Visceral Kaposi´s Sarcoma as a presentation in a newly diagnosed HIV-Infected man. A case report

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Case Report

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Background: Kaposi's sarcoma is an angioproliferative malignancy due to human herpesvirus-8 and is associated with immunosuppression. Although most cases are cutaneous and resolve with treatment of the underlying condition, few cases present with organ involvement and have a fulminant course. We present a case of a 54-year-old male with newly diagnosed human immunodeficiency virus infection who presented with severe respiratory failure and acute dyspnea. Physical examination revealed generalized violaceous macules over the neck, trunk, extremities, and genitals. Diagnostic workup showed profound bicytopenia, a CD4 count of 29 cells/µL, and a massive bilateral serosanguineous pleural effusion. Histopathological analysis of a skin punch biopsy confirmed proliferation of irregular vascular channels infiltrating the dermis with extravasated erythrocytes, diagnostic of Kaposi's sarcoma. Despite continuous medical optimization, the patient suffered an invasive and rapid respiratory decline, dying on day 16 of hospitalization. Visceral Kaposi's sarcoma remains a severe disease typically associated with a poor prognosis, emphasizing the need to recognize this entity in patients with low CD4 counts.

KEYWORDS: Kaposi's sarcoma, HIV.

Kaposi's sarcoma is an angioproliferative tumor associated with the presence of herpesvirus-8 in more than 90% of the lesions. The mode of transmission of herpesvirus-8 remains unclear, but epidemiologic and virologic data suggest that saliva is a source of the infectious virus and may be an important route of transmission as well as sexual contact. Kaposi's sarcoma was first described in 1872 by Moritz Kaposi, an Austro-Hungarian dermatologist, in five patients with multifocal disease. It is a malignancy of the lymphatic endothelial cells that is often highly aggressive in people with human immunodeficiency virus and severe immunodeficiency. In Kaposi's sarcoma, the majority of lesions begin on the skin, but they can involve virtually any lymph nodes or internal organs and poses a difficulty in diagnosis.

Case report

A 54-year-old single male, working as a journalist, presented to the emergency department. He had a recent diagnosis of human immunodeficiency virus infection confirmed a few months ago after evaluation for chronic diarrhea and an 8 kg weight loss over one month. He was prescribed Biktarvy. The patient sought emergency care due to recent-onset exertional dyspnea, which partially improved in a sitting position, accompanied by a dry cough.

Upon admission, his vital signs were normal. Physical examination revealed generalized dermatosis characterized by violaceous macules distributed over the neck, thorax, upper and lower limbs, and genitals. Also, the patient showed the same dermatosis inside the oral cavity. Additionally, a wet necrotic zone was noted on the lateral border of the right lower extremity, and pulmonary auscultation revealed bilateral rales predominantly in the left regions.

Initial blood work revealed severe bone marrow suppression presenting as bicytopenia, with a white blood cell count of 3.3 x10^3/µL, severe anemia with a hemoglobin level of 5.5 g/dL, and a platelet count of 58,000/µL. His CD4 count was 29 cells/µL and his baseline viral load was 634,000 copies/mL, confirming an advanced immunodeficiency stage.

An axial chest computed tomography scan showed a bilateral pleural effusion of approximately 50%, more prominent on the left side, causing passive compression of the lung parenchyma, multiple basal atelectasis, and widespread patchy opacities. Sequential therapeutic and diagnostic thoracenteses were performed, yielding serosanguineous pleural fluid. The cytochemical analysis of the fluid reported glucose of 60 mg/dL, albumin of 1 g/dL, an elevated lactate dehydrogenase level of 1744 U/L, and uncountable red and white blood cells with 60% polymorphonuclear cells. The patient was managed with intravenous solutions optimized with calcium gluconate, trimethoprim/sulfamethoxazol for

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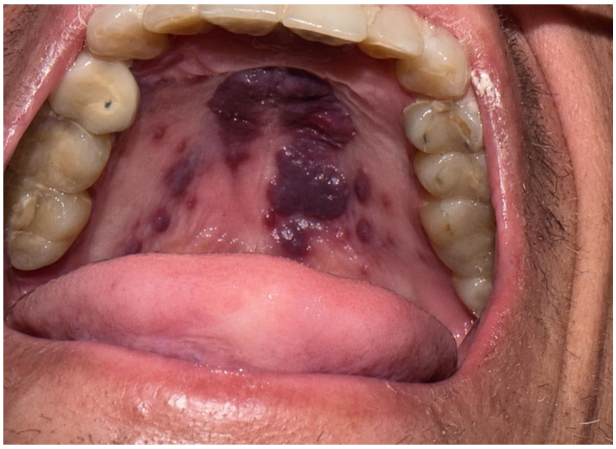

Figure 1. Dark violaceous and blackish elevated, lobulated lesion along the hard palate midline, surrounded by smaller purpuric macules and papules.

suspected opportunistic pneumonia, and fluconazole for fungal prophylaxis. However, the medical team continue working on the etiology of the dermatosis on a suspected Kaposi's sarcoma.

To confirm the diagnosis, a 4 mm punch biopsy was performed on the left femoral region. The microscopic and histopathological diagnostic report revealed a proliferation of irregular vascular channels, partially obliterated, infiltrating the collagenous tissue of the dermis, accompanied by prominent erythrocyte extravasation and frequent hemosiderophages. These findings provided clear histologic data highly suggestive of Kaposi's sarcoma.

