Kaposi's sarcoma: An interesting case report in a human immunodeficiency virus-positive heterosexual male

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Abstract

Kaposi's sarcoma (KS) is an angioproliferative neoplasm that affects skin and other organs. It is one of the acquired immune deficiency syndrome (AIDS)-defining conditions, which tends to occur at low CD4 count. It is the most common neoplasm among patients with AIDS in the Western population. It is rarely reported from India. We report the case of a 38-year-old human immunodeficiency virus-positive heterosexual male, with an unusual presentation of KS.

Key words: Acquired immune deficiency syndrome, human herpesviruses 8, human immunodeficiency virus, Kaposi's sarcoma, promontory sign

Introduction

Acquired immunodeficiency syndrome (AIDS) is a global epidemic affecting many millions of lives, across the world. In 1981, an epidemic form of Kaposi's sarcoma (KS) was reported in homosexual men, which was followed by the discovery of the human immunodeficiency virus (HIV) in 1983.[1] KS was first described by Moritz Kaposi in 1872 as "idiopathic multiple pigmented sarcoma." KS is an angioproliferative neoplasm that affects skin and other organs. It is one of the AIDS-defining conditions and the most common neoplasm among patients with AIDS in the Western population.[2] Even though there is an increase in the incidence of AIDS in India, <30 cases of KS have been reported indicating the low prevalence of the causative organism and heterosexual predominance in modes of HIV transmission.[3]

We report the case of an HIV-positive heterosexual male with an unusual presentation of KS.

Case Report

A 38-year-old married male presented with a 3-month history of asymptomatic multiple dark raised lesions over both legs and a week history of swelling of lower limbs and scrotum, resulting in difficulty in walking and urination. Initially, the lesions started as multiple dark-colored flat lesions over the right leg and then progressed to form nodular lesions in a span of 1 month. Lesions progressed in size and number to form large dark-grouped nodules over both the lower limbs.

Access this article online

Quick Response Code:

Website: www.ijstd.org

DOI: 10.4103/ijstd.ijstd_11_21

Cutaneous examination revealed large, hyperpigmented confluent plaques and nodules over the lower one-third of the bilateral lower limbs and knee joint [Figure 1]. Bilateral nonpitting pedal edema and edema of external genitalia were present [Figure 2]. The oral cavity and other mucosal surfaces were spared. The differential diagnoses of cutaneous vasculitis, hypertrophic lichen planus, KS, bacillary angiomatosis, deep fungal infections, cutaneous tuberculosis, and cutaneous lymphoma were considered.

Blood investigations showed normocytic normochromic anemia (Hb – 8.8 gm%) with leukopenia (3710 cells/mm^3) and elevated liver enzymes (Serum Glutamic Oxaloacetic Transaminase [SGOT] – 58 U/L). The serological test for HIV was positive, and the patient was on antiretroviral therapy (ART) for the past 6 months, with CD4 count <200 cells/mm^3. Other serological tests such as HBsAg, hepatitis C virus, and venereal disease research laboratory were negative. Mantoux test, cartridge-based nucleic acid amplification test, and fungal cultures were negative. Chest X-ray showed a classical coalescent pattern with nodular opacities with minimal pleural effusion [Figure 3].

On histopathology, the epidermis showed lamellate orthokeratosis. In the reticular dermis, a moderately dense infiltrate of spindle cells arranged in small bundles with numerous small vessels protruding them and numerous

How to cite this article: Reddy HK, Raju BP, Methre V, Samagani A. Kaposi's sarcoma: An interesting case report in a human immunodeficiency virus-positive heterosexual male. Indian J Sex Transm Dis 2022;43:179-81.

Submitted: 09-Feb-2021 **Revised:** 05-Jul-2021
Accepted: 05-Jul-2021 **Published:** 01-Aug-2022

siderophages were seen. Mild perivascular lymphohistiocytic infiltrate with few plasma cells were present [Figure 4]. Immunohistochemistry (IHC) revealed CD34 and CD31 positivity, highlighting the blood vessels along with a scattered group of intervening spindle cells [Figure 5].

A diagnosis of KS was made on the basis of clinical presentation, histopathology, and IHC.

