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Case Report

Kaposi sarcoma presenting as "diffuse gingival enlargement": Report of three cases

Jacob K. Stern [a], Ilanit Stern [b], Scott S. De Rossi [b], Saiprasad M. Zemse [c,*], Rafik Abdelsayed [d]

[a] Department of Periodontology, Dental College of Georgia (DCG), Augusta University (AU), Augusta, GA, United States
[b] Division of Oral Medicine, Department of Oral health and diagnostic sciences, DGC, AU, Augusta, GA, United States
[c] Chief of Periodontics, Tinker Air Force Base, Oklahoma City, OK, United States
[d] Division of Oral and Maxillofacial Pathology, Department of Oral Health and Diagnostic Sciences, DCG, AU, Augusta, GA, United States

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ABSTRACT

Background: Kaposi sarcoma (KS) is an angio-proliferative disorder, associated with human herpes virus-8 (HHV-8) infection. Its occurrence may be favored by human immunodeficiency virus (HIV) infection and iatrogenic immunosuppression.
Methods: In this article, we report on three cases of oral KS, presented as diffuse gingival enlargement.
Results: All cases were in men; one was HIV-negative, receiving immunosuppressive therapy. The other two patients were subsequently found to have HIV/AIDS. The clinical, histopathological and immunohistochemical features of the lesions are presented and relevant literature is reviewed.
Conclusions: Patients presenting reddish blue macules, plaques or nodular lesions on the skin or oropharyngeal mucosa, should be suspected for having KS whenever an immunosuppressive therapy is involved. If the patient is not treated by immunosuppressive medications, it is important to determine the HIV status, as it is crucial in the overall management of patients diagnosed with KS.
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1. Introduction

Kaposi sarcoma (KS) is an intermediate, non-metastasizing neoplastic proliferation of vascular origin caused by human herpes virus-8 (HHV-8, also known as KSHV). It was originally described by Moritz Kaposi, a Hungarian dermatologist practicing at the University of Vienna, in 1872 under the descriptive term "*idiopathic hemorrhagic sarcoma*" [1]. At that time, it was described as multicentric cutaneous lesions, particularly on the skin of adult men of Mediterranean and Jewish descent. Kaposi description in this group of patients is what is now known as "classic western indolent type", the first described of four general epidemiologic groups. With the advent of the HIV-AIDS era in the early 1980s in the US, KS was prevalent in AIDS patients. It is a defining lesion of the HIV/AIDS disease and is known as "epidemic or AIDS-associated KS". The two other epidemiologic groups are the "African endemic KS" and the "iatrogenic KS". The iatrogenic KS develops in patients receiving immunosuppressive agents; those who suffer from immune-mediated or inflammatory disorders and those who are organ transplant recipients. The viral cause of this neoplastic disease was discovered in 1994, and was found to be a herpes family member, now known as "HHV-8/KSHV" [2].

Clinically, KS is a systemic disease that can present with cutaneous lesions with or without involvement of internal visceral organs and/or lymph nodes. It is elicited by abnormal vascular proliferation, most commonly observed in lower extremities. The erythematous-to-violaceous cutaneous lesions evolve in progressive stages; macular, patch, plaque, nodular and exophytic. Oral involvement is frequently encountered among seropositive HIV individuals, whereas seldom reported in HIV seronegative subjects. In the first group oral KS may be the first manifestation of undiagnosed HIV/AIDS [3]. HIV-KS is most commonly observed in Middle Eastern, African or Caucasian of Mediterranean origin. HIV-KS affects males more commonly than females [4]. Similar to reports of cutaneous disease, oral lesions develop in rapidly progressive clinical stages. The oral lesions are most commonly reported on the palate, followed by gingiva and tongue [5].

In this article, we report on three cases of oral KS, presented similarly, in the form of diffuse maxillary and mandibular dusky-red gingival enlargement, mimicking the common inflammatory

* Corresponding author at: Diplomate-American Board of Periodontology, Chief of Periodontics, Tinker Air Force Base, Oklahoma City, OK, United States.
Tel.: +14057362234.
E-mail address: saizemse@yahoo.com (S.M. Zemse).

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"diffuse gingival hyperplasia". All cases were in men; one was HIV-negative, but immunocompromised male patient with a long history of systemic lupus erythematosus receiving immunosuppressive therapy. The other two patients were subsequently found to have HIV/AIDS. The clinical, histopathological and immunohistochemical features of the lesions are presented, and relevant literature is reviewed.

2. Report of cases

2.1. Case 1

A 54-year-old Caucasian male patient presented to the periodontic clinic at Georgia Regents University, with a chief complain of swollen and bleeding gingiva at his upper and lower jaws. Patient past medical history was significant for long-term lupus erythomatosus existing for at least 20 years. Other than those findings the patient denied any history of smoking or excessive alcohol use. As a result of his chronic illness the patient had developed a renal insufficiency in the form of nephrotic syndrome due to lupus nephritis. The patient was receiving at that time high dosage of prednisone therapy of 100 mg daily while hospitalized for two weeks. Patient reported that gingival lesions were markedly increased in size toward the end of his hospitalization. Because of his marked gingival swellings he was referred to the department of Periodontics at GRU. Physical head and neck exam revealed a bilateral symmetric facial edema with periorbital involvement. No other pigmentary skin lesions were found at the time of examination including a butterfly type lesion. Patient also denied any infectious disease and reported that HIV testing was negative on two previous examinations.

