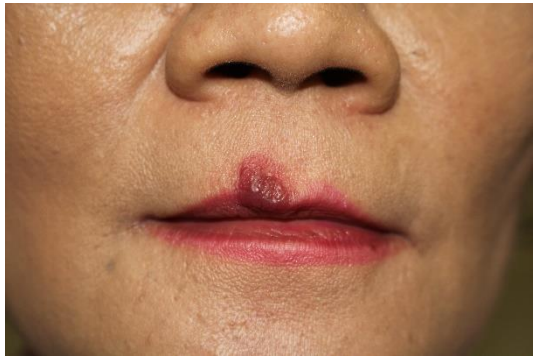
Case 18

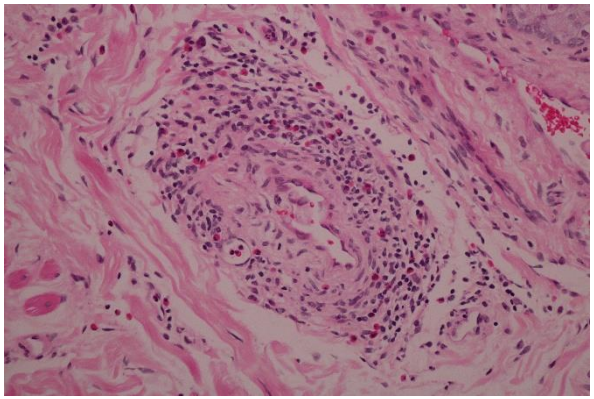
A 59-year-old Thai widowed female from Bangkok
Chief complaint: A red nodule at upper lip for two months



Present illness: The patient developed tiny deep red papule on her upper lip for one year. There was bleeding on scraping but not tender or pruritus. There was no history of prior trauma.
She noticed that her lesion gradually grew into an obvious one in two months.
There was no other looked-alike lesion located on other part of her body.
Past history
She has well-controlled hypertension.
None of her siblings had the looked-alike lesion as hers.
No history of food or drug allergy
Physical examination
HEENT: not pale, no jaundice
Lymph node: no palpable axillary and supraclavicular node
Heart: normal S1S2, no murmur
Abdomen: no hepatosplenomegaly

Skin examination
Solitary erythematous rough-top nodule at size 0.6 cm at medial part of upper lip

Histopathology (S15-023341, upper lip)
- Multiple thick – wall, medium vessels in the upper and mid dermis
- The vascular wall lined by plump and epithelioid endothelial cells
- Dense perivascular and nodular inflammatory cell infiltrate of lymphocytes and some eosinophils in stroma
- **Microscopic diagnosis**: angiolymphoid hyperplasia with eosinophils (ALHE).



Diagnosis: Angiolymphoid Hyperplasia With Eosinophilia (ALHE)

Treatment: V-Beam Laser is considered.

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Discussion:

Angiolymphoid hyperplasia with eosinophilia (ALHE) or epithelioid hemangioma (EH), is an uncommon idiopathic vascular neoplasm characterized by proliferation of histiocytoid endothelial cells with lymphoid and eosinophilic inflammatory infiltration. Although ALHE is considered a benign condition, recurrence is up to one-third of cases in the absence of complete surgical excision. However, reaction to trauma and arteriovenous shunting are considered relevant. ALHE has its many unique characteristics including prominent proliferation of plump endothelial cells, and accompanying eosinophilic and lymphocytic inflammation, often with formation of lymphoid follicles, which can differentiate from other vascular neoplasms.[1]

The presentation of ALHE is most common in young to middle-aged adults as single or multiple, flesh- to plum-colored papules or nodules, from a few to several centimeters ranging in size. It locates in dermal and/or subcutaneous layer.[1] The most involved region is head and neck. Due to the vascular nature of the lesions, tenderness, pulsation, pruritus, or bleeding, also spontaneously or after minor trauma, may occur in some patients.[2] Peripheral blood eosinophilia and regional lymphadenopathy may also be present. ALHE is predominately involve females. The period from onset of lesions to initial medical consultation is from a few months to many years.[3]

The pathogenesis of ALHE is currently inconclusive. There are several hypotheses including a reactive process, a neoplastic process, arteriovenous shunting and local trauma.[2,3] The vascular proliferation in ALHE may explain by the response of the endothelial cells to proliferative stimuli generated by the accompanying inflammatory cells and immunologic allergic reaction. Though considered a benign tumefaction, ALHE has been associated with various lymphoproliferative conditions.[4]

The histopathological feature of ALHE is distinct. The first hallmark is the proliferation of blood vessels of varying sizes lined by plump endothelial cells. The histiocytoid endothelial cells are enlarged and mostly cuboidal with occasional ‘’hobnailing’’, which associated with the presence of cytoplasmic vacuoles in these cells, resulting in cytoplasmic protrusion into lumina. The second characteristic is lymphoid and eosinophilic inflammatory infiltration inflammation. Lymphocytes and eosinophils diffusely confine and infiltrate into the blood vessels. The presentation of lymphocytes may be diffusely or may form distinct follicles with germinal centers. [2,3] Approximately 20% of the patients have blood eosinophilia without elevation of immunoglobulin E (IgE) levels.[1,3] The predominant vascular or inflammatory component may defined the age of the lesion. In early stage or actively growing ALHE, the vascular component predominates and the vessels are immature with prominent epithelioid endothelial cells. While the late stage, lymphocytes become more prominent and the endothelial cells lining the maturing vessels become smaller and less epithelioid.[2,3]

Previously thought to be same entity, Kimura disease is the major differential diagnosis. In the present, it has been found that ALHE is clinically and pathologically distinct from Kimura disease.

Kimura disease is a systemic immune-mediated process that typically presents with lymphadenopathy, eosinophilia, increased serum levels of IgE, and may be associated with renal disease. The hallmark histologically features of Kimura disease include florid lymphoid follicles with germinal center formation, eosinophilic infiltrates, eosinophilic microabscesses, and eosinophilic folliculolysis. Furthermore, histiocytoid or epithelioid cells, which is the characteristic of ALHE, does not present in Kimura disease.[1,5] Other differential diagnosis to be considered for ALHE include angiosarcoma, Epithelioid hemangioendothelioma, Kaposi sarcoma and pyogenic

granuloma.[1,3]

The treatment of choice of ALHE is completely surgical excision because the lesion rarely recurs. One-third of cases that were incompletely excised recur at the same site or distant from it, but still typically along the course of the affected vessel.[2,3] Other destructive procedures such as Mohs micrographic surgery, pulsed-dye laser, carbon dioxide laser, and cryosurgery, have been applied and reported.[1] In medical treatment perspective, conventional intralesional corticosteroids and irradiation is usually ineffective. However, new regimens have been extensively studied and have shown favourable results, including topical imiquimod, tacrolimus[6], isotretinoin, and interferon a-2a.[1]

In conclusion, this patient was diagnosed ALHE, based on the histopathology. We plan to treat her with V-beamed laser. The result will then be followed.

References

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