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SARCOMA DE KAPOSI CON AFECTACIÓN VISCERAL EN PACIENTE CON VIH: REPORTE DE UN CASO

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ABSTRACT

Kaposi's sarcoma (KS) is a rare multifocal angioproliferative neoplasm of endothelial cells with predominantly cutaneous involvement. It was first described in 1872 by Moritz Kaposi. Four groups of KS have been classically described: classic, epidemic or AIDS-associated, iatrogenic and endemic. KS is the most frequent neoplasm associated with AIDS-associated human immunodeficiency virus (HIV) and occurs most frequently in the cephalic region (eyelids, nose and ears). This article describes the case of a 31-year-old male patient diagnosed with HIV and Kaposi's sarcoma with visceral involvement.

Key words: Kaposi's sarcoma, HIV, AIDS, visceral involvement, human herpes virus type 8.

INTRODUCTION

Kaposi's sarcoma (KS) is a systemic disease whose involvement is predominantly cutaneous, although it can also involve lymph nodes and internal organs (1,2). This pathology occurs due to the proliferation of lymphatic endothelial cells as a consequence of infection by the human herpesvirus 8 (HHV-8) (1). It predominantly affects men, mainly those who have sexual relations with the same gender, and initially manifests as multiple vascular

nodules in the skin and other organs (1,3). It is considered the most frequent neoplasm associated with the human immunodeficiency virus (HIV), severely and aggressively affecting patients with acquired immunodeficiency syndrome (AIDS) (4).

The disease was first described in Vienna by the Hungarian Moritz Kaposi in 1872, under the name "multiple pigmented idiopathic sarcoma of the skin" (5). Currently, four clinical forms have been described: classical KS, endemic or African KS, AIDS-associated KS and iatrogenic/post-transplant KS (1).

The classic form occurs mainly in the Jewish and Mediterranean population, appears around the seventh decade of life and affects mostly men with a ratio of 15 to 1 (3,6). It is mostly located at the level of the lower extremities (3).La forma endémica o africana es más frecuente en hombres, puede cursar de manera indolente o agresiva y presenta lesiones de morfología diversa. (6)

The form associated with AIDS most frequently presents in the cephalic region (eyelids, nose and ears) and the initial manifestation of the disease in 15% of these cases is mucosal involvement, mostly oral (3). Gastrointestinal, pulmonary and lymph node involvement may also occur (6).

In the iatrogenic form there are very few published cases, most are found in relation to pharmacological immunosuppression of rheumatologic diseases and can also occur after transplantation (7). It should be noted that the incidence of KS can increase 100 times in transplanted patients (3).

The classic presentation of KS is a lesion that starts as an erythematous-violaceous macule that can evolve into a plaque or nodules resembling granulation tissue, which can erode, bleed and ulcerate (3,8). The most frequent locations of KS are in the oral mucosa, skin and ganglions and at a visceral level the organs that are affected are mostly the lung, genitals, gastrointestinal tract, oral and nasal cavity, liver, bone marrow and spleen, with predominant involvement at the gastrointestinal and oral level in patients with KS associated with AIDS (3).

The diagnosis is histological, so a biopsy is mandatory, in which it is characteristic that spindle cells are observed, which are identified by having elongated cytoplasm and nucleus and, sometimes, by containing hemosiderin (8-10). It is presumed that these cells derive from transactivation changes in lymphatic and blood vessel endothelial cells (8). However, it should be noted that these cells may be difficult to distinguish in early lesions (9). For the identification and localization of HHV-8 within KS lesional cells, a reliable indicator is used, which is the monoclonal antibody against HHV-8 latent nuclear antigen (LANA), being the most useful immunostaining technique for diagnosis that allows differentiating KS from its simulators (10). Also, differential diagnosis should be established with bacillary angiomatosis, hemangioma, pyogenic granuloma, dermatofibroma (11).

There are multiple treatment options, which vary according to the involvement of the disease and the affectation of the patient, so it must be taken into account if the patient has single lesions or disseminated disease, if there is visceral affectation and the immunological status of the patient (3,12). Among the local treatments are cryotherapy, laser and radiotherapy and among the systemic treatments, chemotherapy stands out (3). In patients with KS associ-

ated with AIDS, the use of antiretroviral therapy has significantly reduced the incidence of this disease (3).

CASE REPORT

31-year-old male patient with a history of HIV B3 infection in second line of treatment for 9 months. Among other important antecedents: Man who has sex with man and had liver and skin lesions which required histopathological study.

