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Case Report

Classic Kaposi Sarcoma: An Unusual Presentation in an HIV-Negative Man

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Abstract
Kaposi sarcoma (KS) is a type of cancer that develops in blood vessels of the mouth, lymph nodes, and lungs. Our case report is about a 66-year-old man with purple patches and plaques, as well as nodules distributed in the legs and arms. Biopsy of the skin lesions showed angiomatous proliferation of small vessels with mild perivascular lymphocytic infiltration with plasma cells. We started chemotherapy for this disseminated form of Kaposi sarcoma.
Keywords: KS, HIV, Case

Introduction

Kaposi sarcoma (KS) is a spindle-cell tumor caused by human herpesvirus 8 (HHV-8).[1] Generally, this cancer occurs in HIV-positive people because their immune system is weakened. Lesions of KS usually occur in the skin, oral mucosa, lymph nodes, and visceral organs. Visceral disease may occasionally precede cutaneous manifestations .[2,3] The classic form of visceral disease has a more indolent course than AIDS-related KS, which progresses over 10-15 years or more with highly gradual enlargement of cutaneous lesions.[4] CD4 lymphocyte counts and plasma HIV viral-load studies should be performed in patients with HIV infection. Chest radiographic findings in patients with KS are variable and nonspecific. These findings may include diffuse reticulonodular infiltrates, interstitial infiltrates, pleural effusions, hilar or mediastinal lymphadenopathy, or an isolated pulmonary nodule.[5-7] Thallium and gallium scans may help differentiate pulmonary KS from infection. Pulmonary KS lesions typically display intense thallium uptake but no gallium uptake, whereas the infection is often gallium-avid and thallium-negative.[8] Immunomodulation with interferon-α has clinical activity in KS, which may be mediated by its antiangiogenic, antiviral, and immunomodulatory properties.[9] In the pathology diagnosis, it is not diagnosed as sarcoma but a tumor arising from transformed cells with mesenchymal origin. HHV-8, also known as KS-associated herpesvirus (KSHV), is the primary factor in the pathogenesis of KS. KSHV proteins are uniformly detected in KS cancer cells.[10-12] This discovery spawned a renewed interest in the disease, resulting in a significant amount of research to explain the relationship between HHV-8 and the uncontrolled vascular proliferation found in KS. KS lesions contain tumor cells with a characteristically abnormal elongated shape, called spindle cells. The most typical feature of KS is the presence of spindle cells forming slits containing red blood cells.[13-19]

Case Report

Here, we report a case of a 66-years-old man who referred to dermatology clinic of Sina hospital in Tabriz, Iran, with progressive skin lesions, including purple patches, plaques, and nodules distributed on the legs, trunk, arms, and hands from 3 years ago (Figure 1).

No edema was observed on the lower extremities. There were also multiple red and purple plaques and nodules on the neck, lower face, nose, and ears (Figure 2A). Interestingly, there was a painless fleshy nodule with a size of 1.5 × 1.5 in his upper gingiva (Figure 2B).

Erythematous plaques on the face and neck are common signs of AIDS-associated KS. Biopsy of the skin lesion showed angiomatous proliferation of small vessels and slits with mild perivascular lymphocytic infiltration containing plasma cell and red blood cell (RBC) extravasation in the dermis (Figure 3A). Both the complete blood count (CBC) test and chest X ray were normal. HIV test was negative twice and CD4 count was in normal range. The staining results showed strong immune reactivity of all neoplastic cells to CD31 and CD34 (Figure 3B).

Blood test analyses using PCR showed that HHV-8 was positive. Treatment of the patient was started with doxorubicin and paclitaxel (dose of 340-380 mg/m^2), leading to improvement in his outcomes.

Discussion

KS is a vascular neoplasm which usually has a multifocal origin, with multiple vascular nodules on the skin and other organs. It also involves lymph nodes and visceral organs, especially the respiratory and gastrointestinal tracts.[20] Primary classic KS of the head and neck is rare in any

