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CASE REPORT

Aggressive Kaposi sarcoma in a new case of HIV; case report and review of the literature

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ABSTRACT

Background: Kaposi sarcoma (KS) is an angioproliferative tumor that mainly involves mucocutaneous tissues, but extracutaneous spread to lymph nodes, GI tract, lungs, liver, pancreas, heart, and testes can occur in AIDS-associated KS. Patients with pulmonary KS may be symptomatic or present with an asymptomatic abnormality on chest radiography.

Patient: A 28-year-old man presented with a one month history of rashes, cough, weakness and malaise. He has been an intravenous drug user since 5 years ago. The rashes were first noted in his face and then spread to his oral cavity and trunk. Skin biopsy was compatible with KS.

Conclusion: Kaposi sarcoma of the tip of the nose has been introduced as a sentinel sign for Kaposi sarcoma of the lung. This case is a young man with mucocutaneous and pulmonary KS as a new HIV patient

Keywords: *Kaposi sarcoma, AIDS, Opportunistic infections*.
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INTRODUCTION

The first case of epidemic or AIDS-associated Kaposi sarcoma (KS) was reported in 1981 (1). It has been estimated that malignancy will develop in 40% of all patients with HIV infection at some time during the course of their illness (2). KS is the most common malignancy associated with HIV infection and occurs principally in homosexual or bisexual men with human herpesvirus 8 (3-5). KS is an angioproliferative tumor that mainly involves mucocutaneous tissues, but extracutaneous spread to lymph nodes, GI tract, lungs, liver, pancreas, heart, and testes can occur in AIDS-associated KS (6,7). Patients with pulmonary KS may be symptomatic or present with an asymptomatic abnormality on chest radiography (7). We report a young man with mucocutaneous and pulmonary KS as a new known HIV patient.

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CASE PRESENTATION

A 28-year-old man presented with rashes, cough, weakness and malaise since one month ago. He has been an intravenous drug user (IDU) since 5 years ago. The rashes were first noted in his face and then spread to his oral cavity and trunk (figure 1). He also complained of productive cough and dyspnea during 10 days prior to admission. Past medical history was negative and he had no history of a recent trip or contact with animals and bite.

His initial vital signs were as follow: oral temperature 36.8ºC, blood pressure 115/80 mmHg, pulse rate 82/min, and respiratory rate 20/min.

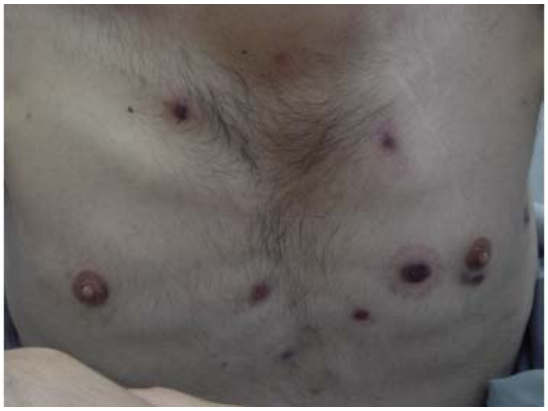

Figure 1. Maculo-papulo-nodular rashes developed on trunk

Physical examination revealed red purple maculo-papulo-nodular rashes in his face, trunk and mucous of the mouth and his tongue. Bilateral basilar fine rales were noted in lungs auscultation. Laboratory findings included white blood cell count: 4500/mm^3 with 23% lymphocyte, hemoglobin 6.3mg/dl (MCV:70.9 and MCH:20) and platelets 445000/mm^3. HIV ELISA was reactive and confirmed by HIV western blot. HCV antibody was positive, but CD4 count was not determined. Skin biopsy revealed proliferation of spindle shaped cells within collagenous fibers and extravasation of red blood cells (figure 2).

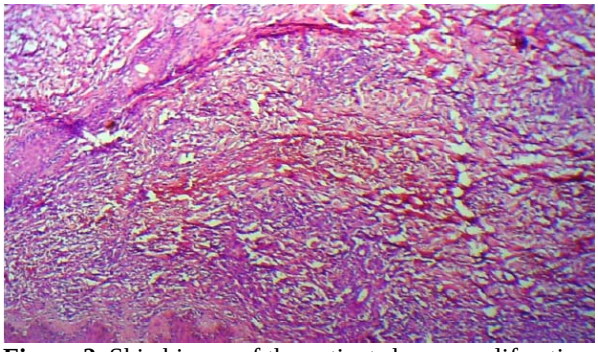

Figure 2. Skin biopsy of the patient shows proliferation of spindle shaped cells within collagenous fibers and extravasation of red blood cells

Chest x-ray demonstrated bilateral paracardiac and para-hilar reticolonodular infiltration. HRCT scan showed non-hemogenous infiltration and air bronchogram in paracardiac and basilar part of both lungs vanishing the right border of heart and diaphragm in left side. Finally, sputum exam showed no evidence of tuberculosis and other bacterial infections.

