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Case Report

AIDS-related Kaposi's Sarcoma of the Nasopharynx

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Abstract

Kaposi's sarcoma (KS) is an acquired immunodeficiency syndrome (AIDS)-related malignancy and may present in the head and neck as an initial sign of AIDS. However, it is rare in the nasopharynx. We report a 28-year-old man who complained of postnasal drip and occasional bloody saliva for 2 months. A purple-red bulging nasopharyngeal tumor was found on examination; a biopsy specimen proved that it was KS with positive human herpes virus type 8 (HHV-8) staining in the tumor cell nuclei. AIDS was diagnosed when enzyme immunoassay and Western blot were positive for serum human immunodeficiency virus. Antiviral therapy was given to treat AIDS, and low-dose radiation was given to treat nasopharyngeal KS with a complete response. (*Tzu Chi Med J* 2009;21(4):342–344)

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1. Introduction

Kaposi's sarcoma (KS) is an angioproliferative malignant neoplasm that arises from mesenchymal progenitor cells infected by human herpes virus-8 (HHV-8) and may be multifocal in origin (1,2). Acquired immunodeficiency syndrome (AIDS)-related KS can affect any area of the skin, aerodigestive tract mucous membranes, and even the internal organs. The most common site reported is the skin on the lower limbs (2). In the head and neck region, the most common sites affected are the oral cavity, oropharynx and conjunctiva (2–4). However, the nasopharynx is rarely a uniquely affected site of KS. Here, we report KS initially presenting as a nasopharyngeal tumor that was diagnosed as AIDS-related and which was successfully managed with low-dose radiotherapy (4,5). The clinical features and treatment modalities of KS are also reviewed.

2. Case report

A 25-year-old man with a history of homosexuality came to our outpatient department in April 2005 complaining of postnasal drip and occasional bloody saliva for 2 months. On examination, his local and physical condition was grossly normal except for a purplish elevated tumor with scattered red plaques in the right lateral and posterior walls of the nasopharynx (Fig. 1), and many small lymph nodes palpated at the periauricular and bilateral neck areas (Fig. 2). A punch biopsy was done immediately through the nose. Pathological examination showed KS (Fig. 3)

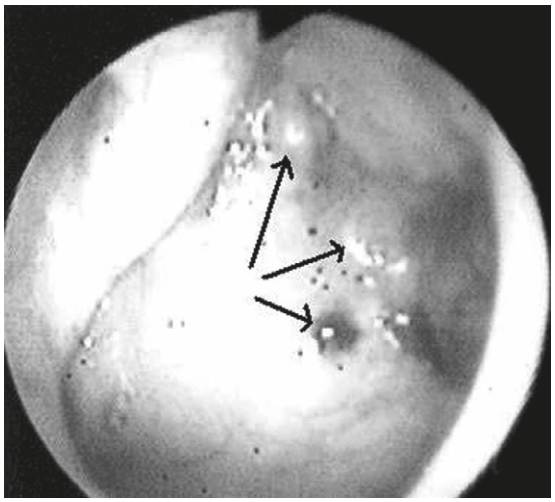

Fig. 1 — Nasopharyngeal fiberscopy reveals a purplish elevated mass in the right lateral and posterior walls of the nasopharynx with scattered red plaques (arrows).

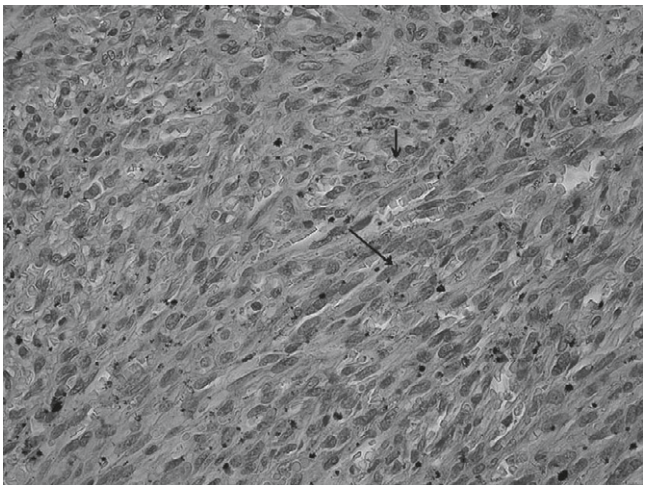

Fig. 3 — Kaposi's sarcoma composed of spindle cells (long arrow) with slit spaces filled with erythrocytes (short arrow) in the submucosa of the nasopharynx (hematoxylin & eosin, 400×).

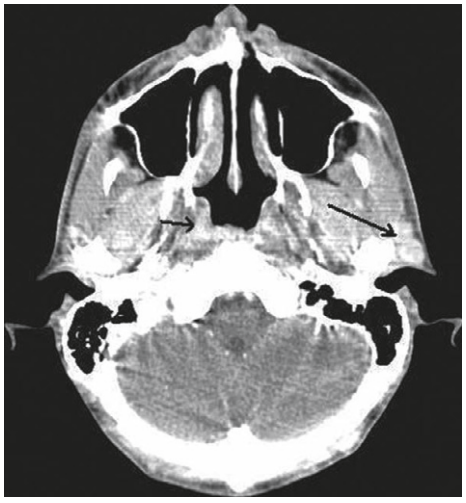

Fig. 2 — Computed tomography shows a contrast-enhanced mass in the right lateral and posterior walls of the nasopharynx (short arrow); left periauricular lymphadenopathy is also noted (long arrow).