Intra oral examination revealed bilateral maxillary and mandibular massive gingival enlargement with deep purplish discoloration (Fig. 1A). With gentle probing perfused bleeding occurred, which required pressure application and vasoconstrictive agents. Similar macular pigmented lesions were noted on the posterior palate bilaterally (Fig. 1B). The preoperative clinical differential diagnosis included Kaposi sarcoma, leukemic infiltration. Any other forms of gingival enlargement were not considered at this point due to the clinical presentation and medical history [5]. Incisional biopsy was performed under local anesthesia. A 5 mm × 3 mm × 2 mm tissue sample was removed, and area was closed with 3.0 black silk sutures. The tissue sample was submitted in 10% formalin for routine tissue processing and microscopic examination.

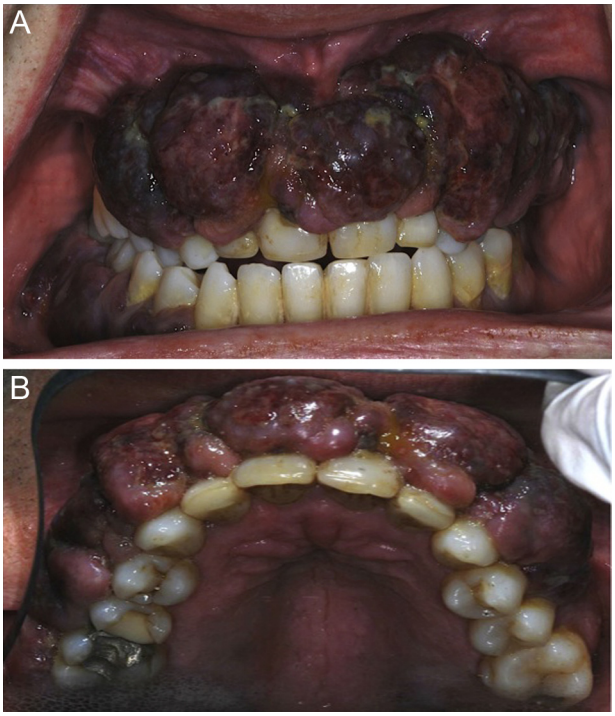

Fig. 1. Case 1 – (A, B) 54-year-old, HIV-negative Caucasian male presented with exuberant gingival purplish masses. Note palatal and mandibular lesions.

Tissue sections examined by light microscopy revealed sections of gingival mucosa showing a markedly cellular neoplastic proliferation covered by intact mucosal epithelium. The neoplastic cells are arranged in intersecting cellular fascicles which replaced the lamina propria in its entirety (Fig. 2A and B). A high-resolution version of this image is available as e-slide: VM00698. Numerous vascular slits packed with erythrocytes were noted (Fig. 2C). The neoplastic cells were spindle-shaped and exhibited palisaded hyperchromatic and pleomorphic nuclei, some of which showed atypical mitotic figures (Fig. 2D). Tissue sections examined with in situ hybridization technique using DNA probe for HHV-8 showed intense reaction (Fig. 3A and B). Based on these features, a final diagnosis of Kaposi sarcoma was made and based on his medical history and a normal "complete blood count" (CBC), it was determined to be of an iatrogenic form. The patient was referred back to his treating physician where a correlation between the development of his Kaposi sarcoma and the high dose of prednisone was made. As a result a clinical decision was made to markedly reduce the prednisone therapy down to 10 mg daily. During a period of 6 month the gingival lesions of Kaposi sarcoma have been resolved without the need for chemotherapy, usually employed for this form of disease. Patient was referred to the clinic of periodontics for follow up, few months later, and the patient had normal gingival health and appearance, with marked reduction of his facial and periorbital edema (Fig. 4).

2.2. Case 2

A 22-year-old African-American male, recently diagnosed with HIV infection presented to the oral diagnosis clinic at the GRU-College of Dental Medicine with a chief complaint of painful and bleeding gingiva in his maxillary and mandibular arches. When the patient was asked, he denied any significant health problems related to HIV. The extraoral head and neck examination revealed symmetric facial swelling, especially around the lips. No significant skin lesions were noted. The intraoral examination revealed remarkable, diffuse gingival enlargement with dark, glistening "rust-colored" appearance involving the maxillary and mandibular attached gingiva and palatal mucosa (Fig. 5). These lesions bled easily when probed. The preoperative clinical impression was Kaposi sarcoma, HIV/AIDS-related. A biopsy was performed under local anesthesia.

The formalin-fixed, hematoxylin and eosin-stained tissue sections revealed a markedly cellular spindle cell proliferation interspersed with "lymphangioma-like" vascular proliferation, hemorrhagic foci and hemosiderin deposits (Fig. 6A). These "lymphangioma-like" vascular elements are composed of endothelial-lined vascular spaces, surrounded by spindle cells arranged in a fascicular pattern and hemorrhagic deposits (Fig. 6B). A prominent feature in this case was the presence of vascular structures exhibiting perivascular lymphoplasmacytic infiltrates (Fig. 6C). Tissue section examined by DNA in situ hybridization for HHV-8 showed positive nuclear reaction with neoplastic spindle cells and with the endothelial cells lining the "lymphangioma-like" vessels (Fig. 6D). The neoplastic cells replaced the entire lamina propria and were present in the form of streaming and intersecting fascicles of spindle-shaped cells with hyperchromatic and pleomorphic nuclei, some of which exhibited atypical mitotic