

**SHORT COMMUNICATION****Case report****Orbital Kaposi's sarcoma in acquired immunodeficiency syndrome**

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*A 28-year-old white male with AIDS - C3 staging, presented with an extensive hemorrhagic dark mass localized in the left orbit. No other ophthalmic findings were disclosed. Ultrasonography and computed axial tomographic scans showed orbital involvement. Orbital Kaposi's sarcoma is a rare finding and only a few cases have been reported. Systemic examination revealed other lesions suggestive of disseminated mucocutaneous Kaposi's sarcoma, oral candidiasis, membranous esophagitis and granulomatous hepatitis. Eyelid incisional biopsy disclosed Kaposi's sarcoma. Despite intensive chemotherapy progression was aggressive with a fatal outcome. (Eur J Ophthalmol 2000; 10: 88-90)*

**KEY WORDS:** *Orbital, Kaposi's sarcoma, AIDS*

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**INTRODUCTION**

Kaposi's sarcoma is a vascular neoplasm of unknown histogenesis (1, 2). Current concepts consider Herpes virus type 8 as a possible causative agent (3).

The tumour tends to involve primarily the skin, also affecting mucous membranes, internal organs and lymph nodes. About 22% of acquired immunodeficiency syndrome patients have multifocal Kaposi's sarcoma, frequently on the eyelids and conjunctiva. Orbital involvement is extremely uncommon (2, 4). We observed an unusual case of AIDS with severe orbital Kaposi's sarcoma.

**Case report**

A 28-year-old white male, a percutaneous drug abuser AIDS-C3 staging, presented with an extensive hemorrhagic dark mass localized on the left orbit (Fig. 1). He also had purplish slightly raised, nonblanching papules, on the nose, chin and upper trunk. On ophthalmic examination, visual acuity was 10/10 in the right eye. The left eye was hidden by a large tumour and no other abnormalities were found.

US scan showed a solid lesion with a sarcoma like

pattern (Fig. 2). CT scans showed anterior orbital involvement of a large tumour with no bone erosion (Fig. 3). Systemic examination brought to light other lesions suggestive of disseminated mucocutaneous Kaposi's sarcoma, oral candidiasis, membranous esophagitis and granulomatous hepatitis.

Eyelid incisional biopsy detected interlacing bundles of spindle cells tending to line blood vessels (Fig. 4). The endothelial cells lining well-formed tumour blood vessels give a strong reaction with immunohistochemical staining for FVIII - RAG with the peroxidase-antiperoxidase technique (Fig. 5). All these features are characteristics of Kaposi's sarcoma.

Despite intensive chemotherapy with adriamycin, bleomycin, vincristine, isoniazid, ethambutol, ethionamide and streptomycin the course was aggressive and there were no conditions for local treatment. The patient rapidly died of atypical mycobacterium dissemination sepsis.

**DISCUSSION**

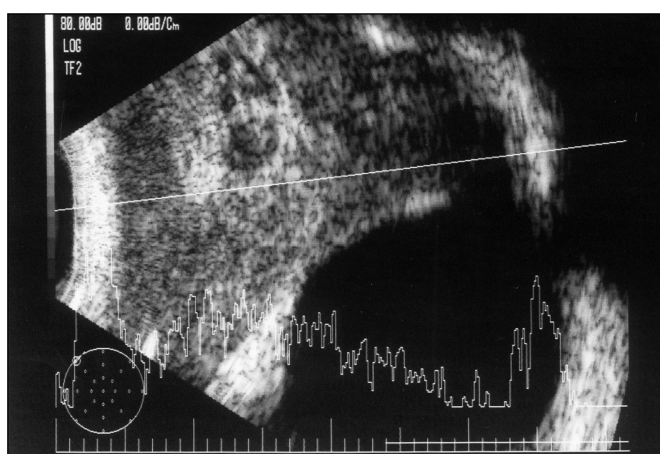
Orbital and periorbital findings in AIDS patients include eyelid involvement by Varicella zoster, Molluscum contagiosum and Kaposi's sarcoma as well as

orbital lymphoproliferative lesions and eosinophilic granuloma (5,6).

