

Atypical Presentation of Conjunctival Kaposi Sarcoma: A Case Report

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Background: Kaposi sarcoma is an antiproliferative spindle-cell tumor derived from vascular endothelium and immune cells infected with human herpes virus type 8 (HHV-8). It is one of the most common ocular malignancies in patients with human immunodeficiency virus (HIV) infection but is also found in immunocompetent patients.

Case presentation: A 55-year-old male patient with a 1-month history of painless maroon-colored nodular mass, surrounded by telangiectatic vessels at the lower palpebral conjunctiva. His serologic test was positive for HIV infection. After performing an excisional biopsy, the histopathological report confirmed the diagnosis of a nodular stage of Kaposi sarcoma.

Conclusion: Although ocular Kaposi sarcoma is extremely rare, especially when manifest as the initial manifestation of occult HIV infection. Recognizing the correct diagnosis is valuable. The advantage of evaluating for HIV infection is early identification and comprehensive highly active antiretroviral therapy (HAART) to prevent further morbidity and mortality sequelae.

Keywords: Kaposi sarcoma, Acquired immunodeficiency syndrome (AIDS), Human immunodeficiency virus (HIV), Conjunctival neoplasms

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Introduction

Ocular manifestations of acquired immunodeficiency syndrome (AIDS) can be caused by opportunistic infections, vascular abnormalities, neoplasms, neuro-ophthalmic conditions, or adverse effects of medications.¹⁻³ In 2020, the World Health Organization (WHO) estimated that almost 4 million people worldwide have been infected with HIV, and the global prevalence is about 0.51%.⁴

Kaposi sarcoma was first described as an unusual multiple idiopathic vascular endothelial malignant tumor by Dr. Moricz Kaposi in 1872.⁵ A century later, it was discovered this tumor was associated with human immunodeficiency virus (HIV), acquired immunodeficiency syndrome (AIDS) and human herpesvirus 8 (HHV-8). The occurrence of Kaposi Sarcoma may indicate worsening of the underlying disorder appearing in patients with CD4 count <500 cells/ μ .⁶ Ocular involvement occurs in 20-24% of patients with AIDS-related Kaposi sarcoma, which commonly affects the conjunctiva, fornix caruncles, lacrimal ducts, and very rarely in the lacrimal gland and orbit.⁷ However, ocular Kaposi sarcoma as an initial presentation of HIV is very rare. To our knowledge, there have been reported only 6 prior cases of conjunctival Kaposi sarcoma as the first manifestation of AIDS.⁸⁻⁹

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Disseminated lesions or positive metastasis can lead to significant morbidity and mortality.¹⁰⁻¹¹ Kaposi sarcoma usually responds well to surgical resection in combination with antiretroviral therapy.¹²

Case report

A 25-year-old lady was referred to our centre. A 55-year-old male patient presented for 1-month history of a purplish growing mass at the left lower palpebral conjunctiva. He complained about mild irritation and minimal eye redness without pain, photophobia, or any visual disturbance. The patient had previously been evaluated by a general doctor for these symptoms. He was treated as having conjunctivitis and prescribed some antibiotic eye drops and artificial tears for a week but failed to respond to the medication. He eventually had blindness in the right eye due to retinal detachment for 10 years without prior ocular surgery. Other medical history was unremarkable. He denied having any underlying medical condition and no history of drug allergy.

On ocular examination, the best corrected visual acuity in the left eye was

20/30. While no light perception and the relative afferent pupillary defect was positive in the right eye. Bilateral intraocular pressures were 10 mmHg. The slit lamp biomicroscopy revealed a single maroon-colored nodular mass size 10x12 mm at a medial third of the left lower palpebral conjunctiva with surrounding telangiectatic vessels, superficial ulceration and subconjunctival petechial hemorrhage. The nodule did not involve the eyelid margin, fornix or lacrimal punctal. The mass effect caused a mechanical ectropion (Figure 1). The other findings were mild ciliary conjunctival injection, prominent punctate epithelial erosion at the inferior corneal area, formed anterior chamber without inflammatory cells and clear lens. The posterior segment was unremarkable (flat retina, no abnormal vascular structure, no infiltration or area of retinitis). The right eye showed a normal anterior segment with a total retinal detached fundus. In general inspection, there was negative for any violaceous skin lesion, hyperpigmentation mass in the oral cavity or lymphadenopathy. After obtaining the patient's counseling and consent, the patient was screened and confirmed with a positive HIV serology test.



