CASE REPORT

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Conjunctival Kaposi's sarcoma in Acquired Immunodeficiency Syndrome (AIDS) patient: a case report

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ABSTRACT

Background: Kaposi's sarcoma (KS) is malignant neoplasm of the vascular endothelium, caused by Human Herpes Virus 8 (HHV 8). Clinical variants consist of classic KS, iatrogenic KS, and AIDS KS. The manifestation of KS in the eye may occur in the eyelids, the lacrimal sac or gland, the orbit and bulbar or palpebral conjunctiva. This case study aims to demonstrate the incidental finding of conjunctival Kaposi sarcoma (KS) leading to acquired immunodeficiency syndrome (AIDS) diagnosis.

Case Presentation: A 34-years old male patient presented with a three-month history of progressively growing reddish mass in bulbar conjunctiva of the left eye. A mass with notable aggressive tumor characteristics heightened the suspicion for KS and comprehensive assessments were done accordingly. The patient was found to suffer from human immunodeficiency virus (HIV)/AIDS. Multistep approach treatment was commenced to treat AIDS-related KS and prevent recurrence in this case.

Conclusion: Patients with undiagnosed HIV infection would benefit from early diagnosis initiated by AIDS-defining illnesses such as KS. Given early identification and comprehensive management, KS abridged the time to diagnosis of HIV infection.

Keywords: Kaposi Sarcoma, Conjunctival Neoplasms, Acquired Immunodeficiency Syndrome

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Introduction

Kaposi sarcoma (KS) is a malignant neoplasm of the vascular endothelial cell caused by human herpes virus 8 (HHV-8). The clinical manifestation of KS comprises classic, iatrogenic, and acquired immunodeficiency syndrome (AIDS)-related KS.[1,2] Epidemiological data in America demonstrated that ocular involvement of KS was the most common finding. Ocular involvement may occur in the eyelids, lacrimal sac or gland, orbit, and bulbar or palpebral conjunctiva, with the greatest frequency in the latter. Kaposi sarcoma management options include cryotherapy, excision, radiation, or intralesional chemotherapy agent.[3]

AIDS-related KS contributed to a considerable proportion of the KS ocular involvement (20-24%). Male preponderance, especially in homosexual individuals, was the predominant demographic feature of these cases. Patients with AIDS-related KS usually have a $CD4^+$ count less than 500 cells/ml, which provides a suitable milieu for the human immunodeficiency virus (HIV) to infect cells and produce extracellular factors that potentiate the growth of KS tumor cells. AIDS-related KS was exceptionally notorious for its aggressive development.[3,4]

Conjunctival Kaposi sarcoma as the key diagnostic feature for HIV is a rare finding. To the best of our knowledge, the report regarding this case was scarce.[5] This case report is expected to improve the depth of knowledge in distinguishing and diagnosing conjunctival Kaposi Sarcoma.

Case Report

An otherwise healthy 34-years old male patient presented with long-standing redness on the left eye which was experienced for more than three months. The delay of presentation was due to his presumption of recurring insect bites. The size of the mass enlarged each time the redness recurred and foreign body sensation was discernible. In spite of the growing mass, the history of pain, tearing, discharge, blurry vision, and glare was denied. The patient had a tattoo and multiple sex partners.

In ophthalmology examination of the left eye, his visual acuity was 6/6, and conjunctival vascular injection and subconjunctival bleeding were observed. The reddish mass (8x5x2 mm) was situated at the inferior portion of bulbar conjunctiva with irregular surface, soft, friable, and fixed characteristics. Funduscopy examination revealed no abnormalities. The patient was then referred for further investigation to the division of infectious diseases and tropical medicine in internal medicine

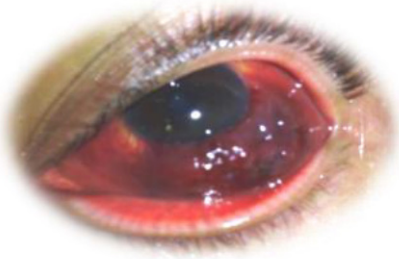

Figure 1. Clinical appearance of the left eye at two weeks after initial presentation

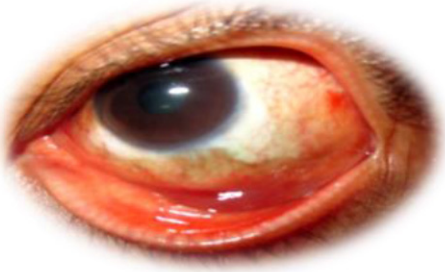

Figure 2. Clinical appearance of the left eye at the one-month follow-up

department. The investigation result confirmed HIV positive status with a $CD4^+$ count of 188 cells/mm^3. The chest x-ray result was within normal limit. Two weeks after his initial eye examination, the mass progressively increases in size to 10x5x2 mm (**Figure 1**).

