A Case Report of Multiple Angiolymphoid Hyperplasia with Eosinophilia

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ABSTRACT:

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Angiolymphoid hyperplasia with eosinophilia (ALHE) is rare benign vasoproliferative disorder that most often affects the head and neck. We present an uncommon presentation of ALHE. A middle-aged Thai man presented with multiple red-brown dome-shaped papules on left shoulder, axilla and extended to scapular area, which were diagnosed as ALHE from typical histopathologic examination. In our case, we used a combination of oral propranolol and surgical excision for treatment.

Key words: Angiolymphoid hyperplasia with eosinophilia, ALHE, propranolol, surgical excision

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Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) was first described by Wells and Whimster in 1969[1]. It is an uncommon benign vasoproliferative disorder. Solitary or multiple pink to red-brown dome-shape papules or nodules on head and neck are normally presentation of ALHE, specifically on the preauricular region[2]. Apart from the appearance of ALHE on head and neck, it was rarely found on trunk, extremities and genitalia[2]. The normal symptoms are pruritus, bleeding and pain[2]. The recent reports have yet to ascertain the pathophysiology of the concerned disease[3].

In the view of management, majority of related reports of therapeutic methods comprise of surgical excision, intralesional/topical/systemic corticosteroid, LASER, oral isotretinoin, oral dapsone and oral pentoxifylline[2]. In some cases, the oral propranolol was demonstrated as an alternative treatment of ALHE[4-7].

This report shows an uncommon presentation of ALHE on left shoulder displayed as a group of arrangement. The management was achieved with a combination of oral propranolol and surgical excision.

Case presentation

A Thai 28-year-old male presented multiple pruritic papules with spontaneous bleeding on his left shoulder for 6 months ago. The patient noticed his first lesion about 1 year ago in which the number of nodules were slowly increasing through time. He also complained about intensely pruritus and spontaneous bleeding from the nodules which was needed to pressure for a long time to stop.

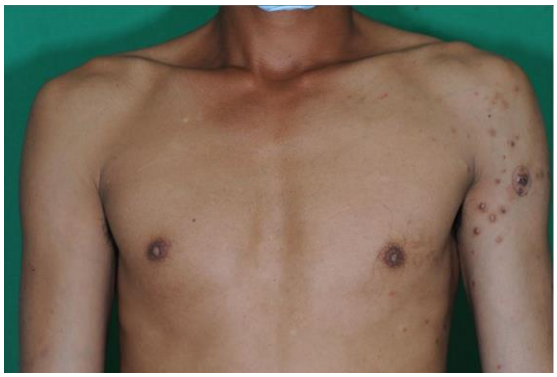

Figure 1 Multiple red-brown firm dome shape papules on left shoulder, axilla and scapular region with multiple excoriation marks

He had been diagnosed as hemangioma with mixed inflammatory cell infiltration predomination with eosinophils from previous biopsy and was received the treatment with oral antihistamine and topical corticosteroid. Nevertheless, the symptoms and lesions had shown lack of improvement. Thus, patient was referred to our hospital for the further investigation and management.

Physical examination revealed multiple red-brown firm dome-shape papules on left shoulder, axilla and scapular region with multiple excoriation marks as shown on Figure 1.

There was no associated with regional lymphadenopathy. His blood examination for complete blood count with differential showed eosinophilia.

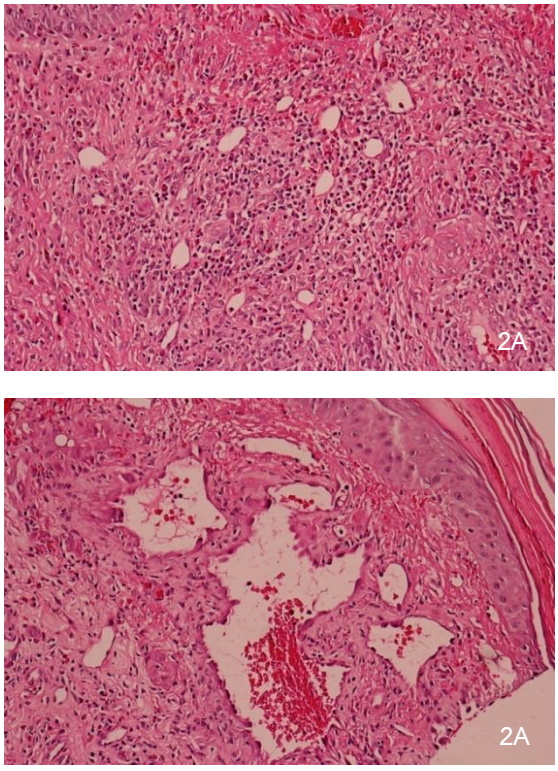

Figure 2 Histologic findings demonstrated dense infiltration with eosinophils and lymphocytes in the dermis admixed with proliferation of vascular component, lining with enlarged pink plum endothelial cells called "Hobnail appearance" (H&E, objective X20)

Histopathologic finding demonstrated, dense infiltration of inflammatory cells predominated with eosinophils and lymphocyte admixed with proliferation of vascular components (Figure 2A) that lining with enlarged pink plump endothelial cells called "Hobnail appearance", and endothelial cells do not have atypia features. (Figure 2B)

From the above findings, our diagnosis is multiple ALHE.

