CASE REPORT

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Disseminated Kaposi's Sarcoma in HIV Infection with Fatal Consequences

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Abstract

We report a case of disseminated Kaposi's sarcoma (KS) in a HIV-infected patient of Nigerian origin. He presented with multiple painful voilaceous plaques and nodules over the right lower limb, trunk and back and, recurrent episodes of haemoptysis. He was managed with chemotherapy with paclitaxel. Due to the rarity of KS in India, especially disseminated disease, we wish to highlight the rapidly progressive course of disseminated KS.

Key words: Kaposi's sarcoma, KS, disseminated, haemoptysis, HIV.

Introduction:

Kaposi sarcoma (KS) is an angio-proliferative, vascular tumour that can involve multiple organs/sites: skin, mucous membranes, lymph nodes, gastrointestinal tract, respiratory system, heart, the testes, bone marrow, bone and skeletal muscle[1]. KS is linked with infection with Human Herpes virus (HHV-8), also known as KSHV[2]. KS has traditionally and historically been associated with AIDS and is an AIDS-defining malignancy. AIDS-related KS has a variable clinical course, ranging from mild disease detected as an incidental finding to a rapidly progressing cancer with significant morbidity and mortality. There are very few reports of KS from India probably due to the low prevalence of the HHV-8 infection – a reported prevalence of 4.7% from South India[3]. We report disseminated KS in a HIV-infected male patient who had a severe clinical course and outcome despite aggressive treatment.

Case history

A 38-year-old HIV-1-infected, homosexual male, of Nigerian descent, presented with multiple, painful, violaceous plaques and nodules over the right leg since 10 months. The plaques gradually appeared in the other limb, trunk and back, and later became lichenified and crusted. The patient complained of cough with expectoration and progressively increasing dyspnoea since 2 months. Over the past 1 month he also reported recurrent episodes of haemoptysis of significant quantity.

There was associated malaise, generalised weakness and significant weight loss. He was diagnosed with HIV infection in 2015 and was initiated on first-line ART (tenofovir/lamivudine/efavirenz) through the National Programme in India as he was incarcerated. On clinical examination, the patient was cachexic, had significant pallor, and the respiratory rate was 22 breaths/minute. There was no lymphadenopathy and oral cavity examination was normal. On cutaneous examination, there were multiple, confluent, painful, crusted nodules present over the right leg extending up to the thigh and fresh erupting well-defined, non-scaly, dusky, violaceous, tender papules and nodules of 1.5 x 2 cm size all over the back Fig. 1. Respiratory examination revealed right infra-scapular coarse crackles with reduced breath sounds at the base. Rest of the physical examination was unremarkable. His blood oxygen saturation by pulse oximetry was 82%.

Upon investigations, there was pancytopenia (haemoglobin

Fig. 1: *Clinical photograph depicting confluent crusted nodules over leg.*

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- 6.2 g/dl, TLC - 1,900 cells/mm^3, platelet count - 95,000/mm^3). His baseline CD4 count (at the time of HIV diagnosis) was 578 cells/mm^3 and the current CD4 count was undetectable. The HIV viral load was 1,22,000 copies/ml. The VDRL, HBsAg, anti-HCV IgG were all non reactive.

In view of haemoptysis, a contrast enhanced CT of the chest was done. It revealed a heterogenously enhancing soft-tissue mass lesion with spiculated margins in the left upper lobe with areas of consolidation in the right middle lobe, nodular opacities in bilateral lung parenchyma in peri-bronchovascular distribution with mediastinal lymphadenopathy and bilateral pleural effusion (right > left) Fig. 2.

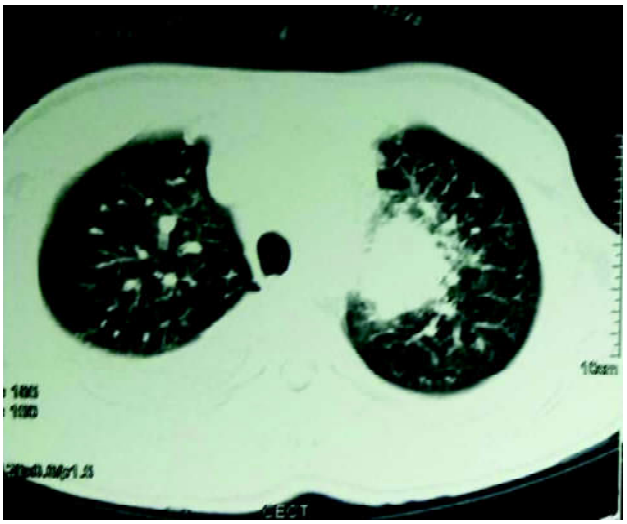

Fig. 2: *CECT chest showing heterogeneously enhancing soft-tissue mass lesion with speculated margins in the upper lobe of left lung.*

A punch biopsy done from the skin lesions on the back and leg showed spindle-cell proliferation with slit-like congested capillaries dissecting between collagen bundles throughout the dermis, present individually as well as in groups with extravasation of erythrocytes – suggestive of KS (Fig. 3). Pleural fluid was grossly haemorrhagic and analysis showed total count – 440 cells, differential cell count – 18% polymorphs, 45% lymphocytes, and 37% malignant cells. The pleural fluid glucose was109 mg/dl and protein 3.2 gm/dl.