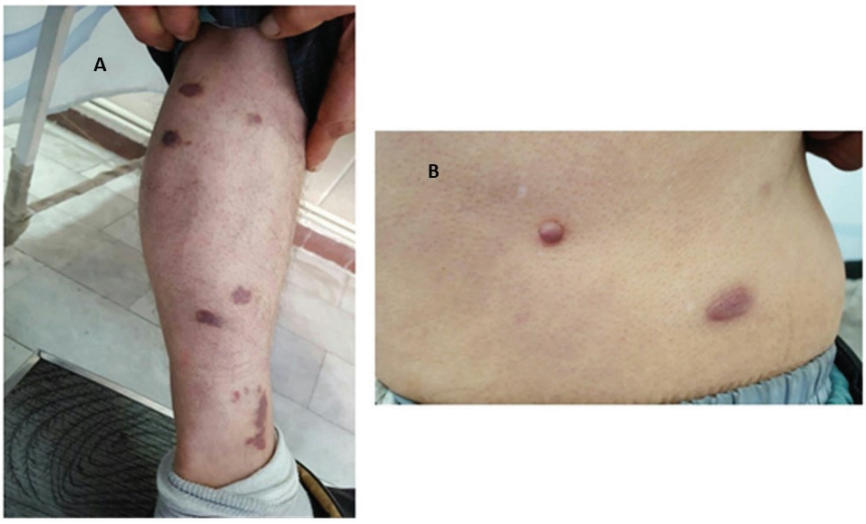

Figure 1. Well demarcated violaceous to brown plaques on the lower extremities and trunk.

Figure 2. (A) Violaceous to erythematous patches and plaques, involving the face and neck, a common site in AIDS- associated KS. (B) Asymptomatic Purple nodule in the gingiva, a less common presentation of classic KS (scale 40×).

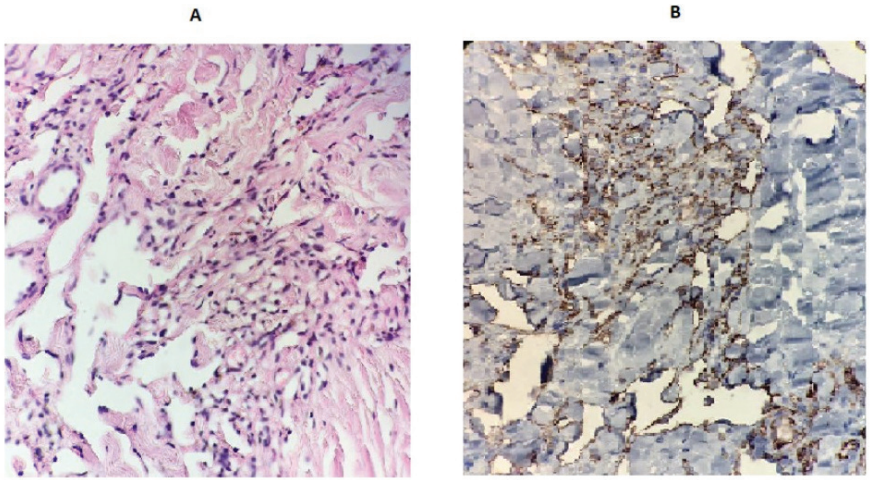

Figure 3. (A) Positive irregular dilated vascular channels associated with hemosiderin deposition and focal inflammation. (B) Immunohistochemistry for the vascular marker CD31

case. Ben et al reported a 71-year-old immunocompetent man who presented with a three-month history of a slowly progressive swelling in the right parotid region. They discussed the peculiarity of the presentation, the differential diagnosis, and the management strategy of such a rare disease.[21] A study carried out by Ascoli et al on a 57-year-old white female patient of Greek origin showed that non-HIV-related KS is an HHV-8-related

KS, an angioproliferative skin cancer that can cause pain, disfigurement, and limb dysfunction.[22] In the present study, we report the case of a 66-years-old man with progressive skin lesions, including purple patches with multiple deep plaques and red to brown nodules disseminated all over the body and oral mucosa. Although no definitive regimen was established, we started with doxorubicin and paclitaxel and shrinkage of lesions was obvious after two sessions. It would be beneficial to investigate the effectiveness of currently-available treatments in these patients. It can be concluded that non-HIV-related KS can have the same signs as HIV-related KS.

Competing Interests
None to be declared.

Authors' Contribution
Conceptualization: Mahsa Eslami.
Data curation: Sanaz Alizadeh.
Formal analysis: Faezeh Alavi.
Funding acquisition: Sina Safari.
Investigation: Sina Safari.
Methodology: Mahsa Eslami.
Resources: Faezeh Alavi.
Supervision: Sina Sfari.
Writing–original draft: Faezeh Alavi.
Writing–review & editing: Sanaz Alizadeh.

Ethical Approval
Informed consent was obtained form the patient for publication of this report.

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