Eyelid Kaposi Sarcoma in HIV-Negative Patient

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Abstract

Introduction: Kaposi sarcoma disease is a proliferative and multifocal disorder with dual components, vascular and fibroblastic cellular, cutaneous and visceral expression. Kaposi Sarcoma can affect the ocular surface and adnexa and can masquerade as other entities, delaying prompt diagnosis can lead to diagnostic wandering delaying treatment. Our aim is to describe a case of KS of the eyelid in an HIV seronegative patient. Case Presentation: A seventy-year-old man developed a bilateral growing tumoral reddish purple vascular mass on both the lower and upper eyelid involving rapidly for 6 months. Both feet and the two shanks show the presence of a brown-violet tumor-shaped formation. The patient was negative for HIV. Histology examination showed a nodular tumor-like mass with a fibro hemangioma-epitheliomatous. Polymerase chain reaction was positive for human herpes virus 8. Initial chemotherapy followed by surgery was proposed to the patient. Unfortunately, the patient rejected treatment and was lost to follow-up. Conclusion: This case reports the difficulty of managing KS in developing countries.

Keywords

Eyelid, HHV-8, Interferon, Kaposi Sarcoma

1. Introduction

Classic Kaposi disease is a proliferative and multifocal disorder with dual components, vascular and fibroblastic cellular, cutaneous, and visceral expression.
Kaposi sarcoma (KS) is an HHV-8-related lympho-angioproliferative with a male-to-female ratio ranging from 2:1 to 5:1. It is evolving in gender males, more than 65 years old, rapidly growing between 3 and 4 months [1]. Kaposi Sarcoma can also affect the ocular surface and adnexa and can masquerade as other entities, leading to diagnostic wandering and delaying treatment [2]. Kaposi Sarcoma can be asymptomatic, but in its progressive phase, it can be very painful and can cause significant aesthetic damage. Many treatments have shown their effectiveness but recurrence is common. Our aim is to describe a case of KS of the eyelid in an HIV seronegative patient.

2. Case Presentation

A Seventy-year-old man developed a bilateral growing mass on both lower and upper eyelid rapidly for 6 months. He was seen in consultation at the Unit of Dermatology; University Hospital Center of Public Health and Care Antananarivo. Gradually, he presented pruritus and swelling of the eyelids and later grew rapidly to purplish nodes. There was no history of trauma, infection, foreign bodies, and no surgery on the eyelids. Dermatological examination showed tumoral reddish purple vascular lesions on both eyes, to the lower and upper eyelids (Figure 1). Best-corrected visual acuity was 20/20 in both eyes. Dermatological examination, in both feet and the two shanks, showed the presence of a brown-violet tumor-shaped formation, with a diameter of 0.5 to 2.0 cm, located on an erythematous-livid base (Figure 2). These lesions are neither painful nor itchy. Oral mucosa was normal. The patient was negative for HIV, hepatitis B and C viruses. Histology examination showed nodular tumor-like mass with a fibrohemangioendotheliomatous structure (Figure 3). Slit-like and irregular vascular spaces, spindle-shaped endothelial cells that do not protrude into the lumen, and hyaline globules; stains positive for human herpes virus 8 with immunoperoxidase staining. Polymerase chain reaction was positive for human herpesvirus 8. Scan images of the brain, chest and abdomen showed no sign of systemic involvement. Initial chemotherapy followed by surgery was proposed to the patient. Indeed, the ocular location, and radiotherapy treatment were not possible. Unfortunately, the patient rejected treatment and was lost to follow-up.

Figure 1. Tumoral reddish purple vascular lesions on the both eyes, to lower and upper eyelids.
3. Discussion

Kaposi Sarcoma was the first to report an idiopathic, multiple-pigmented sarcoma of the skin that predominantly affected elderly men of Mediterranean or Ashkenazi ancestry. Later on, the malignancy was named Kaposi’s sarcoma and can affect the skin, mucous membranes and viscera. Kaposi’s sarcoma is a tumor originating from endothelial cells where there is a suspected infection with human herpesvirus-8 (HHV-8) [2]. It is the most common malignancy among the AIDS patients. Five major subtypes of Kaposi’s sarcoma can be differentiated: classical type of predominantly older Caucasian males; endemic KS of the Sub-Sahara region, which is not HIV-associated; transplantation- and immunosuppression-associated type; AIDS-related type; classical type in HIV-positive patients. This case is about the chronic form. It' is evolving in gender male, more than 65 years old, rapidly growing between 3 and 4 months. Clinical lesions are nodular, reddish purple, vascular lesions. Patient can complain of irritation, pain, swelling, however, his visual acuity in each eye can be correct. The eyelid location is rare, KS can also involve mucosa, lymph nodes or visceral organs. Classic and endemic KS are typically indolent and mainly presents as limb lesions, with less than 10% mucosal, visceral or lymph node involvement [3].
Thirteen percent of KS endemic are located on head or neck [3].

