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**Case Report**

# Isolated Kaposi's Sarcoma of the Bulbar Conjunctiva as an Initial Manifestation of AIDS: A Case Report

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## Keywords

Kaposi's sarcoma · Bulbar conjunctiva · Acquired immunodeficiency virus · Human immunodeficiency virus

## Abstract

Kaposi's sarcoma (KS) is a malignant vascular endothelium-cell-derived tumor caused by human herpesvirus 8. It is one of the most common tumors among human immunodeficiency virus (HIV)-infected patients; however, isolated KS is rarely reported as the initial presentation. This study describes a rare case in which isolated KS of the bulbar conjunctiva was the first presenting symptom leading to the diagnosis of HIV/acquired immunodeficiency syndrome (AIDS) in a 39-year-old man. The patient, who had no prior medical history, presented to the ophthalmology clinic with an isolated large, dark-reddish mass in the left bulbar conjunctiva and subconjunctival hemorrhage. The mass was first identified 6 months prior and had continued to grow since then. KS was confirmed based on the analysis of the incisional biopsy sample, subsequently prompting an HIV test, which was positive. This report highlights the recognition of KS as a relevant ocular complication and potential initial manifestation of AIDS. Additionally, KS should be considered in the differential diagnosis of any vascular lesion, even when present at uncommon sites.



## Introduction

Kaposi's sarcoma (KS) is a subtype of angioproliferative neoplasm that was first described by Moritz Kaposi in 1872 [1]. It is the most commonly diagnosed malignant vascular neoplasm among human immunodeficiency virus (HIV)-infected patients and is believed to be

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caused by human herpesvirus 8 (HHV-8) [2]. KS can affect the vascular endothelium and presents most commonly at mucocutaneous sites, including the lower legs, oral cavity, and genitals [2]. In ocular tissues, KS may manifest in several areas, such as the orbit, lacrimal gland or sac, eyelids, caruncle, and bulbar or palpebral conjunctiva [3]. However, ocular KS as an initial presentation of acquired immunodeficiency syndrome (AIDS) or HIV infection is extremely rare and has been documented in only a few cases [4–11]. Furthermore, the conjunctiva is considered an uncommon site for the initial presentation of this tumor in patients with HIV/AIDS [12]. In the conjunctiva, KS appears as a dark-reddish, raised fleshy mass that can be present similar to subconjunctival hemorrhage [3].

Histopathologically, KS appears as a mass of multiple spindle-shaped cells with ovoid nuclei arranged in bundles interspersed with multiple vascular "slit-like" channels [2]. A definitive diagnosis of KS is made based on diagnostic histopathology following biopsy and molecular testing for HHV-8 [2]. This report describes a case of isolated conjunctival KS that led to the diagnosis of HIV/AIDS in a previously healthy patient.

## Case Presentation

A 39-year-old single Saudi Arabian man with no prior medical history presented to our emergency department complaining of a reddish mass in his left eye that had been slowly growing over the past 6 months. He had no other ocular symptoms, such as pain, photophobia, or visual changes, but did report experiencing a weight loss of 12 kg in 9 months preceding the assessment date. The patient's medical and ocular history was unremarkable, with no chronic medication use and no previous history of trauma. Regarding social history, the patient did not report engaging in sexual activity, illegal drug use, or having a prior blood transfusion.

The patient's height and weight at admission were 169 cm and 88 kg, respectively. Physical examination of the patient confirmed that there were no lesions in the oral cavity, mucocutaneous changes, or lymphadenopathy. Ophthalmologically, the patient had full extraocular range of motion, the best-corrected visual acuity was 20/20 in both eyes, pupil examination was normal, and intraocular pressure was 14 mm Hg and 19 mm Hg in the right and left eyes, respectively. On slit-lamp examination, a large, diffuse, elevated, dark-reddish fleshy mass was visualized over the left bulbar conjunctiva, along with areas of subconjunctival hemorrhage sparing the superior bulbar and palpebral conjunctiva (shown in Fig. 1). The lesion was loosely connected to the underlying tissue. Examination of the right eye and fundoscopic examination of both eyes were normal.

