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Case report

Aggressive conjunctival Kaposi sarcoma as the initial manifestation of acquired immunodeficiency syndrome

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ABSTRACT

Purpose: To report a case of Kaposi sarcoma in a patient with previously undiagnosed Human Immunodeficiency Virus (HIV) infection.

Observations: A 23-year-old male patient presented to the eye clinic with complaints of redness of his left eye for the past month. The patient had been seen one day prior to presentation in the emergency department for a neck mass and pneumonia. Exam was notable for a left hemorrhagic, nodular, bulbar conjunctival mass, a right hemorrhagic conjunctival lesion, and violaceous facial skin lesions. Due to suspicion for conjunctival Kaposi sarcoma, HIV and Acquired Immune Deficiency Syndrome (AIDS) serologies were obtained which returned positive. Biopsies of the ocular mass and axillary lymph nodes confirmed Kaposi sarcoma. The patient was started on anti-retroviral therapy and Doxorubicin. The left conjunctival mass initially progressed to cover most of his cornea but eventually regressed by 6 months.

Conclusions and Importance: Ocular involvement of Kaposi sarcoma as the initial manifestation of HIV/AIDS is rare with only a few reported cases. Since the advent of highly active antiretroviral therapy, conjunctival or adnexal Kaposi sarcoma is not commonly encountered by healthcare providers. Concern for Kaposi sarcoma of the conjunctiva in this patient led to the evaluation for HIV/AIDS. It is vital for all healthcare providers to have Kaposi sarcoma in the differential diagnosis of a hemorrhagic conjunctival or adnexal mass and be familiar with its association with HIV/AIDS. If suspected, appropriate counseling and testing should be performed.

1. Introduction

Kaposi sarcoma (KS) is a vascular spindle cell tumor of endothelial cell origin which can present in the skin, viscera, or mucosa. It was first described in 1872 by Dr. Moritz Kaposi, but the association with human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS) and human herpes virus 8 (HHV8) was not discovered until over a century later.[1,2] Although classic Kaposi sarcoma is considered an indolent disease, the epidemic or AIDS-associated form tends to be more aggressive. With the advent of highly active antiretroviral therapy (HAART) the incidence of KS has declined dramatically. In a large population study based in San Francisco, incidence rate of KS among white men averaged 32.1 per 100,000 people between 1987 and 1992 before declining to 2.8 in 1998.[3] A second large population study found that from 1990 to 1998 the incidence of KS decreased 8.8% per year in the United States, while initiation of triple antiretroviral therapy was associated with a 50% reduction in the incidence of KS.[4] Between 2000 and 2014, incidence rates of AIDS-associated KS amongst men aged 20 to 54 decreased from 1.44 per 100,000 people to 0.95 per 100,000 people in the United States.[5] Additionally, the progression and overall prognosis of KS has improved significantly through modern treatment regiments involving HAART used in conjunction with surgery, chemotherapy, and radiation.[1,6–8] However, as with most cancers the prognosis of AIDS-associated KS is highly dependent on the timing of diagnosis. Thus, prompt diagnosis of this condition can lead to a significant improvement in morbidity and mortality in patients who develop this

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disease. Furthermore, given the close association between KS and HIV/AIDS, early recognition of KS in patients with no known history of HIV/AIDS could lead to earlier and more successful management of the viral infection as well as possible prevention of a large number of HIV/AIDS-associated conditions.

In the height of the AIDS epidemic, AIDS-associated KS was found to involve the eye or ocular adnexa in 20% of patients diagnosed with KS.[9,10] In HIV patients, conjunctival KS has been described as the initial presentation of AIDS.[9,11] However, ocular KS as an initial presentation of HIV is very rare. To our knowledge, there are only 5 prior cases of conjunctival KS leading to a diagnosis of HIV.[12–16] In each of these cases, a patient presented with a single conjunctival mass prompting an excisional biopsy which revealed pathology consistent with KS. Each patient was subsequently diagnosed with HIV/AIDS as a result of their initial ocular pathology. In this report we present a case of aggressive conjunctival KS leading to a diagnosis of HIV/AIDS and the discovery of multifocal KS.