The patient experienced a progressive and torpid clinical decline, maintaining severe hypoxemia despite supplementary oxygen at 5 liters per minute via nasal cannula. Due to recurrent accumulation of the hemorrhagic pleural fluid and suspected extensive pulmonary visceral involvement, the short-term prognosis was determined to be very poor. Unfortunately, the patient died after 16 days of hospitalization.

Discussion

1. Definition and epidemiology

Kaposi's sarcoma is an angioproliferative tumor of lymphatic endothelium-derived cells infected with human herpesvirus-8, typically occurring in patients with immunodeficiency. It has variable presentations that range from indolent, isolated dermatologic manifestations to fulminant disease with visceral involvement. In the United States, it is the most common virus-associated tumor, occurring in approximately one-fourth of all patients with acquired immunodeficiency syndrome. While the incidence of epidemic Kaposi's sarcoma decreased drastically in developed countries after the introduction of highly active antiretroviral therapy, the reduction in sub-

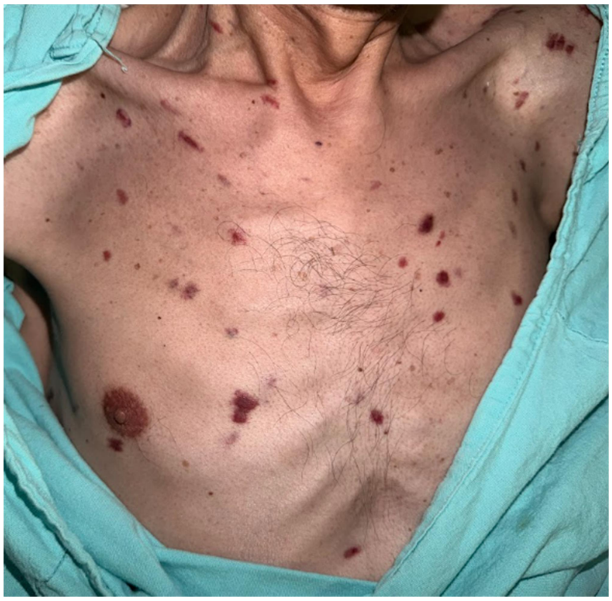

Figure 2. Disseminated, asymmetric dermatosis on the chest region characterized by multiple round to oval wine-red and purpuric macules and papules with well-defined borders.

Saharan Africa has been much lower, where it remains the most common malignancy in men in countries like Uganda and Zimbabwe. There is no information about the prevalence in Latin American countries. The disease demonstrates a strong male predominance, with a male-to-female ratio ranging from 2.5 to 1 up to 9 to 1 across different clinical subsets. Visceral Kaposi's sarcoma can appear in any organ, but it mainly involves the lymph nodes, lungs, and gastrointestinal tract. Although 33% to 77% of patients with cutaneous Kaposi's sarcoma have been found to have visceral lesions on autopsy, such lesions in the absence of skin manifestations are rare.

2. Diagnosis and histopathology

For a definitive diagnosis of Kaposi's sarcoma, a biopsy or excision of the suspicious areas must be performed to look for the characteristic spindle cell proliferation in the dermis. Immunohistochemistry shows expression of CD34, CD31, and D2-40, and the introduction of monoclonal antibodies against the latency-associated nuclear antigen-1 helps confirm human herpesvirus-8 infection. Traditional histopathological staging involves tracking progression from early patch stage, which features the promontory sign where new vessels dissect collagen around pre-existing vascular structures, to the plaque and highly cellular nodular stages. The virus maintains a biphasic lifecycle, where the latent phase allows tumor maintenance through episomal replication, and the lytic phase drives a paracrine oncogenic mechanism. Lytic genes trigger signaling cascades that induce the translation of vascular endothelial growth

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factor and platelet-derived growth factor, promoting angiogenesis and cellular proliferation.

3. Treatment and prognosis

The aim of treatment is usually palliative and primarily targets symptom improvement and prevention of progression. Regarding treatment options, therapy is started according to the severity of the disease on the initial presentation. If the disease is localized or minimally disseminated cutaneous Kaposi's sarcoma, it usually responds to antiretroviral therapy plus either cryotherapy, radiotherapy, or surgical resection of the lesion. Antiretroviral therapy causes resolution of lesions by suppressing viral replication and increasing CD4 cell counts, without acting directly on the herpesvirus. If the disease is disseminated with visceral involvement, a combination of systemic chemotherapy with antiretroviral therapy is typically required. Liposomal anthracyclines like liposomal doxorubicin are considered first-line systemic agents for the treatment of disseminated disease due to their favorable response rates and lower toxicity. Paclitaxel represents an effective alternative, demonstrating high response rates but carries a higher risk of myelosuppression and neurotoxicity. Overall, visceral Kaposi's sarcoma, especially with extensive pulmonary compromise, carries a high lethality and is typically associated with a poor prognosis.

Conclusion

Visceral Kaposi's sarcoma remains a severe disease. Primary therapy is liposomal doxorubicin with antiretroviral therapy, and it is usually associated with a poor prognosis. This case emphasizes the need to recognize and strongly consider visceral Kaposi's sarcoma as a possible cause in any people living with human immunodeficiency virus or acquired immunodeficiency syndrome with low CD4 counts. A computed tomography scan of the abdomen and pelvis can play an important role in the recognition of visceral lesions and lymph nodes and the diagnosis of Kaposi's sarcoma. This case reflects the complexity of human immunodeficiency virus and reinforces the need for greater awareness in screening regardless of whether a clear risk behavior has been identified.

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