Owing to the large size of the mass, an incisional biopsy was first performed as both a diagnostic and therapeutic (debulking) procedure. A 5 mm × 2 mm mass was excised from the inferior bulbar conjunctival lesion, and bleeding vessels were obstructed by cauterization. Given that the tumor site was approaching the inferior fornix, an amniotic membrane graft was used to close the area of excision to decrease the risk of scarring, fibrosis, and symblepharon formation. Subsequently, a bandage contact lens was applied. There were no intra- or postoperative complications.

Histopathological examination of the biopsied tissue revealed proliferation of spindle-shaped endothelial cells, slit-like formations, and numerous extravasated red blood cells and inflammatory cells, consistent with KS (shown in Fig. 2). Immunohistochemical staining (shown in Fig. 3) strongly supported the diagnosis of KS.

Serologic and immunologic tests upon admission confirmed HIV infection, with a viral load of 186,876 copies/mL, and the patient's CD4+ T lymphocyte count was 169 cells/mm$^3$. Oncology and infectious disease consultation data were also obtained. There was no evidence of nodular or distal metastases on whole-body positron emission tomography imaging.

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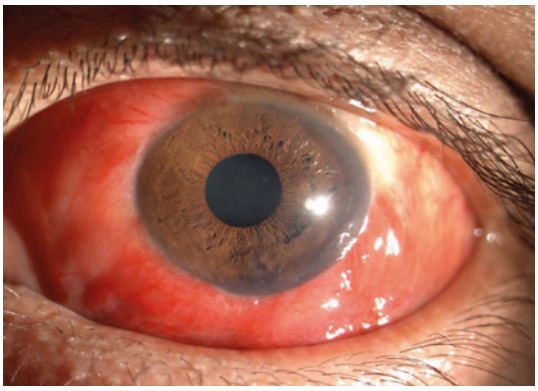

**Fig. 1.** Photograph of the left eye upon admission, showing a large, diffuse, fleshy hemorrhagic mass in the left bulbar conjunctiva.