2. Case presentation

A previously healthy 23-year-old African American male presented for evaluation of 1 month of left eye redness and foreign body sensation. He denied pain, photophobia, or any visual changes. The patient had previously been evaluated by an optometrist for these symptoms and was prescribed tobramycin-dexamethasone drops, artificial tears, and cool compresses without improvement. The previous day, he was seen in the emergency department for cough, fever, chills, malaise, left eye redness, and a right-sided neck mass. He was diagnosed with pneumonia and started on a course of amoxicillin and clindamycin.

On examination, best corrected visual acuity was 20/20 in the right eye and 20/30 in the left eye. The patient was noted to have several violaceous skin lesions on his nose and cheek. A nodular, vascular, hemorrhagic mass was appreciated on the left inferior bulbar conjunctiva with palpebral conjunctival sparing. A sub-conjunctival hemorrhage involved the rest of the left bulbar conjunctiva (Fig. 1). A small sub-conjunctival hemorrhage was also found in the right inferior bulbar conjunctiva. The left cornea was clear except for one nodule overlying the inferonasal limbus. Fundoscopic exam revealed a single cotton wool spot nasal to the left optic nerve.

Due to concern for KS, a discussion was initiated with the patient to obtain more history. He had been seen by a dermatologist prior to presentation and treated with a topical steroid for the skin lesions without resolution. On further questioning he admitted to a 20 lb. weight loss and a sexual history involving occasional unprotected sex with both male and female partners. After obtaining the patient's consent, testing for HIV resulted as positive. Subsequent CD4 count was 104 cells/mL and viral load was 248,326 copies/mL. The patient was admitted to the hospital for worsening systemic symptoms.

Biopsy of the conjunctival mass revealed KS. Left axillary and cervical lymph node biopsies also confirmed the diagnosis. The patient was started on highly active antiretroviral therapy (HAART) and Doxorubicin. The patient was admitted to the hospital several times over the next 4 months for recurrent fevers, chills, malaise, nausea, and diarrhea. CT imaging revealed diffuse pathologic lymphadenopathy, enlargement of the spleen and liver with hypodense lesions, and enlargement of the left parotid gland. Endoscopy found multiple gastric, duodenal, and colonic lesions consistent with KS as well.

The conjunctival mass initially improved with systemic chemotherapy and HAART but then progressed over the next few months, covering most of the cornea. Surgical removal and an Interferon alpha injection were considered, but continued systemic therapy was deemed the appropriate treatment. At the last patient visit 6 months after presentation, the conjunctival and corneal lesions had significantly regressed leaving only a small elevated vascular mass in the superotemporal conjunctiva (Fig. 2).

3. Discussion

With HIV treatments becoming more efficacious and available, early recognition of HIV-related illnesses has become increasingly important in avoiding progression to advanced stages of HIV/AIDS. HIV-related diseases can present in a wide variety of ways in ocular tissues. In addition to the indolent cotton-wool spots seen on fundus exam in HIV-positive patients, ophthalmologists often provide assistance in diagnosing HIV infections by recognizing HIV-related illnesses such as KS, herpes zoster ophthalmicus, fungal infections, Tuberculosis, syphilis, Toxoplasmosis, cytomegalovirus (CMV), pneumocystis, and progressive outer retinal necrosis.[17,18] While certain conditions such as CMV are associated with more advanced stages of HIV/AIDS, some of these illnesses can occur in earlier stages, allowing for very early detection of HIV infection by ophthalmologists.

Ocular KS presents as a painless, red-purple, nodular mass most commonly involving the palpebral or bulbar conjunctiva or adnexa. Although KS should be highly suspected in any patient with a known history of HIV presenting with a red or purple conjunctival or adnexal mass, this presentation can mimic a variety of other conditions such as sub-conjunctival hemorrhage, conjunctivitis, episcleritis, scleritis, melanosis, malignant melanoma, squamous cell carcinoma, pyogenic granuloma, lymphangioma, cavernous hemangioma, and lymphoma. In a patient without a known history of HIV, diagnosing KS from a slit lamp exam alone can be very difficult. Ultimately, these lesions require biopsy in order to definitively diagnose KS. Fortunately for patients diagnosed with ocular KS, modern treatments have proven to be very effective at managing the condition.