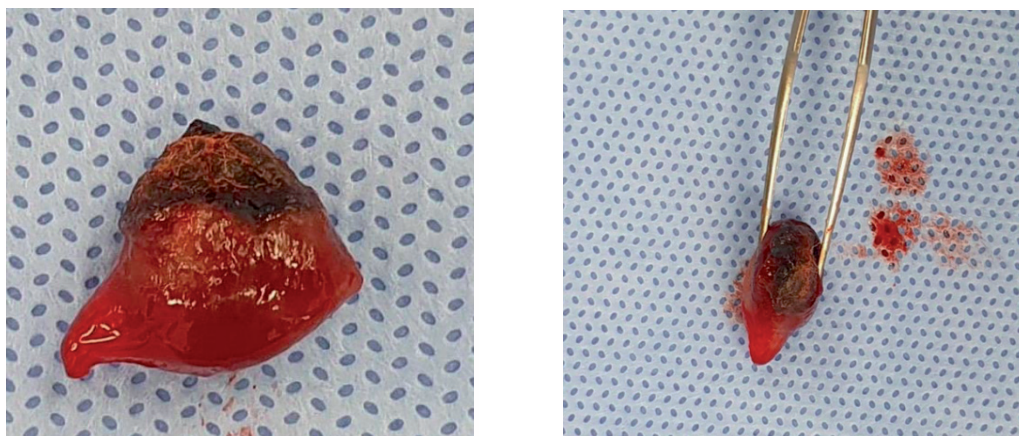
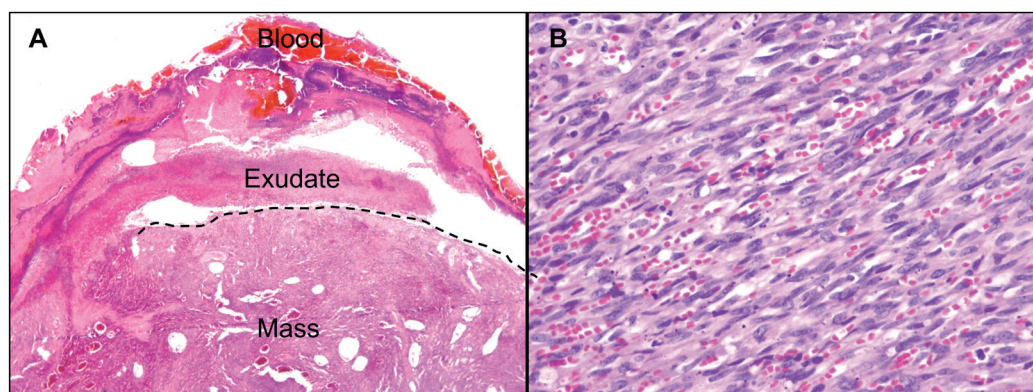


Figure 1: Left lower conjunctival mass. As single maroon-colored nodule size 10x12 mm at the inner third of left lower palpebral conjunctiva with surrounding telangiectatic vessels and subconjunctival petechial hemorrhage without the eyelid margin, fornix or lacrimal punctal involvement. Superficial ulceration is observed.

A 2-mm marginal free excisional conjunctival biopsy and cryocoagulation was performed one week later. The histopathological report, which confirmed the diagnosis of Kaposi sarcoma, described that the mass was composed of a bland spindle cell proliferation with slit-like spaces containing erythrocytes and some extravasated RBCs were observed. The mass also has superficial ulceration topped by exudate and

blood (Figures 2). Immunohistochemistry (IHC) for HHV8 revealed diffuse immunoreactivity in a spindle cell nuclei confirming the diagnosis of nodular stage Kaposi sarcoma (Figure 3A). IHC for CD31 also showed diffuse immunoreactivity in spindle cells cytoplasm supporting a vascular tumor in origin (Figure 3B). The definite diagnosis was Kaposi sarcoma subtype 3 localized at the lower palpebral conjunctiva left eye.



Figures 2: Histopathology of the left lower conjunctival mass. A. Low magnification shows the mass with a superficial ulcer (dashed line) topped by exudate and blood (H&E, 40x). B. High magnification reveals proliferation of bland spindle cells with slit-like spaces containing erythrocytes and some extravasated erythrocytes are observed. (H&E, 400x).

