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# Mucocutaneous Kaposi's sarcoma in an HIV-positive patient: diagnosis and treatment

## *Sarcoma de Kaposi mucocutâneo em paciente HIV positivo: diagnóstico e tratamento*

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### ABSTRACT

Kaposi's sarcoma (KS) is an endothelial neoplasia caused by infection with the human herpesvirus 8 (HHV-8), and the type associated with the human immunodeficiency virus (HIV) is considered the most aggressive and frequent. This paper reports a case of mucocutaneous KS in a patient not formerly aware of being an HIV bearer. A 38-year-old male patient has sought treatment with multiple oral lesions and one in the skin. Serology was positive for HIV and incisional biopsy diagnosed KS. After 11 months of chemotherapy and antiretroviral therapy, there was complete remission of the skin KS and partial remission of oral lesions.

**Key words:** Kaposi's sarcoma; human herpes virus 8; HIV infections.

### INTRODUCTION

Kaposi's sarcoma (KS) is a malignant neoplasia of endothelial cell proliferation<sup>(1)</sup>, considered as rare till the advent of acquired immune deficiency syndrome (Aids) in the 1980s<sup>(2,3)</sup>. It has affected about 37% of male patients, with buccal manifestations in 50% of them<sup>(2)</sup>.

The oncovirus human herpesvirus 8 (HHV-8)<sup>(4)</sup> is involved in the etiology of KS, being found in 90% of the diagnosed cases. It seems to be associated with the production of interleukin-6 (IL-6), which produces a mitogenic effect, as well as the *bcl-2* gene, a counterpart of the proto-oncogene *bcl-2*<sup>(2)</sup>.

From the three<sup>(3,4)</sup> or four<sup>(1)</sup> clinical standards described, the type associated with the immune deficiency syndrome can be observed in young adult patients, often affecting skin, mucous and internal organs<sup>(2,3)</sup>.

The clinical course is aggressive<sup>(1-3)</sup> and its prognosis is somber. The treatment varies between surgery for local lesions and radiotherapy in small doses and/or systematic or intralesional chemotherapy<sup>(2,5)</sup>.

### CASE REPORT

A 38-year-old melanodermic male patient sought dental treatment because of a dental fracture. During anamnesis, he

informed about his homosexual condition, but he denied being a bearer of any infectious disease. During a physical examination, the presence of a hardened nodular lesion was observed; it was painless, about 4 cm long on the patient's left arm, and had been developed over a two-month period (**Figure 1**). The intraoral examination revealed extensive and irregular, multinodular, erythroleukoplasic lesions of fibroelastic consistency and bleeding in the palate, in the upper anterior labial ridge, and the right and left lower retromolar regions (**Figure 2**).

The patient reported that these lesions had been developed over an eight-month period, causing spontaneous pain in the palatal region and some difficulty in eating. Under palpation, impairment of the unilateral submandibular lymph nodes was found. An incisional biopsy of the oral lesions was performed and serology exams were required for human immunodeficiency virus (HIV) and venereal disease research laboratory (VDRL) test. Two weeks later, the patient returned with a positive result for HIV, but a negative result for VDRL. Then, he was sent to a reference hospital for the treatment of infectious diseases, where a purified protein derivative (PPD) test was required, as well as the viral and CD4 T-lymphocyte count. The result of the PPD test was not reactive, the viral load was 450 copies/ml, and the CD4 count was 163 cell/μl.

The histopathological exam of the specimen revealed fragments of malignant neoplasia of vascular origin, showing proliferation of endothelial and pleomorphic mesenchymal cells, hyperchromatic, with prominent nucleoli, besides some figures of mitosis. The

formation of irregular blood vessels was sometimes seen in the shape of cracks (**Figure 3**). The diagnosis was Kaposi's sarcoma.

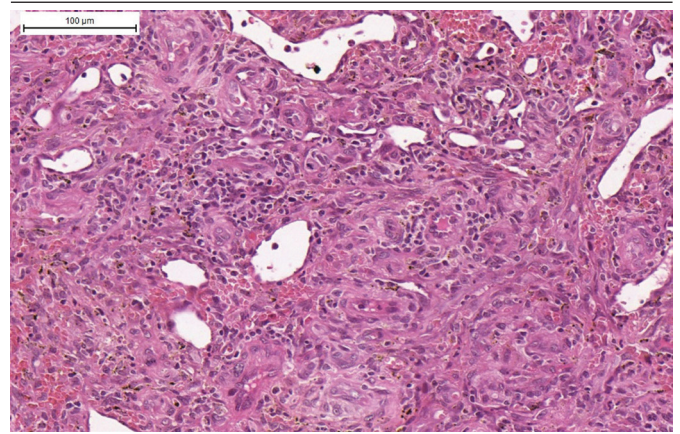
Once the diagnosis of HIV and KS with oral and skin lesions was established, the patient began highly active antiretroviral therapy (HAART), as well as additional chemotherapy. After an 11 months' follow-up, the skin lesion regressed completely, with only a brownish hyperpigmentation of the skin (**Figure 4**) remaining. At the same time, oral lesions regressed almost completely, also with red hyperpigmentation areas (**Figure 5**) being observed. There was improvement of the patient's general health condition.



