

Kaposi's sarcoma (KS)

Summary

A virus that is relatively common among gay and bisexual men called HHV-8 (human herpes virus-8) or KS herpes virus (KSHV) can cause Kaposi's sarcoma (KS).

Kaposi's sarcoma (KS) is a form of cancer. It appears as lesions (abnormal patches or large spots) on the skin or inside the body. KS is relatively common among HIV-positive men who have sex with men (MSM) and can sometimes occur among HIV-negative men. In Canada and other high-income countries KS is rare in women (regardless of HIV status).

Among HIV-negative men who are otherwise healthy, KS may appear as a few isolated skin lesions. In such cases, KS tends to grow slowly and is generally not aggressive.

Among HIV-positive men, KS can cause serious and sometimes life-threatening problems because not only can it affect the skin, but it can grow inside the body and affect the health of internal organs. Fortunately, potent combination anti-HIV therapy (ART) helps to revive the immune system and in many cases this is sufficient to make KS lesions gradually shrink, fade and disappear. In some cases, despite the initiation of ART, KS lesions may not immediately begin to fade or in other cases when KS affects internal organs, your doctor or specialist may recommend additional treatment.

There have also been rare reports of KS skin lesions appearing among HIV-positive men who are taking ART and have high CD4+ counts and an undetectable HIV viral load. In such cases KS is generally not aggressive.

FACT SHEET

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HHV-8 can also be associated with a general inflammatory complication called multicentric Castleman' disease (MCD or more simply, Castleman's disease). Reports suggest that cases of MCD appear to be slowly increasing among HIV-positive men in the present era.

What causes KS?

KS is caused by a herpes virus called human herpes virus-8 (HHV-8) or KSHV (Kaposi sarcoma herpes virus). This virus is relatively common among MSM. Surveys from the U.S. and Western Europe estimate that about 25% of HIV-negative MSM have this virus. Among HIV-positive MSM, HHV-8 is more common, with some surveys finding as many as 38% are co-infected with HHV-8.

Having HHV-8 by itself is usually not sufficient for someone to develop KS. The health of the immune system plays a major role in controlling HHV-8 and in most people KS remains dormant or latent inside infected cells. When the immune system is suppressed (as in the case of untreated HIV infection, or in people who have a transplanted organ, or who receive high doses of corticosteroids), KS lesions can sometimes occur.

How is HHV-8 spread?

Scientists do not understand everything about HHV-8. However, research strongly suggests that HHV-8 appears to be spread via saliva. Therefore wet kissing, oral sex or use of saliva as a lubricant for anal sex between men can all help to spread this virus.

HHV-8 has also been spread by transfusions of contaminated blood and, in the past, by use of contaminated clotting factor used by hemophiliacs.

Patterns of KS

Scientists have found four general patterns or types of KS as follows:

- Classic KS – This type of KS tends to affect otherwise healthy elderly men who live in or who have emigrated from the Mediterranean region. Generally the lesion(s) are slow growing and confined to the skin.
- Endemic KS – This type of KS can occur among young men and women in parts of sub-Saharan Africa. This form of KS is aggressive even in HIV-negative people.
- HIV-related KS – This is the type of KS that is associated with the HIV epidemic. In cases of untreated HIV infection, KS can be aggressive and fatal if left untreated.
- Medicine-induced KS – This type of KS can occur in people who have received organ or tissue transplants and who have to take drugs that weaken their immune system.

KS lesions

KS is a form of cancer that most often affects the skin. HHV-8 causes infected cells to transform and develop a rich network of blood vessels. On the skin, KS causes lesions. These can look like bruises on a person's skin. KS most commonly appears on the feet and legs. KS can also develop inside the body's mucosal tissue, such as the lining of the mouth or the nose; in the lymph nodes; or in internal organs, such as the bowel, lungs, liver or stomach. Whereas other kinds of cancer typically develop in one part of the body and can then spread to others, KS can develop in different parts of the body at once.

Who is at risk for KS?

The immune system tends to keep HHV-8 under control. However, the immune system grows weaker with age and that is perhaps one reason why some older men are susceptible to KS, or KS can appear in people with HIV or those who are taking medicines that weaken the immune system

(such as people with organ transplants or severe inflammatory-related disorders).

There have been cases where people whose HIV was under control were diagnosed with the disease; however, such cases are rare and the KS in people with stronger immune systems tends to be very mild and not pose a serious danger to their health.

Cases of KS have occurred within six months after HIV-positive people have initiated ART. This has most likely happened because prior to instituting ART, the immune system was weak and could not detect or mount a response to HHV-8 or minor KS lesions. Once a person starts to take ART, the immune system becomes stronger. As a result a person's immune system begins to respond to germs, lesions or tumors that it previously did not recognize or attack. These lesions should eventually shrink and fade.

