Disseminated violaceous plaques in a HIV-1-positive patient from eastern India: A manifestation of Kaposi's sarcoma

Sumit Sen, Subhadeep Mallick, Somnath Das, Jyoti Das, Surabhi Sharma, Subhasmita Baisya
Department of Dermatology, IPGME&R and SSKM Hospital, Kolkata, West Bengal, India

Address for correspondence:
Dr. Subhadeep Mallick, 41, Lenin Sarani (West), North 24 Parganas, Kanchrapara - 743 145, West Bengal, India.
E-mail: subhadeep.mallick1@gmail.com

Abstract

Kaposi's sarcoma (KS) is an angioproliferative disorder primarily of viral etiology, though multiple cofactors are also responsible. Human herpes virus-8, a gamma herpes virus, is considered to be the causative agent. Acquired immunodeficiency syndrome-associated KS has different clinical pictures than those seen in other types of KS. As it progresses rapidly, early institution of highly active antiretroviral therapy (HAART) after proper diagnosis is expected. Though HAART has reduced the prevalence of KS in HIV disease, HAART has not eliminated the disease. Here, we report a case who is HIV 1 and hepatitis B surface antigen positive with numerous violaceous plaques over the face, upper extremities, and trunk along with oral mucosal involvement. He had received ten sessions of electron beam radiotherapy on the face, and the facial lesions have healed with residual hyperpigmentation.

Key words: Angioproliferative disorder, Kaposi's sarcoma, seropositive male

INTRODUCTION

Kaposi's sarcoma (KS) is a multicentric proliferation of endothelial cells.[1] The cause seems to be multifactorial apart from human herpes virus-8 (HHV-8) viral infection, of which HIV co-infection is important.[2] It primarily involves skin with or without visceral dissemination. The following four clinicopathological subtypes have been described: classic, endemic, iatrogenic, and acquired immunodeficiency syndrome (AIDS) associated. Clinical features differ among these subtypes.[3] Our case is unique because the patient has only HIV1 infection with lesions of KS in the upper limb, trunk, and oral lesions without AIDS, and he has responded well to external beam radiotherapy (EBRT).

Access this article online

Quick Response Code:

Website: www.ijstd.org

DOI: 10.4103/ijstd.IJSTD_17_20

CASE REPORT

A 35-year-old male patient presented with multiple asymptomatic erythematous raised lesions over the face, neck, trunk, and both upper limbs for the last 2 months. He had a history of intermittent low-grade fever for the last 3 months without chills and rigors. A history of swelling of the face for the last 20 days was present. It first appeared on the neck, hand, and nose, and later it progressed to involve the trunk, both upper limbs, and the entire face. No plaques were found in the penis and lower limb. He had a history of unprotected sexual intercourse with both male and female partners. There was no history

How to cite this article: Sen S, Mallick S, Das S, Das J, Sharma S, Baisya S. Disseminated violaceous plaques in a HIV-1-positive patient from eastern India: A manifestation of Kaposi's sarcoma. Indian J Sex Transm Dis 2021;42:69-72.

Submitted: 29-Mar-2020 **Revised:** 07-Aug-2020
Accepted: 12-Oct-2020 **Published:** 15-Feb-2021

