

Extensive bulbar conjunctival Kaposi's sarcoma as initial symptom of human immunodeficiency virus

Sarcoma de Kaposi extenso na conjuntiva bulbar como apresentação inicial do vírus da imunodeficiência humana

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ABSTRACT | Here, we present a case in which extensive bulbar conjunctival Kaposi's sarcoma was the initial presentation of human immunodeficiency virus in a 36-year-old man. The patient had a 3-month history of recurrent self-limited inferior conjunctiva hyperemia in the right eye, and presented with a painless bullous lesion in the right inferior bulbar conjunctiva persisting for 15 days. Surgical incision biopsy was performed at five locations and revealed a pattern compatible with Kaposi's sarcoma. Serologic testing was positive for human immunodeficiency virus; however, the patient had no other symptoms, or knowledge of human immunodeficiency virus infection. This case highlights the need to consider Kaposi's sarcoma as an early presentation of human immunodeficiency virus even if the patient denies infection.

Keywords: Sarcoma, Kaposi; Acquired immunodeficiency syndrome; HIV; Conjunctival neoplasms; Human; Case reports

RESUMO | Este relato de caso apresenta um sarcoma de Kaposi extenso na conjuntiva bulbar como a apresentação inicial do vírus da imunodeficiência humana em um homem de 36 anos de idade. O paciente tinha história de hiperemia na conjuntiva inferior do olho direito há 3 meses, autolimitada e recorrente e de surgimento de uma lesão bolhosa indolor no mesmo local 15 dias antes da sua apresentação. Uma biópsia incisional cirúrgica foi realizada e revelou um padrão compatível com sarcoma de Kaposi. Teste sorológico posterior revelou positividade para o vírus da imunodeficiência humana, no entanto, o paciente não

apresentou outros sintomas, sinais ou conhecimento prévio sobre a infecção. Como conclusão deste caso, deve se ressaltar que a suspeita do diagnóstico do sarcoma de Kaposi deve ser levantada ainda que na apresentação inicial do vírus da imunodeficiência humana ou mesmo naqueles ainda sem este diagnóstico.

Descritores: Sarcoma de Kaposi; Síndrome de imunodeficiência adquirida; HIV; Neoplasia conjuntival; Humanos; Relatos de casos

INTRODUCTION

Various studies have revealed the association between acquired immunodeficiency syndrome (AIDS) and an aggressive form of Kaposi's sarcoma⁽¹⁾. For example, Macher et al. reported a 38-year-old homosexual male with AIDS with Kaposi's sarcoma of the palpebral conjunctiva, extraocular mucocutaneous areas, and lymph nodes⁽²⁾. Patients with AIDS who initially present with opportunistic infections commonly develop Kaposi's sarcoma at follow-up; conversely, patients presenting with Kaposi's sarcoma may subsequently develop opportunistic infections.

Kaposi's sarcoma is a lesion with multifocal areas that presents as vascular tumors of the skin, oral mucosa, and internal organs; localized violaceous lesions on the lower extremities may also be present⁽²⁾. Histopathology typically shows atypical spindle cells in a fascicular arrangement, interspersed with multiple vascular slit-like channels⁽³⁾. Lesions are more common in AIDS patients, but do also occur in other immunosuppression diseases⁽³⁾. Differential diagnosis includes hemangiopericytoma, capillary hemangioma, pyogenic granuloma, cavernous hemangioma, melanoma, and metastatic tumor. Kaposi's sarcoma usually responds well to surgical resection in combination with antiretroviral therapy⁽⁴⁾.

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Here, we present a case of a young male patient without an AIDS diagnosis who presented with conjunctival Kaposi's sarcoma confirmed by biopsy. This was the patient's first manifestation of human immunodeficiency virus (HIV).

CASE REPORT

A 36-year-old man presented with a painless bullous lesion in the right inferior bulbar conjunctiva that had persisted for 15 days. He reported a three-month history of recurrent self-limited inferior conjunctiva hyperemia in the right eye.

The patient had an unremarkable medical history, including ocular history, and no chronic medication use. He denied any family history of systemic or ocular hereditary diseases.

