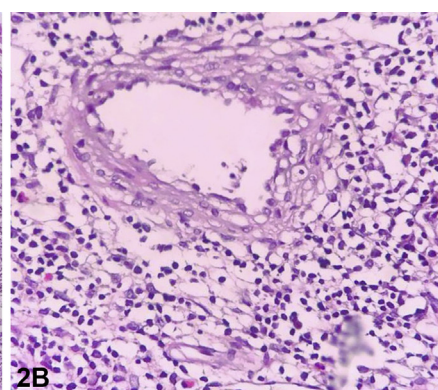
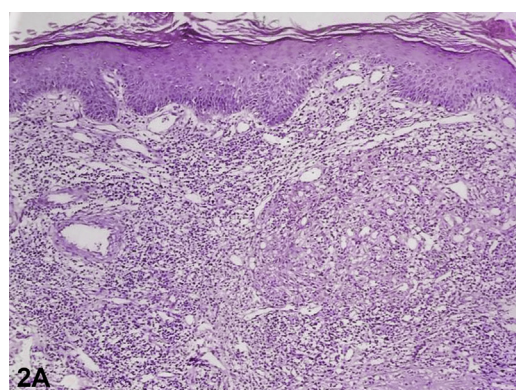


A solitary erythematous papule on the nose



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A 40-year-old healthy white Persian woman presented to the clinic with a 5-month history of a slightly pruritic papule on her nose. The lesion had gradually increased in size and bled with minor trauma. Physical examination found a nontender, indurated erythematous 0.7- × 0.7-cm dome-shaped papule on the dorsum of the nose (Fig 1) with no associated regional lymphadenopathy. The rest of the examination was otherwise unremarkable. Histopathology of the dermis found a vascular proliferation with prominent plump endothelial cells and a mixed infiltrate including lymphocytes and eosinophils (Fig 2, A and B). Biochemical and hematologic studies including absolute eosinophil count were normal.

Question 1: What is the most likely diagnosis based on clinical and histopathologic images?

- A. Kimura disease
- B. Angiolymphoid hyperplasia with eosinophilia (ALHE)
- C. Pyogenic granuloma
- D. Kaposi sarcoma
- E. Granuloma faciale

Answers:

A. Kimura disease — Incorrect. Kimura disease commonly occurs in young Asian males. It usually presents with one or multiple subcutaneous masses on the head and neck often accompanied by regional lymphadenopathy. The disease is often associated with peripheral blood eosinophilia and elevated IgE, which were both normal in the presented patient. On histopathologic examination, hyperplastic lymphoid follicles with prominent germinal centers and eosinophilic infiltrate are typical features of Kimura disease.¹

B. ALHE — Correct. ALHE is an uncommon, benign disorder that commonly presents as solitary or multiple papulonodular lesions located mostly on the ear, forehead, and scalp and rarely on the nose.^{1,2} Histologically, ALHE shows a vascular proliferation with prominent hobnailed endothelial cells and an intense eosinophilic and lymphocytic infiltrate.¹

C. Pyogenic granuloma — Incorrect. Pyogenic granuloma may also present as a vascular papule. On histology, however, a vascular proliferation containing lobular collections of small round capillaries separated by fibrous stroma is seen in pyogenic granuloma, which is different from thick-walled vessels with hobnailed endothelial cells found in ALHE.³

D. Kaposi sarcoma — Incorrect. Although solitary lesions of Kaposi sarcoma can clinically mimic ALHE, histologically it is defined by proliferation of spindle cells with slit-like vascular spaces, which

is in sharp contrast with vascular proliferation with prominent plump endothelial cells seen in ALHE.³

E. Granuloma faciale — Incorrect. Granuloma faciale usually presents as a red-brown plaque located on the face. In addition, it is a leukocytoclastic vasculitis as compared with ALHE, which is a vascular lesion. In fact, granuloma faciale consists of a grenz zone with an underlying diffuse polymorphous infiltrate containing lymphocytes, eosinophils, plasma cells, and neutrophils as well as leukocytoclasia and fibrin deposition.³

Question 2: Which of the following is true regarding this disorder?

- A. The lesion is usually asymptomatic.
- B. The disease has the potential for distant metastasis.
- C. Lymphadenopathy is seen in more than 90% of patients.
- D. It is often seen in young or middle-age adults.
- E. The disease usually presents with multiple lesions.

Answers:

A. The lesion is usually asymptomatic — Incorrect. Most cases of ALHE are symptomatic. Patients may complain of pruritus, bleeding and pain.⁴

B. The disease has the potential for distant metastasis — Incorrect. Metastatic disease in ALHE has not yet been reported.⁴

C. Lymphadenopathy is seen in more than 90% of patients — Incorrect. ALHE is associated with reactive lymphadenopathy in only 5% to 20% of patients. Lymphadenopathy, however, is a characteristic feature of Kimura disease.⁴

D. It is often seen in young or middle-age adults — Correct. ALHE is often seen in young or middle-age adults.⁴

E. The disease usually presents with multiple lesions — Incorrect. Only about 20% of patients present with multiple lesions.⁴

Question 3: Which of the following is the most effective treatment for this patient?

- A. Intralesional or systemic glucocorticoids
- B. Pulsed dye laser
- C. Surgical excision
- D. Cryotherapy
- E. Electrosurgery

Answers:

A. Intralesional or systemic glucocorticoids — Incorrect. With the recurrence rates reaching as high as 79% and 87%, intralesional and systemic corticosteroids are not considered effective treatment options.⁵

B. Pulsed dye laser — Incorrect. The recurrence rate of around 50% makes pulsed dye laser an acceptable treatment option for ALHE. This technique is especially used for elderly, debilitated patients and poor surgical candidates including patients requiring systemic anticoagulation or those with multiple lesions.⁵

C. Surgical excision — Correct. Current literature recommends the use of surgical excision as the most effective treatment modality for ALHE, which will lead to a significantly lower treatment failure rate (ie, 40%) compared with other available treatment modalities.⁵

D. Cryotherapy — Incorrect. Although cryotherapy can potentially be an option in the treatment of ALHE, its 80% reported failure rates makes this modality a less-favorable option among experts.⁵

E. Electrosurgery — Incorrect. Similar to cryotherapy, electrosurgery has been associated with failure rates as high as 88% in published evidence, which makes it an unfavorable option in the treatment of ALHE.⁵

Abbreviation used:

ALHE: angiolymphoid hyperplasia with eosinophilia

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