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Clinical Image
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Disseminated Kaposi´S Sarcoma as Primary Manifestation of AIDS

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Abstract

Kaposi's sarcoma (KS) is a polyclonal multifocal disease and the most common neoplasm associated with Human Immunodeficiency Virus (HIV) infection. AIDS-related KS has a variable clinical course, ranging from minimal and localized disease to the skin to a rapidly progressing neoplasm that can involve the mucosal, lymph nodes and visceral with a significant morbidity and mortality. We describe a case of disseminated KS with mucosal and visceral involvement as primary manifestation of AIDS.

Clinical Image

A 32-year-old male was admitted in our Department of HIV/AIDS disease with a history of 8 months of multiple cutaneous and mucosal tumoral lesions. Lesions located on the nose (Figure 1) and in the oral cavity gradually increased in size. Oral cavity lesions involved the tongue, the gingiva and a large tumoral lesion that compromise the hard and soft palate (Figure 2). Lesions were bluish-red in color and firm in consistency. One month previous to the admission, he referred fever, weight loss, cough and progressive dyspnoea. Diagnosis of Kaposi´s sarcoma was suspected. An Enzyme-Linked Immunosorbent Assay of 4th generation (ELISA) was performed and was diagnosed as reactive to HIV antibodies. Routine laboratory analysis were performed; the CD4 T cell count was of 61 cell/μL (5%) and the plasma viral load was 972 000 copies/mL ($\log_{10}$ 6.1). Mild to moderate anemia (hematocrit 34%) was detected. The rest of blood tests, renal and liver function were normal. Hepatitis B and C antibodies and VDRL were negative. Blood cultures, sputum and bronchoalveolar lavage were negative for bacteria, fungi and mycobacteria. A CT scan of the lungs revealed multiple and diffuse opacities and pulmonary nodules compatible with KS lesions (Figure 3). Skin biopsy of nasal and oral tumoral lesions was performed. Histopathological examination of both, nasal and oral cavity biopsies, showed a proliferation of blood vessels with ovoid and spindle cells, with numerous vascular slit-like spaces containing red cells, lymphocytes and plasma cells with perivascular deposits of hemosiderin were observed (Figure 4). The detection of DNA HHV-8 in biopsy smears was positive by PCR. Final histopathological diagnosis was disseminated Kaposi´s sarcoma associated with human immunodeficiency virus infection. Patient was started on Highly Active Antiretroviral

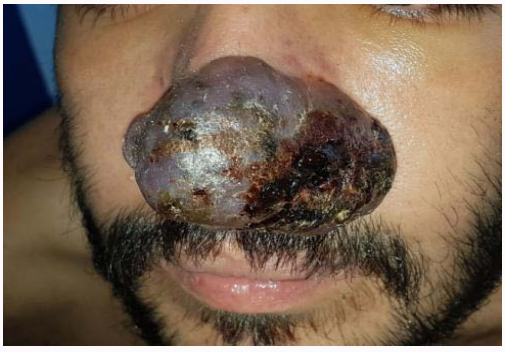

Figure 1: Cutaneous lesion of KS involving the tip of the nose.

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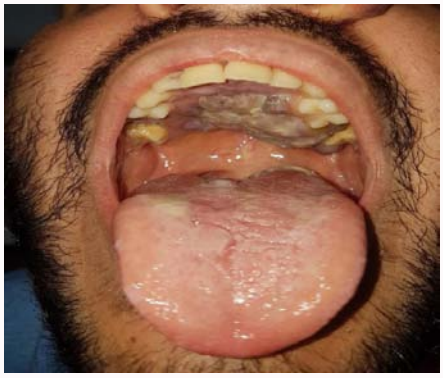

Figure 2: A large intraoral lesion involving the palate with a small lesion over the tongue.

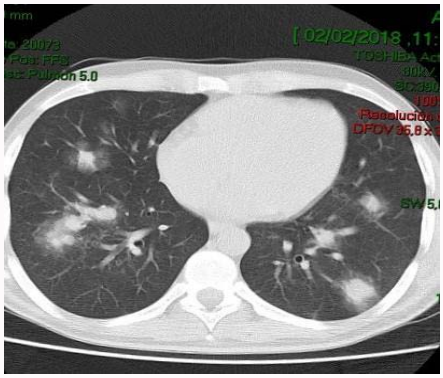

Figure 3: CT scan of the lungs showing multiple nodules and opacities.

Therapy (HAART) based on tenofovir/emtricitabine and efavirenz plus chemotherapy including liposomal doxorubicin in cycles of 20 mg/m^2 with a good clinical, immunological and virological response.

KS is a malignant neoplasm, strongly associated with Human Herpes Virus 8 (HHV-8) in its pathogenesis [1]. The presence of HHV-8 antibodies preceded and is strongly associated with the subsequent development of KS [2]. Lesions of KS can disseminate rapidly in severely immuno compromised patients, as we can see

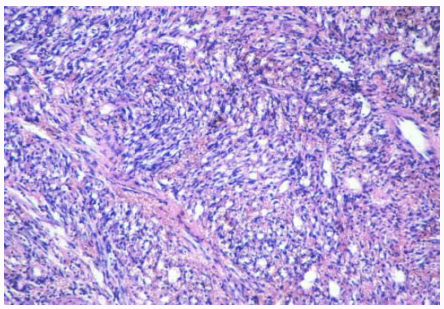

Figure 4: Skin biopsy showing spindle cell proliferation, chronic inflammation, numerous vascular slit-like spaces, lymphocytes and plasma cells.

in the case that we present, with cutaneous, mucosal and visceral involvement [3]. In advanced HIV/AIDS disease, KS has a very aggressive clinical course with frequent involvement of lymph nodes, the lungs and the gastrointestinal tract in 50% of the cases [4]. Lungs involvement occurs in 20% of the patients and is the most frequent cause of mortality. Diagnosis of pulmonary KS can be made by a combination of clinical, laboratory, radiologic and bronchoscopic findings [5].

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