Kaposi’s sarcoma of the oral cavity: Case reports

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Classic Kaposi’s sarcoma is a rare malignant neoplasm of endothelial cell origin. Oral manifestation of Kaposi’s sarcoma is even more uncommon. The purpose of this article is to describe two cases of classic Kaposi’s sarcoma and to discuss the current knowledge regarding its pathogenesis, epidemiology, clinical characteristics, and therapy.

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Introduction

Kaposi’s sarcoma (KS), or multiple idiopathic hemorrhagic sarcoma, is a malignant neoplasm of endothelial cell origin.

Three different clinical patterns of KS have been identified since the tumor was first described by Kaposi in 1872. The first form occurs predominantly in older men living in the Mediterranean basin. The second form is considered an endemic disease of the African continent. It frequently strikes children 10 years of age or younger, as well as older people. The third pattern of KS has been found in patients with acquired immunodeficiency syndrome (AIDS) or other conditions associated with immunodeficiency.

The first form, which is characterized as classic, has a rare incidence in contrast with the other two forms. Oral involvement in patients with the classic form of KS has been reported, but is even more rare.

The purpose of this article is to report two new cases of classic KS and to discuss the current knowledge of the pathogenesis, epidemiology, clinical characteristics, and treatment of the disease.

Case reports

Case 1

In May 1992, a 72-year-old man was referred to the Clinic of Oral Medicine at the University of Thessaloniki, School of Dentistry, for evaluation of an asymptomatic intraoral lesion that had been present for 4 months.

Examination revealed a single tumor, about 2 cm in diameter, involving the gingiva in the maxillary central incisor region. The tumor had a wide peduncle and was covered with bluish red mucosa (Fig 1a). The patient was edentulous and had worn complete dentures for 1 year. Extraoral lesions were not apparent.

The patient underwent complete blood counts, including measurement of platelets and coagulation tests. No deviation from normal was found. Tests revealed that the patient was not infected with human immunodeficiency virus.

The tumor was excised while the patient was under local anesthesia. Histologic examination showed that the tumor consisted of numerous newly formed blood vessels (Fig 1b). Endothelial hyperplasia was evident. The nuclei of the endothelial cells were atypical and mitotic figures were present. In some instances the blood vessels were dilated, while in others their lumina were not clearly apparent. Extravasated erythrocytes were seen in perivascular sites. A diffuse cellular infiltrate consisting of lymphocytes, histiocytes, and plasma cells was observed around the blood vessels. Spindle-shaped cells resembling young fibroblasts were also apparent (Fig 1c).

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Fig 1a  A tumor with a wide peduncle is present in the area of the maxillary central incisor.

Fig 1b  Microscopic view of the tumor in Fig 1a. Numerous newly formed blood vessels and a surrounding cellular infiltrate are evident. (Hematoxylin-eosin stain; original magnification ×10.)

Fig 1c  Higher magnification of Fig 1b. Endothelial hyperplasia and atypical, spindle-shaped cells and their cellular infiltrate are present. (Hematoxylin-eosin stain; original magnification ×40.)

Fig 2a  Plaques with multinodular surfaces are present in the patient's palate.

Fig 2b  Dark macules are present on the upper extremities.

Fig 2c  Dilated blood vessels, endothelial hyperplasia, and atypical extravasated erythrocytes and their cellular infiltrate are evident in the connective tissue. (Hematoxylin-eosin stain; original magnification ×10.)
A diagnosis of classic KS was made; it was based on clinical, laboratory, and histopathologic features.

The patient was reexamined 9 months later. There was no recurrence of the tumor, nor were new lesions present.

Case 2

In June 1992, an 85-year-old man was examined because he had asymptomatic intraoral lesions that had been present for 20 days.

Examination revealed the presence of bluish red plaques with multinodular surfaces involving the border of the hard and soft palates (Fig 2a). In addition to the oral lesions, the patient had preexisting, similar cutaneous lesions, particularly on the upper and lower extremities (Fig 2b).

The patient's blood counts were within the normal values. Results of examinations for human immunodeficiency virus and hepatitis B surface antigen were negative. Computed tomography of the chest and abdomen as well as dynamic ultrasonography of the patient's abdomen did not reveal any pathologic findings.

After local anesthesia was administered to the patient, biopsy specimens were obtained from the oral mucosa (soft palate) and skin lesions. Both tissue specimens showed common histopathologic features that were identical to those described in case 1 (Fig 2c).

A diagnosis of classic KS was established. The patient was referred to an oncologic clinic for systemic treatment.

Discussion

Classic KS is a rare neoplasm. Oral manifestations of KS are even more uncommon. Farman and Uys undertook a literature review and found that, during the period 1928 to 1975, only 50 patients were reported to have oral involvement of classic KS. To our knowledge, eight more cases have been added to the literature since the end of that study. The patients in the present report represent the 9th and 10th cases documented since 1975. Among these 58 cases, 82% were males. Sixty-five percent of the patients were found to be more than 60 years old, 28% were between the ages of 40 and 60 years, and 7% were between the ages of 20 and 40 years. The most frequent site of the lesions was the palate (40%) followed by the tongue (14%) and lips (14%). This type of KS occurs more frequently in individuals living in the Mediterranean basin.

Our patients represented typical cases because they were elderly men with palatal lesions.

The oral lesions of classic KS do not exhibit pathognomonic clinical characteristics. They commonly begin as multiple red-purple macules, and in more advanced cases they tend to enlarge and form clusters of single nodules. This disease tends to involve the skin. The cutaneous and oral lesions are identical. Other lesions that should be considered during clinical differential diagnosis include pyogenic granuloma, hemangioma, melanoma, and erythroplakia. Classic KS has a rather indolent course and a fair-to-good prognosis.

Several factors have been cited as having a possible etiologic significance for the occurrence of the disease. They include genetic predisposition, viral infection, environmental influences, and immune disturbances. Some data, such as the characteristic epidemiology of the African type of the disease, which has endemic and aggressive features, support the concept that KS may be of viral etiology. Cytomegalovirus has been blamed as a potential causal virus for the disease, but data supporting its role in the pathogenesis of KS are not definitive to date. Another factor that seems to have a definite relationship with the occurrence of KS is immune disturbance. Data supporting its significance in KS development include the epidemic occurrence of KS in AIDS patients and the fact that KS often develops in patients suffering from other conditions associated with immunosuppression. In the present cases, we were not able to determine any etiologic factor since the examination of the patients did not reveal any pathologic findings.

The histopathologic features of KS are identical in the three forms of the disease. The histologic picture in the early lesions is not always diagnostic. The presence of large protruding endothelial cells, extravasated erythrocytes, hemosiderin, and inflammatory cells can raise the suspicion of KS. In late stages of KS, the appearance of a prominent spindle-cell component and mitotic figures aids in the diagnosis of this condition. Frequently both phases are found intermingled in the same lesion. All these histopathologic features were identified in the microscopic observations in the present patients. Entities considered during microscopic differential diagnosis should include hemangioma, pyogenic granuloma, angiopericytoma, and angiosarcoma.

The type of treatment in patients with KS usually depends on the multiplicity of the lesions. Surgery has been useful for localized lesions, while low-dose radiation and chemotherapy are useful for larger and multifocal lesions.
References