An unusual presentation of Kaposi's sarcoma in AIDS patient. A case report

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ABSTRACT: A 45-year-old male patient, recently diagnosed with HIV infection, admitted to the emergency department for constitutional syndrome plus acute confusional syndrome, with suspected opportunistic germ neuroinfection. Physical examination revealed incipient skin lesions characterized by violaceous spots on the lower limbs and on the hard palate of 5x5mm in diameter, with no adenopathies. Serology was performed for positive IGG toxoplasmosis, coproparasitic revealed positive fecal occult blood. Computed tomography of the brain and chest revealed opportunistic infection due to cerebral toxoplasmosis, and multiple nodular lesions in the chest. Upper gastrointestinal endoscopy revealed elevated lesions with central ulceration and biopsies were taken. The histopathology result of the biopsies revealed Kaposi's Sarcoma with Gastric involvement.

Keywords:
Kaposi's sarcoma, gastric Kaposi's sarcoma.

Case Report

Dermatology

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Introduction

Kaposi´s Sarcoma KS is a vascular endothelial cell tumor caused by the combination of human herpesvirus 8 (HHV-8) infections and impaired host immunity [1]. It is the second most common tumor in patients with HIV infection [2]. The prevalence is highest in sub-Saharan Africa, where in some populations it exceeds 90% [3]. A 2017 study reported an incidence in Latin America of 244 per 100,000 person-years [4]. It is estimated that 30% of patients who do not receiving highly active antiretroviral therapy (HAART) will develop KS [2].

The clinical progression of KS is generally slow and not very aggressive; however, KS in the context of AIDS, also called AIDS-KS or epidemic KS, has a more aggressive natural history, which can affect the mucous membranes, lymph nodes, the gastrointestinal tract and lungs, particularly in untreated people [5]. In the Western population, the prevalence of AIDS-KS is 6% to 30% [3]. The risk is five to ten times higher in men who have sex with men (MSM) [6].

KS manifests in 78% of cases due to skin disease and/or visceral involvement in 15% [7]. The gastrointestinal (GI) tract is the most common extracutaneous site 40% [8]. When there is clinical suspicion, biopsy is the gold standard to histologically confirm the diagnosis [4]. The basic histopathology characteristics include vascular proliferation in the dermis, the presence of blood extravasation, an inflammatory infiltrate, the proliferation of spindle cells expressing endothelial markers, are considered KS tumor cells [7].

Recent studies have shown that HAART combined with systemic chemotherapy is associated with a significant improvement in morbidity and mortality. Therefore, in patients with suggestive dermatological lesions and with risk factors, endoscopy screening has been suggested for early detection and timely initiation of treatment [11, 12]. The prognosis depends on the CD4 count and opportunistic infections [13]. The objective of this study is to document the case of a patient with incipient skin lesions, without gastrointestinal symptoms, promptly diagnosed with AIDS-KS with gastric involvement.

Case report

A 45-year-old male patient, bisexual, with a recent diagnosis of HIV infection, without treatment, admitted to the emergency service for a 3-month history of symptoms characterized by weight loss (20kg), asthenia and hyporexia, and hours before admission, headache, disorientation and confusion are added.

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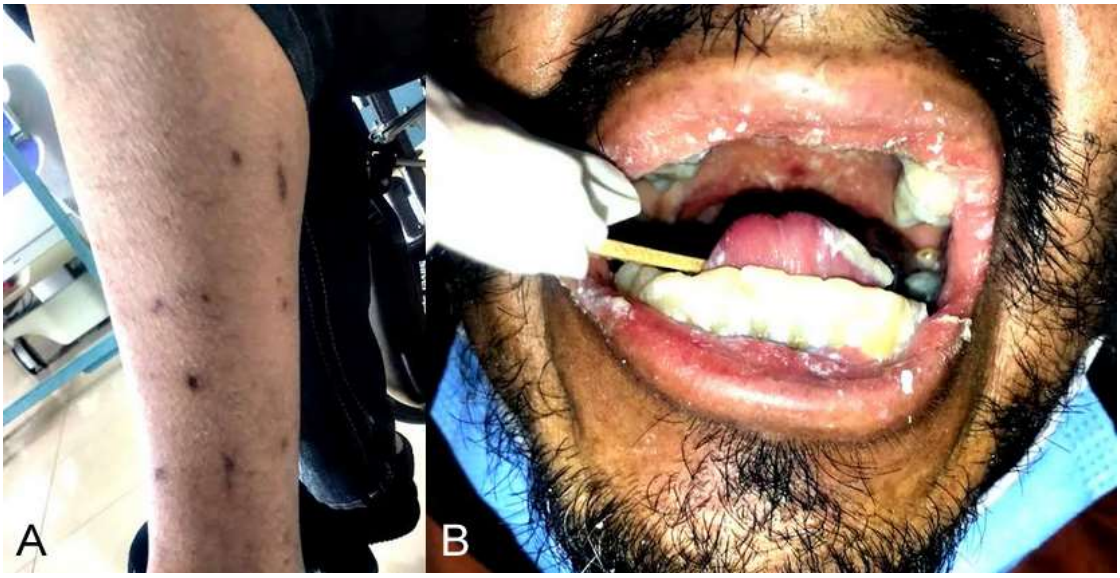

