

**CASE REPORT**

Kaposi Sarcoma Presenting as an Index Sign of HIV Infection in an Indian

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Abstract

A -38 year old, married man presented with multiple asymptomatic livid reddish brown plaques, papules over lateral side of left ankle and dorsum of left foot along with difficulty in walking for last six months. Histopathological examination of biopsy from the lesion showed a Kaposi's sarcoma (KS). Subsequently, his serology for the HIV viruses was done and found to be positive for HIV-1.

Key Words

Kaposi's sarcoma, HIV, Infection

Introduction

Kaposi's sarcoma (KS) is an angioproliferative disease characterized by proliferation of spindle-shaped cells. Kaposi's sarcoma was first described in 1872 by Moritz Kaposi as a disease seen in elderly men of Mediterranean or Jewish descent. In 1981, initial reports described it in homosexual men with AIDS, but recent publications have also reported its incidence in heterosexual males (1). Patients with AIDS-related KS usually present with cutaneous lesions, mucous membrane lesions, or lymph node involvement. Visceral involvement occurs in 50% of the patients, especially of the lungs and gastrointestinal tract. Lung involvement occurs in 20% of the patients and is the most life-threatening form of the disease (2). Recent findings of a herpetic-like viral DNA in Kaposi's sarcoma tissue have suggested an infectious co-factor for the disease (3). We report a case of KS in an HIV-1 positive heterosexual male. This case is reported due to paucity of Kaposi's sarcoma in Indian literature and moreover in this case KS was the index presentation of HIV disease.

Case Report

A -38 year old, married man presented with asymptomatic dark colored flat and noduloulcerative

lesions over lateral side of left ankle and dorsum of left foot along with difficulty in walking for last six months. He was on treatment with anti actinomycotic drugs for last 4 months but the disease continued to progress. Lesions first started on the lateral side of left ankle as purplish macules followed by papules and plaques with gradual progression to involve adjacent skin of lower leg and dorsum of foot. Older lesion became nodular in shape and got eroded in due course of disease. There was no history of trauma, fever with chills, cough, diarrhoea, weight loss, genital lesions or blood transfusion prior to onset of lesions. He admitted history of multiple unprotected heterosexual extramarital sexual exposures. He denied history of any homosexual exposure or use of intravenous drugs. He did not have any other systemic complaints. Clinical examination revealed mild pallor with enlarged axillary, cervical and inguinal lymph nodes which were firm, mobile and non-tender.

Cutaneous examination revealed multiple dark skin coloured to violaceous compressible papules, nodules and plaques over lateral side of left ankle and adjacent area of lower leg and dorsum of foot (*Fig-1*). Some of the papules and nodules showed central ulceration with hyper

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Fig. 1 Shows Dark violaceous Papules, Nodules and Pyogenic Granuloma like Lesions Over Lateral Aspect of Left Ankle

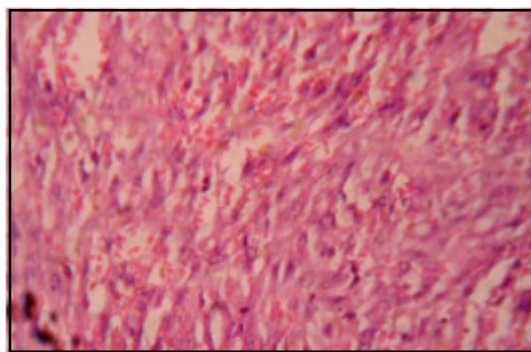


Fig. 2 Histopathology Shows Proliferating Spindle Cells & Slit Like Vascular Channels Containing Erythrocytes (H&E x 100)

proliferative granulation tissue in centre like pyogenic granuloma while other had blackish scab in centre. Lesions were non tender but ulcerated lesions tend to bleed on touch. A clinical diagnosis of Kaposi's sarcoma was suspected. Under all strict precautions biopsy from lesion was taken up for histopathological examination. Histopathological examination of the representative skin lesions showed proliferation of irregular, jagged, lymphatic like vascular channels lined by single layer of plump, pleomorphic endothelial cells (Fig-2). There was extravasation of RBCs forming linear streaks amidst the collagen bundles with a moderate perivascular lymphocytic infiltrate. Large areas of hemosiderin deposition were also seen on Prussian blue staining (Fig-3). Many abnormal mitotic figures were also seen in section. A diagnosis of Kaposi's sarcoma (KS) was made. After pre-test counseling, the patient was advised for ELISA test for HIV which turned out to be positive for HIV-1 for the first time. His HIV status was again reconfirmed by two more positive HIV-ELISA with two different antigens.

