

Pre- and Posttreatment MR Imaging in AIDS-Related Kaposi Sarcoma of the Conjunctiva and Lacrimal Gland

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Summary: AIDS-related Kaposi sarcoma can affect the eye: the (bulbar) conjunctiva and lacrimal gland being rare sites of occurrence. We present a case of AIDS-related Kaposi sarcoma of the conjunctiva in which MR imaging was effective in suggesting the diagnosis and aiding therapeutic strategy. We also discuss advances in imaging of ocular and orbital malignancies presented in the recent literature.

The ocular manifestations of AIDS have received relatively little attention in the literature. The most common lesions seen are retinal. They can be either infectious (mostly as cytomegalovirus retinitis) or noninfectious retinopathy (microvasculopathy on progression of AIDS). Cerebral toxoplasmosis is only rarely associated with ocular involvement (1). A less common involvement of the eye in AIDS-related disease is Kaposi sarcoma (KS) of the conjunctiva (2), and one should be aware that this can be an initial manifestation of HIV infection, although it is rarely the case (3). Isolated KS of the conjunctiva was first reported in 1967 (4).

KS, now known as angioproliferative disease, occurs in several clinico-epidemiologic forms, all of which are associated with infection by human herpesvirus eight (HHV-8) (5). The current concept of KS pathophysiology is complex. In a context of immune dysregulation, a latent HHV8 infection of endothelial cells can occur. Progression to KS is most likely caused by two factors: deregulated expression of oncogenes and oncosuppressor genes by HHV-8 and, for AIDS-related KS, the proliferative and angiogenic effects of the HIV-tat protein (5).

Relatively few reports provide information on the imaging characteristics of KS in bone, pharynx, lung, or liver. In 2000, Collaco et al (6) presented a case of disseminated KS with orbital involvement and reported sonographic and CT findings. To our knowledge, no information on MR findings in KS of the eye

has yet been published. We present an MR imaging study of a patient with histologically confirmed KS of the eye and also include results of follow-up MR imaging after therapy.

Case Report

We report the case of a 38-year-old male patient with a 3-week history of progressive reddening and swelling of his left eye (Fig 1) followed by blurred vision. Microbiological tests did not reveal an infection. Eighteen years earlier, he was diagnosed with HIV infection and has received antiretroviral therapy for 9 years. On presentation, his CD4 count was 4/ μ L after a 7-month period of noncompliance under highly active antiretroviral therapy (HAART, consisting of trizivir [abacavir sulfate, lamivudine, and zidovudine] and viread [tenofovir disoproxil fumarate]). An orbital MR imaging study (Fig 2 A–C) revealed abnormal enhancing tissue with low signal intensity on T1-weighted images and high signal intensity on turbo inversion recovery magnitude (TIRM) images involving mostly the conjunctiva and lacrimal gland of the left eye. As a differential diagnosis, a lymphoproliferative disorder was considered, including a conjunctival manifestation of KS with involvement of the lacrimal gland. Histologic analysis confirmed the diagnosis of KS. Mucocutaneous examinations revealed additional oral lesions, whereas no gastrointestinal disorder could be found. After repeated therapy with liposomal doxorubicin, serial MR imaging (Fig 2D–F) did not show a contrast-enhancing mass in the previously affected tissues of the left eye.

Discussion

KS is the most common tumor arising in HIV-infected patients and is an AIDS-defining illness according to Centers for Disease Control guidelines. The clinical course of AIDS-related KS is highly variable, and skin lesions often coincide with pulmonary or gastrointestinal KS. When KS is suspected, mucocutaneous and gastrointestinal investigation should be initiated. In the case of our patient, KS was present in one eye, the pharynx, and lung. KS of the eye is an entity that has not been widely described in the literature.

Although clinical examination and biopsy are still required to confirm the diagnosis of KS, knowledge about the radiologic presentation could aid diagnosis in the orbita or other intracranial locations with limited access for evaluation. To our knowledge, this is the first publication of information on MR imaging results of KS of the eye. In the case of our patient, an abnormal mass was seen with high signal intensity on T2-weighted (in this case, turbo inversion recovery magnitude) and low signal intensity on T1-weighted

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FIG 1. Clinical aspect of left eye before therapy, conjunctival injection and infiltration.

images with a marked enhancement on postcontrast images due to rich vascularization. The findings were interpreted as an infiltrative process of the bulbar conjunctiva and diffuse involvement of the left lacrimal gland. Although those findings are nonspecific and raise the possibility of a lymphoproliferative disorder, pseudotumor, or infection, the diagnosis of KS is prompted by the context of HIV infection. Bulbar conjunctival KS can be confused with caroticocavernous fistula, hemangioma, or lymphangioma. Caroticocavernous fistula would be accompanied by edematous

swelling or chronic subconjunctival hemorrhage. A fistula would show a pronounced vascular congestion of the superior ophthalmic vein, in contrast to KS. Cavernous hemangiomas occurring in adults are almost always intraconal in location. They are usually hypointense on T1-weighted images and markedly hyperintense on T2-weighted images. Capillary hemangiomas can be located in the preseptal region and show an infiltrative pattern, but they are usually pediatric tumors affecting infants up to 2 years of age. Lymphangioma occurs in children between 1 and 15 years old and involves muscle tissue. In addition to MR imaging, orbital tumors can be further differentiated by clinical history, examination, CT, and angiography. In vascular tumors, Doppler sonography may be helpful.

