

## Case Report

# A lady with angiolymphoid hyperplasia with eosinophilia of the vulva

## 會陰部血管淋巴樣增生伴嗜酸細胞增多症一例

CFY Siu 蕭鳳兒 and KC Lee 李景中

---

A 34-year-old Indian lady presented with a six-month history of itchy erythematous papules over vulva and perianal area with on and off bleeding. A diagnosis of angiolymphoid hyperplasia with eosinophilia was made based on the clinical findings and the histopathological features. Case reports of this rare presentation of the disease in genital areas were reviewed. Treatment options were also discussed.

一名34歲印度女患者的會陰部及肛門周圍出現紅色癢性丘疹六個月，並伴有間歇性出血。經臨床及病理綜合診斷為血管淋巴樣增生伴嗜酸細胞增多症。此病罕見於生殖器，過往病例及治療方法將一一討論。

**Keywords:** Angiolymphoid hyperplasia with eosinophilia, genital area, perianal area, vulva

**關鍵詞：**血管淋巴樣增生伴嗜酸細胞增多症，生殖器，肛門周圍，會陰部

---

## Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon idiopathic condition, which presents as red or purple coloured papules

and nodules in the dermis or subcutaneous tissue of the head and neck region, particularly in the auricular and peri-auricular areas. It is rarely seen in the genital area. We hereby report a case of ALHE over the vulval region.

---

**Social Hygiene Service, Centre for Health Protection,  
Department of Health, Hong Kong**

CFY Siu, MBBS(HK), MRCP(UK), FHKAM(Medicine)

**Department of Pathology, Queen Elizabeth Hospital,  
Hong Kong**

KC Lee, FRCPath, FHKAM(Pathology)

Correspondence to: Dr. CFY Siu

Fanling Integrated Treatment Centre, 6/F, Fanling Health Centre, 2 Pik Fung Road, Fanling, N.T.

## Case report

A 34-year-old Indian lady presented with multiple erythematous itchy papules over the vulva and perianal area for three months with on and off bleeding. She had a history of termination of pregnancy and use of oral contraceptive pills approximately one to two months before the onset of the lesions. Her past medical history was otherwise unremarkable.

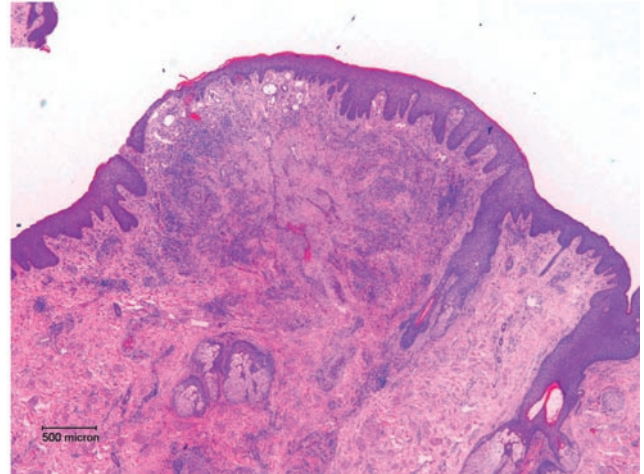
Physical examination showed multiple erythematous vascular papules over the right labium majus and right perianal skin (Figure 1). The papules ranged from 2 mm to 4 mm in diameter. Main clinical differential diagnoses included haemangioma, pyogenic granuloma, angiokeratoma, nodular prurigo, scabies, bowenoid papulosis, molluscum contagiosum, genital wart and condylomata lata. A rare condition, angiolymphoid hyperplasia with eosinophilia, was yet another differential diagnosis that had to be considered.

An incisional biopsy was taken from a lesion at the right labium majus and showed a vascular lesion in the upper dermis with reactive epidermal changes and focal excoriation. In the centre of the lesion, the abnormal vessels possessed a thick

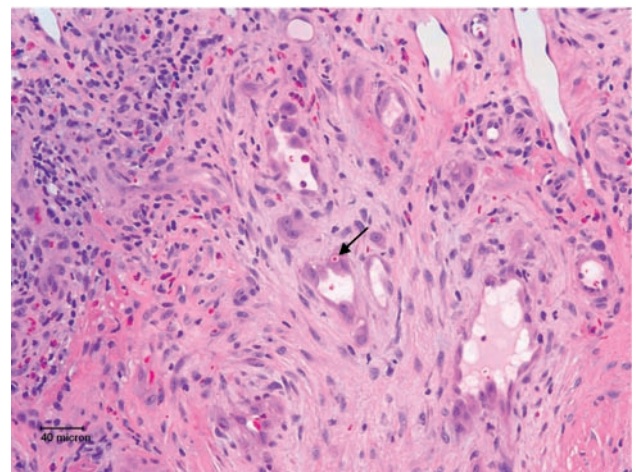


**Figure 1.** Multiple red vascular papules ranging from 2 to 4 mm in diameter over right labium majus.

fibromyxoid wall and the more peripheral ones were smaller with admixed lymphocytes and readily found eosinophils. No germinal centres were present. The blood vessels were lined by typical epithelioid endothelial cells with abundant dense eosinophilic cytoplasm with occasional intracytoplasmic vacuoles (Figures 2 and 3). The



**Figure 2.** Wedge-shaped upper dermal lesion with overlying epidermal hyperplasia and focal excoriation. Note the pale central zone consisting of larger blood vessels with thick fibromyxoid wall and a more inflammatory rim (H&E, marker 500 micron).