Discussion

AIDS-associated KS (AIDS-KS) is one of the clinical forms of KS. Other clinical types include classical or sporadic, endemic or African, iatrogenic, or immunosuppressed types. It is caused by human herpesviruses 8 or KS-associated herpesvirus (KSHV), which is a lymphotropic herpes virus that encodes a number of oncogenes involved in abnormal cell proliferation, anti-apoptosis, angiogenesis, and cytokine activation. It tends to occur at low CD4 counts or in immune reconstitution inflammatory syndrome (IRIS).[4] It has a synergistic interaction with HIV in producing KS. HIV transactivating protein stimulates KSHV leading to widespread infection in patients. In reciprocation, latency-associated nuclear antigen of KSHV enhances the replication of HIV.[5]

The first case of AIDS-KS from India was reported in a female sex worker from Mumbai, in 1995.[6] The incidence of KS is directly proportional to the prevalence of KSHV in that geographical area and the immunosuppressive status of the individuals.[3]

AIDS-KS has a variable clinical course affecting the skin, oral mucosa, lymph nodes, and visceral organs. The cutaneous manifestations are more common and characterized by brownish or violaceous macules, patches, plaques, and nodules which form a large confluent lesion with yellow-to-green perilesional halo, extensively involving the trunk, face, and limbs. Similar lesions can be seen in the oral mucosa (gingiva, hard palate, and oropharynx) seen in >70% of AIDS-KS. Lymphedema in AIDS-KS presents as nonpitting edema of the legs, external genitalia, and the periorbital regions, which are due to the involvement of dermal lymphatics. The common visceral organ involvement includes the lungs and gastrointestinal (GI) tract. Pulmonary KS indicates advanced HIV disease with a poor prognosis.[7]

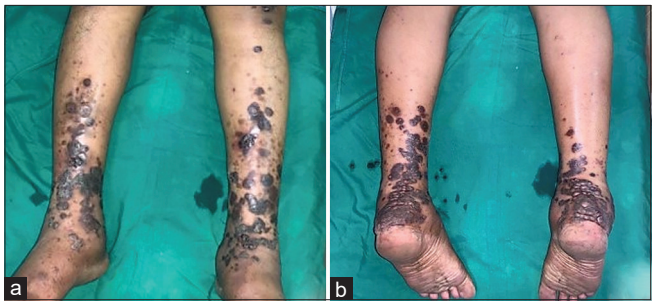

Figure 1: (a and b) Violaceous to hyperpigmented confluent plaques and nodules over the lower one-third of bilateral lower limbs and knee joint

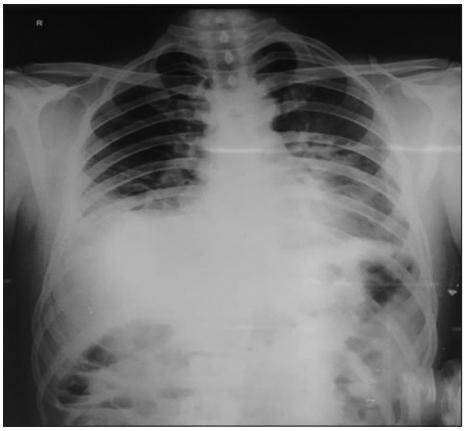

Figure 3: Chest X-ray showing a coalescent and nodular opacities with minimal pleural effusion

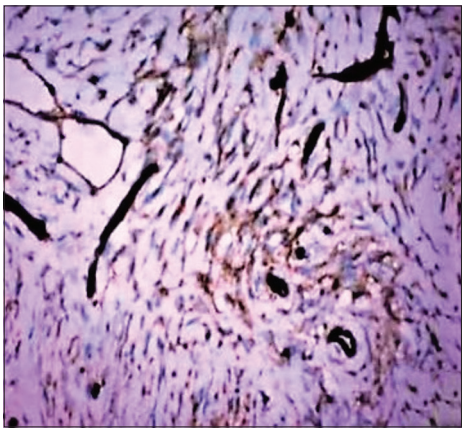

Figure 5: Immunohistochemistry showing CD34 and CD31 positivity for intervening spindle cells and blood vessels

Figure 2: Lymphedema of the external genitalia

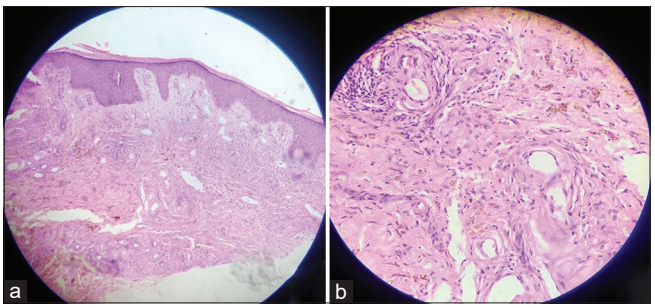

Figure 4: Histopathological examination showing lamellate orthokeratosis in the epidermis. In the reticular dermis, a moderately dense infiltrate of spindle cells arranged in small bundles with numerous small vessels protruding them and numerous siderophages were seen (H and E) (a) ×10 (b) ×40

