Eyelid Kaposi Sarcoma in an HIV-negative Patient

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Kaposi sarcoma (KS) is a low-grade, multicentric vascular neoplasm. Most commonly, it involves the skin, but it can occur at any site on the body. The cutaneous lesions are often located on the lower legs, genitalia, oral mucosa, and face. KS is categorized in four different types: classic, endemic, epidemic or AIDS associated, and transplantation associated. We report a case of HIV-negative, classic KS located on the eyelid. The eyelid lesion was completely excised, and after a 1-year follow-up, no recurrences were observed. Ocular involvement by KS in a patient who is serologically negative for HIV is extremely rare.

Key words: Eyelid, human herpes virus 8, Kaposi sarcoma

Kaposi sarcoma (KS) is a low-grade, multicentric vascular neoplasm. Most commonly, it involves the skin, but it can occur at any site on the body. The cutaneous lesions are often located on the lower legs, genitalia, oral mucosa, and face. The lesions are not painful and can have a variety of colors due to the vascular component, which can differ from pink to brown. The most frequent sites of noncutaneous KS include the gastrointestinal tract and respiratory system.^[1] Ocular involvement is extremely rare. KS is currently categorized into four different types: classic, endemic in young African children, epidemic or AIDS associated, and transplantation associated.^[2]

Case Report

A 75-year-old Spanish man was referred to our department because of a lesion localized on his lower right eyelid that grew over the course of the previous month. Clinical examination showed a reddish, elevated, 1.5-cm lesion on the tarsal

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conjunctiva of the right lower eyelid [Fig. 1]. He had never received immunosuppressive therapy and denied high-risk sexual encounters or intravenous drug abuse.

The hematologic and biochemical tests were within normal limits. There were no abnormalities in the differential blood or in B and T cell subcounts (CD4+ T lymphocytes: 862/µL; CD8+ T lymphocytes: 932/µl; CD4+/CD8+ ratio: 0.93). The HIV test was negative. A chest X-ray showed signs of mild chronic obstructive pulmonary disease. Further examination of the skin showed multiple lesions, not painful, of about 2 cm in size on both arms [Fig. 1]. A biopsy of the eyelid lesion was made and the pathology study revealed a KS. A subsequent examination of the oral cavity, gastroscopy, and colonoscopy did not reveal anomalies.

After informed consent was obtained, the eyelid lesion was excised under local anesthesia. The pathology findings were considered typical of nodular stage KS. The margins of the resected tissue were free of lesion.

Microscopically, the lesion was composed of cellular bundles and fascicles of spindled cells as well as slit containing red blood cells. Cytologically, bland spindled cells, with moderate mitotic activity, and inflammatory cells were closely associated with narrow vascular spaces. The immunohistochemistry study showed positivity for the endothelial markers CD31 and ERG (a subfamily of the ETS family transcription factors) and for human herpes virus (HHV8) [Fig. 2].

The forearm lesions were treated with cryotherapy. There was no recurrence of this lesion or development of other cutaneous lesions after a 1-year follow-up period. No HIV-seroconversions were detected after this period of time.

Discussion

We report a case of HIV-negative, classic KS located on the eyelid. Ocular involvement by KS in a patient who is serologically negative for HIV is extremely rare. In 1994, Ron *et al.*^[3] and Munteanu *et al.*^[4] reported two cases of patients who were HIV seronegative and had a palpebral-conjunctival classic KS together with skin lesions, similar to our case. Kalinske and Leone^[5] described a patient with eyelid and conjunctival KS, who also had a gastrointestinal malignant neoplasm. More recently, Reiser *et al.*^[6] and Dammacco *et al.*^[7] published two cases of eyelid KS in an HIV-negative patient.

The etiopathogenesis of KS is controversial, but it is associated with HHV-8.^[8] It has been reported that an HHV-8

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Figure 1: Eyelid and cutaneous lesions of the case reported here. (a) Solitary eyelid nodule localized on the right lower eyelid. (b) Reddish nodule in the right lower eyelid involving the tarsal conjunctiva. (c) One of the cutaneous Kaposi sarcoma lesions located on the skin of the arms. (d) Postoperative image of the eye

infection is necessary but not sufficient for the development of KS and that more cofactors are probably required to trigger the KS.^[9]

The differential diagnosis of the eyelid involvement of KS includes hordeola/chalazia, pyogenic granuloma, angiosarcoma, and hemangioma.

The reported treatment for KS includes conventional surgery, cryotherapy, laser therapy, intralesional injections of vinblastine or bleomycin, interferon- α , radiotherapy with a multiple-fraction regimen, and systemic chemotherapy with liposomal daunorubicin or paclitaxel.^[7] In our case, we performed surgical excision to avoid further eye and orbital involvement. The eyelid lesion was completely excised, and after a 1-year follow-up, no recurrences were observed.

Conclusion

HIV-negative immunocompetent patients may manifest KS in the eyelid. Complete excision of the tumor may be curative.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

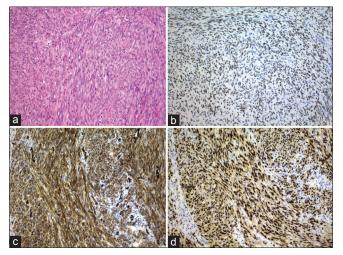


Figure 2: Histopathology of the eyelid nodule. (a) Microscopic appearance of Kaposi sarcoma showing monomorphic spindle cells arranged in ill-defined fascicles in the central area of the section (H and E, \times 200). (b) Immunoreactivity for human herpes virus 8 antigen (\times 200) showing cells positive for this antigen (brown cells). (c) Immunoreactivity for CD31 antigen (\times 200). (d) Immunoreactivity for ERG antigen (\times 200)

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Conflicts of interest

There are no conflicts of interest.

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