CASE STUDY

Immune Reconstitution Inflammatory Syndrome Manifested as a Kaposi’s Sarcoma in the Tongue

Síndrome inflamatorio de reconstitución inmune manifiesto como sarcoma de Kaposi en lengua

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Case Report

Kaposi’s sarcoma (KS) is one of the most common neoplasms in patients suffering HIV/AIDS, especially in patients with CD4+ counts below 200 cells/μL. There is a strong association between infection by human herpes virus 8 (HHV8) and the appearance of this neoplasm. Under the optic microscope, KS is a multicentric, angioproliferative disorder characterised by the proliferation of spindle cells, neoangiogenesis, inflammation and oedema.

We present the case of a 30-year-old male, diagnosed with infection by HIV 1 year and 4 months earlier. The CD4+ count was of 24 cells/μL and the HIV viral load (HIV-VL) of 15,331 copies/ml. The following diagnoses were established during admission: Molluscum contagiosum in upper and lower limbs and genitalia, Condyloma acuminatum in the genital area, oral candidiasis, pulmonary tuberculosis, disseminated infection by cytomegalovirus (CMV) and disseminated disease by Mycobacterium avium complex (MAC).

One month after the start of treatment of the opportunistic infections, we began highly active antiretroviral therapy (HAART) with emtricitabine/tenofovir and efavirenz. No KS lesions had been documented at that point.

One year after the start of HAART, the CD4+ count had reached 105 cells/μL and HIV-VL was undetectable. The treatment for tuberculosis, MAC and CMV infections had concluded and no activity of these diseases was recorded.

The patient was referred to the otolaryngology service after a nodular lesion was detected in the dorsum of the tongue 1 month earlier. Upon questioning, the patient reported mild odynophagia and intermittent dysphagia for solids. We conducted an indirect laryngoscopy (IL) which revealed a rounded tumour with an ulcerous-necrotic surface in the right half of the tongue base, with a diameter of approximately 2 cm (Fig. 1A). Incisal biopsies of the tumour, which was soft and friable, were taken by indirect laryngoscopy. A microscopic study revealed spindle cells with an elongated nucleus and vascular clefts (Fig. 2A). Immunohistochemical analysis was positive for antigen CD34 (Fig. 2B). The histopathological diagnosis was of KS.

We actively sought other KS lesions at a cutaneous level and reviewed imaging studies from 1 week earlier, including thoracic and abdominal scans. KS was only confirmed in the dorsum of the tongue.


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Figure 1  (A) Lesion in the right half of the tongue base formed by a round tumour with an ulcerous-necrotic surface, measuring 2 cm in diameter. (B) The same lesion, now with a pink hue and measuring 0.8 cm in diameter, showing an involution of oral KS.

We opted for maintaining the patient under surveillance and continuing HAART, awaiting an involution of the tumour. In the follow-up period of 1 month we observed a 60% decrease in the size of the tumour (Fig. 1B).

Discussion

The increase of the use of HAART among the population of patients infected by HIV has caused a significant reduction of the prevalence of KS. Exceptionally, KS manifests paradoxically, that is, it appears or worsens after HAART. In these paradoxical cases, as the immune system recovers there can even be a worsening or appearance of KS lesions as a manifestation of immune reconstitution inflammatory syndrome (IRIS).6,7 The rate of prevalence of IRIS is between 8% and 30%, and it is generally associated to tuberculosis, MAC, retinitis by CMV and disseminated Cryptococcus.8 There are several

Table 1  Criteria Proposed for the Diagnosis of IRIS in Patients With HIV Following HAART.1

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
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<tr>
<td>A. Atypical presentation of opportunistic infections or tumours in patients responding to HAART</td>
<td>• Increase of cellular CD4 count following HAART</td>
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<tr>
<td>• Localised disease (lymph nodes, liver and spleen)</td>
<td>• Increase of specific immune response for a relevant pathogen (for example: response like delayed hypersensitivity for mycobacterial antigens)</td>
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<td>• Exaggerated inflammatory reaction</td>
<td>• Spontaneous resolution of disease without specific antimicrobial therapy or tumoral chemotheraphy without continuing HAART</td>
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<td>• Severe fever, without an established cause</td>
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<td>• Painful lesions</td>
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<td>• Atypical inflammatory response in affected tissue</td>
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<td>• Granulomas, suppuration, necrosis</td>
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<td>• Perivascular lymphocytic inflammatory cellular infiltrate</td>
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<tr>
<td>• Progression of organic dysfunction or extension of preexisting lesions, after definition of clinical improvement following germ-specific treatment before starting HAART and exclusion of toxicity of treatment and new diagnoses</td>
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<td>• Development or extension of lesions occupying space in the brain following treatment for cerebral cryptococcosis or toxoplasmosis</td>
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<td>• Progressive pneumonitis or development of organised pneumonia following treatment for pulmonary TB or pneumonia by Pneumocystis</td>
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<tr>
<td>• New instalment or worsening of uveitis/vitreitis following resolution of retinitis by CMV</td>
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<tr>
<td>• Fever and cytopenia following treatment for disseminated MAC</td>
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<tr>
<td>• Extension of KS lesions and subsequent resolution or partial remission without start of radiotherapy, systemic chemotherapy or intralesional therapy</td>
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<td>B. Decrease in plasma levels of HIV-RNA &gt;1 log 10 copies/ml</td>
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Source: based on French et al.11

* The diagnosis of IRIS requires at least 2 major criteria or 2 minor criteria and 1 major criterion.
works on KS related to IRIS, with prevalence around 6.4% of total cases of IRIS. Nevertheless, there are very few cases of isolated, oral KS.

This case report fulfills the criteria for belated IRIS (Table 1), as it appeared in a patient suffering HIV infection who, after more than 1 year of HAART, presented an increase in CD4+ T-cell count (a minor criterion) and a persistently undetectable HIV viral load, followed by formation of a neoplasm, in this case KS (2 major criteria). One month after the appearance of KS, after adopting an expecting attitude and continuing with HAART, the lesion began its involution (a major criterion). 11

In cases presenting KS lesions which do not represent a significant clinical symptom for patients and do not affect the airway, despite being a case of KS secondary to IRIS, according to the clinical experience in these cases, continuing with HAART is expected to lead to a full remission of the lesion. 7,10

Conflict of Interests

The authors have no conflict of interests to declare.

References