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SPECIAL REVIEW

Marathon of eponyms: 11 Kaposi sarcoma

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The use of eponyms has long been contentious, but many remain in common use, as discussed elsewhere (Editorial: Oral Diseases. 2009: 15; 185). The use of eponyms in diseases of the head and neck is found mainly in specialties dealing with medically compromised individuals (paediatric dentistry, special care dentistry, oral and maxillofacial medicine, oral and maxillofacial pathology, oral and maxillofacial radiology and oral and maxillofacial surgery) and particularly by hospital-centred practitioners. This series has selected some of the more recognized relevant eponymous conditions and presents them alphabetically. The information is based largely on data available from MEDLINE and a number of internet websites as noted below: the authors would welcome any corrections. This document summarizes data about Kaposi sarcoma.

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Also known as

Kaposi disease
Kaposi–Spiegler sarcomatosis

The condition

Kaposi sarcoma (KS) is a malignant neoplasm of endothelial cells, a multifocal tumour characterized by deregulated angiogenesis, proliferation of spindle cells and extravasation of inflammatory cells and erythrocytes. Varieties of KS include:

- *Classic KS* – the original type of KS described, a relatively indolent disease affecting elderly men of Eastern European descent or from countries bordering the Mediterranean basin, where there is a higher rate of Kaposi's sarcoma-associated herpes virus (KSHV-human herpesvirus-8) infection than in the remainder of Europe (see below).
- *Endemic KS* – described later in young people originating mainly from sub-Saharan countries in Africa, and a more aggressive disease which infiltrates the skin extensively, especially on the lower limbs. These populations frequently have greater than 50% rate of KSHV infection.
- *Transplant Related KS* – recognized mainly since the 1980s. KS arising either when a KSHV-infected organ was transplanted into a person not previously exposed to the virus or when the recipient already harboured KSHV infection.
- *Epidemic KS* – first recognized during the 1980s as an aggressive disease in patients with AIDS. In this case, KSHV is usually sexually transmitted. KS is over 300 times more common in AIDS patients than in renal transplant recipients. In 1994, a virus isolated from a KS lesion revealed it to be the eighth human herpes virus (HHV-8) – now known as KSHV.

KSHV is responsible for all types of KS and, in addition, is also present in some B cell neoplasms, including primary effusion lymphoma and multicentric Castleman disease. Infection is thought to be lifelong, so that persons infected with KSHV may develop KS years later if they develop AIDS or become otherwise immunocompromised.

The virus encodes genes that stimulate cellular proliferation and migration, prevents apoptosis and counter the host immune response. KSHV is a unique virus that incorporates into its genome, cellular genes that cause tumours (molecular piracy); these genes may help KSHV evade the immune system but, in doing so, it also causes cells to proliferate.

Like other tumour viruses, KSHV infection only leads to cancer in a minority of infected persons. Other factors are required for progression into cancer. Infection of endothelial cells with KSHV leads to rapid suppression of TLR4 (Toll-like Receptor) expression, a mechanism of immune escape as TLR4 mediates innate immunity against KSHV. HIV-infected individuals carrying a mutant TLR4 allele appear more likely to develop Castleman disease. Activation of the interleukin-6 receptor signalling pathway and constitutive signalling of viral G protein-coupled receptor play an important role in the activation, proliferation and transformation

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of KSHV-infected endothelial cells. HIV-tat protein, HIV-induced immune suppression and a hyper-inflammatory state facilitate the oncogenic activity of KSHV.

Kaposi sarcoma is generally not considered a true sarcoma, but rather a cancer of lymphatic endothelium which forms vascular channels that fill with blood cells, giving the tumour its characteristic bruise-like appearance. Endothelial cells harbour the KSHV genome, and are thought to be the precursors of the KS spindle cells which become the predominant cell type in plaque- and nodular-stage KS lesions. KS lesions contain KSHV proteins (latency-associated nuclear antigen) uniformly detectable in the KS cancer cells.

Kaposi sarcoma lesions are macules or nodules that may be red, purple, brown or black, typically found on the skin, but spread elsewhere is common, especially to the respiratory tract, gastrointestinal tract and the mouth. Usually oral lesions are part of more widespread disease but, in AIDS, early KS lesions are red, purple or brown macules, later becoming nodular, extending, ulcerating and disseminating. KS typically involves the palate or gingivae. Diagnosis is confirmed by biopsy. Blood tests to detect antibodies against KSHV have been developed and can be used to determine if a patient is at risk of transmitting infection to his or her sexual partner, or if an organ is infected prior to transplantation. Unfortunately, these tests are available mainly as research tools and thus there is little screening for persons at risk of becoming infected with KSHV, such as transplant patients. Epithelioid angiomatosis, haemangiomas, lymphomas and purpura may need to be differentiated.

Management is firstly treatment of the underlying predisposing condition if possible. In 40% or more of patients with AIDS-associated KS, the Kaposi lesions will shrink upon first starting highly active antiretroviral therapy. Patients with a few local lesions can often be treated with local measures, such as radiation therapy. Surgery is generally not recommended as Kaposi sarcoma can appear in wound edges. More widespread disease, or disease affecting internal organs, is generally treated with systemic therapy with alpha interferon, vinca alkaloids systemically or intralesionally, liposomal anthracyclines or paclitaxel.

The person

Moriz Kohn Kaposi was born on 23 October 1837, in Kaposvár, Hungary, and studied Medicine in Vienna, Austria, qualifying in 1861. He was appointed an assistant to Ferdinand von Hebra, the noted Austrian dermatologist, with whom he worked from 1862 to 1867 and married Hebra's daughter. As the Hebra family was Catholic and Kohn was Jewish, Kohn changed his name to Kaposi – a play on the name of his town of birth. Kaposi was habilitated (achieved the highest academic qualification) as Privatdozent of dermatology in 1866 and helped his father-in-law complete von Hebra's dermatology textbook, published in 1872. He described his sarcoma that year.

After Hebra's death, Kaposi was appointed to the Chair in dermatology at the University of Vienna in 1875 and, in 1879, was appointed Director of the skin clinic in Vienna. He was appointed Hofrat in 1899. Kaposi made several other original contributions to clinical dermatology – on aspects of syphilis, dermatitis herpetiformis (originally described by von Hebra), lymphoderma perniciosa, lichen ruber moniliformis, lupus erythematosus, rhinoscleroma and rhinophyma.

Source internet sites (accessed 21 February 2009) and further reading

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