She was admitted for a clinical picture of 1 month of evolution characterized by visceral abdominal pain of progressive onset that increased its intensity 1 week ago reaching 8/10 according to the visual analog scale, associated with abdominal distension. In addition, she reports unintentional weight loss of approximately 10 kg in the last 6 months, asthenia, adynamia, occasional vomiting unrelated to food intake.

On physical examination with normal vital parameters, regular musculonutritional conditions, icteric sclerae are found, in oral mucosa shows violet and reddish macules type lesions, flat, not well delimited, painless, involving hard palate (Figures 1 and 2). Decreased sounds in both lung bases, globose abdomen, with positive ascitic wave and changing dullness, painful to superficial and deep palpation in all abdominal quadrants, with palpable hepatomegaly 7 cm below the costal ridge. Asymmetric grade III edema predominantly in the right lower limb and scrotal edema. In addition, with skin lesions like painless, pink and purplish multiform skin spots and plaques involving the entire body anatomy and respecting palms and soles (Figures 3 and 4).

In laboratories provided at admission, the following stand out: biopsy report of oral mucosa and palate (July and August 2021) with findings suggestive of neoplasia of vascular origin, corroborated with immunohistochemistry positive for HHV8 suggesting Kaposi's sarcoma.

Taking into account the history and clinical presentation, complementary studies were requested, documenting severe immunosuppression status given by CD4 counts of 32mm/3, in addition to anemia and jaundice with cholestatic pattern. Hepatomegaly with multiple diffuse periportal hepatic lesions of angiomatous appearance was documented in ultrasound and exploratory laparoscopy. Endoscopic study of the gastrointestinal tract showed global erythematous gastropathy and nodular duodenitis suggestive of lesions secondary to Kaposi's sarcoma, in addition to rectosigmoiditis in colonoscopy.

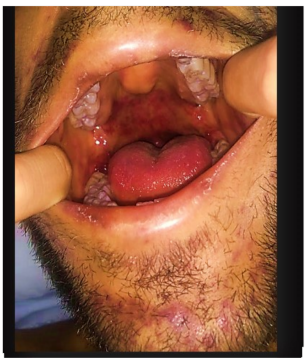
On admission, abdominal pain of etiology to be determined was considered, HIV cholangiopathy was ruled out due to diagnostic images that did not show structural compromise of the biliary tract and Kaposi's sarcoma in oral mucosa was considered by immunohistochemistry study, so in support by oncology and infectology service antiretroviral therapy is continued and opportunistic infection is considered to be ruled out before starting chemotherapy. Co-infection with hepatitis B or C, EBV or CMV, histoplasma, borrelia and varicella is ruled out. Study of non-infectious ascitic fluid with GASA <1.1.

During his stay he presented with clinical deterioration due to pleural and pericardial effusion requiring diagnostic and therapeutic surgical management, with reports of pleural and pericardial studies that ruled out infectious pathology. Histopathological study of pericardium suggests neoplasia of vas-

cular origin with suspicion of Kaposi's sarcoma. Finally, immunohistochemistry of liver biopsy concludes vascular neoplasm HHV8 + consistent with Kaposi's Sarcoma.

It is considered then, a patient with HIV in AIDS stage C3 with generalized Kaposi's Sarcoma Ezinger IV; who having ruled out opportunistic infectious involvement, chemotherapy with Doxorubicin is started. Unfortunately, in spite of timely medical management, he did not have an adequate clinical evolution and died after a few days of hospital stay.

Fig 1 y 2



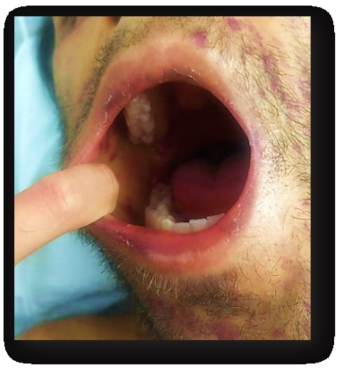
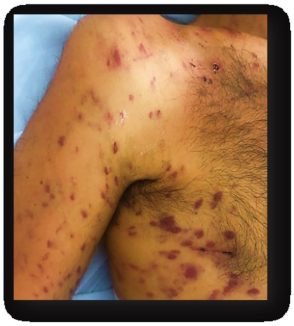
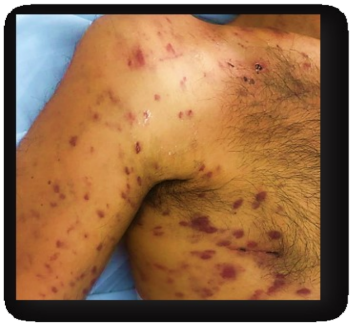


