CASE REPORT

Disseminated oral Kaposi sarcoma lesion with extensive visceral involvement

Kishore Shetty *

University of Texas Health Sciences Center, Medically Complex Patient Clinic, 6516 M.D. Anderson Blvd., Suite 475, Houston, TX 77030, United States

Received 7 August 2005; accepted 9 August 2005

Summary Kaposi’s sarcoma (KS) has been considered the most common malignancy associated with human immunodeficiency virus (HIV) infection because of its propensity to develop in such individuals. This article, reports an unusual presentation of a disseminated oral KS lesion with extensive visceral involvement in an otherwise healthy young African-American male leading to a positive diagnosis of HIV.

© 2005 Elsevier Ltd. All rights reserved.

KEYWORDS Kaposi sarcoma; Disseminated; HIV; AIDS

Case report

A 26-year-old African-American male presented at the Oral Surgery Outpatient Clinic for evaluation of an oral lesion. He first noticed a “gum boil” four months ago which has been steadily increasing in size since its appearance. He failed to seek medical attention for this lesion since it was not painful or has not bled in the past. He also reported unexplained weight loss in the last six months and febrile episodes. His previous medical history was unremarkable. He reported no known drug allergy. At the time of our examination he was taking amoxicillin. He denied tobacco use, alcohol use, illicit drugs use or intravenous drug use.

The patient’s extra oral exam revealed a normocephalic facial symmetry with no signs or symptoms of acute infection, swelling or tenderness. There was a raised well defined reddish nodular mass measuring $2.0 \times 1.8$ cm on the posterior side of his head (Fig. 1). Palpable lymph nodes were appreciated on his left submandibular and posterior cervical chains; the nodes were firm, mobile and painless. Poor oral hygiene was noted with moderate dental plaque and calculus accumulation and generalized gingival edema. Bleeding on periodontal probing was generalized. There was no radiographic evidence of bone loss and the deepest probing was around 4 mm. Additionally, there was a $2 \times 1.8$ cm raised, dark red, soft, exophytic enlargement of the mandibular attached gingiva of lower right canine on the facial and lingual aspect, with a smooth surface, ill defined margins...
and negative blanching (Figs. 2, 3). The lesion was not tender to palpation and showed no erosions or ulcerations. Another 5·4 cm ill-defined raised, diffuse, reddish-blue, soft, nodular enlargement on the posterior right hard palate was observed, involving the soft palate and the uvula and progressing into the oropharynx (Fig. 4). This lesion showed an ulcerated surface and there was no tenderness. The patient repeatedly denied any history of dysphasia or dyspnea or difficulty to chew associated with this lesion. There were no other oral lesions, tumors or masses noted intraorally.

Two other dark-red nodules were observed on his left arm and forehead with the same characteristics described for the oral lesions (Figs. 5, 6). The patient agreed to undergo an incisional biopsy of the exophytic tumor on the facial aspect of attached gingiva. A local anesthesia infiltration of the right mental nerve was obtained using 1.8 mL of lidocaine 2% with epinephrine 1:100,000. The tumor was excised and placed in 10% formalin and submitted for histopathological diagnosis. The histologic examination revealed a cellular spindle tumor within the connective tissue, numerous extravasated erythrocytes and poorly defined vascular slits. These microscopic findings plus the clinical presentation and the medical history were consistent with a diagnosis of Kaposi’s sarcoma.

Additionally, head and neck magnetic resonance imaging (MRI) with contrast solution were ordered for further assessment of respiratory and/or esophageal tract obstruction and did not show any major obstruction of upper airways.
We discussed the results with patients and the risk factors for HIV were elucidated. He agreed to take the enzyme-linked immunosorbent assay (ELISA) test. The test was positive and his status as HIV seropositive was confirmed by the western blot procedure. Subsequent biopsies of the cutaneous and pharyngeal lesions were consistent with a similar diagnosis of AIDS-related Kaposi’s sarcoma. At the time of his hospital admission his CD4+ lymphocytes count was 4.8 cells/cc$^3$ and his CD8+ lymphocytes count was 57.3 cells/cc$^3$ (CD4/CD8 ratio = 0.1) with a viral load of 35000.

**Discussion**

Kaposi’s sarcoma (KS) is a malignant, multifocal systemic disease that originates in the vascular endothelium and has a very variable clinical course. The disease, first described by the Hungarian dermatologist Moritz Kaposi in 1872, was rare and found classically in elderly men of Italian, Jewish, or Slavic ancestry. With the advent of the AIDS epidemic, KS has become one of the most common malignancies associated with HIV infection and was one of the first diseases to define AIDS in 1981.

KS is traditionally separated into four different types: classic, which primarily affects elderly men of Mediterranean and eastern European origin; endemic, which is common in parts of Africa; epidemic or AIDS-associated; and transplantation-associated. The human herpes virus-8 infection and its pathogenic process characterize all four types. Although most Americans remain at low risk for infection of AIDS-HHV-8 KS, homosexual and bisexual men are at strikingly high risk as a result of sexual transmission in some form.

KS sometimes begins as a single lesion of the oral mucosa but more frequently begins as multiple lesions. Fifty percent of the affected patients show oral lesions and 20–25% of these lesions may be the initial site of involvement. KS can invade bone and create tooth mobility when involves the palate or gingiva. The lesion initially is a flat brown or reddish purple macule that does not blanch with pressure. The neoplasm may develop into plaques or nodules, and this progressive malignancy may disseminate to lymph nodes and organ systems. Morbidity of KS oral lesions may be associated with pain, bleeding, and functional interferences caused by the tumor.

Months before manifestation of the tumors, HHV-8 viremia leads to development of specific antibodies. In most cases, epidemic KS causes widespread lesions that erupt in many places soon after AIDS develops. Lesions of epidemic KS may arise on the skin and the mouth and may affect the lymph nodes and other organs, usually the gastrointestinal tract, lung, liver, and spleen. In contrast, classic KS usually involves only one or a few areas of skin, most often the lower legs. At the time of diagnosis of epidemic KS some people experience no symptoms, especially if their only lesions are on the skin. However, many of those with epidemic KS — even those with no skin lesions, will have swollen lymph nodes, unexplained fever, or weight loss. Eventually, in almost all cases, epidemic KS spreads throughout the body. Extensive KS lung involvement can be fatal. More often, however, patients die of other AIDS-related complications, such as infections.

KS afflicts of 10–20% of HIV-positive patients. Some studies suggest that HIV-positive patients with Kaposi’s sarcoma have a shorter survival than might be predicted from their CD4 concentrations.
The authors reported that patients with lesions of the oral mucosa had a higher death rate than those with exclusively cutaneous manifestations of the disease.

KS treatment has been rapidly improving with many new options currently under study or recently proven to be efficacious and licensed for use in patients with AIDS. Various local therapies aim to eradicate small lesions; however, they do not affect the KS in general nor the likelihood of its recurrence. Systemic chemotherapy treats extensive visceral involvement, which no other form of treatment may affect. Newer treatment approaches are aimed at HHV-8 or at the inflammatory cytokine or angiogenic milieu necessary for KS growth.

References