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# Fatal Outcome in a Patient with Kaposi's sarcoma: Case Report and Literature Review

Andrea Guadalupe Navarro Ortega<sup>1</sup>, Eder Fernando Rios Bracamontes<sup>2</sup>, Javier Pérez Gutierrez<sup>3</sup>, Diana Itzel Tene Corona<sup>4</sup>, Ivanna Ocampo Carrasco<sup>5</sup>

<sup>1</sup>Internal Medicine Resident
<sup>2</sup>Doctor of Medical Sciences, Professor of Internal Medicine
<sup>3</sup>Dermatologist, Dermatology Department
<sup>4</sup>Internal Medicine Resident
<sup>5</sup>Internal Medicine Resident Hospital General de Zona No. 1, Instituto Mexicano del Seguro Social

### ABSTRACT

Kaposi sarcoma (KS) is an angioproliferative disorder that is characterized by the appearance of purplish, reddish blue, or dark brown macules, plaques, and nodules on the skin, it can range in size from very small to several centimeters. In the etiopathogenesis, Human Herpesvirus 8 infection and immunosuppression has a fundamental role. The clinical evolution is usually chronic and indolent. We present a case in a patient with Human Immunodeficiency Virus (HIV) infection and complicated Kaposi sarcoma that caused his death.

#### ARTICLE DETAILS

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**KEYWORDS:** Kaposi sarcoma, immunosuppression, Human herpesvirus 8

#### **INTRODUCTION**

KS was first described in 1872 by Moritz Kaposi as "idiopathic multiple pigmented sarcoma of the skin." Initially, it was characterized as an idiopathic multiple hemangiosarcoma with a tendency for chronic evolution, more common in men over 40 years of age, with visceral involvement and vascular origin. These characteristics define classic KS (1).

The etiopathogenesis of KS involves Human Herpesvirus 8 (HHV-8), with infection being necessary for the development of the neoplasm. However, not all infected individuals will develop KS. Another significant risk factor is immunosuppression, whether iatrogenic or acquired (1).

The diagnosis of KS begins with the identification of classic lesions (purplish, brown, or dark macules, plaques, or nodules) and their topographic distribution, which most frequently occurs in the lower extremities. Diagnostic confirmation is made through histopathological characteristics and immunohistochemical staining in the biopsy (2).

This article describes the case of a patient diagnosed with HIV infection and KS with a fatal outcome secondary to complications from the neoplasm.

### CLINICAL CASE

A 44-year-old man with a 20-year history of HIV/Acquired Immunodeficiency Syndrome (AIDS), initially diagnosed due to the presence of purplish macules on the lower extremities typical of KS. At that time, antiretroviral therapy (ART) was initiated, with clinical improvement. However, the patient showed poor therapeutic adherence, intermittently abandoning treatment for at least 7 years, leading to the recurrence of the dermatosis.

On this occasion, he presented to the emergency room due to pain and purulent discharge from lesions on both lower extremities. He was found in septic shock, with acute kidney injury, a viral load of 80.7 copies/ml, and CD4 lymphocytes of 12 cells/ul. Physical examination revealed disseminated dermatosis characterized by multiple purplish macules that did not disappear with pressure, bilaterally distributed on the thighs, perineum, pubis, and lateral abdominal region, with indurated consistency, apparently infiltrated, with multiple scales and yellowish exudate (*figure 1*). On the lower extremities, ulcerated nodules with purulent discharge, scales, as well as color changes, thickening, and nail dystrophy were observed (*figure 2*).

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Initially, he was managed with vasopressor support, broadspectrum antibiotics, and antifungals. Surgical debridement was performed with the placement of a negative pressure wound closure system (VAC). However, he continued to deteriorate clinically, requiring bilateral supracondylar amputation. Following this intervention, he showed clinical improvement, allowing for the withdrawal of vasopressors.

Histopathological study reported a lesion compatible with ulcerated KS with added fungal infection and vascular infiltration, immunohistochemistry positive for HHV-8 (figure 3). Antibiotic therapy was completed with meropenem/vancomycin, antifungal therapy with fluconazole, antiretroviral therapy was restarted with emtricitabine, tenofovir alafenamide, and bictegravir, along with valganciclovir, and prophylaxis with trimethoprim/sulfamethoxazole was initiated. However, he experienced clinical deterioration with dyspnea and increased oxygen requirements, necessitating invasive mechanical ventilation, leading to a poor outcome and his death.

### DISCUSSION

The presented case highlights several critical considerations in the management of this disease in people living with HIV/AIDS. Firstly, it is observed that KS remains a significant complication in HIV patients, especially in those with low CD4 lymphocyte counts, as documented in other studies. This case underscores the importance of early diagnosis and timely antiretroviral treatment to reduce the risk of HIV-associated cancers and other opportunistic infections.