Fig 3 y 4





Paraclinicals

Study	Results
Biopsy of oral mucosa and palate (July and August 2021)	Neoplasm of vascular origin with immunohistochemistry positive for HHV8
CD4	32 mm/3
Esophagogastroduodenoscopy	Global erythematous gastropathy and nodular duodenitis suggestive of lesions secondary to Kaposi's sarcoma.
Colonoscopy.	Rectosigmoiditis.
Ascitic fluid study.	GASA < 1.1.
Hepatitis B o C, VEP y CMV.	Negative.
Immunohistochemistry of hepatic biopsy.	Vascular neoplasm HHV8 +

DISCUSIÓN

The present case allows us to have a broader view of the diagnostic and therapeutic approach to an HIV/AIDS patient in the context of Kaposi's sarcoma. Although since the introduction of antiretroviral therapy the incidence of this etiology has decreased, it is still a challenge for the clinician to know how to identify and approach this disease (13). Therefore, the present case attempts to set a precedent on how to approach an HIV/AIDS patient who consults for abdominal pain associated with skin and mucosal lesions.

The case to be discussed is a patient with Kaposi's sarcoma, a rare multifocal angioproliferative endothelial cell neoplasm caused by Kaposi's sarcoma-related herpesvirus (KSHV) also known as human herpesvirus 8 (HHV-8) (14).

Within the epidemiology of the epidemic type of Kaposi's Sarcoma (associated with HIV/AIDS), which is the one presented by our patient, we found that it is more frequent in young men who have sex with men, in HIV-1 infections and occurs mainly in those who present a decreased CD4 count (10,15,16). It should be noted that AIDS patients are 100,000 times more at risk of developing KS than the general population (17).

Regarding pathogenesis we find lesions characterized by spindle cell proliferation, as well as abnormal vascularity, inflammatory infiltrate and fibrosis (18).

Human herpesvirus 8 (HHV-8) is believed to penetrate endothelial cells by binding to cell surface receptors; this action is followed by induction of a signal transduction cascade that promotes viral entry into the cell and trafficking within the cytoplasm. HHV-8 gene products activate signaling pathways involved in angiogenesis and vascular differentiation (9).

Spindle cells are infected with HHV-8 and express latency-associated nuclear antigen (LANA), a viral protein that binds HHV-8 episomes to chromatin; HHV-8 encodes genes involved in proliferation, cytokine production, and angiogenesis (18).

In the present case, our patient is a young male with a history of HIV under second line antiretroviral therapy who consulted for abdominal pain. Physical examination revealed ascites, hepatosplenomegaly, jaundice, constitutional syndrome and lesions on the skin and oral mucosa. After a complete diagnostic approach, immunohistochemical findings showed hepatic and gastrointestinal tract involvement in relation to the diagnosis of generalized Kaposi's sarcoma enzinger IV, so it was considered to start chemotherapeutic management.

It is important to emphasize that KS should be suspected in patients presenting with non-painful violaceous lesions associated with a state of immunosuppression in order to subsequently biopsy the lesion and thus confirm the diagnostic suspicion.

Regarding the management of our patient the first thing to know is that it is a Kaposi's Sarcoma HIV/AIDS (epidemic) so it can be said that the treatment of choice is antiretroviral therapy (ART) considering the risk of immune reconstitution syndrome (IRIS). Another important pillar of treatment is chemotherapy (19). The recommendation given by the National Comprehensive Cancer Network (NCCN) for HIV/AIDS-associated Kaposi's sarcoma with advanced cutaneous lesions, oral mucosal involvement or visceral involvement is to offer antiretroviral therapy (ART) associated with chemotherapeutic management with liposomal doxorubicin as the first option (20). The prognosis of this disease has improved thanks to antiretroviral therapy (ART); however, our patient presents certain poor prognostic factors such as decreased CD4 count, peripheral edema of the lower limbs, scrotal edema, weight loss and oral lesions that would indicate a high risk of mortality (10).

On the present clinical case we can leave a reflection on how the clinician should be attentive to HIV+ patients who present mucocutaneous manifestations and know that KS is a neoplasm that although it is rare, we can find it with greater regularity in this type of patients. It should be known that timely diagnosis and treatment of this oncologic pathology can greatly improve the prognosis of patients. A joint management should be carried out by both the infectologist and the oncologist, given that the therapeutic pillar will be chemotherapy with doxorubicin.

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