His laboratory investigations for complete blood count (CBC), liver and renal function tests were normal. X-ray chest was also normal. Serology for HbsAg and VDRL was negative. His absolute CD4 count was 156/mm³. Polymerase chain reaction (PCR) for human herpesvirus 8 (HHV-8) could not be done because of lack of facilities.

Discussion

Kaposi's sarcoma (KS) is a multifocal vascular proliferative disorder which starts as a reactive angioproliferative and inflammatory process, but progress to become a monoclonal neoplastic process (4). This is probably the result of close complex interactions between inflammatory cytokines, angiogenic factors, human herpesvirus-8 (HHV-8), and in HIV-positive patients, the HIV virus itself (5). The possibility that the HHV 8 is the

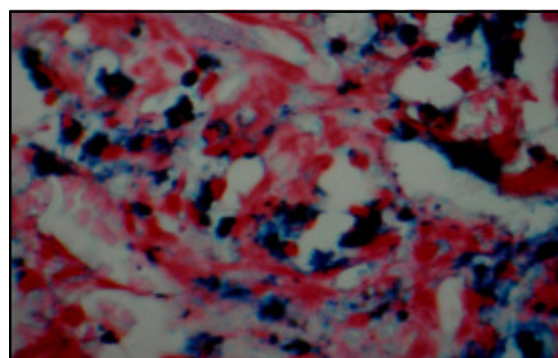


Fig.3 Prussian Blue Staining Showing Hemosiderin Deposits

inciting factor in KS pathogenesis was considered because of the presence of virus in pathological lesions of KS (6). On the basis of clinical and epidemiological features, four types of KS have been recognized: classic, endemic (African), iatrogenic and epidemic (AIDS related). The course of Kaposi's sarcoma ranges from indolent, with only skin manifestations to fulminant with extensive visceral involvement (7). HIV- associated KS was first recognized in 1979 when an epidemic of KS was identified in the homosexual community in New York (8). Upto 30% of those infected homosexual men developed KS (9). In patients with other modes of HIV transmission, the prevalence of KS was less than 5% (10). The World Health Organization (WHO) clinical staging for HIV/AIDS recognizes KS as an AIDS-defining illness. HIV-associated KS is common among homosexual men and it is uncommon in countries where HIV is predominantly transmitted heterosexually. It differs from the classic disease in the rapid evolution of the lesions, and atypical distribution affecting the trunk and mucous membrane. Kaposi's sarcoma usually presents initially as violaceous skin lesions, but oral, visceral, or nodal KS may precede cutaneous involvement (11). Unusual cutaneous forms



of KS include presentation like lichen planus, thrombophlebitic, telangiectatic, ecchymotic, pyogenic granuloma, indurated plaque, keloidal, warty exophytic, and lymphangiomatous (12). The present reported case of KS was heterosexual in nature and had pyogenic granuloma like lesions in the plaque of the diseased skin (Fig-1).

Despite high prevalence of HIV/AIDS in India, very little is known regarding the spectrum of neoplasm in patients with AIDS. The first report of AIDS associated Kaposi's sarcoma in India was described in the year 1993 (13). Since then very few case reports of AIDS associated Kaposi's sarcoma are described in Indian literature. This low prevalence of KS may be attributed to the low prevalence of HHV-8 in our country (14). HIV-associated KS is usually asymptomatic, may be seen at any stage of HIV infection, even at normal CD4+ count (15). Prognosis of patients with epidemic KS depends not only on the extent of KS but also on other factors such as the level of immunocompromization, opportunistic infections and antiretroviral treatment (16). As seen in other HIV-infected without KS, mortality in patients with KS is contributed mostly by other opportunistic infections.

An excellent staging system has been developed by the National Institute of Allergy and Infectious Disease AIDS clinical trials group (ACTG). It distinguishes patients on the basis of extent of tumor (T), immune status (I) and severity of systemic illness (S)(17). Good prognosis is expected when CD4 count is $>200/\text{mm}^3$, if only cutaneous involvement is seen and in absence of "B" symptoms (fever, weight loss, diarrhea). The fundamental basis for the treatment of AIDS-related KS is the suppression of HIV replication by starting antiretroviral treatment and treating the opportunistic infection. HAART can significantly decrease the incidence of KS.(18) It can slow the rate of progression of KS and even result in regression of the pre-existent disease.(19)

Conclusion

We report this case for its rarity in India and the occurrence of KS as the index manifestation of HIV disease in a heterosexual male.

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