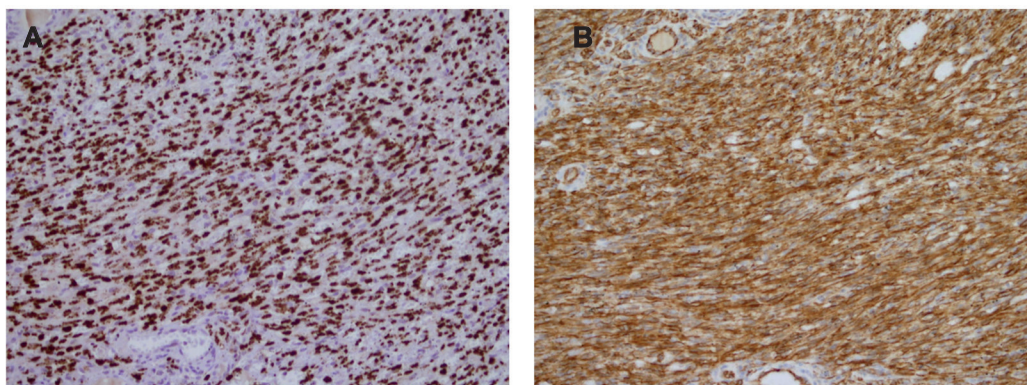


Figure 3: IHC of the left lower conjunctival mass A. IHC for HHV8 shows diffuse immunoreactivity in spindle cells nuclei (HHV8, 100x). B. IHC for CD31 shows diffuse immunoreactivity in spindle cells cytoplasm (CD31, 100x)

After completing surgical removal without post-operative complications, the patient was started on the standard antiretroviral therapy for HIV treatment. At the last visit 3 months after the presentation, the clinical remained stable and there was no sign of tumor recurrence.

Discussion

Kaposi sarcoma is commonly encountered in immunocompromised patients including HIV/ AIDS, under immunosuppression or post-organ transplantation but can also present in immunocompetent patients.¹³ The 4 variant forms of Kaposi sarcoma include classic (sporadic), African (endemic), AIDS-associated, and iatrogenic (or transplant-associated) Kaposi sarcoma.¹⁴ Kaposi sarcoma may affect many ocular structures. Ocular tumor growth can result in severe damage to the ocular adnexa, ocular surface and the orbit.¹⁵

The histopathological staging of Kaposi sarcoma consists of 3 stages. Stage I, thin and enlarged blood vessels lined with flat endothelial cells without mitotic appearance can be identified in the absence of spindle or fissure cells. Stage II shows enlarged and empty blood vessels coated with fusiform endothelial cells with immature spindle cells. Extensive aggregation of dense spindle cells with hyperchromatic nuclei and mitotic features is common in stage III. Clinically, stages I and II are flatter with a size of less than 3mm, while stage III is more nodular

and larger than 3mm.^{12,16-18} Further causes may need to be investigated if conjunctivitis and subconjunctival hemorrhage do not improve after two weeks of treatment. Differential diagnoses of similar lesions should include arteriovenous shunt, pyogenic granuloma, malignant blue nevus, bacterial angiomatosis and vascular malformations.¹⁹ To confirm the diagnosis through histopathologic and immunohistochemical tests, an excisional biopsy of these lesions is required.

Currently, conjunctival and adnexal Kaposi sarcoma are treated with a combination of HAART and surgical excision, cryotherapy, radiation therapy, or intralesional injections of chemotherapy such as interferon and mitomycin, depending on the size and location of the tumor. In many cases, treatment of isolated ocular Kaposi sarcoma is associated with high response rates and frequently, complete remission. Even without surgery, radiation, or chemotherapy, response rates of local Kaposi sarcoma to initiation of HAART alone range from 66 to 86% with complete remission in 35% of patients.^{12,19-23}

Although ocular Kaposi sarcoma is extremely rare, especially as the initial manifestation of occult HIV infection. Recognizing the correct diagnosis is valuable. The advantages of evaluating for HIV infection has benefits of early identification and comprehensive highly active antiretroviral therapy (HAART) to prevent further morbidity and mortality sequelae.

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