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Case report Free Access

Giant benign phyllodes tumor equal to size of patient's abdomen: a case report

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Abstract

Introduction: Kaposi sarcoma is uncommonly encountered in clinical practice. It is described as vascular malignancy associated with HHV-8 infection. It most commonly affects skin, but can also affect mucosal surfaces, lymphatics and visceral organs. In recent times, with availability of HAART and newer chemotherapy agents, prognosis has improved. One of the complications associated with KS is exacerbation secondary to immune reconstitution with initiation of ART and Steroid.

Case presentation: Hereby We example case of 17-year-old male presented as Kaposi sarcoma with steroid induced exacerbation. Through this case, we enlighten the epidemiology, clinical features and management of such patients which might help clinicians in further management.

Discussion: One of the complications associated with Kaposi sarcoma is exacerbation associated with initiation of antiretroviral therapy (ART) which might lead to Immune reconstitution inflammatory syndrome (IRIS) and steroids induced flare, and thus steroids are contraindicated even as management of IRIS.

Conclusion: Use of corticosteroid may cause life threatening exacerbation and prompt initiation of cytotoxic agent may prove to be beneficial.

Keywords: HHV-8, Immune reconstitution inflammatory syndrome, Kaposi sarcoma

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Introduction

Classified as an AIDS-defining malignancy, Kaposi sarcoma is etiologically linked with to HHV8. There are 4 recognized variant of Kaposi sarcoma. These are classic, endemic (african), iatrogenic (immunosuppression related) and (AIDS-associated) (1, 2). It usually presents with cutaneous and mucosal lesions which may be plaques, papules, nodules, or bullae (3). Since the use of anti retro-viral therapy, the incidence has decreased, and overall survival has improved (4).

Case presentation

A 17-year patient assigned male at birth presented to the outpatient department (OPD) of KMIO Bangalore in June 2024 with a four-month history of multiple painful violaceous, reddish papulomacular lesions that had progressively enlarged over the past 15 days (Figure 1&2). The lesions were widespread, affecting the face, trunk, upper and lower limbs, and buccal mucosa. On examination, he had moderate pallor, cervical lymphadenopathy, and severe edema over face, lip and extremities.

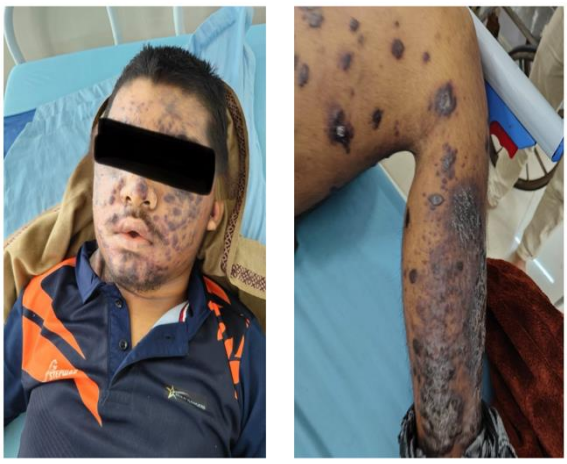

Figure 1. Initial presentation. multiple violaceous, reddish papulomacular lesions on the face(left) and lower limbs (right).

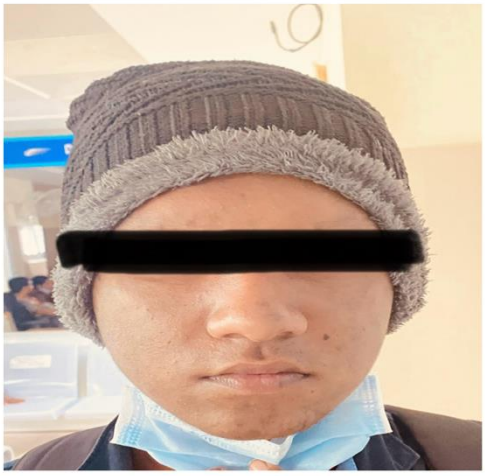

Figure 2. Present status, suggestive of resolution of all visible lesions.

He was priorly diagnosed with HIV infection and had been on antiretroviral therapy (ART) for the last 10 years. Despite ongoing treatment, his CD4 count was 172 cells/mm³, indicating significant immunosuppression. He had been adhering to his highly active antiretroviral therapy (HAART) regimen, and there was no history of tobacco use. The patient gave the history of evaluation elsewhere and he received a few days of corticosteroids after which the lesion increased in size and severe edema developed over the face and extremities for which he was referred to tertiary centre. Upon clinical examination, the patient was well-oriented and stable, with vital signs within normal limits. Laboratory tests revealed anemia (hemoglobin 6.9 g/dL), while his total leukocyte count was 6400/mm³, and other routine blood parameters were unremarkable.

A PET-CT scan showed evidence of hypermetabolic lymphadenopathy both supra and infra-diaphragmatically, along with nodular pleural thickening in the left hemithorax, and hypermetabolicpapules on the skin of the upper and lower limbs, face and trunk.

