Kaposi Sarcoma

Kaposi Sarcoma (KS) is a low grade, malignant vascular neoplasm that develops from endothelial cells. Infection with human herpesvirus 8 precedes the development of Kaposi sarcoma, and patients who are immunocompromised or HIV-positive, particularly CD4+ counts below 500, are at higher risk of developing this disease. Clinically, KS presents as multifocal purple lesions of the skin, mucous membranes, and/or visceral organs. On microscopy, spindle shaped cells and lymphocytic infiltrate may be seen. Management can include observation, chemotherapy, and treatment of underlying disorders.



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Characteristics

Malignant Vascular Tumor

Malignant-man with Vessels

Kaposi Sarcoma is a low grade, malignant vascular neoplasm that develops from endothelial cells of the lymph or blood vessels. In this condition, infection of the endothelial cells inactivates their RB1 and p53 tumor suppressor genes. Transcription of these genes normally helps keep cell division under control or directs apoptosis. When inactivated, the cell is inappropriately allowed to survive and divide.

Human Herpesvirus Type 8 (HHV-8)

Herpes-harp with (8) Ball

Human herpesvirus 8, also called Kaposi Sarcoma-Associated Herpesvirus (KSHV), is an oncogenic virus transmitted through sexual contact. Infection with HHV-8 precedes the development of Kaposi Sarcoma, although the precise pathogenesis remains unclear. Body cavity based lymphoma, also known as primary effusion lymphoma or PEL, and Castleman's disease, have also been linked to this virus. The onset of disease typically occurs several years after acquisition of infection.

AIDS/Immunocompromised

AIDS Bandaid and Moon-Suppressed

The most common type of Kaposi Sarcoma in the United States is epidemic, or AIDS-associated Kaposi Sarcoma. This type of Kaposi Sarcoma develops in people who are infected with human immunodeficiency virus. Kaposi Sarcoma is considered an AIDS defining illness (when an HIV-positive patient develops Kaposi Sarcoma, that person officially has AIDS). When Kaposi Sarcoma develops in people whose immune systems have been suppressed after an organ transplant, it is called iatrogenic, or transplant-related Kaposi Sarcoma. Most transplant patients need to take medications to suppress their immune system and prevent it from rejecting (attacking) the new organ. But by weakening the body’s immune system, these drugs increase the chance that someone infected with HHV-8 will develop Kaposi Sarcoma.

CD4+ < 500

CD with (4) Fork Less-than (500) Race-car

CD4+ cells are a type of white blood cell. Also known as "helper T cells", these lymphocytes normally circulate in the blood and help coordinate the immune response by stimulating other immune cells. Human immunodeficiency virus (HIV) weakens the immune system by destroying CD4+ T cells. CD4 cell count is an indicator of immune function in patients living with HIV. When HIV patients have CD4+ counts that drop below 500 cells/µL, they have an increased risk of developing Kaposi sarcoma.

Presentation

Multifocal Purple Lesions

Purple Leeches with Focal Magnifying Glass

Skin lesions of Kaposi's sarcoma may vary, but most patients have nontender, hyperpigmented, macular or nodular skin lesions. These skin lesions appear mainly in the extremities, but may appear elsewhere as well. Lesion pattern may vary by type of Kaposi Sarcoma. In HIV-associated Kaposi Sarcoma, nodules tend to appear on the upper body, head and neck. In Classic Kaposi Sarcoma hands and feet are usually first affected, and lesions may extend up the limbs. Iatrogenic (transplant-associated, or immunosuppression related) Kaposi Sarcoma has a similar pattern as Classic type, however mucosal involvement may be more extensive. Finally, in Endemic (African) Kaposi Sarcoma, lymphadenopathy tends to occur in mouth and face or in inguino-genital regions.

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Skin

Skin-Suit-Man

Kaposi sarcoma most commonly affects the skin, mouth, GI tract, and respiratory tract; although manifestations vary widely. Most patients have lesions along skin tension lines.

Mucous Membranes

Mucus

In patients with suspected Kaposi sarcoma, you should examine the pharynx for lesions, especially the palate. Oral lesions occur in approximately one-third of patients and are predictors of pulmonary involvement and less favorable treatment outcomes.

Visceral Organs

Lung and GI-Guy with Visceral-Visor

Kaposi sarcoma most commonly affects the skin, mouth, GI tract, and respiratory tract. Patients with visceral involvement may be asymptomatic, or manifest with shortness of breath, painless rectal bleeding or melena, and other non-specific pulmonary and gastrointestinal symptoms. Kaposi sarcoma can cause serious problems when the lesions are in the lungs, liver, or digestive tract. Kaposi sarcoma in the digestive tract, for example, can cause bleeding, while tumors in the lungs may cause trouble breathing.

Diagnosis

Spindle-shaped Cells

Spindle Cell

Biopsy of skin lesions should be performed to confirm a diagnosis of Kaposi Sarcoma. Histological examination of Kaposi Sarcoma reveals predominantly lymphocytic inflammatory infiltrate, which can help distinguish Kaposi Sarcoma from bacillary angiomatosis. Additionally, spindle cell proliferation often occurs. A spindle cell is an elongated, spindle-shaped cell. In Kaposi Sarcoma, the spindle cells infiltrate through the collagen, forming slit-like spaces that may contain erythrocytes.

Management

Observation

Observing Through a Telescope

Sometimes, observation is recommended if a person's immune system is functioning well, and the KS lesions are small and not bothersome to the person.

Chemotherapy

Chemo-Head-Wrap

Chemotherapy (chemo) is the use of drugs to treat cancer. This is a type of systemic treatment for Kaposi sarcoma. Liposomal anthracyclines or paclitaxel are the drugs most often used for chemotherapy. Liposomal anthracyclines, such as Pegylated liposomal doxorubicin, are medications in which the drug molecule is packaged in a liposome made of various lipids (the drugs are enclosed in tiny fat globules). Another systemic treatment option are immunomodulating agents; although their effect on the immune system isn’t entirely clear. Three immunomodulating agents can be used to treat Kaposi sarcoma (KS); thalidomide, pomalidomide (Pomalyst), and lenalidomide (Revlimid). A local treatment such as radiation therapy, cryosurgery, surgical resection, intralesional chemotherapy, or a topical retinoid, may be used when there are just a few lesions in a very visible area (such as the face).

Treat Underlying Disorder

Treating Disorders Under the Table

In patients with AIDS, highly active antiretroviral therapy (HARRT), also known as combination antiretroviral therapy (cART), refers to the combinations of drugs that are used to keep HIV infections under control. HAART may be started to improve the patient's immune fuction and reduce the size of KS lesions. Similarly, in organ-transplant patients whose immune systems are suppressed by drugs, stopping, lowering or changing the drugs may be helpful.

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