A rare case of bilateral conjunctival Kaposi’s sarcoma in a HIV-negative patient

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ARTICLE INFO

Keywords: Kaposi’s sarcoma Eyelid Skin cancer Oncology

ABSTRACT

Purpose: Kaposi’s sarcoma (KS) is a rare multi-centric vascular neoplasm, first described by Moritz Kaposi in 1872. It can appear in four different forms: classic, endemic, HIV-related and post-transplant form. We present an uncommon case of seronegative HIV infection patient with skin KS involving conjunctiva of both eyes.

Observations: Firstly, right eye lesions were surgically removed. Subsequently, the patient was administered with systemic chemotherapy (doxorubicin) to treat both skin and left eye lesions. No signs of recurrence were observed at 20 months’ follow-up.

Conclusions: Both eyes involvement in KS is rarely described in scientific literature. Both surgery and chemotherapy could be considered valid treatment options for conjunctival KS.

1. Introduction

Kaposi’s sarcoma (KS) is a low-grade, neoplastic disorder of vascular origin. It occurs in four different forms: classic, endemic, epidemic or Acquired Immune Deficiency Syndrome (AIDS) associated, and post-transplant form. All four types have the same histological features and are often associated with human herpes virus type 8 (HHV-8) infection. Nowadays KS has a worldwide circulation, especially due to an increase of the human immunodeficient virus (HIV) infection’s incidence. Recently, KS has also been frequently diagnosed in immunosuppressive patients after an organ transplant.

Most commonly, KS involves skin (being often located on lower legs, genitalia, face, oral mucosa), but it can spread in any visceral organs. KS of the eye is included in muco-cutaneous nodal lesions. Several cases of this sarcoma in HIV patients involving eyelids and conjunctiva have been published. The involvement of both eyes in a HIV- negative patient is extremely rare.

We hereby describe a case of HIV- negative patient with skin KS involving both eyes, as well as the treatment and the subsequent follow up.

2. Case report

An 83-years old white man referred to our department with a 3 months history of rapid growing conjunctival nodal lesions in both eyes, causing ectropion. Slit-lamp examination showed firm, pedicled purple-red nodular lesions (respectively 3 in right eye and 2 in left eye) on the infero-medial conjunctival fornix, causing bleeding and itching (Fig. 1A–C). Patient did not mention any pain or visual changes. Best corrected visual acuity was 20/30 for each eye and subsequent fundoscopic exam was normal for the age. Patient denied previous transplant history or intravenous drug abuse. The patient had pacemaker due to a previous myocardial infarction. The hematologic and biochemical tests were out of normal ranges. In particular, granulocytopenia and hyper- eosinophilia were observed. Serologic tests (Western Blot and ELISA tests) for HIV-1/2 were negative, while HCV-RNA was positive. Further skin examination showed multiple, over 2 cm length, painless asymmetrical, not scaly, maculo-papular, hyperpigmented focally nodular cutaneous lesions involving face, trunk (anteriorly and posteriorly) and both legs. Subsequent oral cavity examination, chest X-ray, gastroscopy and colonoscopy did not reveal any abnormalities, while Computed Tomography (CT) scan showed a hyperdense soft tissue mass in the medial canthus region (Fig. 1D). After the acquisition of the informed consent, three conjunctival nodular lesions of right eye were surgically removed.
removed under local anesthesia. Histological examination showed a cellular nodular tumor composed of slit and sieve-like spaces containing red blood cells (Fig. 1E). Immunohistochemical examination revealed positive staining for CD31 on vessel walls (Fig. 1F). Both features were considered typical of nodular stage of KS. In addition, immunohistochemistry for HHV-8 was negative. Margins were tumor free.

At the beginning, KS lesions were treated with cryotherapy and vinblastine in Dermatological Department. Two months later, due to recurrence of skin lesions and the persistence of left eye conjunctival lesions, patient was assessed by oncologist who decided to start chemotherapy with INN-Doxorubicin (CAELYX; 20mg/mq every 21 days for 3 months). Three months later the patient referred to our Department for a checkup examination. No recurrent lesions on the right eye were detected and both left eye conjunctival nodular lesions disappeared. At 20 months follow-up no recurrent neither new lesions were identified (Fig. 1G). Moreover, no HIV-seroconversion was detected.