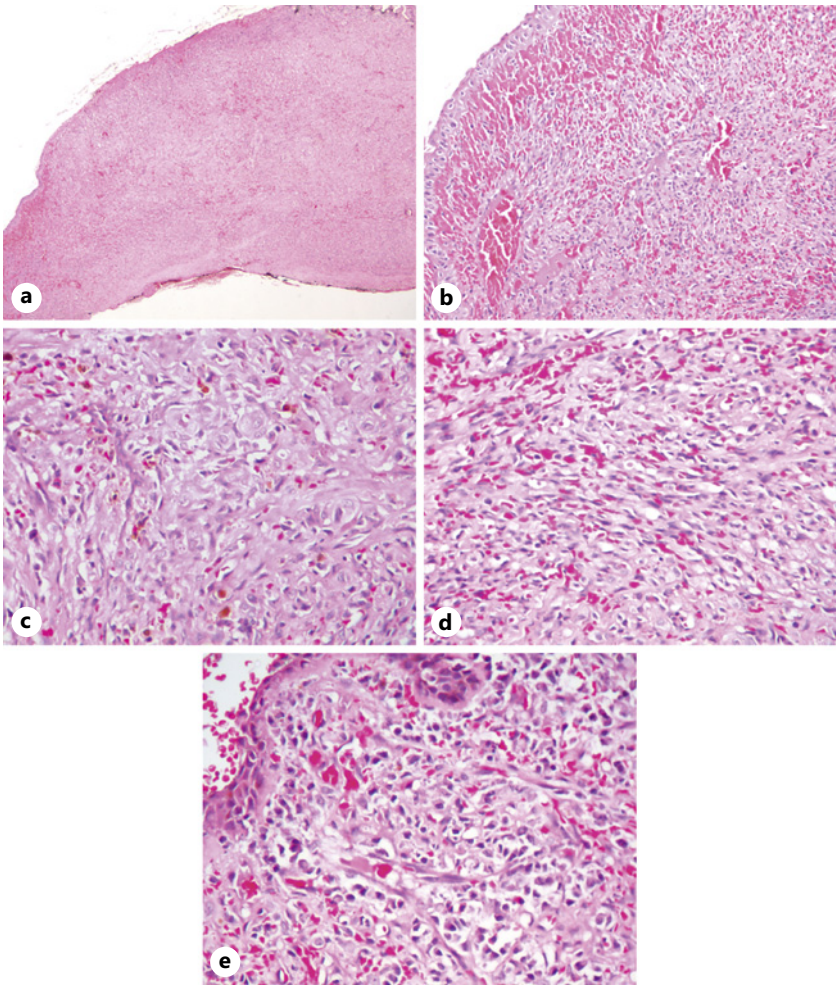


**Fig. 2.** Hematoxylin-eosin-stained sections of the KS biopsy tissue. **a** Low-magnification microscopic image showing a spindle cell variant of conjunctival squamous epithelium with dispersed red blood cells (original magnification, ×40). **b**, **c** Higher magnification images showing mildly atypical spindle cells forming slit-like, blood-filled channels and red blood cells extravasating into the surrounding stroma (original magnification, ×100 and ×200, respectively). **d** Section with plump spindle cells and extravasated red blood cells, a few inflammatory cells, and hemosiderin deposition (original magnification, ×400). **e** Spindle cell proliferation with intervening vascular spaces and prominent lymphoplasmacytic infiltration. Stratified squamous epithelium of the conjunctiva is seen in the left upper corner of the image (original magnification, ×400).

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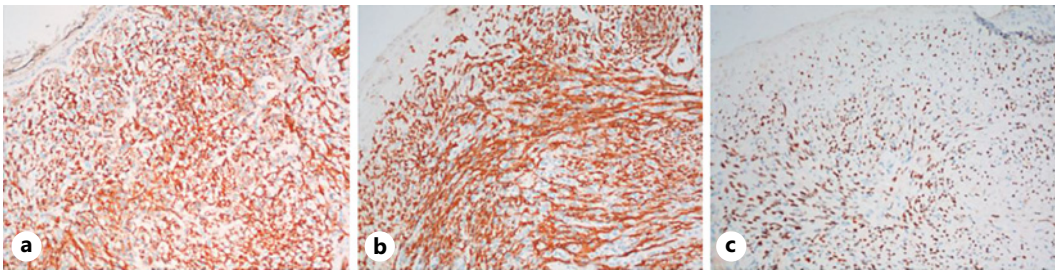


**Fig. 3.** Immunohistochemical staining of the KS biopsy sections. **a** Positive CD31 immunoreactivity. **b** Positive CD34 immunoreactivity. CD31 and CD34 immunoreactivity confirmed the endothelial cell origins of the KS. **c** Positive HHV-8 immunostaining distinguishing the KS from other vascular tumors.

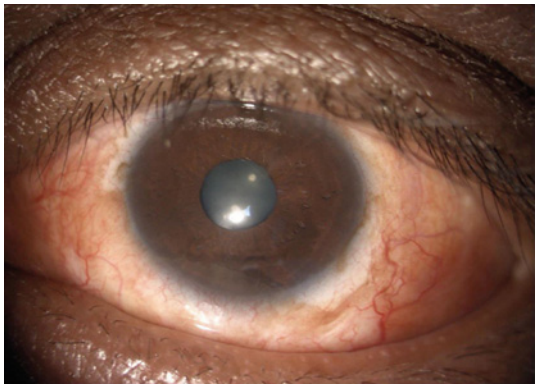

**Fig. 4.** Photograph of the left eye showing total resolution of KS following HAART and local radiotherapy.

Subsequently, owing to the local presentation and significant lesion extension noted in this case, highly active antiretroviral therapy (HAART) was initiated along with local irradiation of the tumor. The patient received radiotherapy at a dose of 20 Gy over 10 fractions. All fractions were delivered within 10 days. The lesion resolved completely in response to the local irradiation, and only mild redness, without any fibrosis or ocular range of motion restrictions, was observed at the 8-month follow-up (shown in Fig. 4). Multidisciplinary follow-ups were scheduled for the patient, who has had favorable responses with no relapses till the time of the last follow-up.

