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**Case Report**

# Kaposi's sarcoma in a different concept from HIV-AIDS: report of a case in a patient with Behçet's disease and immunomodulatory drugs

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## ABSTRACT

Kaposi's sarcoma (KS) is a malignant vascular neoplasm that typically appears opportunistically in patients with acquired immunodeficiency syndrome (AIDS); it can also arise in organ transplanted subjects and exceptionally in carriers of autoimmune diseases, such as Behçet's disease. A 23-year-old man with Behçet's disease who debuts with clinically and histologically compatible dermatosis with KS. Conclusions. KS is not exclusive to HIV-AIDS, it also prevails in situations of primary or secondary immunocompromision that favor its appearance, as is the case of Behçet's Disease and its immunomodulatory therapy.

**Keywords:** Behçet's disease, Immunomodulatory drugs, Case report

## INTRODUCTION

Kaposi's sarcoma (KS) is a malignant vascular tumor, of universal distribution, related to the infection of Herpes virus no. 8 with diverse clinical behavior that has a clinical impact on the skin, gastrointestinal tract and respiratory system. For its development it is necessary to possess a status of severe immunocompromission, flourishing frequently in the context of infection by the human immunodeficiency virus (HIV) that conditions AIDS.[1-4]

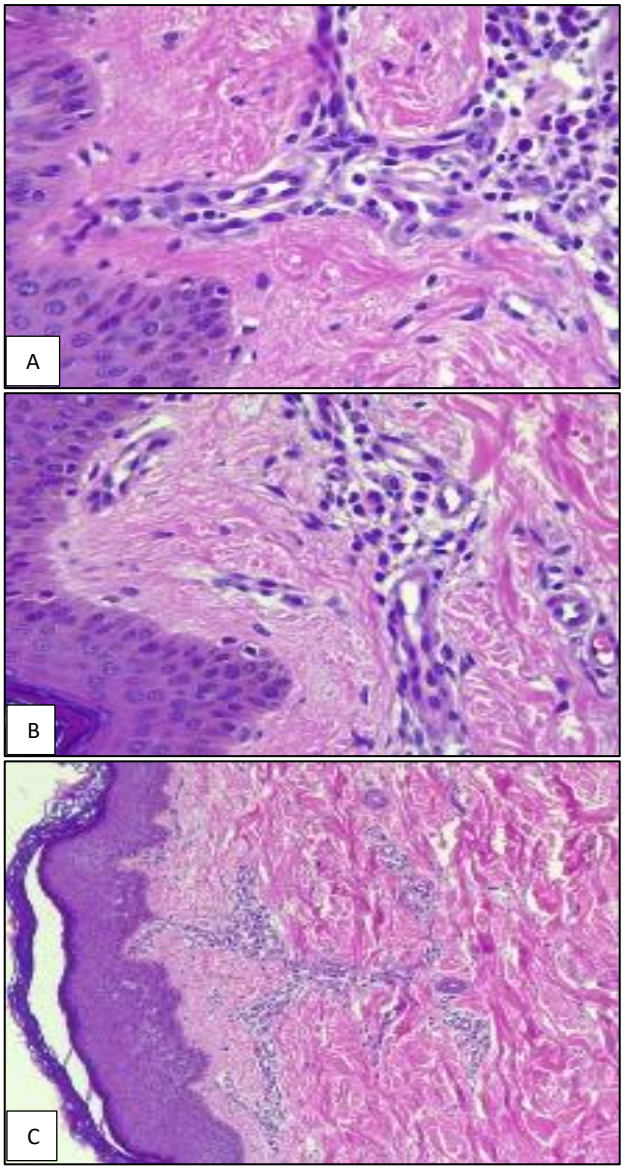
Although most of the time it appears as a result of this infectious environment, it is not only the only situation in which it is expressed; they also arise in environments of sustained immune deterioration, standing out in transplant recipients and less repeatedly in autoimmune diseases; that according to the latest bibliographic reports there are less than 35 reported cases of different rheumatological pathologies.[3] Due to its appearance in a concept different from AIDS and very few cases reported in rheumatological conditions, case of a 23-year-old man with Behçet's disease who debuts with this histologically confirmed dermatosis is presented.

