Non-HIV Oral Kaposi Sarcoma: A Case Report and Review of the Literature

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A 28-year-old previously healthy male presented with 3 months of swelling of the right upper gingiva and lower gingiva and right palate (Figures 1 and 2). He initially presented to dentistry with concern for a tooth abscess and subsequently failed 2 rounds of clindamycin. The patient reported unprotected sexual activity with males and females and endorsed a 13 pack-year smoking history and marijuana use, but denied IV drug use. He underwent biopsy with oral surgery which revealed vascular spindle cell proliferation with local hemosiderin deposition and reactive lymphoplasmacytic infiltration, as well as CD31, CD34, and HHV-8 positivity.

During subsequent workup, he was found to be HIV-negative via antigen, antibody, and polymerase chain reaction testing but was found to be hepatitis B positive. An immunodeficiency workup was negative. He was sent to an outside hospital otolaryngologist who repeated the biopsy and confirmed the diagnosis of Kaposi sarcoma (KS) per review by the National Institute of Cancer. He received a course of amoxicillin/clavulanate with slight improvement in the size of the lesions. A complete blood count, chemistry, flow cytometry, as well as upper and lower gastrointestinal endoscopies were within normal limits.

The lesions were progressively increasing in size and associated with intermittent bleeding and pain. Computed tomography and positron emission tomography scans showed a hypermetabolic, expansile lesion of the right maxilla with extension to the hard palate and inferior anterior aspect of the right maxillary sinus. No distant spread was noted. Surgical resection was considered at multidisciplinary tumor board, but ultimately primary radiotherapy (RT) was recommended. A dose of approximately 40 Gray (Gy) in 20 fractions limited to the primary site was indicated. The patient desired care to be transferred to a center closer to his residence. He received 25

Figure 1. Exam reveals poor dentition and a violaceous lesion with right palatal and upper gingival involvement.

Figure 2. A 3.5-cm violaceous, soft, exophytic mass of the right maxilla extending from the first molar to the front incisor is seen.

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fractions over 5 weeks and experienced oral thrush, nausea, vomiting, and fatigue. These side effects resolved upon completion of RT. He had a complete response to RT and continues to exhibit no evidence of disease with a follow-up period of 16 months.

This case represents a unique presentation of KS in that the patient was young, previously healthy, not immunosuppressed, HIV-negative, and had isolated oral cavity involvement. We believe this patient was susceptible due to his positive HHV-8 status. Although the literature correlating non-HIV KS and HHV-8 is limited, one seroepidemiological study concluded that 100% of patients with African endemic KS and 96% of American patients with AIDS-associated KS in their cohort tested positive for the lytic HHV-8 antigen.1 Per a comprehensive literature review using the MEDLINE database via PubMed, this patient is also among the youngest to present with isolated, HIV-negative oral KS. The majority of the other reported cases of HIV-negative oral KS are in males older than the age of 45.2-9 Only one other study with a similar presentation.2

Although literature regarding management for HIV-negative oral KS is sparse, one prospective study using RT alone for KS showed a local control rate of 83% with 40 Gy delivered in 20 fractions.3 This dose regimen correlates with what was recommended for our patient. The decrease in purple pigmentation was greatest, and lesion failure was lowest with this regimen when compared with the dose regimens of 20 Gy in 10 fractions and 8 Gy in 1 fraction. It is thought that this regimen was superior because it allowed for the highest total delivered dose, and thus led to a longer time interval of tumor control. Although radiation toxicity to the skin is a concern with deliverance of a high RT dose, toxicity-related side effects were reported to be minimal in this study. Surgical resection remains a viable option for patients whose KS has only local involvement; in one surgically treated case, because of clear surgical margins the decision was made not to use adjuvant radiation.2

Although rare, KS should be considered in the differential for young immunocompetent individuals presenting with an oral cavity lesion. This is only the second description of an HIV-negative patient younger than the age of 45 presenting with an oral KS and the first in the United States. Treatment for KS depends on the level of dissemination of disease, and the management with primary RT has been shown to be an effective treatment option due to the radiosensitivity of KS.2-4,5 In particular, RT appears to work best when delivered in high-fractionated dose regimens, although treatment must be tailored to each patient.

Authors’ Note
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