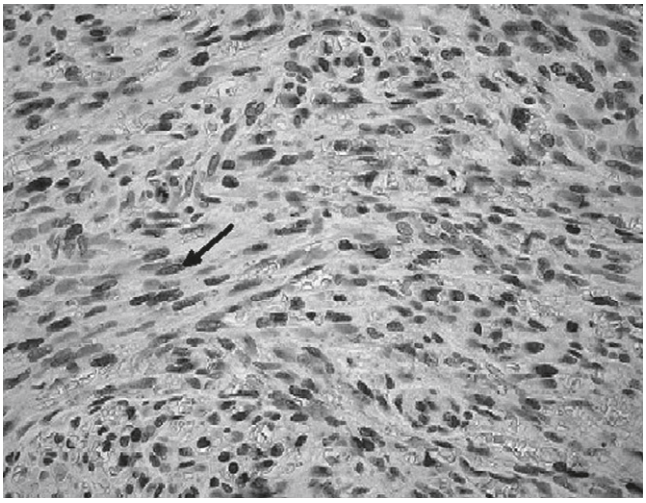

Fig. 4 — Immunohistochemical stain shows human herpes virus-8 in the tumor cell nuclei (arrow) which are stained brown (latent nuclear antigen-1, 400×).

with positive HHV-8 staining in the tumor cell nuclei (Fig. 4). Positive results for serum human immunodeficiency virus (HIV) were revealed on enzyme immunoassay and Western blot. Thereafter, computed tomography scan of the head and neck showed an elevated contrast-enhanced mass in the posterior wall of the nasopharynx with multiple small lymph nodes at the left periauricular, bilateral periparotid and bilateral neck areas, which were compatible with KS and AIDS-related lymphadenopathy (Fig. 2).

The patient was referred to the Department of Infectious Diseases, where antiviral treatment and local radiotherapy were given. Complete response and local symptom relief were noted after radiotherapy with 28 Gy (2 Gy/fraction, 5 times/week). Subsequently, KS was also noted on his chest wall and skin of the scrotum. CO_2 laser treatment was administered for the former and local radiation for the latter. The patient received highly active antiretroviral therapy (HAART) and remained in stable condition as of July 2008.

3. Discussion

KS was first described by Moritz Kaposi, a Hungarian dermatologist practicing at the University of Vienna in 1872. The incidence of KS is 310 times higher

among AIDS patients than in the general population [1]. The first case of epidemic or AIDS-associated KS was reported in 1981 [2]. It is more aggressive in patients with AIDS. KS is now the malignant neoplasm that is most frequently associated with AIDS—it has been seen in up to 80% of AIDS patients in some series, and is more prevalent in male homosexuals than in other HIV risk groups [1,2].

In 1993, Riderer et al reported that 163 (70%) of the 233 HIV-positive patients diagnosed or treated in their otolaryngological department had AIDS [3]. Of these, 46 (28%) had KS in the head and neck region. KS was most often located in the mouth (32/46, 67%), oropharynx (30/46, 65%) and skin (18/46, 39%), while the larynx (5/46, 10.9%), hypopharynx (4/46, 8.7%), lymph nodes (3/46, 6.5%) and nasopharynx (2/46, 4.3%) were rarely involved. In 40 of the 46 KS patients (87%), more than one anatomical site of the head and neck was involved. In 15 of the 233 HIV-infected patients (6.4%), KS of the head and neck region was the initial symptom of HIV infection. Unifocal KS in the nasopharynx is very rare and KS in the head and neck area as an initial symptom of HIV-infected patients is also rare [3].

Clinically, KS must be differentiated from other nasopharyngeal angiogenic tumors such as angiofibroma. The latter is a lobulated, firm, nonencapsulated, pink-gray mass arising from the fibrovascular stroma in the posterolateral wall of the roof of the nose; a punch biopsy is not done for this tumor because of the possibility of massive bleeding. In contrast, KS presents grossly as small well-delineated plaque lesions or larger infiltrative nodules arising from the posterolateral wall of the nasopharynx; a punch biopsy can be done for mucosal and skin KS for definitive diagnosis without causing intensive bleeding. Still, if any hypervascular tumor is suspected, an endoscopy-guided biopsy should be done in the operating room to see the tumor clearly, with facilities well-prepared to control possible bleeding [3].

KS is not considered curable with standard therapies, and treatment decisions are instead guided by the presence and extent of symptomatic or extr acutaneous KS [6]. It is now accepted that most, if not all, patients with KS should be treated with antiretroviral drugs such as the HAART regimen. The benefits of HAART include inhibition of HIV replication, diminished production of Tat protein, amelioration of the host's immune response to HHV-8, and direct antiangiogenic activity by some protease inhibitors. HAART taken together with systemic chemotherapy such as liposomal doxorubicin, daunorubicin, or paclitaxel may be used to treat advanced or progressive disease [6,7].

Local therapy is most useful for localized KS lesions for local symptomatic control and/or cosmesis, but will not prevent the development of new lesions at untreated sites. Alitretinoin gel 0.1% is the only topical patient-administered therapy approved for the treatment of KS, but it is only suitable for skin KS [6,7]. Alternative local treatments include intralesional chemotherapy using vinblastine, radiation therapy, laser therapy, and cryotherapy. Low-dose radiation therapy can be employed to palliate symptoms [5]. Complete response to radiation therapy is found in 50–80% of patients. Antiangiogenesis therapy is a new modality that is currently being developed [6,7].

In summary, AIDS-related KS varies from minimal to fulminant and from unifocal to multifocal disease. Head and neck presentations and diseases in HIV-infected patients are new challenges for otolaryngologists in the diagnosis and treatment of these conditions [3,6,7].

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