The patient was diagnosed with HIV-infection and treated with highly active antiretroviral therapy (HAART) and cotrimoxazole. An excisional biopsy was conducted under local anesthesia. Pantocaine (2%) eye drop was instilled on the left eye, followed by disinfection with povidone-iodine and draping. Perilesional injection of 2% lidocaine was done prior to mass excision. Conjunctival closure with 8-0 vicryl absorbable suture was performed. The excised sample was fixated in 10% neutral buffered formalin and sent for histopathological examination. Postoperative medication prescribed included neomycin and polymyxin B sulfate and dexamethasone eye drop with additional artificial tears, and mefenamic acid orally.

Histopathological examination result reported a morphology consistent with the plaque stage of KS. The outcome was good and there was no clinical recurrence at one-month follow-up (**Figure 2**).

DISCUSSION

Kaposi sarcoma is a spindle cell malignancy of endothelial cells, first discovered by Mariecz Kaposi in 1872.[3,6] The incidence of KS increases with concurrent immunosuppressive conditions such as organ transplantation and AIDS.[7,8] AIDS-related KS is mostly caused by HHV-8. HIV infection leads to immune dysregulation by increasing cytokines secretion (interleukin [IL]-1,6; TNF-α), in which cascade resulted in increasing angiogenic growth factors which are the fundamentals of fibroblast growth factor. These factors combined with the presence of HIV protein factors stimulate cells to proliferate into neoplastic cells. Integrins and apoptotic processes are essential for the proliferation and neovascularization of tumor cells.[8]

Ocular involvement of KS can be found on the skin of the palpebral, bulbar, tarsal and conjunctival fornix, semilunar plica, caruncles, lacrimal ducts, and very rarely in the lacrimal gland and orbit. Conjunctival KS generally appears in the lower fornix, followed by the conjunctival bulbar and upper fornix. Lesions are usually elevated, mobile, bright red in appearance, and easily bleed. Ocular symptoms consisted of pain, photophobia, recurrent redness, irritation, foreign body sensation, epiphora, dry eyes, mucopurulent discharge, swollen eyelids, inability to close the eyes completely, impaired visual acuity, and blurred vision.[7,9]

Histopathological staging of KS consists of three stages. In 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 are common in stage III. Clinically, stage I and II are flatter with a size of less than 3 mm, while stage III is more nodular and sized larger than 3 mm.[7,10] In this case, both clinical and histopathological findings supported the diagnosis of stage III KS.

A comprehensive assessment is needed to confirm the diagnosis and detect any metastases in other organs. Thorax and abdominal computed tomography (CT) scan, with or without contrast, is preferred.[7] However, it is not indicated in cases where no significant development is identifiable after local treatment. Several studies have shown complete resolution of KS after HAART administration for four months.[11] Excision was first attempted by Dugel et al., where 14 patients with conjunctival KS were treated with surgical excision only. No recurrence was detected in patients with KS stage I and II, while recurrences after 8-31 months were reported in KS stage III. This served as a fundamental for our treatment decision with surgical excision and HAART. Residual tumor in the tarsal conjunctiva was treated with cryotherapy. Cryotherapy is reserved for severe conjunctival lesions affecting the bulbar conjunctiva and palpebral conjunctiva in nasal and temporal area.[9]

Topical and intralesional administration of interferon (IFN) is the modality of choice for AIDS-related KS. The recommended dosage for conjunctival AIDS-related KS using IFN-α 2a is 3 million IU/0.5 ml intralesional injection and 3 million IU topical eye drops every 6 hours for nine weeks or Mitomycin C 0.15 mL 0.2% MMC intralesional in the inferior or nasal area of the lesion followed by normal saline irrigation on the conjunctival and corneal surface.[9,12] Radiotherapy offers as the alternative therapeutic modality which is more effective on new KS lesion. Photocoagulation with argon laser and low-voltage photon radiation also provides excellent results.[1,13] AIDS-related KS requires HAART preceded by the administration of cotrimoxazole 960 mg/day single dose as primary prophylaxis two weeks before antiretroviral therapy.[14,15]

CONCLUSION

Patients with undiagnosed HIV infection would benefit from early diagnosis initiated by AIDS-defining illnesses such as KS. Given early identification and comprehensive management, KS abridged the time to diagnosis of HIV infection.

CONFLICT OF INTEREST

There was no competing interest regarding manuscript.

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None

AUTHOR CONTRIBUTION

All authors equally contribute to the study from the case selection, appropriate intervention to the patient, until evaluate the clinical outcome of patient.

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