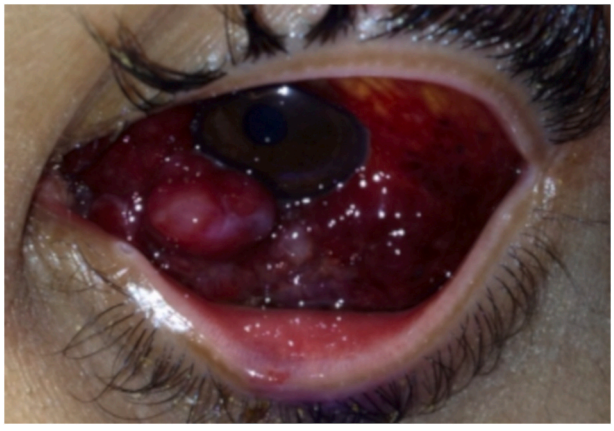

Fig. 1. Photograph of the left eye from initial visit showing a nodular, vascular, hemorrhagic mass on the left inferior bulbar conjunctiva.

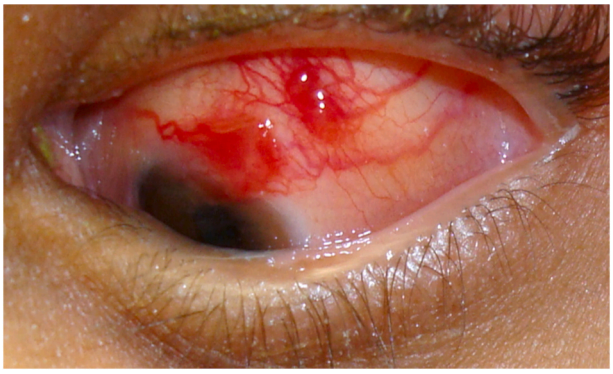

Fig. 2. Photograph of the left eye following surgical resection and initiation of HAART and doxorubicin showing a small elevated vascular mass in the superotemporal conjunctiva.

Currently, KS is treated with a combination of HAART and chemotherapy and/or radiation. For patients with disseminated KS, HAART plus systemic chemotherapy is used. First line chemotherapy regimens include liposomal anthracyclines such as doxorubicin or duanorubicin for initial treatment with paclitaxel, docetaxel, gemcitabine, vincristine, vinblastine and vinorelbine used for resistant cases. While treatment of disseminated KS is considered palliative, these regimens have proven to be very effective.[19] Liposomal anthracyclines have shown to produce a 76–82% response rate with a 26–40% complete remission rate, while paclitaxel has shown a 59% response rate. In patients with local disease, treatment regimens typically involve initiation of HAART along with either surgical excision, radiation, or intralesional chemotherapy. Although surgical excision with clean margins has the highest rates of full remission, radiation therapy has been shown to be a very effective option with response rates of 90% and complete remission rates of 70%. Even without surgery, radiation, or chemotherapy, response rates of local KS to initiation of HAART alone range from 66 to 86% with complete remission in 35% of patients.[20] For KS of the conjunctiva and adnexa, treatment options used alongside HAART involve surgical excision, radiation, cryotherapy, and intralesional injections of chemotherapy such as interferon and mitomycin. In many cases, treatment of isolated ocular KS is associated with high response rates and frequently, complete remission.[9,19,21–25]

Given the severity of undiagnosed HIV infection, benefits of timely diagnosis of underlying HIV infection, and favorable prognosis of patients diagnosed with local or early-stage KS, it is vital for all healthcare providers to have KS in the differential diagnosis of a hemorrhagic conjunctival or adnexal mass. In addition, providers must be familiar with its association with HIV/AIDS. If KS is suspected, appropriate counseling and testing should be performed so proper treatment can be initiated in a timely manner.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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