Physical examination showed no lymphadenopathy, and no other skin or hard palate changes were detected. At ophthalmologic examination, the patient presented best-corrected visual acuity of 20/30 in the right eye and 20/20 in the left eye; ocular motility was normal, pupillary reflections were preserved, and intraocular pressure was 14 mmHg for both eyes. The slit-lamp examination revealed a red, bullous conjunctival mass measuring approximately 24 mm x 10 mm originating from the inferior fornix (Figure 1). Fundoscopic examination was normal in both eyes.

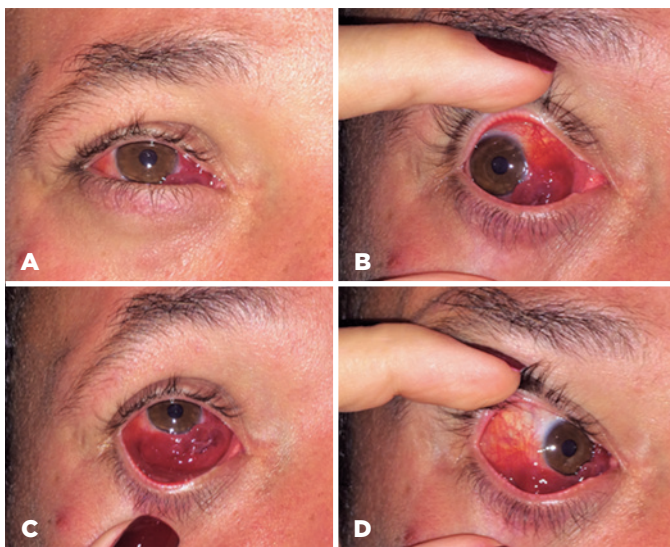


Figure 1. Ectoscopy of the patient's right eye at presentation. (A) Ectoscopy in supraduction. (B) Ectoscopy with retraction of the eyelids in abduction. (C) Ectoscopy with retraction of the eyelids in supraduction. (D) Ectoscopy with retraction of the eyelids in adduction.

Surgical incision biopsy was performed at five locations (the four cardinal points of the lesion and the central area) of the conjunctival lesion followed by hemostasis with bipolar cautery. There were no complications after the biopsy.

Anatomopathological exam revealed that the lesion consisted of conjunctival and subconjunctival chorion (Figure 2A), which comprised a proliferation of endothelial cells with mostly spindle nuclei (Figure 2B) arranged in fascicles with red blood cells in the lumen (Figure 2C). Some lymphocytes permeated the neoplasia

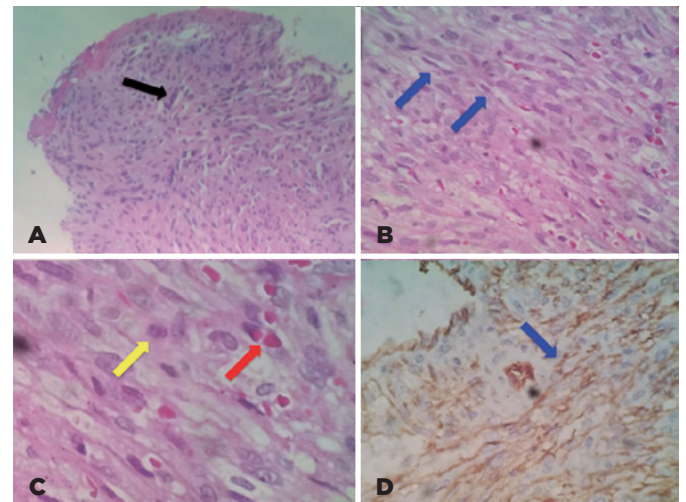


Figure 2. Pathology analysis. (A) Conjunctival epithelium largely consumed by neoplasm (Kaposi's sarcoma); (hematoxylin and eosin (H&E) staining, x40); Black arrow: vascular lumen limited by atypical endothelial cells (hyperchromatic and irregularly shaped nuclei). (B) Blue arrow: Proliferation of atypical spindle-like endothelial cells limiting vascular lumen (H&E staining, x200). (C) Red arrow: Detail revealing neovessels with red blood cells in the lumen; Yellow arrow: rare lymphocytes (H&E staining, x400). (D) Immunohistochemistry with HHV8 antibody revealing multifocal immunopositivity; Blue arrow: spindle-like cells showing HHV8 antibody positivity.