## CASE REPORT

A 23-year-old male from Motul, Yucatan, with a personal history of Behçet's disease of 4 years of diagnosis, in pharmacological management with azathioprine 25 mg every 24 hours, colchicine 1 mg every 24 hours, paracetamol 500 mg every 24 hours. Previous hospitalization 24 months ago due to Steven Johnson syndrome, secondary to sulfonamide administration with a stay of 10 days in the intensive care unit, rest without importance. He began his condition 4 months ago, after presenting exacerbation of symptoms of basic pathology; characterized by pain in the lumbar region and at the level of bilateral sacroiliac territory, without predominance of schedule, with nocturnal awakening, improving with the change of position with a pillow on the feet or in the back, worsening with the increase in activity, as well as spinal stiffness of predominance in dorsal spine and neck

associated with decrease in caloric intake; handled with rocainol and hot water cloths, ketorolac, tramadol, and betamethasone with partial improvement. Edema of both feet, hands, ankles, morning joint stiffness and crunchy sounds in both knees is added 2 weeks later, and a large number of oral, painful and bleeding ulcers, in addition to gingivitis and gingivorrhagia; as well as pain in both eyeballs, conjunctival erythema, photophobia, decreased visual acuity and occasional blurred vision. It is evaluated by a physician who starts in the first instance prednisone 5 mg every 24 hours, and azathioprine 25 mg every 24 hrs with partial results, deciding to increase the dosage of azathioprine up to 100 mg daily, with progressive decrease in symptomatology until its final eradication. Two months later he presents dermatosis characterized by well-delimited maculas in both thoracic extremities, meriting biopsy, which reported chronic perivascular and perianexial dermatitis with nexostosis, infiltration of focal lymphocytes in pigmentary continence, photoprotective management is given; 2 weeks later patient comes again after referring new appearance of disseminated dermatosis to extremities characterized by hyperpigmented well delimited plaques some with few stretch marks (Figure 1 A-C), suspecting lichen planus, meriting second histopathological review, yielding data (Figure 2 A-C), compatible with KS. Viral serology is performed for HIV, resulting in negative, and it is concluded that this state is merely iatrogenic, deciding suspension of immunomodulator and granting support measures with disappearance of skin lesions after 3 months.


**Figures 1 (A-C): Disseminated dermatosis to extremities characterized by well-delimited hyperpigmented plaques some with few stretch marks.**




**Figures 2 (A-C): Skin histology, proliferation of small vessels branched and parallel to the epidermis with endothelial hyperplasia and findings compatible with KS.**

## DISCUSSION

Kaposi's sarcoma (KS) is a vascular neoplasm of a malignant nature and endothelial origin that appears in situations and conditions that condition immune exhaustion. It is widely recognized for its potential to proliferate in a sustained manner in connective tissue as well as to create new vasculature at the capillary level.[1,3-5]

Its term was coined at the end of the eighteenth century by dermatologist Moritz Kaposi after finding red-violet lesions in geriatric patients and was described as a very rare entity with little clinical and epidemiological impact; however, it was until the eighties that it began to gain more strength and popularity when after the declaration of an increase in cases and deaths related to AIDS, observed that

these skin lesions were in the final stages of this.[1,2,6] In the early nineties, Chang and colleagues associated a virus (then unrecognized, currently described as the human herpes virus 8) in multiple fluids of patients who shared these characteristic skin lesions.[7,8] Later they realized that not only was this dermatosis exclusive to this state of post-infectious immunosuppression, but also that multiple diseases had the potential to trigger it, concluding that for its appearance a state of immunocompromision is indispensable regardless of its underlying cause.[1,9]

Its genesis is not fully understood, it is still unknown; despite multiple investigations, no reproducible results have been found to confirm its development and origin. It is well known that human herpes virus 8 is involved in all cases, however it is not yet known if it is an etiological, detonating or aggravating agent.[1,5,8,10]