We prescribed oral propranolol starting with low dosage and considered to increase in dosage depending on toleration of the patient and did not have any serious side effects; for example, hypotension, bradycardia and bronchospasm.

After two months of treatment with oral propranolol, there was no improvement with clinical presentations and symptoms. Therefore, we have decided to combine treatment with surgical excision.

Discussion

Form previous researchers, ALHE is rare benign vasoproliferative disorder, also named as epithelioid hemangioma. The exactly prevalence is unknown yet[3]. Most of the patients are the Asians (27.4%), without sex predominance and the mean age presentation is 37 year[2].

The pathogenesis of ALHE is still unclear, while recent literatures revealed various assumptions; for instance, trauma, hyperestrogenemia (e.g. pregnancy, oral contraceptive use)[2], environmental factor (e.g. insect bites, parasites)[8], infectious agent (HHV-8)[9], atopy, reactive hyperplasia and benign neoplasia[2]. In addition, some authors had

hypothesized that ALHE may represent a $CD4^+$ T-cell lymphoproliferative disorder because their finding showed that T-cell receptor gene rearrangement and monoclonality had been previously observed in ALHE case[3]. Furthermore, there were two reported cases of ALHE being associated with peripheral T cell lymphoma[10,11].

As state above, the typical case is characterized by red-brown papules that commonly appear on the head and neck, particularly in the preauricular region[2]. However, lesions were also found on entire body surface and rarely found on internal organs such as heart, colon, kidney, bone and lung[2,3]. The systematic review of retrospective ALHE cases showed that only 1.9% of total cases (908 patients) were identified to have a lesion on the shoulder[2]. In our interesting case, it was unusual presentation of the disease that the patient had several gradually developed lesions on his left shoulder, axilla and extended to scapular region without history of trauma.

ALHE commonly present with some symptoms, which the most common symptoms were pruritus and followed by spontaneous bleeding and pain respectively[2]. This is consistent with our patient that had intense pruritus and bleeding.

Our patient was diagnosed by the characteristic features of histology compatible with the disease. The laboratory analysis includes a complete blood count which resulted peripheral eosinophilia. From the previous literatures, laboratory findings in ALHE were found peripheral eosinophilia at approximately 20% of the total cases[12].

Although the disease is benign condition, its management is challenging as the post-treatment recurrence rate of the concerned disease is more than 40%[3]. The prognosis of the disease in multiple lesions had found a higher recurrent rate when compared with single lesion. The mean time to recurrence was 4.2 years, associated with earlier age of onset, longer duration, pruritus and bleeding[2].

Therapeutic modality for treatment ALHE includes surgical excision, topical and systemic corticosteroid, laser (pulse dye laser, Argon, CO2 laser), cryotherapy, radiotherapy, electrosurgery, systemic treatment with oral isotretinoin, dapsone, pentoxifylline[2,3] and propranolol[4-7]. However, from previous case reports, surgical excision is considered as the most effective treatment due to the lower rate of failure comparing to other reported therapeutic medthods[2,3]. Moreover, some reported cases showed the interesting successful treatment by oral propranolol[4-7].

The use of oral propranolol, which is effective for vascular tumors has been well established in the management of infantile hemangioma[5,7]. Although the definite mechanism

remains unclear, the hypothesis has been proposed that beta-blocker-induced localized vasoconstriction, inhibition of angiogenesis and apoptosis of capillary endothelial cells[5,13].

We had initially tried the alternative management with oral propranolol. However, there was no improvement in clinical presentations and symptoms. Hence, with the high recurrence rate of multiple lesions, we decided to combine surgical excision.

Conclusion

We report an unusual presentation of ALHE which presented with multiple red-brown dome shape papules on left shoulder, axilla and extended to scapular region. In respect of management, the combination with oral propranolol and surgical excisional is provided in our case.

Learning-points

- ALHE usually found on head and neck region which characterized by itching and spontaneous bleeding.

- Typical histology for ALHE is dense infiltration of eosinophils and lymphocytes with proliferation of vascular component and a hobnail appearance.

- There are several therapeutic options for treatment ALHE, but we provided a combination of oral propranolol and surgical excision.

Acknowledgment

The patients in this manuscript have given written informed consent to publication of their case details.

Declaration of conflicting interest

The authors declare that there are no conflict of interest.

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