A biopsy from one of the lesions on the left forearm revealed a vascular neoplasm, characterized by proliferating blood vessels of varying sizes, surrounded by spindle cells arranged in fascicles. These spindle cells exhibited no necrosis or mitotic activity, but there was extravasation of red blood cells (RBCs), a hallmark of Kaposi sarcoma. Immunohistochemical

staining (IHC) was positive for CD31, CD34, CD3, ERG, and FLI1, confirming the diagnosis of Kaposi sarcoma (Figure 3).

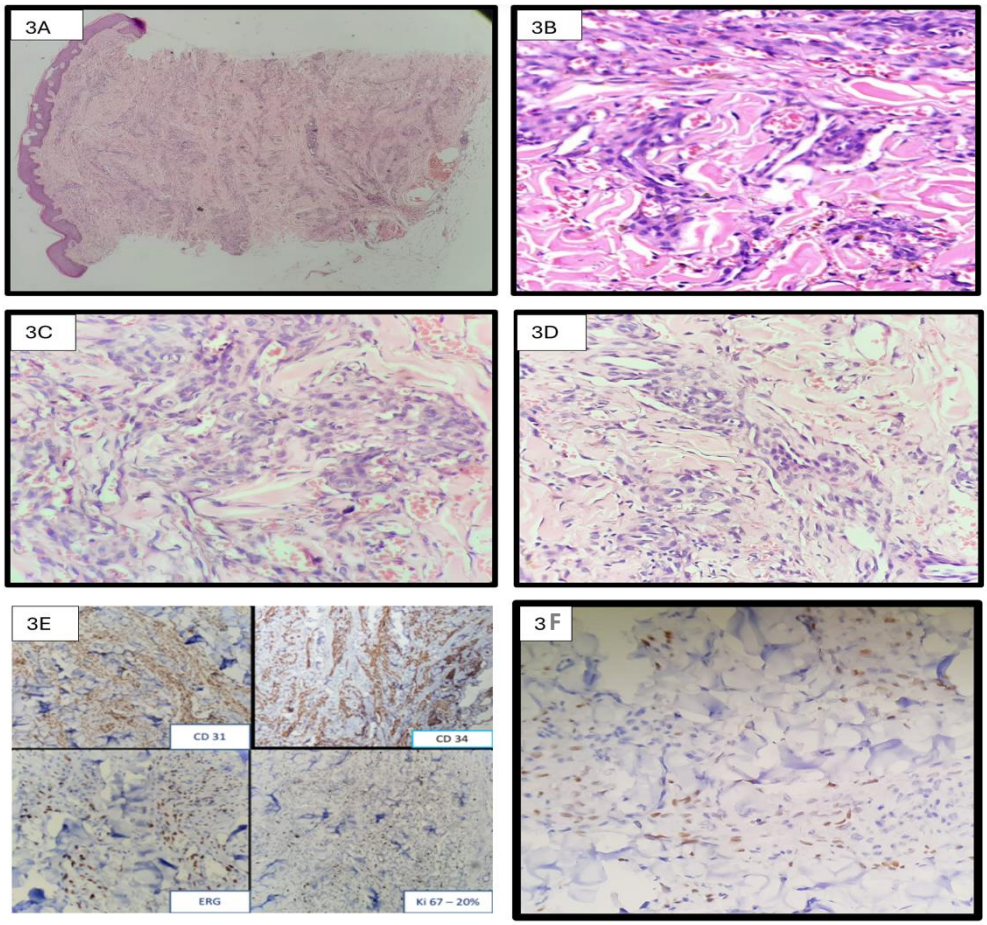

Figure 3. Histopathological and immunohistochemistry evaluation. Figure 3A showing punch biopsy of skin from left arm (whole mount view). Figure 3B showing promontory sign (protrusion of pre existing vessels into laminate of new vessels. Figure 3C Showing Proliferating blood vessels of varying caliber with surrounding spindle cells Dissecting through dermal collagen. Figure 3Cshowing spindle cell proliferation and sieve like and slit like vascular spaces. Figure 3E showingIHC markers(CD31,CD34, ERG, Ki67%) and figure 3F showing LANA antigen.

LANA (Latency-associated nuclear antigen) was done revealing positivity for HHV8. These markers support the diagnosis, as they are associated with the endothelial origin and vascular proliferation and HHV8 association characteristic of this neoplasm. The combination of clinical presentation, imaging findings, histopathology, and IHC results led to the diagnosis of Kaposi sarcoma. In view of diagnosis of Kaposi sarcoma with steroid-induced exacerbation, patient was quickly initiated on liposomal doxorubicin alongside the continuation of the current antiretroviral regimen (TLD). It was given as two weekly schedule of 20mg/meter square. This integrated approach targeted both the underlying HIV infection and Kaposi sarcoma, aiming to control the malignancy and improve immune function. Clinically and radiologically, the patient demonstrated remarkable improvement, including the resolution of facial and lip edema and a marked reduction in skin lesions across the body. A PET-CT scan conducted in September, following four cycles of chemotherapy, revealed a partial response: complete resolution of mediastinal, axillary, abdominal, retroperitoneal, and pelvic lymph nodes; significant reduction in the size, number, and metabolic activity of cervical and inguinal lymph nodes; normalization of the thickened pleura in the left hemithorax; and resolution of hypermetabolicpapules on the skin of

both upper and lower limbs. The chemotherapy regimen was extended to 10 cycles, and a follow-up PET-CT in November indicated stable disease, with no evidence of further progression. Because of financial issues and stable disease despite 10 cycles, the chemotherapy agent was changed to paclitaxel 175 mg/m2 q3weekly. Response assessment after 3 cycles revealed a complete metabolic response with no evidence of disease. Paclitaxel was continued for 6 cycles and currently patient is under regular follow-up with continuation of anti retroviral therapy with CD4 of 196 cells/mm3.