Figure 1. A. Purplish spots on lower limbs. B. Purplish spot on hard palate.

On physical examination: incipient skin lesions: scattered violaceous spots 5x5mm in diameter, on the hard palate and lower extremities, without regional adenomegaly (**Figure 1**). The CD4 lymphocyte count with 21 cells compatible with AIDS, a multidisciplinary management was performed. The patient started HAART and adequate antibiotic prophylaxis for opportunistic infections.

Serology was requested for IGG toxoplasmosis with a positive result, brain tomography showed lesions compatible with cerebral toxoplasmosis. A lumbar puncture was performed with compatible fluid for tuberculous meningitis, antituberculous treatment was started, and for cerebral toxoplasmosis.

With no improvement in the clinical picture, a tomographic scan of the chest, abdomen, and pelvis with intravenous contrast was performed, revealing multiple scattered nodular images in both lung fields in the lung parenchyma, compatible with unknown primary metastases, the rest normal. In search of primary neoplasia, a coproparasitic test was requested, where positive fecal occult blood was found. It was decided to perform upper gastrointestinal endoscopy, in the gastric mucosa erythematous lesions were observed, elevated with central ulceration of 10x20mm in diameter with alteration of the mucosal and vascular pattern, with normal gastric submucosa (**Figure 2**). Correlating with the skin lesions, biopsies were taken.

After a long hospital stay, the biopsies of the stomach lesions revealed an atypical vascular lesion and the histopathological result showed a dense spindle cell lesion with a mesenchial appearance that infiltrates the lamina propria of the mucosa, dissecting the glandular structures with the focal presence of nuclear pseudoinclusions (**Figure 3**), immunohistochemistry revealed the positive immunoexpression of the HHV8 marker that corroborates the etiopathogenesis of Herpes Virus type 8 associated with Kaposi's Sarcoma (**Figure 4**). With histopathology results, the patient was referred to the Cancer Institute to receive specific chemotherapy treatment.

Discussion

KS is an angioproliferative disorder that comprises several types of diseases, all etiologically related to HHV-8 infection [3]. One of the widely recognized epidemiological forms is KS-AIDS, which is still considered the most common cancer in people living with HIV [4]. GLOBOCAN data in 2018 show that there were 41,799 new cases of KS and 19,902

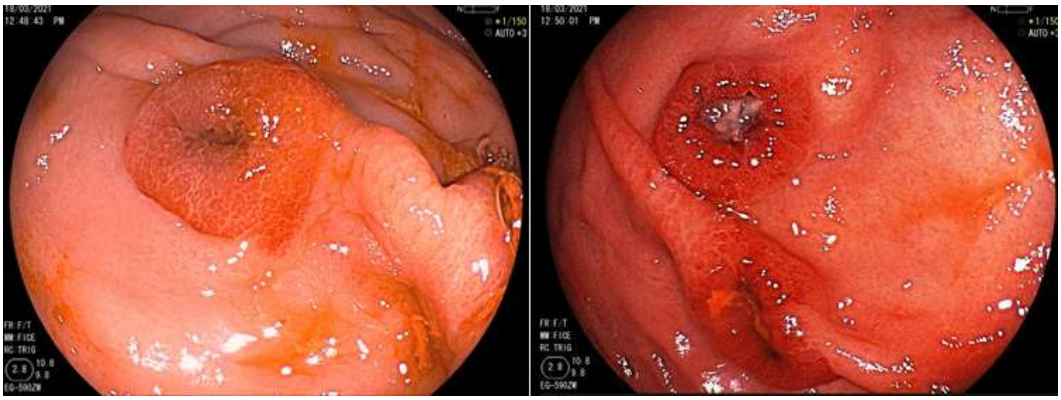

Figure 2. Raised lesions with central ulceration, crater-shaped in the gastric mucosa.

