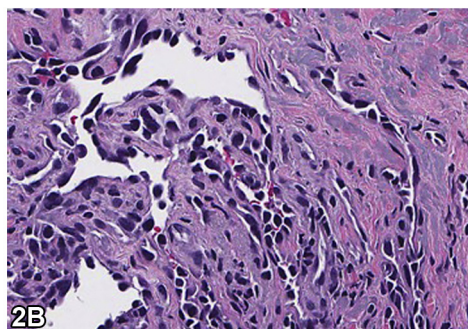
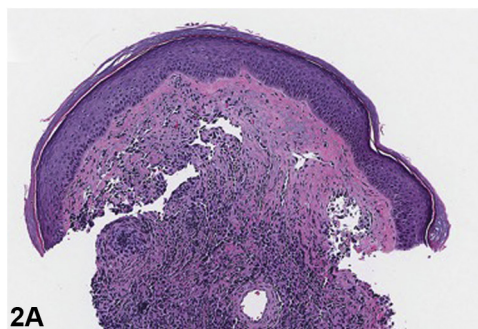


A red plaque on the face of a healthy man



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A 65-year-old man presented with a red, soft plaque on his left nasal sidewall of 2 months' duration (Fig 1). Per the patient report, the plaque initially developed 2 days after dental surgery. Previous treatments included several courses of antibiotics, including clindamycin, amoxicillin-clavulanate, and cefalexin, ranging in duration from 1 week to 4 weeks of therapy. A computed tomography scan of the face with intravenous contrast revealed

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a soft-tissue swelling without evidence of an abscess. On presentation to the dermatology clinic, the patient denied prior history of radiation exposure and chronic swelling of the involved area and was without a personal or family history of skin cancer. Biopsies were obtained for hematoxylin-eosin staining (Figs 2 and 3). Tissue culture was negative for bacteria, fungi, and mycobacteria.

Question 1: What is the most likely diagnosis?

- A. Cutaneous dental sinus tract
- B. Angiosarcoma
- C. Erysipelas
- D. Kaposi sarcoma
- E. Angiolymphoid hyperplasia with eosinophilia

Answers:

A. Cutaneous dental sinus tract — Incorrect. Cutaneous dental sinus tracts are communications that allow the drainage of purulence from the oral cavity onto the skin surface and are typically caused by chronic periapical abscesses. Common presentations include a smooth, erythematous, slightly tender nodule on the skin overlying the mandible.¹

B. Angiosarcoma — Correct. Cutaneous angiosarcoma (CAS) is a rare, malignant tumor of vascular mesenchymal origin. Due to the variability and nonspecificity of clinical presentations, the diagnosis of angiosarcoma remains a challenge.² Common histopathologic features include poor circumscription with irregularly dilated matrices of vessels lined by atypical endothelial cells that dissect through the dermis. CAS typically expresses endothelial markers, with CD31 considered the gold standard of diagnosis.²

C. Erysipelas — Incorrect. Erysipelas is a superficial bacterial infection of the skin, most commonly caused by the β -hemolytic group A streptococci.³ Erysipelas is a clinical diagnosis and usually presents as well-demarcated tender plaques involving the face.

D. Kaposi sarcoma — Incorrect. Kaposi sarcoma (KS) is a malignant neoplasm of lymphatic endothelial cell origin secondary to human herpesvirus 8.⁴ The clinical presentation may vary based on the disease subtype, and histopathologic evaluation typically reveals a vascular proliferation composed of varying proportions of pleomorphic spindle cells and irregular, slit-like blood vessels that surround the pre-existing vessels. Immunohistochemistry highlighting human herpesvirus 8 positivity is key to diagnosis.⁴

E. Angiolymphoid hyperplasia with eosinophilia — Incorrect. Angiolymphoid hyperplasia with

eosinophilia is an uncommon, benign vascular proliferation that typically presents with flesh-colored, pink, dull red, or brown papulonodules on the head and neck. Histopathologic findings include well-circumscribed dermal or subcutaneous blood vessels lined by enlarged endothelial cells with a variable inflammatory infiltrate of lymphocytes, eosinophils, and mast cells.⁵

Question 2: What is the best treatment for this localized condition?

- A. Surgery and/or adjuvant radiation
- B. Paclitaxel
- C. Pulse-dye laser
- D. Cryosurgery
- E. Systemic antibiotics

Answers:

A. Surgery and/or adjuvant radiation — Correct. The initial treatment of angiosarcoma typically involves surgery; however, negative surgical margins are achieved in only 21% to 47% of cases.² Thus, treatment often relies on a multimodal approach, most commonly utilizing surgery with radiation for localized CAS.²

B. Paclitaxel — Incorrect. Paclitaxel, a taxane, has demonstrated clinical benefits in the treatment of metastatic or unresectable angiosarcoma.² Given the localized nature of the patient's CAS, surgery with the consideration of adjuvant radiation is considered the best treatment option.