**Figure 3.** High power view showing typical epithelioid endothelial cells with dense eosinophilic cytoplasm. Note the presence of cytoplasmic vacuole containing red cell (arrow). Eosinophils are readily found in the left upper field (H&E, marker 40 micron).

picture was that of angiolymphoid hyperplasia with eosinophilia.

She was treated with liquid nitrogen in our unit, and was referred to gynaecologists for excision of around 20 raised lesions and cauterisation of the flat lesions under general anaesthesia. The treatment response was satisfactory (Figure 4). Her symptoms of pruritus improved and no more bleeding was reported.

## Discussion

Angiolymphoid hyperplasia with eosinophilia (ALHE) is also known as epithelioid haemangioma. It was originally described by Wells and Whimster in 1969 and was considered to be a late stage of Kimura's disease, a disorder described in the Japanese literature 20 years earlier. Nowadays,



**Figure 4.** Improvement of the condition after cryotherapy, surgical excision and electrocautery.

however, most researchers believe that ALHE and Kimura's disease are two separate disease entities, with different clinical and pathological features.<sup>1</sup>

The aetiology of ALHE is unknown. It is considered to be an unusual reactive process rather than a malignant process. Underlying arteriovenous shunting may play a role in the pathogenesis, as evidenced by the histopathological findings of damaged blood vessels. Trauma, hyperoestrogenaemia in pregnancy or with use of oral contraceptive pills, infection, allergy and other immunological factors are suggested in its pathogenesis.

Clinically ALHE presents as red to purple coloured papules, plaques or nodules of 0.1 to 2 cm in size in the dermis or subcutaneous tissues of the head and neck, particularly in the auricular and peri-auricular areas, while less commonly affecting the trunk, limbs, colon, lip and oral mucosa. Rarely is the genitalia involved. The commonest symptoms are pain, pulsations, pruritus and spontaneous bleeding.<sup>2</sup> It is more common in woman aged 20 to 50 years, with a mean age of onset of 30-33 years.

ALHE over the genital area is very rare. The vulval papules and pruritus vulvae described in other case reports were similar to those found in our case.<sup>3,4</sup> Surgical excision was reported to be successful in treating ALHE. Table 1 summarises the features of reported cases of genital ALHE. Treatment is usually required for ALHE as spontaneous remission is rare. Complete surgical excision, laser ablation, electrocautery, cryotherapy and radiotherapy<sup>5</sup> have been reported in various case reports. A recurrence rate of 33% associated with surgical excision was reported in one review with incomplete removal of the tumour.<sup>2</sup> Topical and intra-lesional steroid, topical tacrolimus ointment,<sup>6,7</sup> topical imiquimod,<sup>8,9</sup> oral isotretinoin,<sup>10</sup> oral pentoxifylline,<sup>11</sup> oral indomethacin farnesil,<sup>12</sup> intralesional interferon alfa-2b,<sup>13</sup> intravenous anti-interleukin-5 antibody<sup>14</sup> and oral suptatostilate<sup>15</sup> have also



**Table 1.** Summary of case reports of biopsy proven angiolymphoid hyperplasia with eosinophilia over the genital area

Author	Age and sex	Symptoms	Treatment	Remarks
Aguilar et al 1990 <sup>3</sup>	F/27	4-month history of vulval pruritus, with red-purple 2 mm to 8 mm vascular painless lesions on external aspect of the labia majora	Excisional biopsy	No response to topical corticosteroid cream before excision
Scurry et al 1995 <sup>4</sup>	F/23	1-year history of vulval pruritus, with around 15 discrete papules on both labia majora and minora, mainly on the right side. Further 6 similar papules on the perineum and adjacent to the anus	Excision	The lesions began before starting of oral-contraceptive pills, however, patient reported distressing symptoms after starting oral contraceptive pills
Sezer et al 2007 <sup>16</sup>	M/58	4-year history of multiple hyperpigmented brownish to pink plaques 5-30 mm in diameter over the inguinal folds, scrotum and perineal region	Treated with liquid nitrogen with partial response	The lesions resembled bowenoid papulosis clinically, however, histopathology confirmed ALHE
Dewan et al 2008 <sup>17</sup>	M/47	8-month history of light red marks on glans penis, the lesions expanded into painful nodules and ulcerated	Topical clobetasol propionate (Dermovate) with oral pentoxifylline	Biopsy also suggestive of coexistent lichen planus
Park & Lee 2009 <sup>18</sup>	M/17	15-day history of papules and nodules on penis and scrotum	Injection of intralesional triamcinolone acetonide	Patient also had left varicocele

been reported to be effective. The mechanism of action for most of these agents is not fully understood, and may probably act through mechanisms of anti-angiogenesis, anti-inflammation or inhibition of eosinophils.