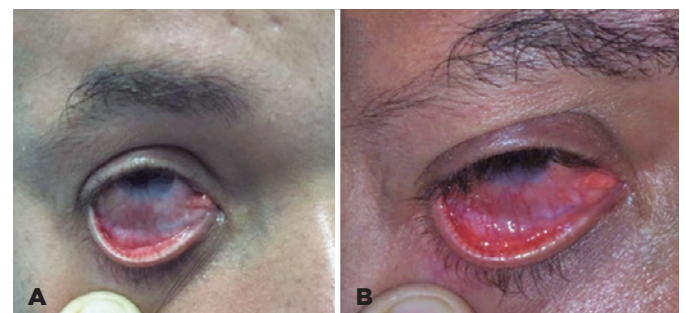


Figure 3. Ectoscopy of the patient's right eye (OD) with inferior bulbar conjunctiva exposure after incisional biopsy at the 12-months postoperative visit. (A) Ectoscopy showing no restriction to OD elevation. (B) Close-up ectoscopy image.

(Figure 2C). In immunohistochemical testing, the lesion exhibited diffuse immunopositivity with anti-CD20 and anti-CD34 antibodies as well as multifocal immunopositivity with the anti-HHV8 antibody (Figure 2D).

All the biopsied areas had the same anatomopathological pattern, compatible with Kaposi's sarcoma. Then, serologic and immunological status were requested, which resulted in HIV positivity and low CD4 and CD8 rates. Antiretroviral therapy was instituted. The patient presented on the 15th postoperative day and was then lost to further ophthalmology follow-up. At nine-month follow-up with an infectious disease specialist, the patient presented with resolution of the conjunctival Kaposi's sarcoma, with local fibrosis, but without ocular motility restrictions (Figure 3). At one-year follow-up, the patient had no recurrence of the Kaposi's sarcoma in the conjunctiva or elsewhere and was receiving regular anti-retroviral therapy.

DISCUSSION

The present case describes an extensive bulbar conjunctival Kaposi's sarcoma as the initial presentation of HIV in a 36-year-old man. Reports of Kaposi's sarcoma in patients without AIDS are very sparse in the literature^(1,4,5).

Kaposi's sarcoma was first described in 1872 as an unusual multifocal idiopathic hemorrhagic sarcoma. Ocular manifestation of Kaposi's sarcoma may present in the lid, lacrimal sac, or conjunctiva^(6,7). This tumor type has been reported in additional areas, including cutaneous sites, mucous membranes, lymph nodes, and viscera. Disseminated lesions can lead to significant morbidity and mortality. The disease typically presents in AIDS patients, usually those who are young and male, with other opportunistic infections such as *Pneumocystis carinii* pneumonia, disseminated *Mycobacterium avium-intracellulare*, or *Cryptococcus neoformans* meningitis. As stated earlier, the patient in this case report denied any other symptoms suggesting systemic infection.

The physiopathology of Kaposi's sarcoma remains controversial: viral, neurogenic, and vascular etiologies have been described. Recent evidence suggests that Kaposi's sarcoma is not a true tumor; rather, it is a dysregulation of inflammatory response. Additionally, it is associated with human herpes virus 8 infection, and lesion growth depends on various cytokines and growth factors, including the *tat* gene from the HIV genome⁽⁸⁾.

Although rare, Kaposi's sarcoma has been described in patients without AIDS or HIV. Mikropoulos et al. described a case of a HIV negative, immunocompetent 70-year-old

male with a 3-month history of a gradually growing mass on his superior bulbar conjunctiva⁽⁹⁾. The lesion was surgically excised with no additional treatment, and histological examination confirmed a diagnosis of Kaposi's sarcoma. There was no recurrence or metastasis five years after surgery. This case demonstrates that conjunctival malignancies may occur as isolated incidents and are not necessarily related to AIDS or immunosuppression^(9,10).

Proposed treatments for Kaposi's sarcoma include local excision, cryotherapy, radiation therapy, or chemotherapy. Nevertheless, recurrence is common⁽²⁾. It is crucial to routinely examine the mucosal surface in all patients with AIDS to identify Kaposi's lesions. Early diagnosis is essential for successful outcomes, as the underlying depression in cellular immune function appears to be irreversible.

In conclusion, this is an unusual case of a patient whose sole manifestation of HIV infection was Kaposi's sarcoma of the conjunctiva which, in itself, is an uncommon tumor. This case highlights the need for physicians to be cautious when identifying vascular tumors in the skin or mucosa and the need for histopathological investigation.

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