Due to persistent hypoxia and poor general condition, the bronchoscopy was not performed.

A final diagnosis of disseminated Kaposi sarcoma was made – involving skin, lungs and pleura-stage (T1I1S1 ACTG staging- overall poor risk).

Patient was switched to second-line ART regimen (raltegravir and lopinavir/ritonavir) for ART failure. As patient had disseminated severe KS, systemic chemotherapy was started with paclitaxel 100 mg/m^2. The patient showed marginal improvement symptomatically with 2nd-line ART and 2 cycles of chemotherapy. However, on day 23 of his admission, he developed massive haemoptysis and succumbed to his illness.

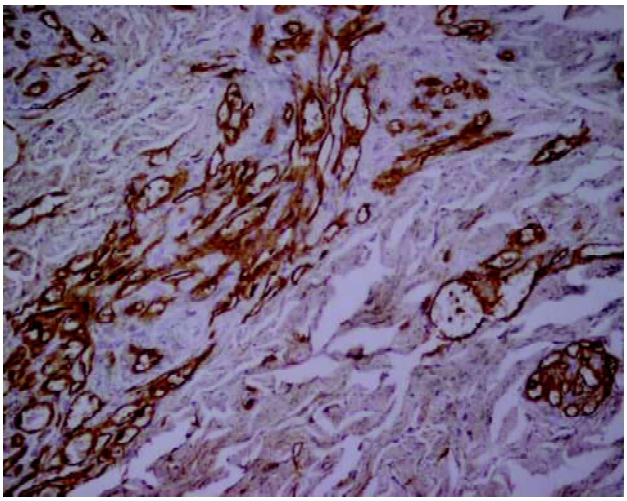

Fig. 3: *Histopathology: IHC (Immunohistochemistry) (200 x) showing CD 34 positive endothelial cells in KS.*

Discussion

Although KS can involve virtually any site in the body, cutaneous disease is most common and is the usual initial presentation. The most frequent sites of non-cutaneous disease are the oral cavity, gastrointestinal tract, and respiratory system. However, visceral involvement as the initial manifestation of KS is relatively uncommon[4]. Pulmonary involvement is the most life-threatening form of KS. In 80 - 90% of cases, pulmonary involvement with KS occurs in conjunction with more extensive mucocutaneous disease[5]. Unusually, pulmonary involvement can be the initial manifestation of KS and occurs in the absence of mucocutaneous disease in 15 per cent of patients[6]. Parenchymal lung involvement is usually manifest clinically by dyspnoea, hypoxaemia, and dry cough developing over a few weeks . Haemoptysis, fever, fatigue, and occasionally respiratory failure can also occur.

The most commonly utilised staging system for AIDS-related KS is ACTG system which considers 3 factors: extent of the tumour (T), status of the immune system (I), as measured by the number of CD4 cells and extent of systemic illness (S) within the body .This system divides patients into good or poor risk prognostic categories, taking into account both the KS and HIV infection. Patients with poor risk KS having extensive cutaneous lesions, oral or visceral disease, CD4 count < 100 cells/mm^3, systemic diseases and opportunistic infections have worst prognosis. This patient had poor risk KS disease. Management of KS includes early initiation of

ART, surgery, radiotherapy, cryotherapy, intralesional chemotherapy for skin lesions. Systemic chemotherapy is indicated for disseminated KS. Pegylated liposomal doxorubicin is recommended as the first-line regimen in the absence of a cardiac contraindication. Other available options include a single-agent taxane, oral aetoposide, vinblastine, vinorelbine, or gemcitabine. No one regimen can be recommended over any other. The decision must be individualised, taking into consideration the patient's age, accompanying co-morbidity, and clinician and patient preference. Our patient had poor ejection fraction on 2-D echocardiography and was hence given paclitaxel.

Even in the age of advanced, universal, and early HIV treatment, disseminated KS continues to be a life-threatening disease with high mortality rate though the prevalence and aggressiveness of KS has decreased with early administration of ART. In HIV-infected patients with atypical pneumonia, KS should be high on the list of differentials so that prompt diagnosis can be made for timely initiation of chemotherapy.

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