We recommend use of the following sequences: axial T2-weighted images (3 mm), axial T1-weighted images without fat saturation (3 mm), postcontrast axial and coronal T1-weighted images with fat saturation (3 mm), and postcontrast T1-weighted parasagittal images if spatial information is required. KS may not present with distinct imaging criteria but may have characteristic tissue distribution (glands, mucosal tissues).

If precise spatial information on the dimensions of ocular KS is required, advanced MR imaging techniques may be employed. Ultra thin-section 3D fast spin-echo (3D-FSE) T2-weighted imaging, a new MR

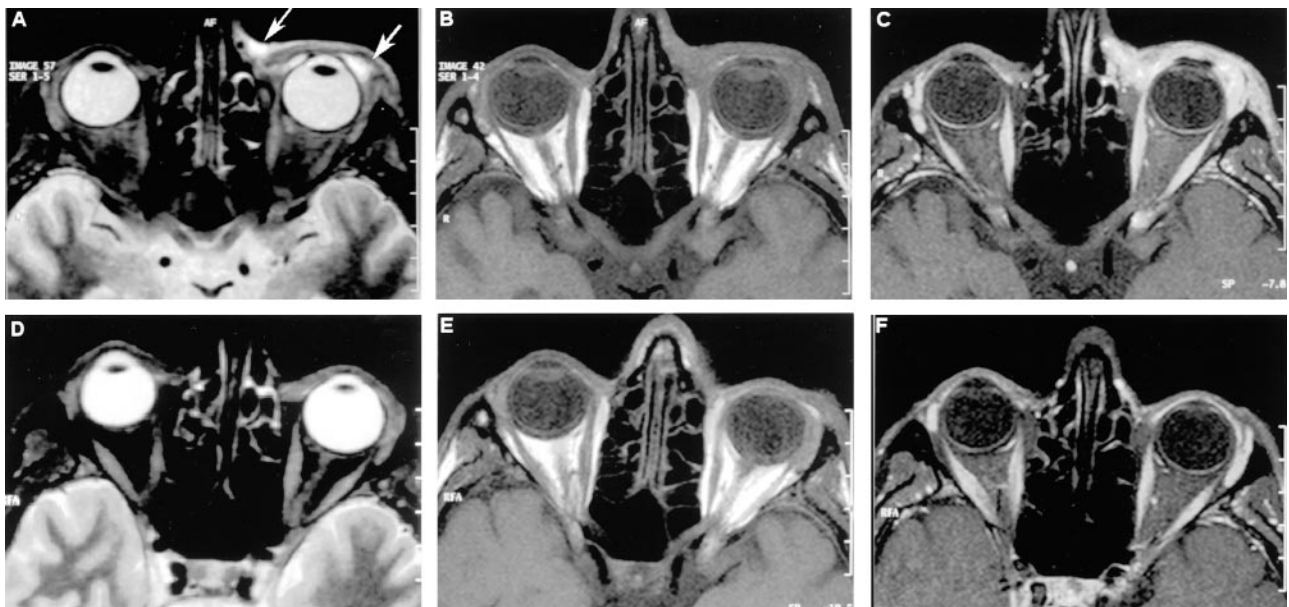


FIG 2. MR images obtained a 38-year-old man who developed AIDS-related Kaposi sarcoma of the conjunctiva and lacrimal gland. All axial sections were tilted to the level of the optic nerve; coronal sections were perpendicular to axial sections. Posttreatment images were obtained 3 months after pretreatment images.

A, Axial turbo inversion recovery magnitude (4000/30 [TR/TE]) MR image of the orbita shows an abnormal mass (arrows) with high signal intensity that involves the conjunctiva and lacrimal gland.

B, Axial T1-weighted (735/12) MR image of the orbita shows an abnormal mass that is isointense relative to muscular tissue (arrows) and involves the conjunctiva and lacrimal gland.

C, Corresponding postcontrast T1-weighted (735/12) MR image shows a marked enhancement of the abnormal tissue of the lower eyelid.

D, Corresponding posttreatment axial turbo inversion recovery magnitude (4000/30).

E, Corresponding posttreatment axial T1-weighted (735/12).

F, Corresponding posttreatment postcontrast T1-weighted (735/12)

imaging technique (7), can be used as a supplementary method and has been proposed recently for the study of ocular and orbital malignancies. The 3D-FSE T2-weighted sequence offers superior resolution of intraocular and orbital structures compared with that of conventional MR imaging. The main advantage of the 3D-FSE T2-weighted sequence is the option to perform multiplanar reconstructions. This method allows a subtle analysis of anatomic structures and thus can be helpful in localizing pre- or postseptal tumors and checking for bulbar infiltration. Although this new technique is particularly useful in the evaluation of intraocular tumors and the nerve-sheath complex, it may also improve diagnosis and therapeutic management in patients with ocular and orbital malignancies as KS.

Conclusion

In our patient, a regression of ocular KS was seen after a therapy with liposomal doxorubicin. Thus, we observe that a marked reduction of KS can be seen at follow-up MR imaging, but this observation is not

specific to any particular therapeutic or pathogenetic mechanism. Nevertheless, knowledge of the MR imaging features of KS are useful not only in diagnosis but also in noninvasive assessment of the impact of emerging therapeutic strategies (8) for KS.

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