Journal of Clinical Case Reports

Case Report Open Access

A Case Report on Disseminated Non-Epidemic Kaposi Sarcoma in Non-HIV Non-Organ Transplant Heterosexual Yemeni Man: A Discussion of Possible Etiology

Taha R[1]*, Al-Osaimi R[2], Omar A[2] and Al-Fares M[2]

[1]*Department of Infection Diseases, King Abdul-Aziz Hospital, Jeddah, Saudi Arabia*
[2]*Department of Internal Medicine and Infection Diseases, King Abdul-Aziz Medical City, Jeddah, Saudi Arabia*

Abstract

Kaposi's sarcoma (KS) is the most prevalent malignancy in patients with HIV infection. few reports describe KS in HIV-negative, non-immune compromised patients in the United States, but the incidence of the disease in the area of middle east in non-HIV, Immunocompetent is not well recognized. It affects the endothelial cells of the skin and mucous membranes. The cases seen in HIV-negative men were less aggressive and rapidly progressing than those in people with HIV. However, although, they tended to be more malicious than in the classical type of KS which is seen mostly in men of Mediterranean origin and occurred at an earlier age. We are presenting a rare case of *Mucocutaneous* KS diagnosed based on histopathology in an 82-year-old Yemeni male patient, who is seronegative HIV, non-immunocompromised with no history of organ nor bone marrow transplantation.

Keywords: Kaposi sarcoma; Non-epidemic Kaposi sarcoma; Apparently immunocompetent host

Abbreviations: AIDS: Acquired Immunodeficiency Syndrome; HIV: Human Immunodeficiency Viruses

Introduction

Kaposi's sarcoma (KS) is a systemic malignant disease, involve neoplastic cutaneous lesion with or without internal organ involvement. It was first described by Moritz Kaposi a Hungarian dermatologist. It has four clinical types such which is classic (Mediterranean), endemic (African), post-transplant, and epidemic or AIDS-related. All four forms of KS have similar histo-pathogical features; in addition KS is strongly associated with HHV-8 virus which can be found in most cases Regardless of the variable clinical presentations and fragility of immune system [1] what we know that most cases are strongly associated with partial or complete immune suppression. Very few publications on the diagnosis of Kaposi's sarcoma has been described in immunocompetent population in the area of Saudi Arabia and southwestern area. In the present case report, we describe a classic case of non-AIDS/non-immunocompromised KS. He did not present with any co-morbidities associated with KS or HIV infection. So, this case report improves the knowledge about non-AIDS KS and adds to the already available literature.

Case Report

An 82-year-old male rural Yemeni known case of hypertension, bronchial asthma, dyslipidemia, ischemic heart disease presented with progressively erythematous violaceous macules which had progressed to plaque like lesions that involve whole body for sex months. lesion started in lower extremity progress with time to involve upper extremity, trunk, mouth. He denied any history of Fever, convulsion, numbness, weight loss, loss of appetite or night sweats. Direct questioning regarding his occupational history revealed that he is a farmer and a sheep shepherd. Patient gave history of Khat chewing for the last twenty years. The patient had no history of immunosuppression or HIV. No family history of similar illness and no family history of hematological malignancy.

Upon admission, patient was fully conscious, oriented. He was hemodynamically stable. There are several well-defined, papules, nodules, and violaceous (purple) skin lesions measuring between 3-10 mm, multi-pigmented plaques, friable, tender, nodulo-ulcerative lesions involving entire body involving trunk, hands, feet and forearm (Figures 1 and 2). There were also violaceous nodular lesions involving the hard palate and oral mucosa. No regional lymph nodes no oral or genital involvement. Remainder of the physical examination was within normal limits.

Investigations revealed Leukocytic count 11.5 per mm [2,3], with a normal differential, reduced hematocrit 24. Renal and hepatic profile, ANA/Anti ds DNA, HBs Ag/Anti HCV Ab, U1 RNP, c and p ANCA, cryoglobulins, HIV serology and HIV PCR, Immunoglobulin level were normal. Chest radiograph was within normal limits. CT chest was not revealing any masses or nodule. CT abdomen and pelvis were within normal limits. Patient underwent diagnostic skin biopsy which showed the entire reticular dermis is filled with a vascular neoplasm, that is made up of nodules of spindled cells that enclose small capillary sized and slit-like vascular spaces (Figures 1 and 2). Several atypical mitotic figures seen in proliferating spindled cells. It wass CD34 positive in the immunohistochemical analyses. Besides, the human herpes virus-8 latent nuclear antigen-1 (HHV-8 LNA-1) and nuclear staining was detected which was found compatible with KS (Plaque stage). Skin biopsy is shown in Figures 3 and 4. Unfortunately patient developed hospital acquired pneumonia and died before starting radiotherapy, despite no clear immunodeficiency causing his disease.