of blood transfusion or intravenous drug abuse. Systemic symptoms present were nausea, vomiting, and fever. On examination, (after the patient has given informed consent), the patient had generalized lymphadenopathy without hepatosplenomegaly. Cutaneous examination revealed multiple, discrete, violaceous plaques measuring 0.5 cm × 0.5 cm to 3 cm × 2 cm over the trunk, face [Figure 1, left one], neck, and both upper extremities [Figure 2]. On oral examination, there were erythematous plaques over the left tonsillar fossa and posterior pharyngeal wall [Figure 3]. He had low hemoglobin (8.4 g/dL), low platelets (38,000/mm^3), and normal liver and renal function; serology for HIV-1 was positive by enzyme-linked immunosorbent assay and negative for HIV-2, and his CD4 count was 337/mm^3. Hepatitis B surface antigen was reactive. Ultrasonography of the whole abdomen showed a space-occupying lesion in segment 7 of the liver suggestive of hemangioma. Histopathological examination showed irregularly distributed vascular channels, some of which were surrounding preexisting vessels, with jagged outline throughout the dermis and were lined by a single layer of endothelial cells, cords, and clumps of spindle cells between dermal collagen bundles, with cells having plump vesicular nuclei [Figure 4]. Patchy lymphoplasmacytic infiltrates were seen in the dermis around the blood vessels and adnexal structures and beneath the epidermis. Immunostaining was performed for CD34. Endothelial cells and spindle cells were strongly immunoreactive. A contrast-enhanced computed tomography scan of the thorax was performed, which showed patchy air space opacities in both lung fields and a well-defined hypodense lesion in the liver, suggestive of KS. A final diagnosis of HIV-associated

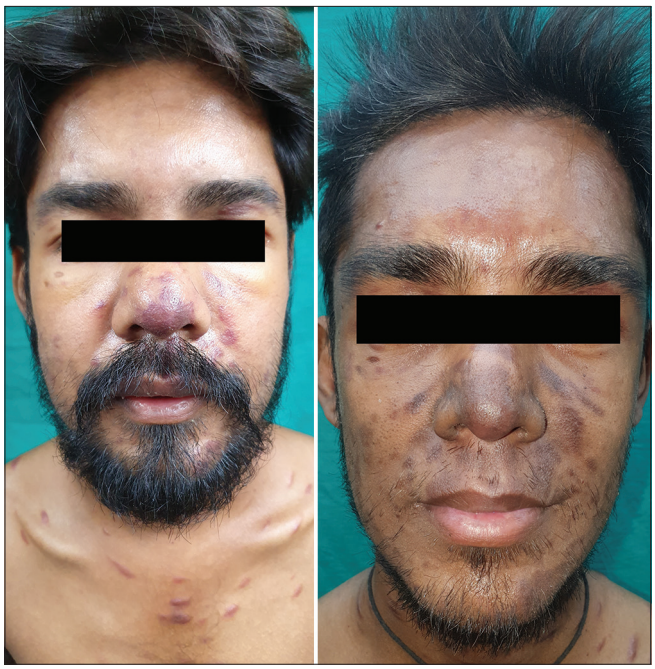

Figure 1: Left image showing violaceous plaques before external beam radiotherapy and right image showing hyperpigmentation following external beam radiotherapy

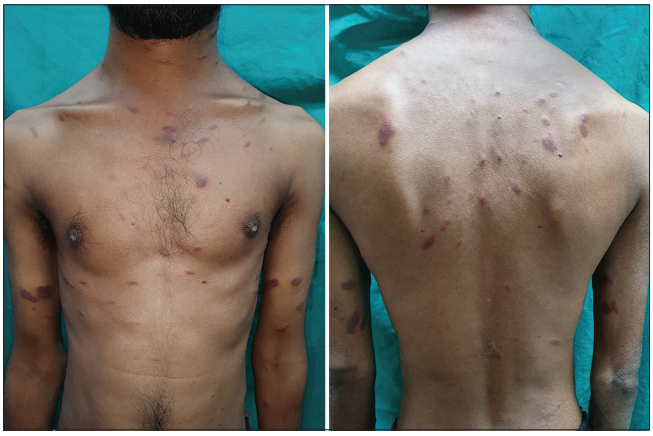

Figure 2: Violaceous plaques measuring 0.5 cm × 0.5 cm to 3 cm × 2 cm over the neck, trunk (anterior and posterior), and both upper extremities

