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Case Report

Oral, conjunctival and Auricular Kaposi's sarcoma in a HIV-negative patient : an unusual presentation

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ABSTRACT

Kaposi's sarcoma is an unusual multicentric malignant neoplastic disease, that characterized by the proliferation of spindle-shaped cells. This tumor is habitually involving in the skin of the lower extremities. However, it is rarely developing in the mucosa or skin of the head and neck. Herein, we report an uncommon presentation of Kaposi sarcoma affecting auricle,oral and ocula rmucosa in an immunocompetent patient. The diagnosis was made on clinical and histopathological finding.

Keywords: Kaposi sarcoma, Oral cavity, Conjunctival, Auricular

INTRODUCTION

In 1872, the first description of Kaposi sarcoma (KS) was published by Moritz Kaposi. His publication described as idiopathic pigmented sarcoma.[1] Indeed, it is an angioproliferative disease of endothelial origin. Four clinical types have been depicted: KS classic (sporadic), African (KS endemic), iatrogenic (immune-suppression associated) and AIDS-associated (KS epidemic).[2] Typically, KS affects the extremities, and infrequently the mucosa or skin of the head and neck. We hereby report a case of immunocompetent patient with KS involving the oral, ocular mucosa and the external ear. The aim of this report is to underline the rarity of these locations and to discuss the different therapeutic modalities.

CASE REPORT

A 77-year-old man was referred to our dermatology department for the evaluation of a multiple violaceous, slow-growing, nodular lesions on the left helix (Figure 1). The patient reported that the skin lesions had evolved over the last nine months and were painless. He denied past medical history or associated immunosuppression.

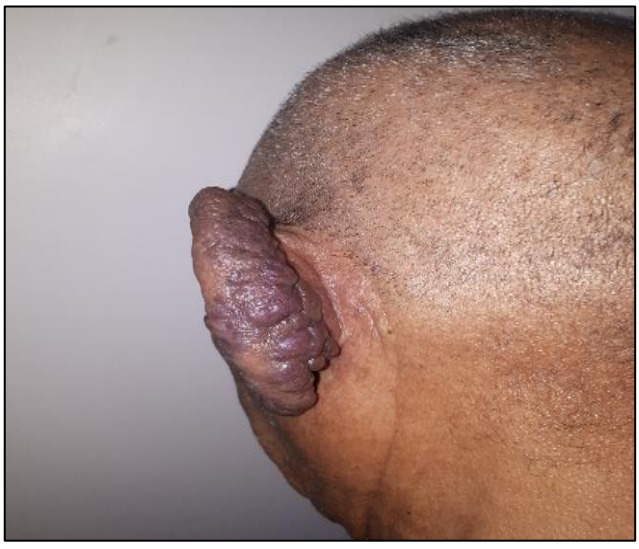

Figure 1: Nodular lesions of the external ear involving the helix.

Upon physical examination, he had also two red macular presents on the hard palate and the conjunctiva of the left eye (Figure 2, 3). The rest of the skin examination was uneventful. There were no lymphatic involvements.

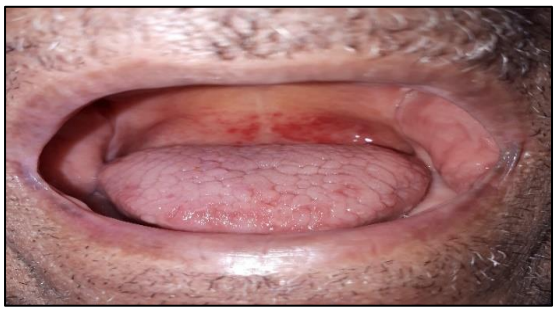

Figure 2: Red macular on the hard palate.

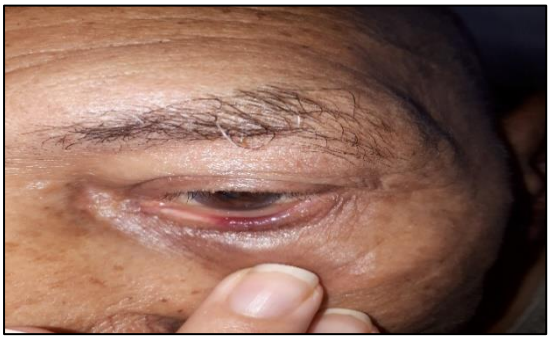

Figure 3: Red macular conjunctival.