In conclusion, angiolymphoid hyperplasia with eosinophilia in the vulva is rare. This lady was successfully treated with surgery and electrocautery for the larger lesions, with additional cryotherapy for smaller lesions. However, recurrence is possible in the future.

## References

1. Seregard S. Angiolymphoid hyperplasia with eosinophilia should not be confused with Kimura's disease. *Acta Ophthalmol Scand* 2001;79:91-3.
2. Olsen TG, Helwig EB. Angiolymphoid hyperplasia with eosinophilia. A clinicopathologic study of 116 patients. *J Am Acad Dermatol* 1985;12:781-96.
3. Aguilar A, Ambrojo P, Requena L, Olmos L, Sanchez Yus E. Angiolymphoid hyperplasia with eosinophilia limited to the vulva. *Clin Exp Dermatol* 1990;15:65-7.
4. Scurry J, Dennerstein G, Brennan J. Angiolymphoid hyperplasia with eosinophilia of the vulva. *Aust N Z J Obstet Gynaecol* 1995;35:347-8.
5. Conill C, Toscas I, Mascaro JM, Jr., Vilalta A, Mascaro

- JM. Angiolymphoid hyperplasia with eosinophilia of the nail bed and bone: successful treatment with radiation therapy. *J Eur Acad Dermatol Venereol* 2004;18:584-5.
6. Mashiko M, Yokota K, Yamanaka Y, Furuya K. A case of angiolymphoid hyperplasia with eosinophilia successfully treated with tacrolimus ointment. *Br J Dermatol* 2006;154:803-4.
  7. Wang S, Li W. Angiolymphoid hyperplasia with eosinophilia successfully treated with tacrolimus ointment. *J Eur Acad Dermatol Venereol* 2010;24:237.
  8. Gencoglan G, Karaca S, Ertekin B. Angiolymphoid hyperplasia with eosinophilia successfully treated with imiquimod. A case report. *Dermatology* 2007;215:233-5.
  9. Redondo P, Del Olmo J, Idoate M. Angiolymphoid hyperplasia with eosinophilia successfully treated with imiquimod. *Br J Dermatol* 2004;151:1110-1.
  10. Oh CW, Kim KH. Is angiolymphoid hyperplasia with eosinophilia a benign vascular tumor? A case improved with oral isotretinoin. *Dermatology* 1998;197:189-91.
  11. Person JR. Angiolymphoid hyperplasia with eosinophilia may respond to pentoxifylline. *J Am Acad Dermatol* 1994;31:117-8.
  12. Nomura K, Sasaki C, Murai T, Mitsuhashi Y, Sato S. Angiolymphoid hyperplasia with eosinophilia: successful treatment with indomethacin farnesil. *Br J Dermatol* 1996;134:189-90.
  13. Oguz O, Antonov M, Demirkesen C. Angiolymphoid hyperplasia with eosinophilia responding to interferon-alpha2b. *J Eur Acad Dermatol Venereol* 2007;21:1277-8.
  14. Braun-Falco M, Fischer S, Plotz SG, Ring J. Angiolymphoid hyperplasia with eosinophilia treated with anti-interleukin-5 antibody (mepolizumab). *Br J Dermatol* 2004;151:1103-4.
  15. Harada K, Kambe Y, Takeda H, Nakano H, Hanada K. Angiolymphoid hyperplasia with eosinophilia: successful treatment with the antiallergic agent suplatast tosilate. *Dermatology* 2004;208:176-7.
  16. Sezer E, Erbil H, Koseoglu D, Filiz N, Kurumlu Z. Angiolymphoid hyperplasia with eosinophilia mimicking Bowenoid papulosis. *Clin Exp Dermatol* 2007;32:281-3.
  17. Dewan P, Francis ND, Lear JT, Bunker CB. Angiolymphoid hyperplasia with eosinophilia affecting the penis. *Br J Dermatol* 2008;159:755-7.
  18. Park JS, Lee MJ. A case of angiolymphoid hyperplasia with eosinophilia (ALHE) in the genital area accompanied by varicocele. *Int J Dermatol* 2009;48:1264-6.