Demonstration of Herpes Virus 8 in a Lymphangioma-Like Kaposi's Sarcoma Occurring in a Immunolmmunosuppressed Patient

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Key Words

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In April 1993, a 42-year-old Libyan man presented in the dermatology department with several lesions of the right foot. The lesions consisted of slightly raised reddish-brown plaques, 1.5 cm in diameter. The patient was in good health otherwise, and the physical examination was unremarkable, in particular there was no ankle edema or lymphadenopathy. The results of laboratory investigations were within the normal limits and HIV serology was negative. A biopsy of one of the lesions was taken. Histologically, it was composed of a proliferation of irregular lymphatic-like endothelial lined channels permeating the dermis. These channels were generally bloodless, and separated by dense collagen bundles (fig. 1). Spindle cells were rare or absent. Thus the diagnosis of lymphangioma-like Kaposi's sarcoma (KS) was suggested, and herpes virus [8] (HHV-8), a virus that has been suggested to play a role in various forms of KS, was demon-

Fig. 1. Typical pathological findings of lymphangioma-like KS: Irregular lymphatic-like endothelial lined channels, separated by dense collagen bundles. Note the absence of any area of spindle cell proliferation. HE $\times$ 20.

strated using the hot-start PCR technique from paraffin-embedded specimen, as previously described [1]. The lesion was positive for HHV-8, as shown by amplification of a 233-bp product (fig. 2) characteristic of the virus.

Lymphangioma-like KS is a rare and poorly understood entity. To date only several cases have been reported in the literature [2-7]. It is characterized by dilated lymphatic-like channels mimicking other primary lymphangiomatous tumors such as progressive lymphangioendothelioma, lymphangiectasis or lymphangioma or vascular tumors such as hemangioma, angiosarcoma or spindle cell sarcoma. Since spindle cells typical of KS are rare, as in our patient, this variant of KS remains conceptually debated. For some authors, it represents not true KS but a lymphatic tumor, whereas others consider it a distinctive variant of KS. We demonstrate here, for the first time, the presence of HHV-8 in lymphangioma-like KS. We as well as others have described this virus in various forms of KS including classic KS, endemic KS and KS occurring in immunosuppressed or HIV-infected patients [1, 7-12]. Naturally, our findings must be carefully investi-

Fig. 2. Ethidium-bromide-stained agarose gel of PCR products of KS samples. Lane 1 = Molecular-weight DNA marker (PBR 322 DNA Hae III); lane 2 = negative control; lane 3 = nodular KS HHV-8 positive, HIV patient; lane 4 = lymphangioma-like KS HHV-8 positive, in an HIV-negative Mediterranean patient. The 233-bp fragment corresponds to specific herpesvirus-like DNA sequences and the 268-bp product to the β -globin gene.

gated; the PCR technique does not provide quantitative data and, despite the paucity of spindle cells, must give intense signals. However, it is interesting to note that our results are consistent with the PCR in situ data recently published by Boschhoff et al. [13], who have described HHV-8 sequences both in endothelial and in spindle cells. These findings suggest that lymphangioma-like KS does represent a rare subtype of KS and not a particular lymphatic-type tumor. Indeed, the majority of vascular tumors are HHV-8 negative, except for an angiosarcoma [14, 15]. In order to clarify the unsolved problems of the epidemiology of this new virus [16], further large, control epidemio-logical studies are required in the future. Such studies are in progress.

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