Case Report

Kaposi's sarcoma as a presenting manifestation of HIV

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Abstract

Kaposi's sarcoma is a multi-focal vascular tumor involving skin and the other organs. HIV associated Kaposi's sarcoma is one of the AIDS defining condition. It is rarely reported from India. We report a 40-year-old heterosexual married male with widespread cutaneous lesions of Kaposi's sarcoma without any oral lesions or systemic association as a presenting manifestation of HIV.

Key words: AIDS, HIV Infection, kaposi's sarcoma

INTRODUCTION

Kaposi's sarcoma (KS) is a well-known multifocal vascular tumor first described by Hungarian pathologist Moritz Kaposi in 1872, involving skin and other organs.[1] It is the most common neoplasm in patients with AIDS.[2] Although there is high prevalence of HIV in India, KS is rarely reported. HIV associated (Epidemic) KS is one of the AIDS defining conditions. We report a young heterosexual HIV positive male with KS as a presenting manifestation.

CASE REPORT

A 40-year-old married male presented with a 2-year history of multiple asymptomatic skin eruptions. Lesions first appeared over lower limbs and then gradually spread over trunk and upper limbs. In addition, painful lesions were also noticed on leg for past one month. Dermatological examination showed multiple, well-defined, non-scaly, discrete, violaceous to hyperpigmented plaques of variable size over trunk and extremities [Figures 1a and b]. Multiple, tender, skin colored to erythematous plaques present on left lower leg [Figure 2]. Oral cavity was normal. Looking to the morphology of the skin lesions we thought of KS's as our diagnosis. On further enquiry, there was no history of fever, diarrhea, or weight loss. Patient admitted having multiple unprotected heterosexual exposures in past but denied having any homosexual contact. There was no history of blood transfusion or intravenous drug abuse. There was no history of any drug intake prior to the onset of lesions. Patient otherwise was in good general health and systemic examination was normal.

Laboratory investigations including complete blood count, renal function, and liver function tests were normal. ELISA for HIV was positive. Sputum was negative for acid fast bacilli (AFB). Serology for hepatitis B and C was negative. Serum VDRL was non reactive. CD4 count was 173 cell /mm^3. Chest X-Ray and ultrasound sonography of the abdomen were normal.

Skin biopsy showed innumerable jagged irregular thin walled vascular spaces infiltrating the collagen with foci of moderately dense plasmacytic infiltrate in upper and mid reticular dermis. At places promontory sign (proliferation of new vessels is seen surrounding preexisting vascular structures) was present [Figure 3]. A diagnosis of KS was made on the basis of clinical presentation and histopathological examination.

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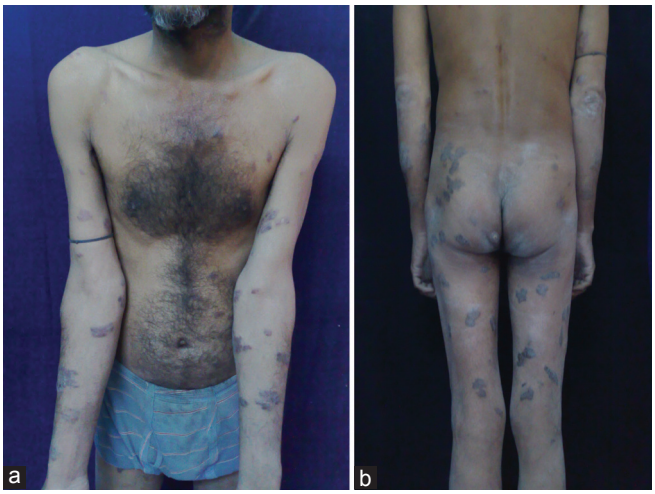

Figure 1a and b: Showed multiple, well-defined, non-scaly, discrete, violaceous to hyperpigmented plaques of variable size over trunk and extremities