The ocular location must eliminate others’ diagnosis. First, pyogenic granulomas, which are developed in response to underlying chalazion, or often associated with trauma and surgery [4]. There are reports of pyogenic granuloma formation after foreign bodies. Our patient didn’t have any traumatism on his eyes. Secondly, bacillary angiomatosis, which is an infectious disease caused by 2 gram-negative bacilli, *Bartonella henselae* and *Bartonella quintana* [5]. Most commonly, the disease is seen in immunocompromised patients, in particular, in those with HIV with CD4 T-cell counts below 200 per microliter. The biggest difference between KS is its reddish-purple papule about 1 cm in diameter that bleeds easily when traumatized [5]. Chromoblastomycosis, actually, Madagascar represents the most important focus of chromoblastomycosis described to date in the world. It is caused by two species *Cladophialophora carrionii* and *Fonsecaea nubica*, by traumatic inoculation with the fungus through wounding by the plant or soil contamination of an existing wound. The face location is rare 2% of case. It appeared as nodular, warty hyperkeratosis plaques or scabby plaques and lesions may produce eyelid retraction [6]. Basal cell carcinoma accounts for 90% of malignant tumors of the eyelid. Although, basal cell carcinoma of the eyelid is found most frequently in the lower eyelid, more than 20% of the basal cell carcinoma of the head and neck region emerge around the eye, with 50% on the lower lid, 15% on the upper lid. It is characterized by a pearly edge and a pink color [7].

There are different means of treatment of KS, such as chemotherapy (doxorubicin, docetaxel), immunomodulatory, and radiation. The choice of treatment is based on lesion locations, number, and patient’s immune status. In a large cohort of classic/endemic KS, the risk factors for introducing treatment included more than 10 lesions, head and neck localizations, and edema. All these risks were found in our patients. No efficacy difference was observed between standard-of-care treatment, chemotherapy and immunotherapy. The choice of treatment depends mainly on the patient’s condition, but above all, in a developing country like Madagascar, on the patient’s financial possibilities [3].

On the conjunctiva or eyelid KS, the aim of the treatment is to restore eyelid anatomy and preserve the vision of the patient. Treatment is indicated in presence of discomfort and loss of eyelid function with compromised aesthetic and corneal damage complications [8]. Patients presenting with symptomatic superficial or isolated skin lesions are treated locally, while more extensive, disseminated, like our patient, or with visceral locations are treated systemically. In our case, we have chosen chemotherapy because of large and several lesions on the eyelid, with KS on both upper and lower limbs. Unfortunately, our proposal treatment was rejected by the patient and he was lost to follow-up. Systemic treatment protocols are based on interferon or chemotherapy. The overall response rate is very positive reaching up to 80%, despite this, the curative is not totally and recurrence is frequent representing 66% of cases [8] [9].
Regarding the literature, many local treatments have been successfully tried. The excision alone is indicated on stage I and stage II of Dugel’s classification to avoid recurrence. Cryotherapy alone used at −80˚C with three applications once a week for 4 weeks until regression, was successful and had no recurrence. Radiation Therapy, the literature concluded that for conjunctiva KS, a single treatment of 800 cGy is as effective as multiple fraction regimens. No serious complications were reported, but minor reactions occurred in 21% of patients, but 29% of recurrences occurred.

Intralesional injection of chemotherapy, injection of Mitomycin C (MMC) was successfully proven with 2 concurrent injections of 0.15 mL of 0.2% MMC in the eyelids, no clinical signs of recurrence were noted, and no adverse reactions to treatment were reported [8]. Occlusive treatment with imiquimod 5% cream also showed therapeutic success at a dose of once application per day for three months. Rapid remission occurred, no local or systemic side effects of imiquimod, and no recurrences [10].

4. Conclusion
In this study, we report a clinical case of endemic eyelid KS with limb involvement. Despite the presence of several successful treatments, the management of this disease is still difficult for low-income countries.

Conflicts of Interest
The authors declare no conflicts of interest regarding the publication of this paper.

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