Kaposi's Sarcoma, AIDS Associated

Kaposi's sarcoma is a cancer of connective tissue. It is caused by a virus called human herpes virus 8. The tumors of Kaposi's sarcoma appear as red or purple patches on the skin, mouth, lungs, liver and gastrointestinal tract. This tumor was discovered years ago. It was thought to be harmless until it became the most common cancer in HIV infected patients.

Four types of Kaposi's sarcoma are known:

- Classic Kaposi's sarcoma: A rare, slow growing skin tumor. It usually affects males of Italian or Eastern European Jewish ancestry.
- African Kaposi's sarcoma: Though extremely rare in the rest of the world, Kaposi's sarcoma occurs more frequently among young males in certain countries in equatorial Africa. Usually it is a slow-growing tumor. In some cases, the skin tumors can be aggressive, invading bone and tissue under the skin.
- Immunosuppressive-treatment-related Kaposi's sarcoma: People who are taking immunosuppressive drugs after an organ transplant may develop this form of Kaposi's sarcoma. It sometimes improves if the immunosuppressive medication is reduced or changed.
- AIDS-related Kaposi's sarcoma: This disease affects people with late-stage HIV/AIDS. It usually appears as a rapidly progressing tumor. It affects the skin, lymph nodes, gastrointestinal tract, lungs, liver and spleen. The incidence of this tumor is decreasing with the advent of highly active retroviral therapy for HIV/AIDS.

SYMPTOMS

The first symptoms of Kaposi's sarcoma are usually red, purple or brown patches, plaques, or nodules on the skin, often with a bruise-like appearance. In the classic African and immunosuppressive forms of Kaposi's sarcoma, these lesions usually grow slowly and develop over years. As the disease gets worse, the lower legs may swell. In some cases, the disease will spread to other organs.

In the AIDS-related form, the tumor is rapid growing. It often covers larger areas and forms tumor like masses. These tumors start out soft and spongy, but over time usually become hard and solid. The surface of the tumor may form open ulcers which may become infected. The disease rarely is confined to the skin. It often affects the mouth, lymph nodes, lungs, liver, spleen, and gastrointestinal tract. When the tumor involves the lungs, it often causes coughing, shortness of breath, and wheezing. In the lungs, the disease often progresses rapidly. It can result in death from respiratory failure. However, when the disease involves the bowel, it rarely causes problems unless the disease is very advanced. In that case, people may develop symptoms of intestinal obstruction (nausea, vomiting and abdominal pain) or bloody stools. If the lymph nodes become involved, severe swelling of the legs or face can occur.

DIAGNOSIS

- If your caregiver suspects Kaposi's sarcoma, he or she will ask you if you are HIV positive or if you have another disease that may be suppressing your immune system. If you do not know if you are infected with HIV, but have HIV risk factors, your doctor will recommend an HIV test.
- If you are HIV positive, your doctor will ask you many questions about your experience with this disease:
  - When were you diagnosed?
  - What medications have you taken or are you currently taking?
  - What other illnesses have you had that are related to the HIV virus?
  - What are the results of your most recent lab tests (such as CD4 count and viral load)?
- Kaposi's sarcoma can be confirmed with a biopsy. In this test, a small piece of tissue is removed and examined under a microscope.
- If you have Kaposi's sarcoma, your caregiver will try to determine how far it has spread by asking you several questions and by a physical exam.

Are you coughing or short of breath? This could indicate that the cancer has reached the lungs.

Are your legs swelling? This suggests the cancer has reached lymph nodes.
Do you have nausea, vomiting, abdominal pain or blood in your stool? This suggests the cancer is affecting the gastrointestinal tract.

EXPECTED DURATION

There is no cure for Kaposi's sarcoma. It is a lifelong condition. However, the symptoms will improve with treatment.

PREVENTION

There is no way to prevent the milder forms of Kaposi's sarcoma (classic, African, and immunosuppression related). The most effective way to avoid developing Kaposi's sarcoma is to prevent the transmission of HIV. People who are HIV positive can decrease their risk of Kaposi's sarcoma by taking highly active antiretroviral therapy (HAART).

TREATMENT

The goal of treatment is to ease symptoms, shrink the tumor, and prevent the disease from getting worse. The type of treatment recommended depends on the type of Kaposi's sarcoma, the size of the tumor, the extent of internal organ involvement, and the person's CD4 count and general medical condition. Treatments include:

- Surgery-Removing the cancer cells.
- Cryotherapy- Freezing the cancer cells to kill them.
- Chemotherapy-Using drugs to kill the cancer cells.
- Radiation Therapy-Using radiation to kill the cancer cells
- Biological Therapy-Using the body's immune system to fight the cancer cells.

In people with AIDS, the lesions will improve with treatment of the AIDS itself. Typically, this involves antiretroviral medications prescribed to improve the functioning of the immune system.

WHEN TO CALL A PROFESSIONAL

See a caregiver if you have skin changes that fit the description of Kaposi's sarcoma. If you are HIV-positive and have Kaposi's sarcoma, you will need immediate treatment. Your caregiver may need to change or add medications to treat the HIV.
If you have not been tested for HIV, and you think you may have Kaposi's sarcoma, it is important to talk to your caregiver right away. He or she will counsel you about risk factors for HIV and ask if you have symptoms suggestive of HIV/AIDS. These include swollen lymph nodes, night sweats, fevers, fatigue, and weight loss. If you are at risk of HIV, a blood test for the AIDS virus may be recommended. Early diagnosis is essential in HIV disease. The prognosis improves dramatically with treatment.

PROGNOSIS

The outlook for Kaposi's sarcoma depends on the form of the disease. Milder forms are rare and develop slowly. People with classic Kaposi's sarcoma usually die of other causes or develop a second type of cancer. This occurs in up to one-third of people with classic Kaposi's sarcoma. Immunosuppression related Kaposi's sarcoma often improves with a decrease or change in drug doses.
The most important factor in the prognosis for AIDS-related Kaposi's sarcoma is the level of immune-system impairment. The development of Kaposi's sarcoma suggests that the immune system is highly impaired. People with this disease usually die of a different HIV-related infection. People who take highly active antiretroviral therapy (HAART) have a better prognosis.

ADDITIONAL INFORMATION

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