

Case Report

Disseminated Kaposi sarcoma in a HIV negative patient

Guoshu Lin*, Hongyan Wang*, Xing Fan, Hui Li, Zaixing Wang, Da Lin, Sen Yang, Xuejun Zhang

Department Dermatology, Institute of Dermatology, The First Affiliated Hospital, Anhui Medical University Hefei China. *Equal contributors.

Received October 26, 2014; Accepted December 22, 2014; Epub March 1, 2015; Published March 15, 2015

Abstract: Kaposi sarcoma (KS) is a neoplasm of the endothelial cells. It often manifests with multiple vascular nodules on the skin and other organs. It is a systemic, malignant and multifactor disease and has a variable course. We describe an elderly Chinese man who had a rapidly growing maroon nodule on his right foot, both arms and cheekbones. KS in HIV-negative patients has only been reported sporadically.

Keywords: Kaposi sarcoma, disseminated, classic, HIV-negative, nodule

Introduction

Kaposi's sarcoma is a frequent skin cancer in HIV-positive patients, but is relatively uncommon in HIV-negative and non-immune compromised patients. Kaposi's sarcoma is the malignant proliferation of the endothelial cell vessels. Its genesis is still unclear; however, it seems to be related to the herpes virus infection (HHV-8). This neoplasia usually affects the lower limbs and the affected persons are mostly from the Mediterranean region. We present a case of disseminated Kaposi's sarcoma in a HIV-negative male.

Case report

A 77-year-old man from China was admitted to our clinic with maroon nodule on his right lower extremities, dorsum of feet. Similar lesions were also present on the extremities and cheek. These lesions have been existed for two months. In the course of disease he had a fever for ten days. He appeared under gastrointestinal bleeding, left pleural effusion six months ago and was referred to our clinic with a prediagnosis of herpes zoster and sarcoidosis two months ago. He had quitted alcohol and cigarette smoking ten years ago. The patient denied homosexual activity. Physical examination showed no abnormalities. Lymphadenopathy was not detected. Dermatological examination

revealed several purplish-brown nodules and plaques lesions sized 6 × 5 cm on the right dorsum of foot and lower extremities, similar lesions were present on the both arms and cheek as well (**Figure 1A-D**). Hemogram and biochemical tests were unremarkable except for mildly high total white blood cells of blood routine test. Serologic tests for RPR, TPPA and HIV were negative. Cd4 + cells were 16.9% (range: 27.4-42.1%), Cd8 + cells were 38.6% (range: 22.3-34.0%), and Cd4 +/Cd8 + ratio was 0.44 (range: 1.02-1.95). Natural killer cells (NK) were 28.3% (range: 8.1~23.7%) HbsAg, HbeAb and HbcAb were positive. Multiple nodular, patchy high density shadow was in thorax CT (**Figure 1**). Complete work-up (including abdominal and lymph node ultrasound examinations) disclosed no signs of visceral involvement of his KS. Biopsies and immunohistochemical stains from one nodule and hyperkeratotic lesion were assessed as KS. Immunoperoxidase staining for CD34, CD31, SMA, P53 and Ki-67 were positive (**Figure 1F-I**). Cryotherapy for papulonodular lesions and local excision for hyperkeratotic lesion were performed and follow up was planned. But the patient refused to continue treatment.

Discussion

Kaposi's sarcoma is a vascular neoplasm, described by Morris Kaposi in 1872. Kaposi's