Additionally, the clinical presentation of KS in this patient, which included cutaneous and visceral involvement, is consistent with previous findings indicating that KS can manifest aggressively in immunocompromised patients. The reviewed literature also suggests that co-infection with HHV-8 is a crucial factor in the pathogenesis of KS, which was evident in this case.

Another relevant aspect is the variability in treatment response. Despite advances in ART and treatment options for KS, some patients continue to have unfavorable outcomes, indicating the need for more effective and personalized therapeutic strategies. This case also highlights the importance of continuous monitoring and comprehensive management of HIV/AIDS patients to improve clinical outcomes.

The patient initially presented with purplish macules on the extremities typical of KS, which later evolved to disseminate, adding ulcerated nodular lesions with fungal superinfection. It is well known that this neoplasm is characterized by macules, plaques, and nodules of purplish, reddish-blue, or dark brown coloration on the skin, ranging in size from very small to several centimeters, which can remain stable without changes or with accelerated growth in a few weeks and disseminate. Additionally, nodular lesions can ulcerate and bleed easily (2).

The definitive diagnosis was made through biopsy. It was reported as an infiltrating proliferation of vascular spaces in the form of clefts in the superficial and deep dermis accompanied by erythrocyte extravasation, with spindleshaped endothelial cells with minimal atypia, scant cytoplasm, and hyperchromatic spindle-shaped nuclei, surrounded by a lymphoplasmacytic inflammatory infiltrate and foci of hemosiderin. All these changes are consistent with what is reported in the literature, being highly suggestive of KS, in addition to the positive immunohistochemistry result for HHV-8.

Histology can identify multiple characteristics such as proliferation of small, irregular, and jagged spaces lined by endothelium surrounding normal dermal vessels, accompanied by an infiltrate of plasma cells and lymphocytes. In the plaque stage, spindle cell proliferation is observed throughout the dermis. While in the nodular stage, it is characterized by fascicles of spindle cells with frequent cytological atypia, often accompanied by a chronic inflammatory infiltrate composed of lymphocytes, plasma cells, dendritic cells, and a network of vascular spaces in the form of clefts (3).

The clinical course is usually chronic and indolent and rarely influences survival, with other causes of death unrelated. It was reported in a series of 438 American patients with classic KS who were followed for an average of 4.8 years, 24 percent died from second malignancies, 22 percent died from other medical conditions, and only 2 percent died from generalized disease (3).

In this case, the patient's cause of death was directly related to KS, highlighting that he had poor adherence to ART, in addition to not having adequate medical follow-up, which led to progressive clinical deterioration over 20 years, resulting in fungal superinfection of the lesions, leading to septic shock and ultimately his death.

Treatment should be individualized, according to the characteristics of the lesions, but the fundamental pillar is based on ART (4). It has been observed in multiple cohort studies that the use of ART is associated with a decrease in incidence in patients with HIV infection, as well as a reduction in the risk of opportunistic infections with increased survival (5).

There is no consensus on the optimal therapy, however, there are various therapeutic options such as radio or chemotherapy, cryotherapy, laser, intralesional or topical therapy, and in some cases, surgery may be necessary, especially when dealing with a single highly symptomatic lesion (2).

In the therapeutic management, antibiotics and antifungals were administered, along with the restart of ART. Bilateral amputation was the last resort to attempt to stop the state of shock, obtaining a favorable response temporarily, however, due to the general condition of the patient, it resulted in a fatal outcome.

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#### CONCLUSIONS

In conclusion, the reported case highlights several key points: Early detection and immediate initiation of antiretroviral therapy are essential to reduce the incidence of KS and improve the prognosis of HIV/AIDS patients. KS can present aggressively, with cutaneous and visceral involvement, especially in patients with low CD4 lymphocyte counts. Coinfection with HHV-8 plays a crucial role in its development. Despite advances in treatment, some patients do not respond favorably, underscoring the need to develop more effective and personalized therapeutic strategies. A comprehensive approach and constant monitoring of HIV/AIDS patients are essential to improve clinical outcomes and prevent serious complications such as KS, with the aim of improving their quality of life and reducing mortality associated with complications.

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Figure 1. Lesions found during the physical examination of the patient. A) Plaques with a hardened consistency, apparently infiltrated, with multiple scales and yellowish exudate discharge on the thighs and lateral abdominal region. B) Purpuric macular lesions on the upper extremities



Figure 2. Lesions found on the lower extremities.

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Figure 3. Histopathological study of skin from the lower extremities: Epidermis with keratosis and extensive areas of ulceration. In the dermis, vascular clefts with endothelium surrounded by spindle-shaped cells with hyperchromatic nuclei, necrosis, and erythrocytes are observed. These cells invade the endothelium of the vessels. Fungal hyphae and septa are observed. Eight mitoses per high-power field are identified, extending to the deep dermis.