Discussion

This case highlights the behavior and the potential of Kaposi sarcoma is frequently seen in patients with HIV/AIDS, particularly in with significant immunosuppression (CD4 count < 200 cells/mm³). Moritz Kaposi recognized it first in 1872 (5). It is universally associated with the presence of HHV8. There are 4 types- classical, AIDS-related, iatrogenic and endemic. Prior to development of ART, AIDS associated KS was seen in up to 30% of AIDS patients. When immunosuppression is severe, the incidence is more common, however patients with normal CD4 also have increased risk as compared to general population. With the development of ART, incidence has declined with improved overall survival. However, few patients may experience IRIS after initiation of anti retroviral therapy, leading to worsening of KS. Iatrogenic KS is seen in patients on long-term immunosuppression like post-transplant. Lesions usually appear after 1-2 months of immunosuppression and are more common in males. Classical KS is more common in males with the mean age of diagnosis being 74 years. Patients with classical KS usually present with indolent cutaneous lesions which may appear and disappear or slowly progress, usually starts on the lower extremities. Endemic KS occurs in children and young adolescents in Africa. As compared to classical KS, it is more aggressive (6, 7).

KS usually presents with violaceous skin lesions which may be plaques, papules, nodules, or bullae. It can also involve oral mucosal lining, gastrointestinal tract, lymphatic, and lungs. Diagnosis requires histopathological and immunophenotyping study. Workup includes history and physical examination, blood counts, biochemistry, HIV screening, CD4 counts and viral load assay. Further investigation may be needed to differentiate it from KS-multicentric Castleman disease, KS-inflammatory cytokine syndrome and KS-associated lymphoma. Chest imaging and stool hemoccult test may be required in cases with advanced cutaneous, nodal involvement or symptomatic patients. KS is staged based on the TIS system which incorporates tumor, immune system and systemic disease with zero points awarded for good risk and one for poor risk7. Some investigators have voted against the incorporation of the immune system (I) because of poor prognostic value. 3 yr OS for T0S0, T1S0, T0S1 and T1S1 is 88%, 80%, 81% and 53%, respectively (8).

The goal of therapy in advanced Kaposi sarcoma is mainly reducing symptoms, and mitigating end-organ damage, however, with the availability of HAART and systemic therapy prolonging survival can be a goal, especially with localized disease. In patients with asymptomatic, cosmetically acceptable and limited cutaneous disease observation is advised with ART. In patients with symptomatic cutaneous disease localized therapy like radiation (20-24 Gy in 5-12 fractions), topical application of alitretinoin/ imiquimod, cryotherapy, intralesionalvinblastine or local excision can be advised. In refractory patients to local therapy or advanced disease, systemic therapy is the treatment. Preferred systemic therapy is liposomal doxorubicin 20mg/m2 q2weekly with an overall response rate of 56% (9). Other agents are paclitaxel, lenalidomide, pomalidomide, bortezomib, Gemcitabine and imatinib. Sirolimus can be used in transplant-related KS, while immunotherapy can be used in relapsed or refractory classical/endemic KS.

Complications related to treatment include IRIS syndrome which is seen within 3-6 months of initiation of ART in 6-39% of cases. It is seen more commonly in patients with low CD4 and high viral load. Corticosteroids are contraindicated in KS as they may cause life-threatening exacerbation (10). This flare is attributed to stimulatory effect of steroids on spindles cells of KS. Also glucocorticoid use had been associated with increased mortality. Both in the case of

IRIS or flare secondary to steroid, Prompt initiation of systemic therapy is required.

In our case, the patient presented with T1S1 stage with severe edema secondary to corticosteroid flare. Patient was started on liposomal doxorubicin as soon as the diagnosis was made. The patient achieved partial response to that and was later changed to paclitaxel which he attained CMR. Currently, the patient is on regular follow-up.

Conclusion

Use of corticosteroid may cause life threatening inflammatory state and thus to be used cautiously. Flare may be seen in the form of sudden increase in lesion number and size, edema and sudden deterioration of general condition. In case of IRIS, prompt initiation of cytotoxic agent may prove to be beneficial.

Author contribution

LKN and **SB** Conceptualization and project administrator.

LKN methodology and supervision.

YP data curation and investigation, resources and software.

RAH, LKR, SCS and **GVG** formal analysis.

LKN, YP, GS, DRS, KA Manuscript writing and figures.

Conflicts of interest
There are no conflicts of interest.

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