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Review article

Consensus of the Brazilian Society of Infectious Diseases and Brazilian Society of Clinical Oncology on the management and treatment of Kaposi's sarcoma

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ABSTRACT

Kaposi's sarcoma is a multifocal vascular lesion of low-grade potential that is most often present in mucocutaneous sites and usually also affects lymph nodes and visceral organs. The condition may manifest through purplish lesions, flat or raised with an irregular shape, gastrointestinal bleeding due to lesions located in the digestive system, and dyspnea and

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hemoptysis associated with pulmonary lesions. In the early 1980s, the appearance of several cases of Kaposi's sarcoma in homosexual men was the first alarm about a newly identified epidemic, acquired immunodeficiency syndrome. In 1994, it was finally demonstrated that the presence of a herpes virus associated with Kaposi's sarcoma called HHV-8 or Kaposi's sarcoma herpes virus and its genetic sequence was rapidly deciphered. The prevalence of this virus is very high (about 50%) in some African populations, but stands between 2% and 8% for the entire world population. Kaposi's sarcoma only develops when the immune system is depressed, as in acquired immunodeficiency syndrome, which appears to be associated with a specific variant of the Kaposi's sarcoma herpes virus.

There are no treatment guidelines for Kaposi's sarcoma established in Brazil, and thus the Brazilian Society of Clinical Oncology and the Brazilian Society of Infectious Diseases developed the treatment consensus presented here.

Introduction

General aspects of Kaposi's sarcoma

Kaposi's sarcoma is a multifocal vascular lesion of low-grade potential that is most often present in mucocutaneous sites and usually also affects lymph nodes and visceral organs.[1] Kaposi's sarcoma was first described in 1872 by Hungarian dermatologist Moritz Kaposi. From that time to the identification of human immunodeficiency virus (HIV) associated with acquired immunodeficiency syndrome (AIDS), Kaposi's sarcoma remained a rare tumor. While most of the cases identified in Europe and in North America occurred in elderly men of Italian descent or Eastern European Jews, the neoplasia also occurs in several other different populations: young black African men, children in pre-adolescence, receivers of allergenic renal transplant and other patients treated with immunosuppressive therapy. The disseminated and fulminant form of Kaposi's sarcoma associated with AIDS is referred to as epidemic Kaposi's sarcoma to distinguish it from the classical, African and transplant-related forms. In addition, Kaposi's sarcoma was identified in homosexual men without HIV virus.[2,3]

Although the histopathology of different types of Kaposi's tumors is essentially identical among the various affected groups, the clinical manifestations and course of the disease differ dramatically.[2] A key to understanding the pathogenesis of Kaposi's sarcoma was the discovery in 1994 of a gamma herpes virus, human herpes virus type 8 (HHV-8), also known as herpes virus of Kaposi's sarcoma.[4] HHV-8 has been identified in tissue biopsies of Kaposi's sarcoma of virtually all patients with different forms of the disease (classical, African, transplant-related and AIDS-associated), but was absent in the tissue not involved by the neoplasia.[2]

Considered a rare disease, Kaposi's sarcoma in its classical form occurs more often in males, with a ratio of about 10–15 men for every woman affected. Among Americans and Europeans, the usual age of onset is between 50 and 70 years of age.[2]

In the 1950s, Kaposi's sarcoma was recognized as a relatively common endemic neoplasia in native populations of equatorial Africa, comprising about 9% of all cancers seen in males in Uganda. In Africa, indolent or locally more aggressive forms of Kaposi's sarcoma occur at a man/woman ratio comparable to that observed for the classical tumor seen in North America and Europe. However, patients in Africa are significantly younger than European patients. A lymphadenopathic form is also seen in Africa, primarily in children in preadolescence, at a male/female ratio of 3 cases to 1,[2,5] and mortality rate of nearly 100% in 3 years.[5,6]

In 1969, the first case of Kaposi's sarcoma associated with immunosuppressive therapy in a patient with renal transplantation was described. Since then, it has been observed that several patients receiving renal transplants and other allergenic transplants who were treated with prednisone and azathioprine developed Kaposi's sarcoma shortly after initiation of immunosuppressive therapy.[2,7] Estimates of the incidence of Kaposi's sarcoma among renal transplant recipients subjected to immunosuppressive therapy are between 150 and 200 times higher than the expected incidence of the tumor in the general population. The average time to develop Kaposi's sarcoma after transplantation is 16 months.[2]

Epidemiological aspects of epidemic Kaposi's sarcoma

In 1981, a disseminated and fulminant form of Kaposi's sarcoma was described in homosexual or bisexual men and was first reported as part of an epidemic now known as AIDS.[8] The etiology of AIDS is a retrovirus with tropism for T lymphocytes known as HIV.[9] The immune deficiency that characterizes AIDS is a profound disorder of cell-mediated immune functions. This immune dysfunction and deregulation of the immune system predispose patients to the development of a wide range of opportunistic infections and unusual neoplasm such as Kaposi's sarcoma. HIV can play an indirect role in the development of Kaposi's sarcoma.[9] Approximately 95% of all cases of epidemic Kaposi's sarcoma in the United States were diagnosed in homosexual or bisexual men. In the past, approximately 26% of all male homosexuals with HIV presented with or developed Kaposi's sarcoma over the course of AIDS. As a comparison, less than 3% of all heterosexual injection drug users with HIV developed Kaposi's sarcoma. The proportion of AIDS patients with Kaposi's sarcoma has declined dramatically since the outbreak of the disease was identified in 1981.[10] About 48% of patients diagnosed with AIDS in 1981 presented with Kaposi's sarcoma at diagnosis. By August 1987, this proportion had declined to less than 20%. The introduction of highly active antiretroviral therapy (HAART) delayed or