

## A Varied Presentation of Kaposi's Sarcoma

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### ABSTRACT

Kaposi's sarcoma (KS) is a multifocal cutaneous and extra cutaneous vascular proliferative disorder. In India, there are only a few cases of HIV-associated KS in published literature. A 65-year-old married man presented with asymptomatic elevated skin lesions over the scalp, face, right arm & trunk with duration of seven months. Skin biopsy showed proliferation of thin walled capillaries with formation of slit like spaces, spindle cell proliferation, abundant extravasation of RBCs and moderately dense inflammatory infiltrate in the dermis. Thus, a clinical diagnosis of cutaneous KS was confirmed. On testing with ELISA for HIV, the patient was for the first time diagnosed as HIV I reactive. So we wish to report this case due to rarity of its presentation.

**Key Words :** HIV, Kaposi Sarcoma, Cutaneous

### Introduction :

Kaposi's sarcoma (KS) is a multifocal cutaneous and extra cutaneous vascular proliferative disorder. KS was initially described by the Hungarian dermatologist, Morris Kaposi in 1872. There are four recognized clinical subsets of KS-Classical, Endemic (African), KS associated with non-HIV induced immunosuppression and with HIV infection (epidemic). HIV-associated KS was first recognized in 1979 when an epidemic of KS was identified in the homosexual community in New York.<sup>1</sup> The World Health Organization (WHO) clinical staging for HIV/AIDS recognizes KS as an AIDS-defining illness. In India, there are only a few cases of HIV-associated KS in published literature. So we wish to report this case due to rarity of its presentation.

### Case Report :

A 65 year old male, farmer by occupation, resident of Chhindwara (MP), presented with multiple skin colored nodules over scalp (**Fig. 1**), face (**Fig. 2**), right arm (**Fig. 3**) and trunk (**Fig. 4**) since last 7 months. He had history of multiple unprotected

heterosexual but no homosexual exposure. He did not have any mucosal involvement, haemoptysis, haemetemesis and malena. He gave no history of blood transfusion or intravenous drug abuse. On general examination patient had cachectic look. Vital parameters were stable. Systemic examinations were normal except mild hepatosplenomegaly. Cutaneous examination revealed discrete bilateral assymetrical nontender skin colored nodular lesions with some of them showing violaceous hue. Bilateral nontender mobile non matted soft cervical lymphadenopathy.

With a clinical differential diagnosis of Kaposi sarcoma and Bacillary angiomatosis, we referred patient to the Integrated Counseling Testing Centre (ICTC) for HIV testing and was for first time diagnosed as HIV I Reactive. Absolute CD 4 count was 270.00 cells/cu mm. Other laboratory investigations revealed Haemoglobin of 9.5 gm/dl, Total white blood cell count of 7,000 cells/mm<sup>3</sup> and a differential count of Polymorphs - 47%, Lymphocytes - 43%, Monocytes - 02% & Eosinophils - 08%. Liver and Renal function tests were within normal reference range. Serology for HBsAg and RPR were non-reactive. Radiograph chest was normal. Sputum for acid fast bacilli was negative. Skin biopsy report (**Fig. 5**) revealed proliferation of thin walled capillaries, along the blood vessels of superficial plexuses. The capillaries were arranged in a clustered pattern and could be seen as rounded spaces filled with red blood cells,

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which extend between the collagen bundles formed by the proliferation of spindle cells arranged in short fascicles. Few inflammatory cells, histiocytes & lymphocytes were also seen. Thus, a clinical diagnosis of cutaneous kaposi sarcoma was confirmed.



**Fig. 1 :** Multiple skin colored nodules on scalp



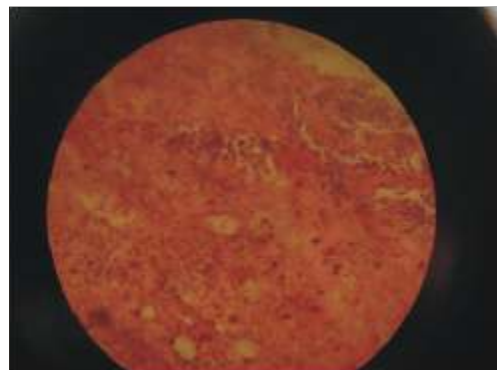
**Fig. 2 :** Multiple skin colored nodules on face



**Fig. 3 :** Multiple skin colored nodules on right arm



**Fig. 4 :** Multiple skin colored nodules on trunk



**Fig. 5 :** Proliferation of thin walled capillaries, along the blood vessels of superficialvascular plexuses. X100

**Discussion :**

KS was initially described by the Hungarian dermatologist, Morris Kaposi in 1872. There are four recognized clinical subsets of KS- Classical, Endemic (African), KS associated with non-HIV induced immunosuppression and with HIV infection (epidemic). HIV-associated KS was first recognized in 1979 when an epidemic of KS was identified in the homosexual community in New York<sup>1</sup>. 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; it is uncommon in countries where HIV is predominantly transmitted heterosexually. Because of this, despite high prevalence of HIV/AIDS in India, only 10 cases of KS exist in the published literature. This low prevalence of KS may be attributed to the low prevalence of HHV-8 in our country<sup>2</sup>. HIV-associated KS is usually asymptomatic, may be seen at any stage of HIV infection, even at normal CD4+ count and CD4+ count is not a consistent prognostic indicator.

In contrast to the other variants of KS, HIV associated KS can appear on any part of body with initial lesions frequently developing on the face, especially on the nose, eyelids, and ears- and on the trunk. Lesions of KS usually start as macule, progress to form papule, plaque and nodule. Sometimes pronounced lymphedema is observed in association with KS on the extremities, scrotum, penis, and face, especially when the eyelids are affected. Unusual cutaneous forms of KS include presentation like lichen planus, thrombophlebitic, telangiectatic, ecchymotic, pyogenic granuloma, indurated plaque, keloidal, warty exophytic, and lymphangiomatous<sup>3,4</sup>. The lesions of AIDS-related KS, frequently involve the mucous membrane, lung, lymph node, and gastrointestinal tract. The oral mucosa is the initial site of localization in 10-20% of all HIV-associated KS and is frequently located on the palate. Diagnosis of cutaneous KS is made on clinical ground and confirmed by histopathological examination.

Prognosis of epidemic KS is related to the extent of KS, underlying immunosuppression, opportunistic infections, and treatment of HIV infection<sup>3</sup>.

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 tumor extent, immunological function and the presence or absence of systemic disease<sup>4</sup>. Good prognosis is expected when CD4 count is  $> 200/\text{mm}^3$ , only cutaneous involvement seen and no "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, slow the rate of progression of KS and even result in regression of the preexistent disease<sup>5</sup>.

Local treatment modalities include cryotherapy, intralesional vinblastine or vincristine, laser and radiation therapy. It is useful when skin or mucosal lesions are few and there is no systemic involvement. Indications for systemic therapy include (1) visceral involvement, (2) extensive KS associated with lymphedema, (3) extensive and rapidly progressing KS and (4) failure to respond to local therapy.

US FDA has approved liposomal anthracyclines (doxorubicin and daunorubicin) as the first line agent for KS. Paclitaxel appears to be more effective than liposomal anthracycline but because of the high toxicity paclitaxel is second line therapy<sup>6</sup>. Other systemic therapy includes interferon-alpha and a combination of chemotherapy. We report this case for its rarity in India and the occurrence of KS as the presenting manifestation of HIV disease.

**Conflicts of Interest :** None Reported By authors

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