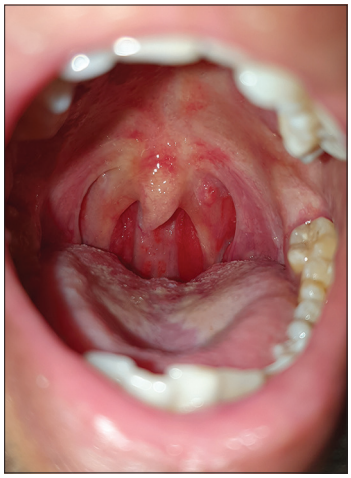

Figure 3: Erythematous plaque over the left tonsillar fossa and posterior pharyngeal wall

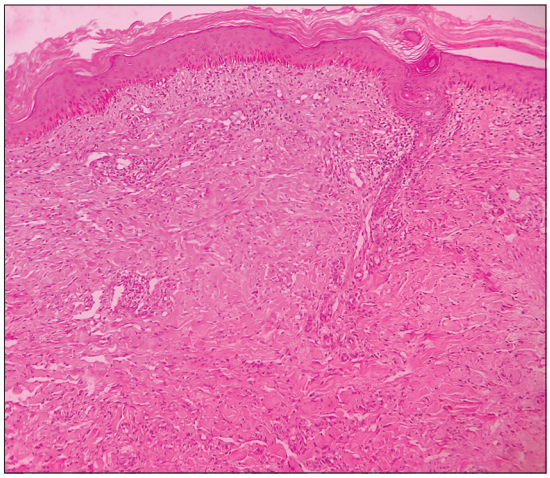

Figure 4: Irregularly distributed vascular channels, low-power view (×10), hematoxylin and eosin stain

Kaposi sarcoma (plaque stage) was made. The patient was on antiretroviral therapy (ART) (tenofovir, lamivudine, and efavirenz regimen) and had received ten cycles of EBRT for facial lesions and developed erosions over the lips and tongues. The patient's facial lesions had resolved with hyperpigmentation including oral lesions, but lesions over the trunk and limbs were present [Figure 1 right one]. The patient had received pegylated liposomal doxorubicin in six cycles.

DISCUSSION

KS, first described by Moritz Kaposi in 1872, is a multifocal endothelial proliferative disease of low-grade potential affecting skin and other organs.[4] Although HHV-8 is considered the causative agent, multiple other cofactors are needed including HIV co-infection which increases the risk 20,000 fold.[5] The low incidence of KS in India is thought to be due to very low incidence of HHV-8. To date, close to 26 cases of KS have been reported from India since 1993, but only one of those was HIV negative.[6] AIDS-associated KS can be rapidly progressive, involving the head, neck, trunk, and mucous membranes; fulminant disease with a widespread nodal and visceral involvement is expected, particularly in the absence of highly active ART (HAART). Oral mucosal involvement, especially hard palate, soft palate, gingiva, and dorsal tongue, is seen.[7] The gastrointestinal tract, particularly the small intestine, is the most common site of visceral involvement with massive hemorrhage as a serious complication.[8] Lungs, heart, and liver are other commonly involved sites including hemangioma of liver. Laboratory investigations include immunohistochemical staining with HHV-8 and other markers such as CD31 and CD34. Histopathological changes of KS typically parallel the clinical progression of patch, plaque, and tumor stages. Patch-stage KS manifests as a mild increase in the number of vessels, which are classically arranged in a horizontal fashion, dissecting through collagen bundles, around the adnexa and the surrounding preexisting vessels (promontory sign). A chronic lymphoplasmacytic infiltrate may be present, with extravasated erythrocytes and hemosiderin deposition. The plaque stage has more obvious and extensive vessel expansion, lined by single-layered, plump endothelial cells. Surrounding them are more spindle cells. The chronic inflammatory infiltrate remains. Finally, in tumor-stage or nodular KS, there is a circumscribed mass of spindle cells with unlined slit-like spaces with extravasated erythrocytes. KS is highly radiosensitive with complete responses in up to 93% of patients and for AIDS-related Kaposi sarcoma radiotherapy it is used in patients with limited cutaneous disease that is symptomatic and/or cosmetically unacceptable along with anti-retroviral therapy, but for advanced disease it is used as palliative therapy.[9] Regimens range from a single 8 Gy irradiation to 30–40 Gy in 10–20 daily fractions.[10] Radiotherapy reduces pain and swelling in the KS lesions, also used for skin lesions that look morbid. For AIDS-associated KS, the institution of HAART to treat HIV infection often results in regression of KS, but up to 50% of cases never achieve total remission.[11] Stopping immunosuppression in iatrogenic KS can lead to regression of lesions, and classic KS is more often treated with surgery, radiation therapy, or another local treatment. Disseminated KS having response rates of over 70% has been reported with liposomal doxorubicin, vinca alkaloids, etoposide, and taxanes. Liposomal doxorubicin and paclitaxel are approved by the US Food and Drug Administration as first-line and second-line treatments, respectively, for advanced KS. In the present study, the patient's main complaints were severe pain and edema over the face, which were preventing him from opening his eyes. Keeping this in mind, EBRT was given earlier to alleviate pain, otherwise doxorubicin could have been given earlier.