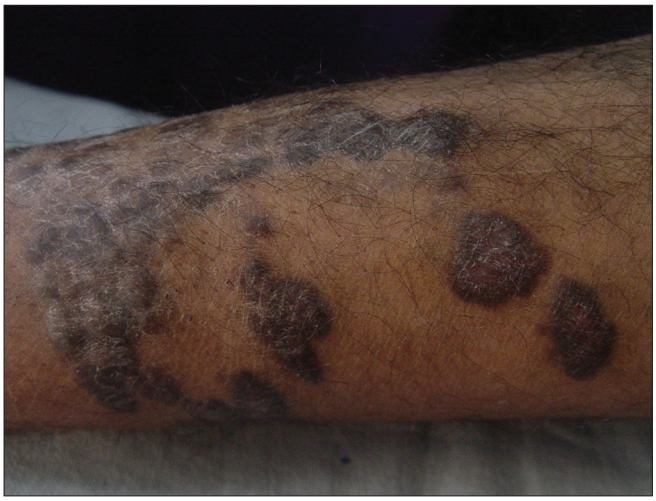

Figure 2: Close-up photo on left lower leg

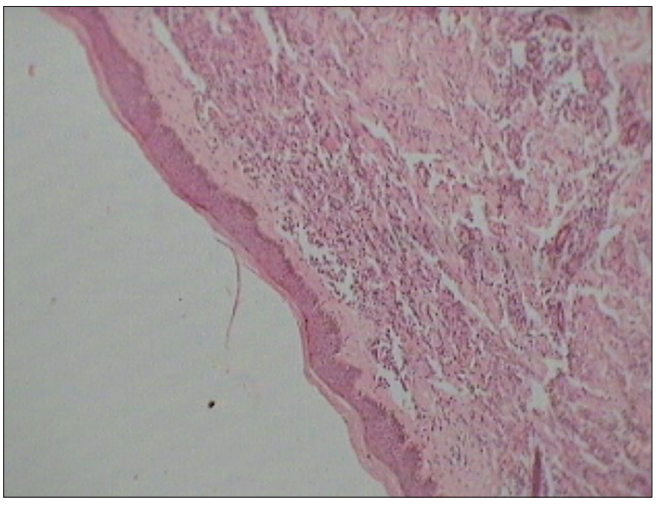

Figure 3: Histopathology showed dermal proliferation of innumerable jagged irregular thin walled vascular spaces infiltrating the collagen with foci of moderately dense plasmacytic infiltrate with promontory sign (H and E, 10×).

DISCUSSION

KS is a multifocal, systemic tumor of endothelial origin. It has four clinical variants, classical Kaposi's sarcoma, endemic African KS, KS associated with immunosuppressive therapy, and KS associated with AIDS. AIDS related KS is known as epidemic KS.[3] KS is the most common malignancy associated with HIV.[2]

HHV8 is thought to be the initiating factor in the pathogenesis of KS. There have been reports of the presence of virus in the lesions.[4] It can be transmitted sexually and by other means more commonly in homosexuals than heterosexuals. Receptive anal intercourse is a significant risk factor. Most of the initial reports of epidemic KS were described in homosexual men and very few cases have been reported amongst heterosexual males. The predominant mode of HIV transmission in India is heterosexual and this might explain relatively low prevalence of KS in India.

The first case of AIDS associated KS from India was reported in 1995 in a 35-year-old female sex worker from Mumbai.[5] Since then very few cases of AIDS-associated KS have been reported.[4,6,7]

Clinically, AIDS related KS differs from its classical form in terms of its rapid clinical course with widespread dissemination.[8] It generally affects skin, mucous membranes, gastrointestinal tract, lymph nodes, and lungs. The oral mucosa is the initial site of localization in 10–20% of all HIV associated KS, frequently involving palate.[6] Our patient, inspite of having multiple cutaneous lesions, had no oral lesions. There was no systemic affection with KS.

KS can present at any time during the course of HIV infection. It accelerates the clinical course of HIV infection, and generally occurs at CD4 count <200 cells/mm^3.[9] Lesions of KS are asymptomatic and usually start as macule, progressing to form papule, plaque, and nodules. Lesions can at times be painful.

The clinical differential diagnosis includes bacillary angiomatosis, lichen planus, drug eruptions, coccidioidomycosis, pyogenic granuloma, angiodermatitis or pseudo-KS, and oral hemangioma.[4,7]

Histopathology usually confirms the diagnosis and is characterized by profuse vascular proliferation showing slit-like spaces with presence of solid cords and fascicles of spindle cell arranged between vascular channels. Immunohistological detection of CD31, CD34 antigens, FVIII-Rag, and sialic acid expression are important for correct diagnosis of KS.[10]

Progression of AIDS-related KS depends upon the tumor extent, as well as level of immune-suppression, opportunistic infections, and treatment of HIV infection.[11] The staging system developed by National Institute of Allergy and Infectious disease AIDS clinical trials group (ACTG) predicts the prognosis of KS on account of tumor extent, immune status and the presence or absence of systemic disease. Good prognosis is expected with CD4 count >200/mm^3, only cutaneous involvement and no "B" symptoms.[12]

The absence of oral lesions, extensive cutaneous affection without systemic involvement, good general health despite low CD4 counts and occurrence in heterosexual individual were some of the unusual features noted in our patient. This inspires us to share this case.

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