## Discussion and Conclusion

HIV/AIDS can affect various ocular structures, including the ocular adnexa (e.g., molluscum contagiosum and herpes zoster ophthalmicus), anterior segment (e.g., infectious keratitis and iridocyclitis), and posterior segment (e.g., retinal microangiopathy and other opportunistic infections), resulting in serious ocular complications. Of these, retinal microangiopathy in the posterior segment is the most common ocular manifestation of HIV infection. Manifestations may present at the early or late stage of HIV/AIDS, depending on the CD4+ T lymphocyte count. However, they rarely appear in the early, asymptomatic stage of HIV infection [13].

KS is an AIDS-defining lesion, with a crude incidence of approximately 481.5 cases per 100,000 person-years in patients infected with HIV. The occurrence of KS has significantly declined with the advent of HAART. Notably, KS can also affect HIV-negative individuals, with

an incidence of 1.53 cases per 100,000 person-years in the general population [14]. Although KS is usually associated with HHV-8 infection, immune system dysregulation, and specific inflammatory cytokines (e.g., high interleukin-6 levels), its pathophysiology remains unclear [2, 12].

In most cases, KS lesions initially develop at cutaneous sites, typically the skin of the lower extremities, trunk, face, or genitals. KS also commonly is present in the mucous membranes lining the oral cavity and the visceral organs [2]. Although ocular presentations are noted in 20% of patients with AIDS [3], the bulbar conjunctiva is considered an unusual initial site for KS [12]. There have been only a few reported cases of conjunctival KS as the first presenting sign that led to the diagnosis of HIV in previously healthy individuals [4–11].

Clinical diagnosis alone can also be challenging because the appearance of KS resembles that of various types of ocular lesions, including pyogenic granuloma, subconjunctival hemorrhage, cavernous hemangioma, and lymphoma. Hence, tissue biopsy is crucial for confirming the diagnosis of any suspicious conjunctival lesion [3]. Early diagnosis of KS can improve clinical outcomes, increase survival rate, and reduce the need for further adjuvant therapy. In the USA, the median survival rate of patients diagnosed with early-stage KS (T0 confined tumor) is 1 year higher than that of patients diagnosed with advanced KS (T1 extensive tumor). In Italy, among 211 patients who presented with AIDS-related KS, those with stage T1 tumor at the time of diagnosis had a 2.6-fold higher rate of death than those with early-stage T0 tumor in the HAART era [2].

Ocular KS is generally responsive to treatment, and available treatment modalities have proven efficacious [3]. However, there is a lack of standard therapeutic protocols – treatments can differ based on the size, location, and extension of the lesion, and the patient's immune system status and other AIDS-related complications can also play a role [15]. In patients presenting with localized ocular KS, the typical regimen involves initiating HAART alone or in combination with surgical excision/debulking of the tumor. Adjuvant radiation or intralesional chemotherapy, such as interferon and mitomycin treatment, can also be used, especially in refractory cases and for cases with large lesions [3]. Although several therapeutic options are available, a remarkable response with complete remission can be achieved with HAART alone in some cases [6, 15].

Owing to the socioreligious factors in Saudi Arabia, obtaining an accurate sexual history from a patient can be challenging; this also represents a limitation of this study. Accordingly, KS should be considered as a differential diagnosis for any hemorrhagic conjunctival mass, even when an unremarkable sexual history is reported. Polymerase chain reaction assays for HHV-8 detection can support the diagnosis of KS, particularly in equivocal cases [2]. However, this requires specialized molecular diagnostic facilities currently unavailable where we practice. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000532106).

In conclusion, early diagnosis of HIV/AIDS is crucial for early initiation of HAART to prevent the devastating complications of KS. This case report proves the importance of recognizing the ophthalmic manifestations of AIDS. It highlights the relevance of considering KS for the differential diagnosis of any vascular lesion, even at unusual sites. Further studies are also needed to investigate the conjunctiva as an important probable location for the initial presentation of KS.

## Statement of Ethics

This case report was conducted in accordance with the ethical standards of the institution and the Declaration of Helsinki. Ethical approval was not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

## Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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## Author Contributions

Hend Alsawadi: conception and acquisition, analysis, and interpretation of data. Abdulaziz Al-Somali: drafting and revising. Hatim Najmi: data collection, analysis, and drafting. Fatimah Al-Mubarak: substantial contributions to conception, acquisition, analysis, and interpretation of data. All authors read and approved the final version of this manuscript.

## Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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