**FIGURE 1** – Painless nodular lesion, about 4 cm long on the patient's left arm



**FIGURE 2** – Initial clinical aspect of the patient's intraoral lesions before the use of antiretroviral therapy: multinodular, erythroleukoplakotic lesions of fibroelastic consistency and bleeding in the palate, in the upper anterior labial ridge, and the right and left inferior retromolar regions



**FIGURE 3** – Proliferation of endothelial and pleomorphic mesenchymal cells. Formation of irregular blood vessels in the shape of cracks



**FIGURE 4** – Complete regression of the skin lesion after 11 months of treatment with antiretroviral therapy, with only an area of hyperpigmentation remaining



**FIGURE 5** – Patient's oral aspect after HAART

A) significant regression of intraoral lesions is observed after 11 months of treatment with antiretroviral drugs; B) patient's oral aspect after HAART.

HAART: highly active antiretroviral therapy.



## DISCUSSION

The risk of developing cancer is much higher in HIV bearers and it has been recognized as a complication of the infection by the virus<sup>(6)</sup>. KS the most prevalent type of cancer in HIV+ patients that have not received treatment, and it is associated with the infection by HHV-8<sup>(7-9)</sup>.

In the reported case, the presence of extensive oral lesions raised suspicions of KS associated with the HIV infection.

The oral cavity is the first area where KS appears in 22% of HIV+ patients, and up to 71% of HIV-carrying patients can develop oral KS together with skin and visceral involvement<sup>(1)</sup>. Supporting this information, the patient in this reported case showed oral lesions associated with a skin nodular lesion that arose after the emergence of oral lesions.

The Aids-related KS lesions tend to be spread, multiply in number, become more nodular or coalesce in association with immunological deterioration and the drop in the count of CD4 cells<sup>(1)</sup>. In this reported case, the patient was not aware of being

infected by the HIV virus and for this reason was not undergoing medical therapy. For this reason, he presented dissemination of oral lesions associated with severe immunosuppression.

Literature reports that CD4 T-lymphocyte counts below 200 cell/ $\mu$ l are associated with the emergence of neoplasia in HIV+ patients<sup>(10)</sup>. Pain occurs in later phases, when lesions reach nerve structures<sup>(6,11-13)</sup>. In the present case, the patient complained about pain and had some spontaneous bleeding, besides some difficulty in eating.

The occurrence of oral KS in non-treated HIV+ patients means a worse prognosis, in which the mortality rate is much higher than in patients affected by skin KS<sup>(1,5,6)</sup>. In the discussed case, by ignoring the fact of being an HIV bearer, the patient had KS spread in the mouth and skin due to the lack of treatment for the infection. There is evidence that KS often goes into remission with HAART, and HIV patients undergoing antiretroviral therapy suffer a less aggressive form of KS than those who do not have this therapy at the time of diagnosis<sup>(9,11)</sup>. After 11 months of patient follow-up in use of antiretroviral therapy, a complete regression of the patient's skin lesions and a significant regression of intraoral lesions were observed.

## RESUMO

*Sarcoma de Kaposi (SK) é uma neoplasia endotelial causada pelo herpes vírus humano tipo 8 (HHV-8), e o tipo associado ao vírus da imunodeficiência humana (HIV) é considerado o mais agressivo e frequente. Relata-se um caso de SK mucocutâneo em indivíduo não anteriormente ciente de ser portador de HIV. Paciente do sexo masculino, 38 anos, procurou atendimento com queixa de múltiplas lesões orais e uma lesão em pele. A sorologia foi positiva para HIV, e a biópsia incisional das lesões teve como diagnóstico SK. Após 11 meses de terapia antirretroviral e quimioterapia, houve remissão completa do SK cutâneo e parcial das lesões orais.*

*Unitermos: sarcoma de Kaposi; herpes vírus humano tipo 8; infecções por HIV.*

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