Laboratory tests were normal and HIV serology was negative. An abdominal and pelvis computed tomography and gastrointestinal endoscopy revealed no visceral lesions. A skin biopsy of the lesion on the helix was performed and histopathologic examination demonstrated a proliferation of spindle cells and vessels, associated with erythrocytes extravasation (Figure 4). Hence, the diagnosis of classical KS was retained. After discussion of therapeutic modalities, we opted for bleomycin intramuscularly (5 mg) per day for three days repeated every four weeks.

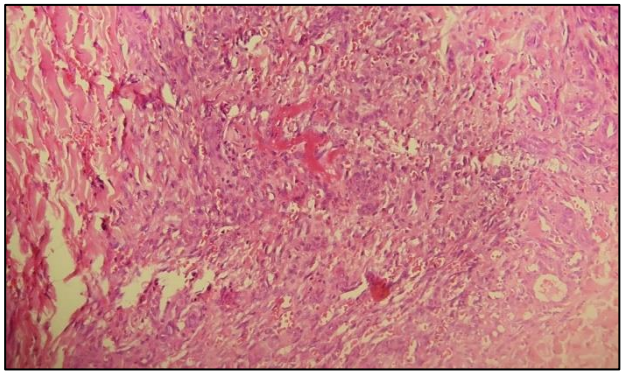

Figure 4: Histological examination revealed proliferating spindle cells and vessels, associated with erythrocytes extravasation.

DISCUSSION

KS is a multicentric vascular tumor with four well-defined clinico pathologic types sharing the same histological features: classic, endemic, iatrogenic and epidemic.[2] The last one is the most frequent and is more aggressive than the other forms. The pathogenesis of this tumor remains unclear. Nevertheless, it has been suggested that several epidemiological, environmental and viral factors (HHV-8) may be involved.[3]

Typically, KS lesions affect the skin of the legs, feet and to a lesser degree the trunk arms, and hands. However, mucosal and cephalic involvement is an unusual manifestation particularly the oral cavity which remains relatively rare[4]. To the best of our knowledge only a few cases of oral KS are reported in dermatologic literature since the first description by Feit in 1928.[5] The oral KS occurs most commonly in the hard palate, gingiva, and dorsal tongue and more rarely in the uvula and oropharynx. While this oral mucosal damage may be seen in all clinical variants of KS, it is especially prominent in epidemic KS and less common in endemic KS.[6] On the other side, auricular and conjunctival localisations of KS are extremely rare and involve the inferior conjunctival fornix and the external ear as seen in our patient.[7] Moreover, they had been mainly described in the epidemic form.

All types of KS share the similar clinical presentation, with different seriousness. Classically, KS manifests as red- bluish macules, papules or nodules, single or multiple that may coalesce into plaques with smooth, keratotic or ulcerated surface. The oral KS presents as a painless, well-limited, purplish-red macule or papule that gradually increases in size to form a nodule or tumour that may interfere with mastication.[8] In addition, the morbidity can be associated with bleeding, pain and suppuration. Although rare, oral KS can also invade the bone and cause tooth mobility.[9]

Histopathologically, KS is characterized by a proliferation of spindle-shaped, epithelioid and endothelial cells with a remarkably greater degree of nuclear and cellular pleomorphism, atypical mitoses and an increased mitotic index.[8]

The clinical differential diagnosis of this mucocutaneous tumor can vary from bacillary angiomatosis, oral nevius, and pyogenic granuloma to vascular lesions.[8] The only possible way to differentiate is a biopsy for histologic evaluation that leads to make a definitive diagnosis.

Management of these tumours is dependent on various factors including the type, extent of the lesions and the organs involved. It seems that in localized forms, therapeutic abstention or local treatment can be proposed namely cryotherapy, intralesional bleomycin/ vinblastine injections, radiotherapy or surgical and laser excision.[8,9] However, systemic monochemotherapy with bleomycin

or vinblastine is recommended in aggressive mucocutaneous or lymphnode forms. On the other hand, in rapidly progressive and visceral forms polychemotherapy is often suggested.[8]

CONCLUSION

In an immunocompetent patient, the oral, auricular and conjunctival KS is an unusual neoplasia, consequently, its diagnosis is rarely evoked in front of a primary lesion in these areas. The only possible method to make the definitive diagnosis is a biopsy.

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