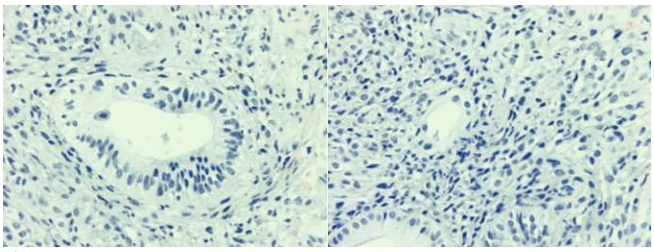

Figure 3. Left. focal viral nuclear changes characterized by viral-like nuclear pseudoinclusion. Right. Dense fusocellular lesion infiltrating towards residual glands.

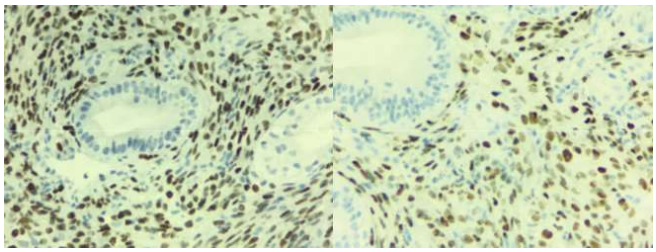

Figure 4. Positive immunoexpression of the HHV8 marker corroborates the etiopathogenesis of Herpes Virus type 8 associated with Kaposi's Sarcoma.

deaths from KS worldwide [7].

AIDS-KS was identified for the first time in 1980 with a more aggressive behavior than the other variants, with potential visceral involvement and high mortality [11]. Cutaneous lesions usually present more frequently on the trunk, extremities and oral cavity as multiple lesions, pigmented, raised or flat, painless, non-blanching [7]. The first skin lesions are usually pigmented macules or small asymptomatic papules on the distal segment of the lower extremities, ranging in color from pale pink to deep purple, which subsequently become in papules, nodules or plaques, being more aggressive in patients with AIDS [7].

Gastrointestinal involvement represents disseminated disease; however, it is asymptomatic in 80% of cases until serious complications arise [4, 11]. There are three endoscopic phenotypes of GI-KS: maculopapular lesions, polypoid lesions, and crater- or volcano-shaped lesions such as those seen are present in our patient [8, 10]. The classic endoscopic finding is represented by subepithelial, reddish, ulcerative lesions [8, 14].

The initial KS screening is clinical in the detection of lesions on the skin or mucosa of the oral cavity [11]. Nagata et al., in their retrospective study, revealed that there are clinical predictive factors of GI-KS such as low CD4 cell count <100 cells/µL, MSM, presence of cutaneous KS, viral load >10,000 copies/ml and absence of HAART [12, 15]. In the presence of incipient skin lesions together with these factors, endoscopy should be considered to detect GI-KS before the development of clinical symptoms and prevent complications and improve the prognosis of these patients.

Histopathology confirmation by biopsy is the gold standard [7]. The malignant cell characteristic of KS is the spindle cell infected by HHV-8, the expression of this marker in spindle cell nuclei is considered 99% sensitive and 100% specific for KS [5]. Currently, the modified staging of the AIDS Clinical Trials Group (ACTG) is used to stage AIDS-KS [2, 16]. According to this classification, our patient presents an advanced stage, which requires early treatment.

HAART is essential for the optimal management of AIDS-KS [16, 17]. In developed countries, pegylated liposomal doxorubicin and paclitaxel have been shown to be very effective in inducing the regression of this disease, being considered by the NCCN (national comprehensive cancer network) as the first line of treatment [18, 19, 20]. In low-resource settings, combinations of bleomycin and vincristine, or oral etoposide are used [21].

Conclusion

KS is one of the most common causes of morbidity and mortality in people living with HIV worldwide. KS-GI is the most common visceral form, mostly asymptomatic and remains undiagnosed. A high index of suspicion should be maintained in the presence of incipient skin lesions added to risk factors, where endoscopy is advisable for screening, being an early treatment associated with a better prognosis.

Conflicts of interests

The authors declare no conflict of interest.

TIS staging of KS	Early Stage/Good Prognosis (T0)	Advanced Stage/Poor Prognosis (T1)
Tumor	Confined to skin and/or lymph nodes, or minimal oral disease.	Tumor-associated edema or ulceration, extensive oral KS, GI-KS, or KS in other non-ganglionic viscera.
Immune status	CD4 cell count greater than 150mm3	CD4 cell count less than 150mm3
Systemic disease	Karnofsky performance status* greater than 70	Karnofsky performance status* less than 70, or other HIV-associated disease

*The Karnofsky scale, which ranges from 100 (best) to 0 (worst), assesses functional impairment.

Figure 2. Modified staging of the group of AIDS-KS clinical trials [2, 16]

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