C. Pulse-dye laser — Incorrect. Although pulse-dye lasers may be considered a treatment modality for angiolymphoid hyperplasia with eosinophilia, lasers are not a treatment of choice for CAS.⁵

D. Cryosurgery — Incorrect. The treatment of KS depends on the extent of involvement and the patient's underlying immune status. For patients with localized KS, cryosurgery may be utilized, with the therapeutic objective being ulceration of the epidermis to allow for scarring to replace atypical vessels.⁴ Cryosurgery is not a treatment modality for CAS.

E. Systemic antibiotics — Incorrect. Systemic antibiotics are not a treatment for CAS. Instead,

systemic antimicrobials, specifically penicillin, may be used to treat erysipelas, which is most commonly due to β -hemolytic streptococci. Clindamycin may be used for patients with true penicillin allergies.³

Question 3: Which of the following is true regarding the prognosis of this condition?

- A.** The prognosis is good, and this condition can be managed with oral antibiotics in the outpatient setting
- B.** The 5-year relative survival rate for a specific stage of this condition is 80%
- C.** This condition is considered a benign disease but may recur after treatment in more than 40% of cases
- D.** Although commonly misdiagnosed, this condition has an excellent prognosis once proper endodontic therapy is performed
- E.** There is an overall poor prognosis of this condition, with the 5-year survival between 10% and 50%

Answers:

- A.** The prognosis is good, and this condition can be managed with oral antibiotics in the outpatient setting – Incorrect. This refers to the prognosis of erysipelas, which is usually managed in the outpatient setting with oral antibiotics. Importantly, hospitalization for intravenous antibiotics may be considered in severe cases affecting those who are immunocompromised, infants, and the elderly.³
- B.** The 5-year relative survival rate for a specific stage of this condition is 80% – Incorrect. There are 4 widely recognized types of KS; however, they differ in epidemiology and prognosis. The 5-year relative survival rate for KS, which compares people with the same type and stage of KS to people in the overall population, is 80%.²
- C.** This condition is considered a benign disease but may recur after treatment in more than 40% of cases – Incorrect. The prognosis described here is in reference to angiolymphoid hyperplasia with eosinophilia. Although spontaneous regression may occasionally occur, angiolymphoid hyperplasia with eosinophilia tends to persist for years, with recurrence after treatment in more than 40% of cases. Early age of onset, long disease duration, multiple lesions, and symptomatic disease have been associated with higher recurrence rates.⁵
- D.** Although commonly misdiagnosed, this condition has an excellent prognosis once proper

endodontic therapy is performed – Incorrect. Cutaneous dental sinus tract is frequently misdiagnosed due to its rarity of occurrence and lack of associated symptoms. Once diagnosed, however, definitive treatment through oral therapy to eliminate the source of infection, whether it is through endodontic treatment or extraction, is effective and provides a cure.¹

E. There is an overall poor prognosis of this condition, with the 5-year survival between 10% and 50% – Correct. Angiosarcomas make up less than 1% of all sarcomas, with approximately 2 to 3 new cases per 1 million diagnoses yearly.² Importantly, many angiosarcomas are highly aggressive, with a 5-year survival between 10% and 50% and a median overall survival ranging from 30 to 50 months.² The clinical variability of these tumors contributes to delayed diagnosis, and that, coupled with their genetic heterogeneity, makes them difficult to treat. Surgical excision with wide margins is considered the mainstay of therapy in localized disease; however, the infiltrative and satellite foci often lead to difficulty in obtaining negative margins and frequent local recurrences. Thus, a multimodal approach, which often includes radiation, is often necessary.² Our patient was treated with wide local excision followed by adjuvant radiation given positive margins. He remains without evidence of disease at 2 years.

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Abbreviations used:

CAS: cutaneous angiosarcoma
KS: Kaposi sarcoma

Conflicts of interest

None disclosed.

REFERENCES

1. Cantatore JL, Klein PA, Liebllich LM. Cutaneous dental sinus tract, a common misdiagnosis: a case report and review of the literature. *Cutis*. 2002;70(5):264-267.
2. Florou V, Wilky BA. Current and future directions for angiosarcoma therapy. *Curr Treat Options Oncol*. 2018;19(3):14.
3. Bisno AL, Stevens DL. Streptococcal infections of skin and soft tissues. *N Engl J Med*. 1996;334(4):240-245.
4. Etemad SA, Dewan AK. Kaposi sarcoma updates. *Dermatol Clin*. 2019;37(4):505-517.
5. Adler BL, Krausz AE, Minuti A, et al. Epidemiology and treatment of angiolymphoid hyperplasia with eosinophilia (ALHE): a systematic review. *J Am Acad Dermatol*. 2016;74(3):506-512.e11.