Discussion

Kaposi's sarcoma (KS), is a form of mesenchymal neoplasm commonly associated with AIDS, it is malignant in nature and

***Corresponding author:** Taha R, Department of Infection diseases, King Abdul-Aziz Hospital, Jeddah, Saudi Arabia, Tel: +966545922187; E-mail: taharabab@gmail.com

Received October 16, 2019; **Accepted** October 24, 2019; **Published** October 31, 2019

Citation: Taha R, Al-Osaimi R, Omar A, Al-Fares M (2019) A Case Report on Disseminated Non-Epidemic Kaposi Sarcoma in Non-HIV Non-Organ Transplant Heterosexual Yemeni Man: A Discussion of Possible Etiology. J Clin Case Rep 9: 1285.

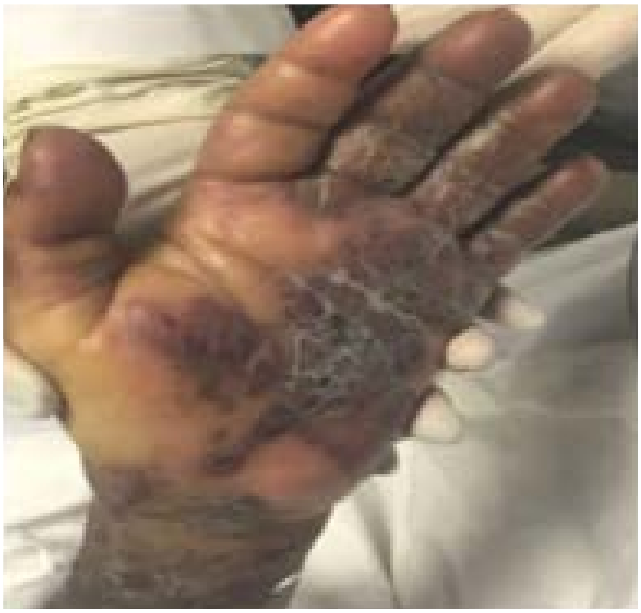

Figure 1: Multi pigmented plaques on the palm.

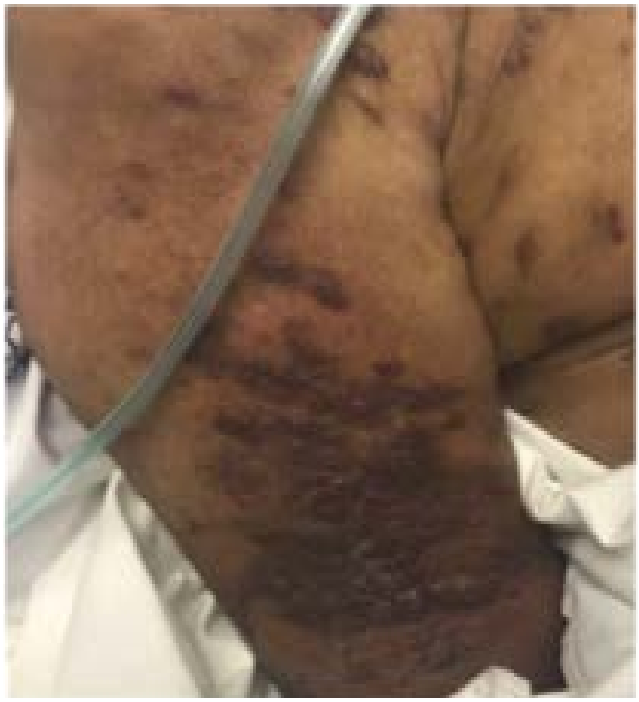

Figure 2: Multi pigmented plaques on the arm.

progresses slowly. It is often characterized by the proliferation of the connective tissue and capillaries. KS manifests as nodules and red-purple plaques [2]. KS is broadly classified into four types [4-6]: (i) classic or endemic, (ii) African, (iii) Epidemic, and (iv) KS associated with renal transplantation. The classic or endemic form is most prevalent in the males with an affected male to female ratio of 15:1 reported in Mediterranean countries, generally in the age group of 50 to 70 years. Clinically, nodules and plaques are seen in the areas near the trunk and inferior limbs. The classical or endemic form of KS has a long clinical course (10 to 15 years) [7]. The African form of KS is also frequently observed in males between 25 and 40 years of age. There are two subtypes of the African form of KS: (i) a less aggressive type, and (ii) a more aggressive type [8]. The epidemic form of KS is typically observed in AIDS patients and accounts for about 90% of malignant neoplasms in these patients. Dispersed mucocutaneous lesions with visceral and lymph node involvement are the characteristic features of the endemic form of KS. The clinical course of this type of KS is very short as the patients die very early due to disease progression or secondary complications [6-9]. The form of Kaposi's sarcoma which is associated with renal transplantation has an incidence of 0.4% in the USA. It progresses slowly but the visceral involvement results in fatality [5,10]. All forms of KS typically are characterized by similar histopathological patterns. The mostly observed histological patterns in KS are rich cellular component without atypia and with vascular lacunae [10]. KS is caused by human herpes virus 8 (HHV-8) and is prevalent worldwide [11,12]. The HHV-8 virus infection alone is not enough to cause KS [13]. Mostly, KS is presented in association with AIDS and in immunocompromised conditions [14].

