. She was referred to the Institute of Dermatology.

Physical examination revealed unilateral multiple discrete well demarcated skin-color and purplish red

firm non-tender papules and nodules with some hemorrhagic crusted. Multiple pedunculated lesions distributed from right wrist to shoulder (Figure 1). Radial, ulnar arterial pulses and capillary refill were normal. No regional lymphadenopathy was detected. Complete blood counts, urine analysis, blood chemistries were normal but the serum IgE level was high (1098 IU/ml). Anti-HIV was negative.



Figure 1 Unilateral multiple discrete well demarcated skin-color and purplish red firm non-tender papules and nodules with some hemorrhagic crusted on the right upper extremity.

Histologic examinations from the pedunculated erythematous nodule and purplish red firm papule on right shoulder showed similar result. There were proliferation of small to medium sized blood vessels in lobular architecture and lumens were lined with epithelioid endothelial cells. Perivascular infiltration composed of lymphocytes, plasma cells, numerous eosinophils and extravasated RBC were found without eosinophilic abscess. The proliferation of blood vessels

were embedded in myxofibrotic stroma involved throughout dermis without subcutaneous involvement. No germinal center of lymphoid follicle was detected (Figure 2). The special stainings were positive for capillary markers (CD31, CD34 and Factor VIII) (Figure 3) but negative for lymphatic markers and HHV 8. Doppler ultrasound of the right upper extremity showed no gross arteriovenous shunting.

Multiple lesions were removed but new lesions gradually appeared.

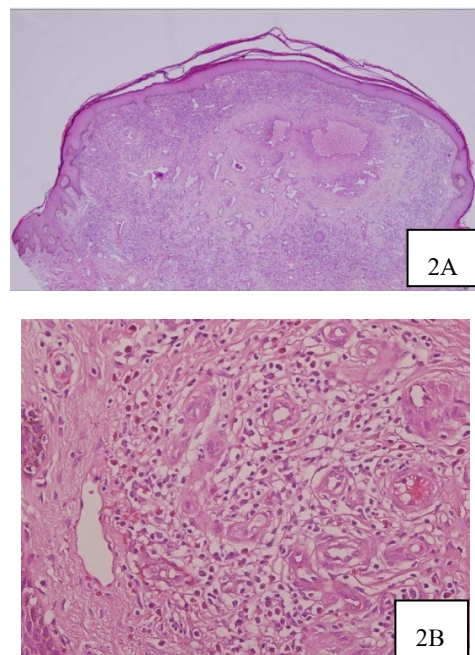


Figure 2 Proliferation of small to medium sized blood vessels in lobular architecture. The lumens were lined by epithelioid endothelial cells (A) (H&E, original magnification x40). Perivascular infiltration with lymphocytes, dense eosinophils and plasma cells in myxofibrotic stroma (B). (H&E, original magnification x200).

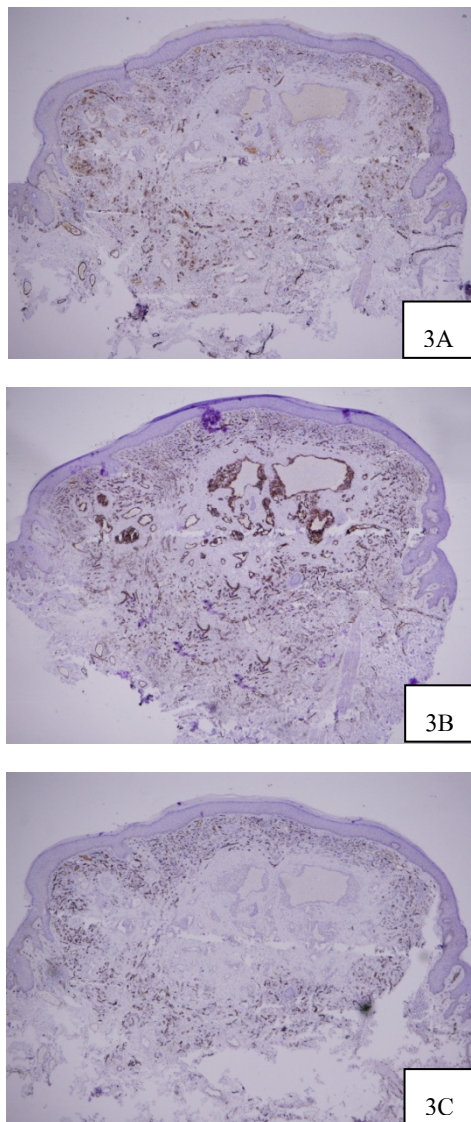


Figure 3. Immunohistochemical staining for CD31 (A), CD34 (B) and Factor VIII (C).

Discussion

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vascular proliferative disease of skin present as a purplish red solitary or multiple superficial nodules with a predilection for head and neck regions,

especially retroauricular area with slightly more common in women. Lesions on extremities, especially the arms and hands, vulva, penis, oral mucosa and tongue were rarely reported.^{4,6-11} Age of onset varies from young to middle age. Most of lesions are asymptomatic but sometime pruritus and bleeding can be occurred.⁴ The pathogenesis of ALHE is still unclear. It may be related to trauma, viral infection (HHV 8 and EBV) or high estrogen status and arteriovenous shunt (AV shunt).^{2,4}

The histopathology of ALHE characterized by a proliferations of small to medium sized blood vessels in a lobular architecture with prominent epithelioid or histiocytoid endothelial cells. The vessels are surrounded by a variable and mixed inflammatory cells infiltrate composed of lymphocytes and eosinophils with rare germinal center formation.

The first line of treatment for ALHE is complete excision.⁴ The alternative treatments include indomethacin farnesil, vascular laser, laser therapy e.g. carbon dioxide, argon, copper vapor, radiotherapy, corticosteroids (topical, intralesional and systemic), cryotherapy, electrodesiccation, pentoxifylline, intralesional chemotherapeutic agents (bleomycin, vinblastine and fluorouracil), oral isotretinoin, tracolimus ointment and imiquimod.¹²⁻¹⁷ Despite of treatment, ALHE has a tendency to recur but there is no reports of malignant transformation.

Arnold M et al⁴ had reported a 20-year-old woman with multiple painful ALHE lesions only on her left arm

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and hand with diminished radial pulse. Ultrasonography and nuclear magnetic resonance demonstrated a tumor-like lesion with concentric formation around the brachial artery. The histopathology of the tumor was similar to ALHE.⁴ Our patient had normal and equal pulse on her upper extremities and the ultrasonography showed neither tumor-like lesion nor gross arteriovenous abnormality. Cham E et al, reviewed four cases of multiple unilateral angiolymphoid hyperplasia with eosinophilia on extremities. One of them had a clinical and histopathology closely resemble to our patient.⁷ Usually ALHE patients had a normal range of serum IgE and the high levels were rarely reported.¹⁸ Our patient seems to be an unusual case of ALHE due to high serum IgE level.

In conclusion, the authors report an unusual patient of ALHE who presented with multiple polypoid and nodular lesions exclusively on the right upper extremity with high serum IgE level. The immunohistochemical staining was positive for capillary markers but negative for lymphatic markers. HHV 8 marker and AV shunt could not be demonstrated.

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