Violaceous Nodules on the Hard Palate

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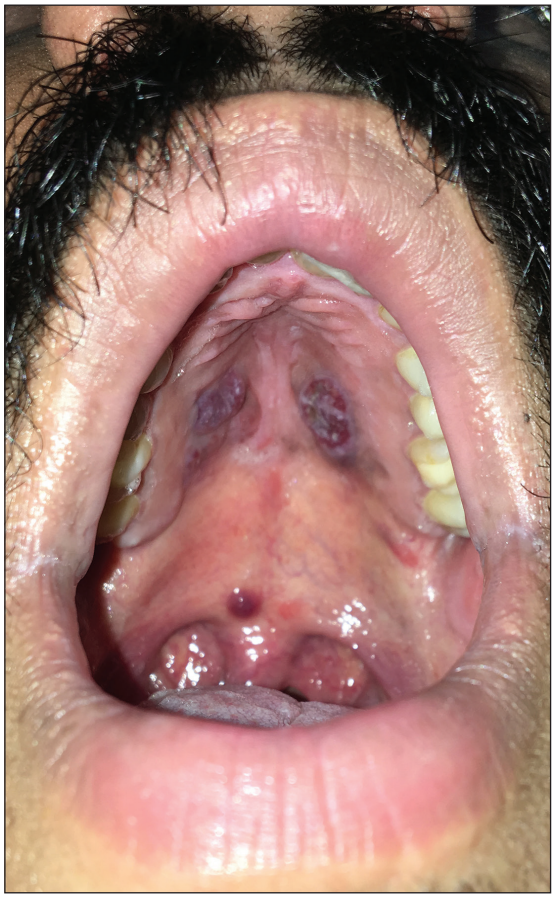

A 30-year-old man presented to our outpatient clinic with rapidly growing, ulcerated, violaceous lesions on the hard palate of 4 months' duration. Physical examination revealed approximately 2.0×1.5-cm, centrally ulcerated, violaceous, nodular lesions on the hard palate, as well as a 4-mm pinkish papular lesion on the soft palate.

WHAT'S THE **DIAGNOSIS?**

a. bacillary angiomatosis
b. Kaposi sarcoma
c. lymphangioma
d. lymphoma
e. melanoma

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The authors report no conflict of interest.
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THE **DIAGNOSIS:** Kaposi Sarcoma

A 4-mm punch biopsy from the border of an ulcerated nodular lesion on the hard palate demonstrated diffusely distributed spindle cells, cleftlike microvascularity with extravasated erythrocytes, and widespread human herpesvirus 8 immunoreactivity on histopathology (Figure 1). Serologic tests were positive for human immunodeficiency virus (HIV) infection; HIV RNA was 14,584 IU/mL and the CD4 count was 254/mm^3. The patient was diagnosed with Kaposi sarcoma (KS) and referred to the infectious disease department for initiation of antiviral therapy. Marked regression was detected after 6 months of highly active antiretroviral therapy (HAART) without any additional treatment (Figure 2).

Kaposi sarcoma is a human herpesvirus 8–associated angioproliferative disorder with low-grade malignant potential. There are 4 well-known clinical types: classic, endemic, iatrogenic, and AIDS associated.[1] Involvement of the oral cavity may be seen in all types but mostly is associated with the AIDS-associated type, which also could be a signal for undiagnosed asymptomatic HIV infection.[2] Oral KS most often affects the hard and soft palate, gingiva, and dorsal tongue, with plaques or tumors ranging from nonpigmented to brownish red or violaceous. AIDS-associated KS is known to be related to cytokine expression, which is induced by HIV infection causing immune dysregulation by altering the expression of cytokines, including IL-1, tumor necrosis factor α, and IL-6.[1] An in vitro study showed that cytokines secrete a number of angiogenic growth factors that, along with HIV proteins, induce and proliferate cells to become sarcoma cells. Integrins and the apoptosis process also are important in proliferation and neovascularization of KS tumor cells.[3]

Bacillary angiomatosis (BA) is a rare manifestation of infection caused by *Bartonella* species, which leads to vasoproliferative lesions of the skin and other organs. Bacillary angiomatosis affects individuals with advanced HIV or other immunocompromised individuals and may clinically mimic KS, which is similarly characterized by red-purple papules, nodules, or plaques. Differentiating BA from KS largely depends on histopathologic examination, with BA demonstrating protuberant endothelial cells surrounded by clumps of bacilli that are visible on Warthin-Starry silver stain.