Symptoms

KS on the skin causes lesions that:

- can appear in a wide range of colours—from pink to red to purple on light-skinned people, and from dark purple to brown to black on dark-skinned people
- can range in size from a few millimetres to a few centimetres in diameter
- may be raised or flat
- are usually painless and do not itch

At first, these lesions may be small and may cause no problems except for concern about their appearance. KS lesions may change very little from month to month or they may spread and become raised or lumpy. Lesions on the feet and legs can make it difficult to walk.

KS in the mouth can make it difficult to eat, swallow and speak. However, some people who develop KS in the mouth experience no symptoms at all.

Although KS in the digestive tract usually causes no symptoms, it can cause pain, internal bleeding and blockages.

KS in the lymph nodes can potentially cause swelling of the limbs, face or scrotum.

KS in the lungs can lead to fluid buildup, which may cause coughing and difficulty breathing.

Diagnosis

Some doctors may diagnose KS on the skin by simply examining a person's body. KS skin lesions tend to be flat and, unlike a bruise, when you push on one it does not lose its colour. A biopsy of the skin is required to confirm the diagnosis. A biopsy involves removing a small piece of the affected tissue. That sample is then sent to a lab so it can be examined under a microscope.

To diagnose KS of the internal organs, a thin, flexible tube with a viewing device that allows a doctor to see the inside of organs may be inserted down the esophagus, stomach and small intestine, or into the rectum and colon (this procedure is called an endoscopy); or into the windpipe and lungs (bronchoscopy). During the procedure, samples of tissue may be removed for analysis. KS in the rectum can sometimes be diagnosed by digital exam—a doctor inserts a gloved finger into the rectum to feel for abnormalities. Chest X-rays may be taken to help diagnose KS in the lungs.

There are assays that can detect and measure the amount of HHV-8 in the blood. However, these tests are not standardized and a study with more than 700 people has found that such testing provides little value. Furthermore, the use of these tests is restricted to a research program.

Treatment

After a person is diagnosed with KS, their doctor will advise them to begin taking ART as soon as possible if they aren't already on ART. This will help improve the immune system. As the immune system becomes stronger it will cause the KS lesions to gradually fade and shrink. If there are a lot of lesions, it may take between six and 12 months for all of them to disappear. In cases when there are a few lesions, they may clear up faster because of ART. If ART does not cause the KS lesions to quickly shrink, they will likely cause the symptoms associated with the lesions to improve.

In cases where KS lesions may persist, in addition to ART your doctor may refer you to a dermatologist (a doctor who treats skin conditions) or an oncologist (a doctor who treats cancers) so that you can discuss possible additional options.

There are also various treatments used to treat KS. Whether or not one of these treatments is needed and which type of treatment is most suitable depends on the location, size and extent of the KS. A dermatologist or oncologist can tell you more about possible treatment options.

For KS on the skin, treatments include medicated gels and creams, surgery, radiation therapy, and other procedures used to remove the lesions. If the KS on your skin is widespread, your doctor(s) may recommend anti-cancer drugs (chemotherapy) in addition to anti-HIV drugs. If you have only a few small lesions, you may not require any additional treatment.

KS affecting internal organs can be treated with chemotherapy in addition to using ART.

Side effects of treatments

Local therapy

Local therapies, or therapies that treat a specific area (as opposed to systemic therapy), can cause temporary irritation, discomfort, redness and swelling at the site of treatment. Some local treatments may also leave the area where the lesion was looking noticeably lighter or darker than the rest of the skin. Your specialist can tell you more about possible side effects.

Systemic therapy

Therapy that treats your whole body rather than just one or two spots is called systemic therapy. Commonly used systemic treatment for KS include drugs (chemotherapy) packed into tiny balls of fat called liposomes. Liposomal therapy is generally more effective and associated with fewer side effects compared to standard formulations of the same drug. The class or group of drugs often used

for chemotherapy of KS is called anthracyclines. The liposomal forms of these drugs are as follows:

- DaunoXome (liposomal doxorubicin)
- Caelyx, Doxil (pegylated liposomal doxorubicin)

Side effects of chemotherapy are usually temporary and may include the following:

- hair loss
- nausea
- vomiting
- diarrhea
- chills and/or fever

Speak to your doctor or nurse if these occur.

Prevention

If you are HIV positive, the best way to prevent KS is to take ART to keep your immune system strong. There are currently no vaccines available to prevent a person from getting HHV-8, the virus associated with KS.

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Disclaimer

Decisions about particular medical treatments should always be made in consultation with a qualified medical practitioner knowledgeable about HIV- and hepatitis C-related illness and the treatments in question.

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