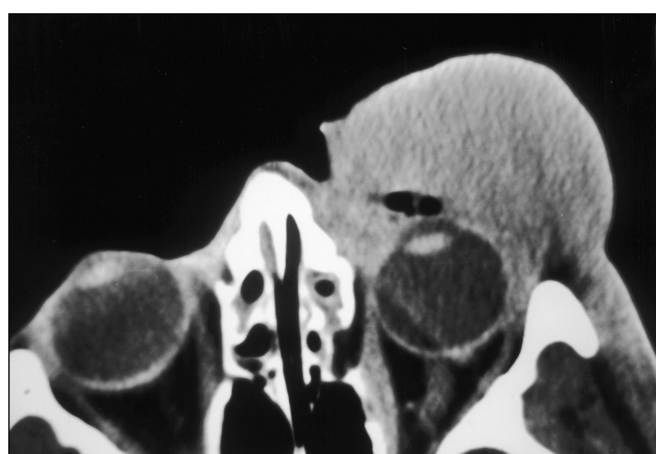
The ophthalmic manifestations of Kaposi's sarcoma are usually limited to the conjunctiva and eyelids (7,8). Characteristically, it begins as a bluish-red macule that coalesces and eventually spreads to internal organs (9). Orbital Kaposi's sarcoma is rare and only a few cases have been reported (1,4,6). The exuberant appearance of our case suggests a severe immunodeficiency partially related to the patient's initial irregular approval for treatment.

The neoplasm is usually a multifocal progressive lesion. The clinical history should alert to the obvious diagnosis. Although large tumours may require local treatment, excision or focal irradiation of isolated oph-

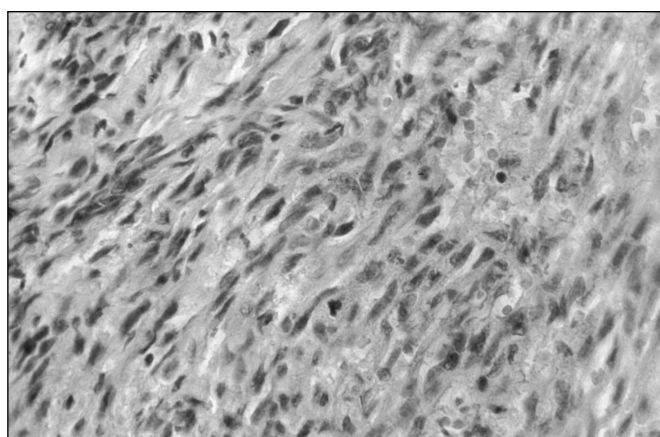
**Fig. 1** - Photograph showing an exuberant hemorrhagic dark mass on the left orbit. Purplish skin papules are also present on the nose and chin.



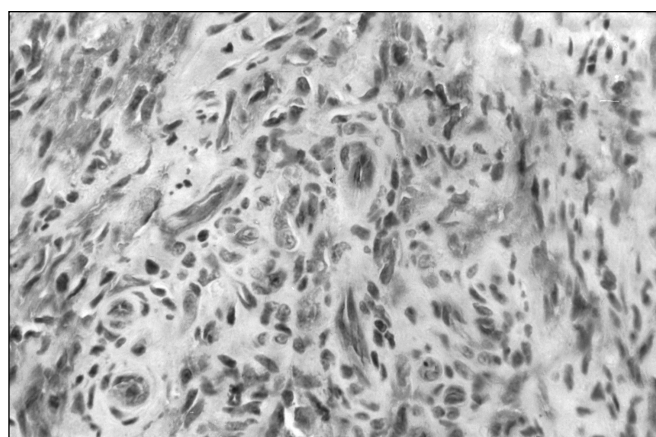
**Fig. 2** - Axial transocular C-scan echogram shows a very large lesion extending into the anterior orbit, next to the globe. The tumour has a somewhat irregular internal structure with moderate sound attenuation.



**Fig. 3** - Axial computed tomographic scan reveals a large tumour infiltrating the anterior aspect of the left orbit. The bone is intact.



**Fig. 4** - Light microscopy of an incision biopsy of the tumour: bundles of spindle-shaped cells tend to line blood vessels forming clefts. (hematoxylin and eosin, x 400).



**Fig. 5** - Immunohistochemical staining for FVIII-RAG (CD34 x 400).

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thalmic lesions will not prevent the eventual development of tumours elsewhere. Therapy must be directed toward systemic control of the disease, particularly when associated with AIDS. Current chemotherapeutic regimens are associated with a dismal prognosis, but the introduction of highly active antiretroviral therapy (HAART) has changed the course of the disease (10).

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