Kaposi sarcoma in a HIV negative patient

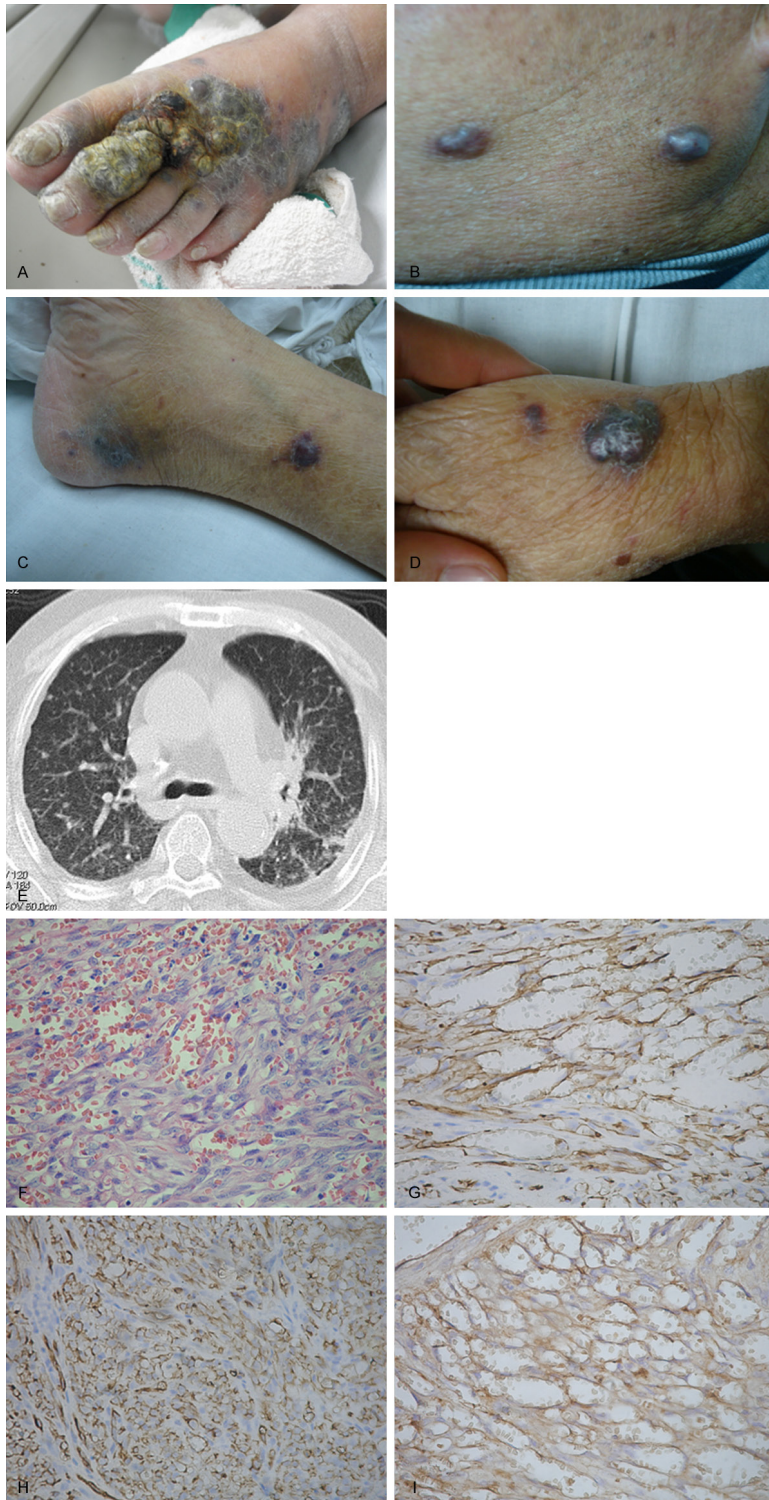


Figure 1. Multiple nodular, patchy high density shadow in thorax CT. A. Purplish-brown hyperkeratotic plaque and nodule on the dorsum of feet; B-D. Purplish-brown nodules on the cheek, right lower extremities, dorsum of hands and heels; E. Multiple nodular, patchy high density shadow in thorax CT; F. Spindle endothelial cells with mild atypia forming slits (H, E, $\times 400$); G-I. Immunohistochemical staining; G. CD31 endothelial cell cytoplasmic staining ($\times 400$); H. CD34 endothelial cell cytoplasmic staining ($\times 400$); I. FVIII endothelial cell cytoplasmic staining ($\times 400$).

sarcoma can be classified into four distinct forms: classic, endemic, iatrogenic and AIDS-associated [1, 2]. When epidemiological features are considered, some races and certain age groups such as middle aged and elderly Mediterranean or Jewish men and male gender are prone to have more KS and very rare in the rest of the world. However, KS patients with AIDS and African cases tend to be younger and the ratio of male/female is also reversed. Clinical presentation may be highly variable as well [3-5].

Classic KS is a spindle-shaped cell malignancy of endothelial cell origin that has a more benign and indolent course, and the affected organs are mainly skin, lower limbs and feet and rarely internal organs [6, 7]. The primary presentation on the face and visceral involvement is rarely described in the HIV-negative and nonimmunosuppressed individual. But the differential diagnoses for facial lesion include pyogenic granuloma, histiocytoma, hemangioma and angiosarcoma [8]. Immunohistochemical staining may be done for CD34 antibody. It yields positive results for endothelial lining of slit like spaces and for spindle cells [2]. The pathogenesis of KS is uncertain. Current data support the notion that KS is a vascular hyperplasia with a tight link to HHV-8 infection. We described a disseminated presentation of classic Kaposi sarcoma in a HIV negative patient from

China. This affected visceral involvement also. Disseminated KS found in the AIDS-associated and immunosuppression individual.

We describe a case of disseminated Kaposi sarcoma in HIV negative, face and visceral involvement Kaposi's sarcoma are rare in immunocompetent individuals and but must be included in the differential diagnoses of suspicious facial lesions. Lesions are usually not associated with systemic involvement and can be treated with surgical excision. But the lesions of patient were extensive. And he refused the continual treatment and died after two months.

Acknowledgements

This work was funded by a grant from the 'CMA-L'OREAL China Skin/Hair' (S2013101026).

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Xuejun Zhang or Dr. Sen Yang, Institute of Dermatology, Anhui Medical University, 69, Meishan Road, Hefei 230032, Anhui, PR China. E-mail: ayxjz@vip.sina.com (XJZ); yang-sen@medmail.com.cn (SY)

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