Histology is diagnostic in AIDS-KS, to distinguish from other pigmented skin conditions such as bacillary angiomatosis, non-Hodgkin's lymphoma, and cutaneous fungal or bacterial infections. The histological features vary depending on the progression of clinical lesions. The classical and earliest presentation is "promontory sign" - protrusion of regular vascular structures into the lumens of neoplastic channels.[8] Other atypical histological variants such as hyperkeratotic, keloidal, micronodular, pyogenic granuloma-like, ecchymotic, and intravascular types are seen.

Other than skin biopsy, nonspecific endothelial markers such as CD31, CD34, lymphatic endothelial marker D2-40, or podoplanin and IHC technique using the antibody anti-latent nuclear antigen-1 of the KSHV can be employed. Other investigations such as chest radiography (pleural effusions or hilar lymphadenopathy) and endoscopy are useful in identifying pulmonary and GI KS.[7]

AIDS-KS has a poor prognosis. The most common cause of mortality is uncontrolled pulmonary hemorrhage.[9]

ART of HIV is equally effective in AIDS-KS. For milder and stable cases, cryotherapy, radiation therapy, intralesional chemotherapy with vinblastine or interferon-α (IFN-α), and topical retinoids (alitretinoin 0.1% gel) are found to be effective. The systemic therapies include IFN-α, which has an antiproliferative, antiangiogenic, immune-modulating, and antiviral (anti-HIV) properties. The cytotoxic chemotherapy regimen of actinomycin D, bleomycin, and vincristine is found to be effective. Thalidomide, bevacizumab, oglufanide disodium, foscarnet, ganciclovir, and rapamycin are under trial.[7]

Our patient was continued on ART with cryotherapy and was planned to start on chemotherapy, but he was lost to follow-up due to the pandemic situation.

We report this case due to the rarity and paucity of KS cases reported in the Indian literature. The extensive cutaneous manifestations over bilateral lower limbs (classical-like), bilateral pedal and lymphedema of the external genitalia, absence of oral lesions, occurrence in a heterosexual individual, and post-ART initiation (probably due to KS-IRIS) were some of the unusual presentations in our patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Harindra V. HIV: Past, present, and future. Indian J Sex Transm Dis 2008;29:1-6.
2. Brodt HR, Kamps BS, Gute P, Knupp B, Staszewski S, Helm EB. Changing incidence of AIDS-defining illnesses in the era of antiretroviral combination therapy. AIDS 1997;11:1731-8.
3. Mehta S, Garg A, Gupta LK, Mittal A, Khare AK, Kuldeep CM. Kaposi's sarcoma as a presenting manifestation of HIV. Indian J Sex Transm Dis AIDS 2011;32:108-10.
4. Volkow P, Cesarman-Maus G, Garciadiego-Fossas P, Rojas-Marin E, Cornejo-Juárez P. Clinical characteristics, predictors of immune reconstitution inflammatory syndrome and long-term prognosis in patients with Kaposi sarcoma. AIDS Res Ther 2017;14:30.
5. Greene W, Kuhne K, Ye F, Chen J, Zhou F, Lei X, *et al.* Molecular biology of KSHV in relation to AIDS-associated oncogenesis. Cancer Treat Res 2007;133:69-127.
6. Shroff HJ, Dashatwar DR, Despande RP, Waigman HR. AIDSassociatedKaposi's sarcoma in an Indian female. J Assoc Physician's India 1993;41:241-2.
7. Govindan B. Recapitulation of acquired immuno deficiency syndrome associated Kaposi's sarcoma. Indian J Sex Transm Dis AIDS 2016;37:115-22.
8. Grayson W, Pantanowitz L. Histological variants of cutaneous Kaposi sarcoma. Diagn Pathol 2008;3:31.
9. Nwabudike SM, Hemmings S, Paul Y, Habtegebriel Y, Polk O, Mehari A. Pulmonary kaposi sarcoma: An uncommon cause of respiratory failure in the era of highly active antiretroviral therapy-case report and review of the literature. Case Rep Infect Dis 2016;2016:9354136.