HHV-8 virus expresses several proteins that affect the immunity of the host, prevents apoptosis and increases the rates of cellular proliferation and cytokine production, and promote angiogenesis,

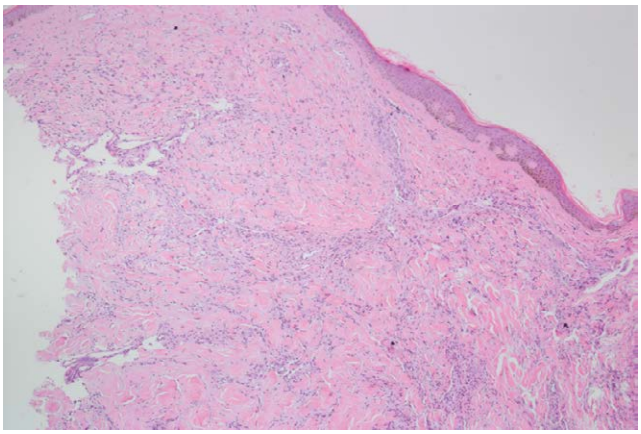

Figure 3: H and E stain 4x dermal-epidermal tissue in sub epithelial region, there is Spindle cell tumor infiltrating the dermis

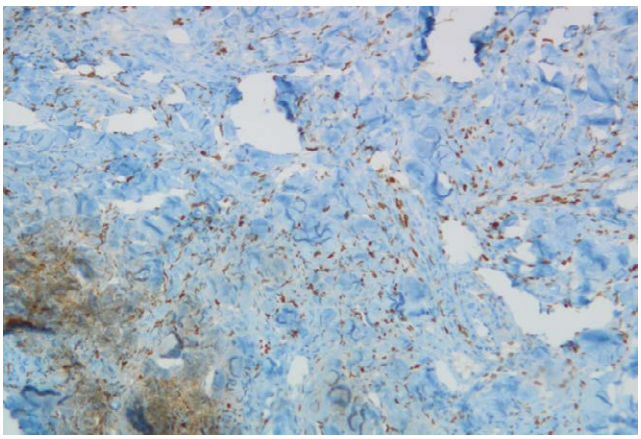

Figure 4: Immunohistochemistry IHC 10x, the neoplastic cells is positive for HHV8.

consequently, results in the pathological condition of KS. KS is the second most prevalent form of cancer in HIV-infected patients [15]. However, several cases of KS without associated AIDS also have been reported [16]. The first non-AIDS associated KS patient was reported in 1872 who was of eastern Mediterranean origin [17]. Fourteen cases of Kaposi's sarcoma in negative HIV patients were diagnosed at King Faisal Specialist Hospital and Research Center, Saudi Arabia, surprisingly that the incidence of 5.3 percent compared with an incidence of 0.4 percent in renal transplant recipients from Western countries. seven out of fourteen were from southwestern region of Saudi where Khat chewing Is common [18]. Low CD 4 count is independent risk factor for KS. some studies inferred that smoking and the lower BMI the higher risk of low CD4 count in non-HIV patients [19]. Since HHV-8 itself is not capable enough to cause KS, the appearance of KS in patients without immunosuppression/AIDS imply the existence of other mechanisms. So, detailed studies about the pathogenesis of KS is warranted in these patients. Factors like lifestyle habits, genetic component, gene environment interaction, and host-HHV-8 interactions should be given utmost importance to elucidate the patho-mechanism of KS. Furthermore, our patient reported history of exposure to Khat which is a unique plant that has a stimulant effect. it is often used either through chew or by adding it to a hot beverage. It is widely used in Yemen along with other countries in East Africa. Khat has been implicated in many studies for its direct effect as a carcinogenic substance. For instance, in a study which included 5938 Yemenis which were diagnosed with different malignancies, the oral cavity cancer was more in men who exposed to Khat than women who did not in a ratio of 2.06:1 [20]. Unusual recalcitrant classic Kaposi's sarcoma in a Non-AIDS Yemeni Man published in 2016 [21]. Cherinet et al. raised a question about possibilities of Khat causing low immunity. However, up to our knowledge, there are limited studies which assess the immunological effect of Khat in association with AIDS negative diseases such as Kaposi sarcoma. In the present case, we were not able to provide radiotherapy to the patient as he died of hospital-acquired pneumonia. The line of treatment or management of KS depends on various factors such as the location and the type of KS, the rate of clinical course of the disease, symptoms of the patients, the count of lesions, host immunity, and the presence of other comorbidities.

Conclusion

Only few sporadic cases of nonepidemic KS have been reported without clear causes. This report may open a new path for the research of KS and agents that affect CD4 count. The early diagnoses are the better prognosis. The prevention of the disease by improving lifestyle and avoiding possible risk factors might improve the prognosis of the disease.

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