PHOTO QUIZ

Swelling of the eyelids

R.G.M. Weijma[1]*, E. Kerver[1], R. van Zijderveld[2] G.E.L. van den Berk[1]

Departments of [1]Internal Medicine and [2]Ophtalmology, Onze Lieve Vrouwe Gasthuis, Amsterdam, the Netherlands, *corresponding author: tel.: +31 (0)20-5991173, fax: +31 (0)20-5993523, e-mail: r_weijma@hotmail.com

A 48-year-old male was referred by the ophthalmologist because of a two-month history of progressive swelling of his eyelids. In the last three weeks he also developed several purple lesions on his legs, neck and groins. The lesions increased in size, were not painful, but did itch occasionally.

Besides fatigue for five months he had no other complaints. His medical history was otherwise unremarkable. On admission physical examination showed a blood pressure of 144/82 mmHg, a pulse of 88 beats/min and a temperature of 37.5 °C. On both eyelids there was a purple raised lesion of 1.5 cm in diameter (*figure 1*). Further examination showed multiple lesions on the face, in the retroauricular region and on both legs. The mouth showed white patches and on the palate there were multiple purple lesions as well.

The first blood work showed a mild normocytic anaemia (7.4 mmol/l) with no other abnormalities.

Figure 1. *Before treatment*

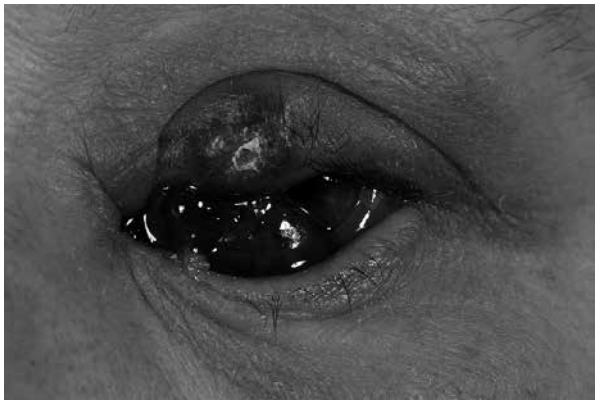
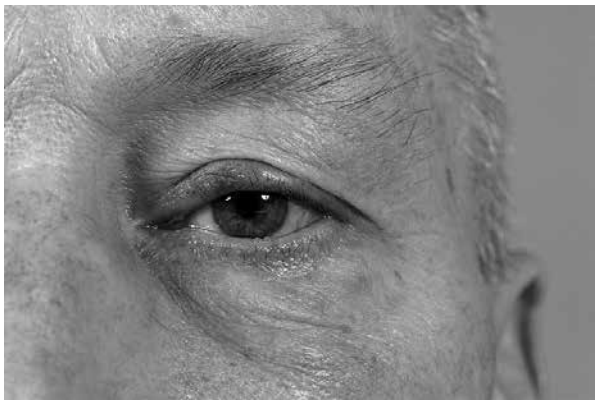


Figure 2. *After treatment*



WHAT IS YOUR DIAGNOSIS?

See page 98 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 94)

SWELLING OF THE EYELIDS

DIAGNOSIS/DISCUSSION

The differential diagnosis of the eye lesions included hordeola/chalazia, pyogenic granuloma, angiosarcoma and haemangioma. However, because of the widespread lesions we mainly considered Kaposi sarcoma and bacillary angiomatosis.

Additional history revealed that our patient had a HIV-positive male sexual partner. His HIV test turned out to be positive. The CD4 count was 220 x 10^6/ mm^3 and the HIV viral load 92,600 c/ml. The diagnosis of Kaposi sarcoma was made after a biopsy, which showed an angioma with positivity for human herpes virus 8 (HHV 8).

Kaposi sarcoma is an angioproliferative disorder which is associated with HHV 8.[1] There are four types of KS with a variable course. The uncommon classic type, which is usually indolent, predominantly occurs in older males with Mediterranean roots. The endemic type, which is prevalent in Africa, is independent of HIV infection. The iatrogenic type occurs in immune suppressed patients.[2] The most well-known is the AIDS-associated type.[2,3] Kaposi sarcoma most commonly involves the skin, but it can occur at any site of the body. The cutaneous lesions are often situated on the lower legs, face, oral mucosa and genitalia. The lesions are not painful and can have an assortment of colours due to vascularisation, which can vary from pink to brown. Lymphoedema is often present.

The most frequent sites of noncutaneous Kaposi sarcoma include the gastrointestinal tract and respiratory system.[3]

The incidence of Kaposi sarcoma has declined from 6.7% to 2% since the introduction on highly active anti-retroviral therapy (HAART) in 1996.[4]

The treatment of Kaposi sarcoma should always include the introduction of HAART. For some patients this is sufficient. In extensive cutaneous, symptomatic visceral involvement or cutaneous Kaposi sarcoma, which is unresponsive to treatment with HAART, the start of local or systemic chemotherapy is indicated. Radiation therapy is suggested for those with larger lesions which are unresponsive to chemotherapy.[5] Despite therapy, Kaposi sarcoma remains a morbid and occasionally life-threatening condition.[6]

In our patient, we started with HAART, after which the CD4 count increased to 500 x 10^6/ mm^3. Because of the extent of the disease our patient was treated with systemic chemotherapy. With liposomal doxorubicin all lesions except those on the feet went into remission. Radiation therapy was recently started for the lesions on his feet.

REFERENCES

1. Pica F, Volpi A. Transmission of human herpesvirus 8: an update. Curr Opin Infect Dis. 2007;20:152-6.
2. Schwartz RA. Kaposi's sarcoma: an update. J Surg Oncol. 2004;87:146-51.
3. Dezube BJ. Clinical presentation and natural history of AIDS--related Kaposi's sarcoma. Hematol Oncol Clin North Am. 1996;10:1023-9.
4. Engels EA, Pfeiffer RM, Goedert JJ, et al. Trends in cancer risk among people with AIDS in the United States 1980-2002. AIDS. 2006;20:1645-54.
5. Bower M, Collins S, Cottrill C, et al. British HIV Association guidelines for HIV-associated malignancies 2008. HIV Med. 2008;9:336-88.
6. Sullivan RJ, Pantanowitz L. New drug targets in Kaposi sarcoma. Expert Opin Ther Targets. 2010;14:1355-66.