

Kaposi's sarcoma

reprinted from dermnetnz.org, new zealand; author, jane morgan md chb mrcp facshp

Kaposi's sarcoma is a cancer of blood vessels that was considered very rare before the start of the AIDS pandemic. It is now the most frequent cancer to develop in people with AIDS, affecting about 20% overall.

- In the United States, Kaposi's sarcoma is most common amongst HIV-positive men who have sex with men. It occurs less frequently in intravenous drug users and is rare in women, hemophiliacs or their sexual partners.
- In some parts of Africa, Kaposi's sarcoma is more common in HIV positive women.

A milder form of non-HIV associated Kaposi's sarcoma may occur in elderly men (and sometimes women) of Mediterranean and Middle European descent and in men in Sub-Saharan Africa. Kaposi's sarcoma is associated with:

- Infection with human herpes virus 8 (KSHV) in men who have sex with men. It can also occur in heterosexuals. Data is emerging that non-sexual modes of transmission can occur.
- Genetic factors
- Hormonal factors

Kaposi's sarcoma presents as red to purplish spots (macules) and raised bumps (papules and nodules). They are generally first seen on the skin, commonly on legs or feet. They also occur in the mouth. Initially, the lesions are small and painless but they can ulcerate and become painful. Their visible presence may cause considerable anxiety.

Kaposi's sarcoma lesions can also occur internally; in the gut, lungs, genitals and lymphatic system. These internal lesions may cause symptoms e.g. discomfort with swallowing, bleeding, shortness of breath, swollen legs, etc.



Diagnosis

The appearance of Kaposi's sarcoma lesions is often typical but a skin biopsy of a lesion allows a definite diagnosis.



Prognosis

Kaposi's sarcoma has a variable course. Some patients develop only a few minor skin lesions whilst others have much more extensive external and internal disease. The latter lesions may even result in fatal complications. Kaposi's sarcoma is not curable, but it can be treated and its symptoms controlled.



Presentation

Kaposi's sarcoma may develop at any time during the course of HIV infection. Generally, the greater the immunosuppression (e.g. with CD4 cell counts less than 200) the more extensive the Kaposi's sarcoma will be.

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Treatment

If the lesions are not widespread or troublesome, often the best approach is simply to treat the underlying HIV infection with potent anti-HIV drug combinations that suppress HIV replication. These drugs reduce the frequency of Kaposi's sarcoma and may also prevent its progression or the development of new lesions. It is not yet clear why this approach works; one opinion is that the improvement in immune function results in reduced levels of tumor growth-promoting proteins. The choice of more specific treatment depends largely on the extent of the disease.



Treating localized lesions

Small, localized lesions are generally only treated if they are painful or they are causing cosmetic problems. It should be noted that lesions tend to recur after local treatments. Treatments include:

- Freezing with liquid nitrogen (cryotherapy)
- Treatment with radiation
- Surgical removal
- Injection with anti-cancer drugs such as vinblastine
- Topical application of alitretinoin gel (Panretin).
- Treating extensive or internal lesions with systemic therapy

A combination of anti-cancer drugs are given but at lower than usual dosages because of the pre-existing HIV-related immunosuppression.

Other chemotherapy treatments that are used in some centers include paclitaxel and liposomal

forms of the standard anti-cancer drugs, doxorubicin or daunorubicin. "Liposomal" means that the drugs are coated in small fat bubbles, or liposomes which allows better absorption, hopefully resulting in fewer side effects.

Interferon-alpha inhibits some of the growth factors associated with Kaposi's sarcoma but response tends to be better in those with CD4+ cell counts greater than 200.

Clinical trials into a wide range of other therapies are ongoing. Some examples of these are:

- Photodynamic therapy (a combination of a photosensitiser and light energy)
- Cytokine inhibitors
- The pregnancy hormone, human chorionic gonadotropin (HCG); Kaposi's sarcoma lesions disappear in some women when they become pregnant.
- Ganciclovir and foscarnet (antiviral medications) have been recently reported to result in lower rates of Kaposi's sarcoma amongst those being treated for CMV retinitis (inflammation of the retina caused by cytomegalovirus) and are currently being studied. Acyclovir, another antiviral, has been tried, but does not appear to work.

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