3. Discussion

Kaposi’s sarcoma is defined as an idiopathic multiple pigmented sarcoma and it was first described by Moritz Kaposi in 1872.1 KS is currently categorized into four different types: classic, endemic, epidemic and post-transplant form.2 The form of KS originally identified by Kaposi became known as classic KS or sporadic KS. This form, that occurs mostly in elderly man of Mediterranean or Jewish ancestry, is usually diagnosed on the lower extremities as violaceous macules and plaques and shows an indolent course. Several cases, including a lymphadenopathic involvement, of the endemic form has been described since 1947 in Central Africa before HIV pandemic.10,11 The epidemic form, the most aggressive one, has spread all over the world due to HIV circulation. The post-transplant form is frequently diagnosed in patients after a solid organ transplantation and immunosuppressive therapy; this type of KS is known as iatrogenic KS.6

KS etiopathogenesis is controversial, but it is associated with Human Herpes Virus-8 (HHV-8).12 Moreover, CD31, platelet-derived growth factor receptor alpha (PDGFR-A) and vascular endothelial growth factor receptor-1 (VEGFR) are considered helpful diagnostic adjuncts for ocular adnexal KS.13 Our patient was HCV positive, while blood tests revealed no other viral infections, including HIV and HHV-8, strengthening the evidence that ocular KS may occur in HIV-negative patients.14 There are only few reports in the updated literature on adnexal KS in HIV-negative patients.15,16 Fernández-Montalvo et al. described the case of a 70-year-old male, who presented with bilateral eyelid masses successfully resolved after five cycles of chemotherapy with liposomal doxorubicin.15

Coblentz et al. surgically treated a conjunctival KS extending into the orbital space in a 93-year-old Caucasian male.19

Commonly, KS lesions are not painful and involve skin, being often located on lower legs, trunk, both limbs, genitalia and face. The most frequent sites of non-cutaneous KS include the gastrointestinal tract and respiratory system. Ocular involvement is extremely rare, especially in a HIV-negative patient.1 The tumor rarely spreads to the orbit; therefore, timely treatment is necessary. KS of the eye usually appears as a muco-cutaneous reddish or brown, elevated nodal lesion with conjunctival involvement.2 It is often misdiagnosed as angiosarcoma, hemangioma, pyogenic granuloma.

In the literature different types of KS treatment have been described such as cryotherapy, conventional surgery, interferon-α topic, radiotherapy, systemic chemotherapy and laser therapy.18 Highly Active Antiretroviral Therapy (HAART) combined with weekly docetaxel has been tested as an effective and well tolerated therapeutic option for ocular KS, prior to cryotherapy or surgical removal.19

In our case, we firstly performed surgical excision of the right eye, due to the major conjunctival involvement, in order to limit KS growth and to prevent a rapid spread to the eye as well as the possible orbital invasion. A simple conventional surgical excision of the three right eye lesions was curative. Histological and immunohistochemical examination showed typical features of Kaposi sarcoma with negative margins. After 2 months, due to a worsening of systemic KS (recurrence of skin lesions), and the persistence of left eye conjunctival lesions, we decided, in agreement with the oncologist to start chemotherapy with INN-Doxorubicin (CAELYX; 20mg/mq every 21 days for 3 months), avoiding a further local surgery in a cardiopathic patient. Three months later KS left eye lesions disappeared, highlighting that, in this case, chemotherapy was as effective as the surgery. Both eyes lesions were completely removed, with two different treatments and after 20 months follow-up, no signs of recurrence were observed.

4. Conclusion

KS of the eye is extremely rare in patients with seronegative HIV infection. Both eyes involvement is rarely described in scientific
literature. This case should represent a very important example for the type of treatment, since both surgical and oncological therapy have proven to be effective in this pathology.

Patient consent

A written informed consent was obtained from the patient for the preparation of this work.

Funding

No funding or grant support

Authorship

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

Declaration of competing interest

The authors report no conflicts of interest.

Acknowledgments

None.

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