Lymphangioma is a benign hamartomatous hyperplasia of the lymphatic vessels. The majority of lymphangiomas are superficial, but a few may extend deeply into the connective tissue. Intraoral lymphangiomas occur more frequently on the dorsum of the tongue, followed by the palate, buccal

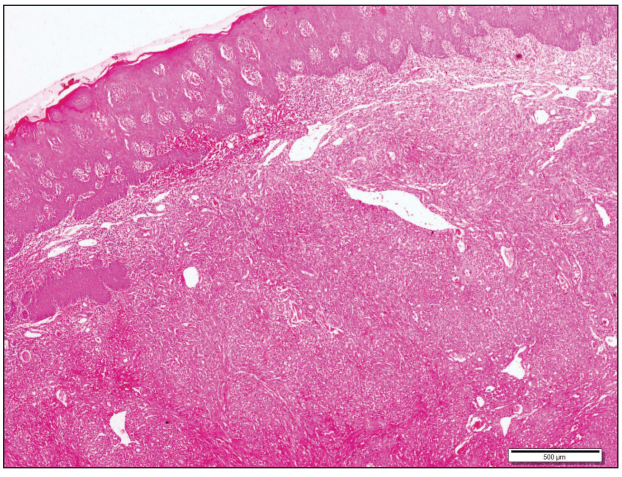

FIGURE 1. A biopsy from the border of an ulcerated nodular lesion on the hard palate showed diffusely distributed spindle cells, cleftlike microvascularity with extravasated erythrocytes, and widespread human herpesvirus 8 immunoreactivity (H&E, original magnification ×4).

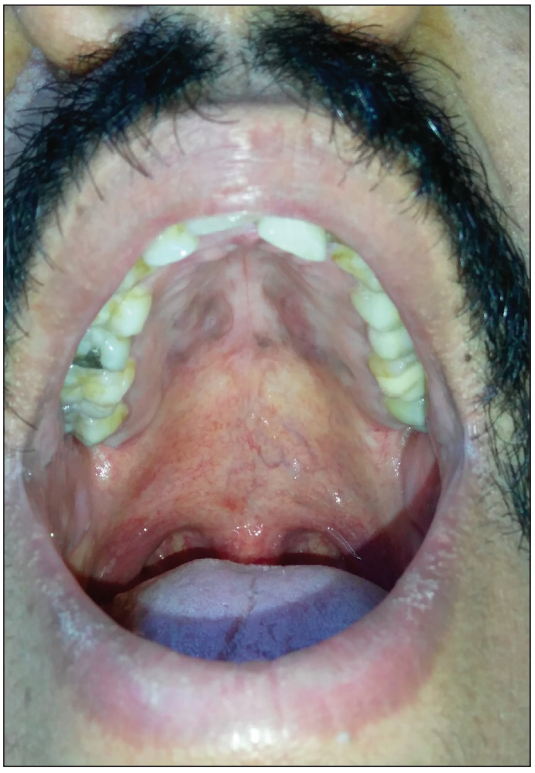

FIGURE 2. Marked regression of Kaposi sarcoma was detected after 6 months of highly active antiretroviral therapy without any additional treatment.

mucosa, gingiva, and lips. They may be differentiated with their soft quality, pebblelike surface, and translucent vesicles.

Malignant tumors of the oral cavity are rare, representing only 5% of tumors occurring in the body.[4] Among malignant tumors of the oral cavity, squamous cell carcinomas are the most frequent type (90%–98%), and lymphomas and melanoma are the most outstanding among the remaining 2% to 10%. Both for lymphoma and mucosal melanoma, the most common sites of involvement are the soft tissues of the oral cavity, palatal mucosa, gingiva, tongue, cheeks, floor of the mouth, and lips.[4] Although mucosal melanoma lesions usually are characterized by pigmented and ulcerated lesions, amelanotic variants also should be kept in mind. Histopathologic examination is mandatory for diagnosis.

Intralesional chemotherapy with vinblastine or bleomycin, radiotherapy, electrochemotherapy, systemic antiretroviral therapy (ie, HAART), and chemotherapy with daunorubicin and pegylated liposomal doxorubicin are the main treatment options.[5,6] The immune system activator role of HAART leads to an increased CD4 count and reduces HIV proteins, which helps induction of the proliferation and neovascularization of KS tumor cells.[3] This effect may help resolution of KS with localized involvement and allows physicians to utilize HAART without any other additional local and systemic chemotherapy treatment.

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