DISCUSSION

HIV infection constitutes a main health problem worldwide. The oral and perioral manifestations are common in HIV infected patients, and often influence the debilitating general health status, a worse prognosis of the disease, as well as a diagnostic factor in the monitoring of the immune status of the patient (8-10).

The vast majority of the HIV infected subjects have presented at least one manifestation in the head and neck area in any state of the disease (11), representing these oral lesions as the oral signs of the disease (12,13). In addition, the occurrence of these lesions indicates a great susceptibility for opportunistic infections and a great possibility of rapid progression to AIDS (14,15). The CD4 cell count and viral load have been used lately as the most important laboratory parameter to evaluate the evolution of the disease (16). Several studies have been focused in the correlation between oral lesions prevalence and the laboratory parameters, such as CD4 cell count and viral load in HIV/AIDS patient serum, evidencing a strong correlation between the oral lesions, lower CD4 cell count and high viral load, concluding that these are involved with monitoring and progression of the disease, as well as the antiretroviral therapy (17-20).

Kaposi sarcoma occurs throughout the course of HIV infection at CD4 cell counts of anywhere from 0 to 800/µL. It remains an open question whether antiretroviral therapy, the first-line therapy for KS, should be started in a patient with KS but higher CD4 cell counts than those counts currently serving

as indicators for starting antiretroviral therapy. KS occurs even in patients with profound suppression of HIV replication. Currently, KS tends to present as subtle purple patches rather than large fixed plaques characteristic of the disease in the pre-potent antiretroviral therapy era (21). Kaposi sarcoma is an angioproliferative neoplasm caused primarily by infection with Kaposi sarcoma–associated herpesvirus (KSHV) (21,22). Four distinct clinical forms are described, including classic, endemic, iatrogenic, and AIDS-associated forms, each following an incidence pattern that parallels KSHV seroprevalence (23-27). Histologically, Kaposi sarcoma lesions are similar for the four distinct clinical forms and are characterized by the presence of spindle-shaped tumor cells of vascular endothelial origin in addition to heterogeneous endothelial, fibroblast, and dermal dendritic cell populations and infiltrating inflammatory leukocytes (28-30). KS is an angioproliferative tumor that mainly involves mucocutaneous tissues, but extracutaneous spread to lymph nodes, GI tract, lungs, liver, pancreas, heart, and testes can occur in AIDS-associated KS (6,7). Patients with pulmonary KS may be symptomatic or present with an asymptomatic abnormality on chest radiography (7). In one study, (31) 33% of all KS patients were reported having clinically evident pulmonary KS and 50% had pulmonary involvement at autopsy. Pulmonary KS may occur without evidence of mucocutaneous disease (31,32). Dyspnea and cough are the most common presenting symptoms. Fever and night sweats usually suggest a concomitant infection. Hemoptysis, fever, and respiratory failure can also complicate pulmonary KS (33). Radiographic findings in pulmonary KS are varied and include isolated pulmonary nodules, pleural effusions, as well as hilary or mediastinal lymphadenopathy. Chest radiographic findings in pulmonary KS consist of interstitial thickening and ill-defined nodules. The thickening becomes more nodular with tumor progression, with eventual confluence of ill-defined nodules leading to dense airspace consolidation (34). The CT findings correlate with chest radiographic appearances and consist of peribronchovascular interstitial thickening and poorly marginated nodules predominantly in peribronchovascular distribution. The nodules are multiple, have lower-lobe predominance, and measure 1 to 2 cm in diameter (34). In one series by Khalil et al, CT findings in 53 patients with pulmonary KS revealed nodules in 42 (79%), bronchovascular thickening in 35 (66%), tumor masses in 28 (53%), and pleural effusions in 29 cases (55%). The association of two or more of these findings was found in 40 patients (75%) and was suggestive of KS (34). Masih noticed the association of a striking, bulbous, tip-of-the-nose KS lesion with pulmonary KS (35).

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