Conjunctival involvement in classic (indolent) HIV negative Kaposi's sarcoma

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Kaposi's sarcoma can occur in four clinical settings. In the first, the classic or European type, Kaposi's sarcoma is manifested as an indolent cutaneous malignancy primarily involving the lower extremities and feet, and is particularly prevalent among elderly men of Mediterranean or East European Jewish origin.1 The second major presentation is the aggressive lymphadenopathic/visceral form found among black Africans (HIV status uncertain).2 In the third AIDS related form, the process is both visceral and cutaneous and often aggressive with a poor outlook.3 The fourth group is HIV negative, but exogenously immunosuppressed, as in the case of transplant patients. The disseminated, subtype of classic Kaposi's sarcoma is extremely rare. We report herein, to the best of our knowledge, the first case of HIV negative classic type Kaposi's sarcoma involving the conjunctiva, with successful use of cryotherapy.

Case report
An 83-year-old Jewish male patient of East European origin presented with characteristic crops of violaceous nodular lesions on the skin of his right foot. His general physical examination was unremarkable as was the whole laboratory examination. Blood tests for HIV antibody, and CMV antigen, using enzyme linked immunosorbent assay (ELISA) were negative. Epstein-Barr virus antigen, checked by immunofluorescence, was likewise negative. Testing of serum immunoglobulins, CD4 lymphocyte count, and helper/suppressor ratio was done, with all results within normal limits. The patient had never received immunosuppressive treatment in the past.

Excisional biopsy of one of his lesions was performed and the histopathological examination revealed the presence of Kaposi's sarcoma stage II of the skin.

Several months later, new lesions appeared on the skin of his right thigh and both scrotums. At the same time, several dark red coloured 3×3 mm nodular elevated lesions were found in the right bulb and palpebral conjunctiva (Fig 1). Subsequently, the patient complained of marked discomfort and tearing. The ophthalmic examination of the right eye revealed a clear cornea, pseudophakia, and normal fundus. Examination of the left eye was unremarkable. The patient was treated by cryoapplication with −80°C probe. Each lesion was treated with three applications of 20 seconds each, once a week for 4 weeks until significant regression was achieved (Fig 2).

At the end of a 2 year follow up, no evidence of systemic involvement was found and the conjunctival lesions did not recur following the cryotherapy (Fig 3).

Comment
Kaposi's sarcoma is a multicentric vascular neoplasm affecting the skin, mucous membranes, internal organs, and lymph nodes. It is usually associated with AIDS, involving 15% to 24% of AIDS patients.4

Figure 1 Stage III classic type Kaposi's sarcoma in right conjunctiva.

Figure 2 Significant regression of the Kaposi's lesions at the end of cryotherapy treatments (photograph taken 10 days following last treatment).

Figure 3 Treated right eye showing no recurrences of Kaposi's lesions in the conjunctiva, at 2 year follow up.
AIDS related Kaposi's sarcoma follows a much more fulminant course and is less responsive to therapy than the more rare indolent HIV negative Kaposi's sarcoma, which affects elderly males of Mediterranean or East European Jewish origin or the type which appears in young adult black African males.1 The therapeutic options for adnexal ocular Kaposi's sarcoma are: (a) observation of asymptomatic lesions; (b) surgical excision; (c) cryotherapy; (d) irradiation.14

Dugel et al.15 classified conjunctival Kaposi lesions into three clinical stages: stage I and II are patchy and flat (less than 3 mm in height); stage III lesions are more nodular and more than 3 mm in height. They found a better rate of success for surgical excision and/or cryotherapy for stage I and II tumours.5

Because our patient's conjunctiva was affected by several lesions, necessitating excision of a large area of the conjunctiva, and because of the benign course of this type of Kaposi's sarcoma, we chose the cryotherapy option of treatment. It is also obvious that the risk/benefit ratio of this method is much lower than that of radiotherapy of the adnexa oculi.

This patient is unique as his disease course was benign, his immunological profile was normal, he had never received immunosuppressive therapy, and his blood tests were negative for HIV antigens.