We report this case due to a paucity of KS cases reported in the Indian literature and our case had widespread cutaneous lesions along with lung and liver involvement, CD4 count 337/mm^3, and fever for 1 week. Score was T1I0S0 according to AIDS Clinical Trial Group which means the patient had a good prognosis.[9] The patient responded well to both chemo and radiotherapy.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Munawwar A, Sharma SK, Gupta S, Singh S. Seroprevalence and determinants of Kaposi sarcoma-associated human herpes virus 8 in Indian HIV-infected males. AIDS Res Hum Retroviruses 2014;30:1192-6.
2. Antman K, Chang Y. Kaposi's sarcoma. N Engl J Med 2000;342:1027-38.
3. Ruocco E, Ruocco V, Tornesello ML, Gambardella A, Wolf R, Buonaguro FM. Kaposi's sarcoma: Etiology and pathogenesis, inducing factors, causal associations, and treatments: Facts and controversies. Clin Dermatol 2013;31:413-22.
4. Tsai YK. Kaposi sarcoma. In: Griffiths CE, editor. Rook's Textbook of Dermatology. 9th ed. West Sussex: John Wiley & Sons Ltd.; 2016. p. 3845-50.

5. Dezube BJ. AIDS-related Kaposi sarcoma: The role of local therapy for a systemic disease. Arch Dermatol 2000;136:1554-6.
6. Prabhakaran N, Gupta D, Chandrashekhar L, Thappa DM, Rajesh NG. Disseminated Kaposi's sarcoma as a presenting sign of HIV in an Indian male: A case report with dermoscopic findings. Indian J Sex Transm Dis 2020;41:102-5.
7. Behara B, Chandrashekar L, Thapa Mohan D, Toi Ch P, Vinod VK, Disseminated Kaposi's sarcoma in an HIV-positive patient: A rare entity in an Indian patient. Indian J Dermatol 2016;61:348.
8. Chang Y, Moore P. Twenty years of KSHV. Viruses 2014;6:4258-64.
9. Yildiz F, Genc M, Akyurek S, Cengiz M, Ozyar E, Selek U, *et al*. Radiotherapy in the management of Kaposi's sarcoma: Comparison of 8 Gy versus 6 Gy. J Natl Med Assoc 2006;98:1136-9.
10. Lukawska J, Cottrill C, Bower M. The changing role of radiotherapy in AIDS-related malignancies. Clin Oncol (R Coll Radiol) 2003;15:2-6.
11. Nguyen HQ, Magaret AS, Kitahata MM, Van Rompaey SE, Wald A, Casper C. Persistent Kaposi sarcoma in the era of highly active antiretroviral therapy: Characterizing the predictors of clinical response. AIDS 2008;22:937-45.