Its clinical picture is diverse in terms of manifestations, mainly suspected after evidencing skin damage; showing predilection in extensor areas, characterized by spots of red-violaceous coloration raised accompanied by hemorrhagic lesions with subsequent appearance of plaques and tumors that simulate nodular tissue whose size is very variable. Despite being clinically evident, they present with absence of symptoms in most cases. It may or may not be followed by edema in the extremities, especially in the lower body, which in situations of greater invasion culminates in elephantiasis.[1,3-5] Although the skin is the most clinically visible site, this entity also affects internal organs, the most affected being the gastrointestinal tract, lymph nodes, hepatic and pulmonary parenchyma, where its clinical translation will depend on the tissue that has been invaded.[1,5]

Four clinical scenarios are seen for the presentation of KS. 1) Classic SK; 2) Endemic or African KS, 3) KS related to HIV and 4) iatrogenic KS. The latter is widely described in subjects receiving solid transplants, especially renal tissue, and to a lesser extent emerges in rheumatological or hematological malignant conditions that are subject to immunomodulatory therapy.[1,3-5]

Currently there are different publications that allude to the association between this vascular neoplasm and autoimmune processes, finding about 35 reported cases (8 for rheumatoid arthritis, 5 for polymyositis/ dermatomyositis, 3 for systemic lupus erythematosus, 1 for Wegener's disease, 1 for allergic arteritiset), with 2 cases established by Behçet's disease, this case being the third described of KS since the appearance of this neoplasm began to be exposed.[3.11]

According to the reports of the iatrogenic clinical variety of the KS, it is established that these diseases are two to six times lower than that of solid organ transplantation and even the latency of appearance of the lesions are usually slower and progressive.[3,11]

Of course, there are several pathologies that appear as differential diagnoses; such as angiomas, hemosiderotic hemangioma, interstitial annular granuloma, cutaneous leiomyosarcoma, bacillary angiomatosis, lichen planus, etc. All of them with pathophysiology, clinical manifestations and different therapeutic tools.

Although interrogation and physical examination potently support the diagnostic suspicion, histological processing is necessary to confirm it, where it reveals the presence of spindle cells with elongated nuclei accompanied by clefts around pre-existing vessels and irregular endothelial cells.[1,3,4]

Its therapeutic approach is aimed at preventing the progression of skin lesions, improving symptoms (if any) and preventing its invasion into extracutaneous tissues. Currently there are no therapeutic modalities that eradicate this neoplasm in its entirety. In the context of iatrogenic KS, it is necessary to immediately interrupt the agent that is feeding the state of immunosuppression, thus recommending the decrease of immunomodulatory therapy, until reaching the minimum effective dose to keep the initial pathology in question latent. For the rest of the variants, there are multiple therapeutic options ranging from chemotherapeutic agents (liposomal anthracyclines), to laser therapy, photodynamic therapy, antiretrovirals, etc.[1,3,5]

The prognosis for this variant is excellent, since this unlike the other types of KS can remit in parallel if the initial focus that led to sustained immunosuppression is eradicated. Generally speaking, most of the cases described culminate in spontaneous resolution without the need for adjuvant treatment; and in case of greater invasion, the modalities for the other variants are also allowed [1,3,5,11]

## CONCLUSION

It is recognized that iatrogenic KS is a vascular tumor that develops in the clinical scenario of a skin lesion accompanied or not by systemic involvement with a clinical and/or therapeutic history that conditions immunosuppression. Although HIV is one of the main causes that favor the debut of this opportunistic disease, there are pathologies of a non-infectious nature that have the potential to lead to immune failure and its subsequent genesis; being of a priority character in not only affiliate it with a state of retrovirosis or reception of organ transplantation for its development but also an autoimmune disease may be responsible. After the control of the underlying disease (Behçet's disease in our case) and the modification of the immunomodulator dosage, all the lesions disappeared without the need to provide antineoplastic treatment.

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