Kaposi's Sarcoma: Case Report and Treatment Options

W C Tan, MRCP, L C Chan, M Med

Department of Dermatology, Hospital Pulau Pinang, Penang, Malaysia

SUMMARY

Kaposi's sarcoma (KS) is strongly associated with Human Herpes Virus 8 (HHV8) and Human Immunodeficiency Virus infection (HIV). It was the first malignancy to be linked with Acquired Immunodeficiency Syndrome (AIDS).

We report a case of Kaposi's sarcoma in a newly diagnosed retroviral homosexual patient with CD4 count of 21. He had multiple firm discrete violaceous plaques and nodules scattered over the face, scalp, hard palate, trunk and genitalia. Biopsy of a skin nodule over the trunk and a biopsy of a lesion from the gastric mucosa confirmed Kaposi's sarcoma. He was started on Highly Active Antiretroviral Therapy (HAART) and cryotherapy (liquid nitrogen) was given for the lesions over the skin. He responded well to treatment.

Liquid nitrogen is a useful adjuvant treatment for Kaposi's sarcoma.

KEY WORDS:
Human Herpes Virus 8 (HHV-8); Homosexual; Kaposi's sarcoma; AIDS

CASE REPORT

Mr J is a 39 year old man who first presented in a hospital in London with severe pneumonia and seizure requiring mechanical ventilator support. He has been unwell for about three months with fever on and off and chronic weight loss. Computed tomography (CT) of his brain showed multiple ring enhancing lesions which was confirmed later to be cerebral toxoplasmosis by brain biopsy.

His retroviral test was positive with CD4 count of only 21 and viral load of 4.6 million copies He was treated for cerebral toxoplasmosis and also covered for Pneumocystis jiroveci pneumonia infection. This was complicated with upper gastrointestinal bleed. Oesophageal gastroduodenoscopy (OGDS) was performed which revealed polypoidal, purplish lesions on the lesser and greater curvature of the stomach. Examination of the skin showed multiple (more than 30 lesions) firm discrete violaceous plaques and nodules scattered over the face, scalp, hard palate, trunk and genitalia (Figure 1). Biopsy of a skin nodule over the trunk and biopsy from the gastric mucosa confirmed Kaposi's sarcoma (KS). Biopsy slides show unremarkable epidermis but underlying dermis showing angiosarcomatous component which composed of numerous slit like congested vascular channels. Extravasation of red blood cells was noted. Immunohistochemical stains for Factor VIII antigen is positive.

When his condition was stabilized, he was transferred back to Penang General Hospital for further management. Subsequently, he was started on Highly Active Antiretroviral Therapy (HAART – Indinavir / Ritonavir / Efavirenz) and the cutaneous KS treated with cryotherapy (liquid nitrogen). Treatment regimen consists of three freeze-thaw cycles (FTC) with freezing times ranging from 20 to 30 seconds. Treatment was repeated at 3 week intervals and was follow-up up to 16 weeks in our clinic. On average, he received two treatments per lesion. Cryotherapy was well tolerated with only mild adverse reactions like blisters and pain at treatment site. The skin lesions responded well to therapy. A complete response was observed in about 80% of the treated KS lesions after 2nd course of cryotherapy. The remaining treated KS lesions resolved after 4th course of cryotherapy. Some lesions however healed with hypopigmentation (Figure 2).

In view of logistic problem, he was then referred back to his hometown, under the care of infectious disease physician in a tertiary hospital. He makes a good recovery from his illness. Currently, he is stable on anti-retroviral treatment with CD4 count of 180. No relapse was noted up to 6 months post treatment.

DISCUSSION

Kaposi's sarcoma was first described by Dr Moritz Kaposi in 1872. It is one of the AIDS defining skin diseases and is strongly linked to HHV 8 and male homosexual behaviour (as noted in our patient). KS is characterized by few or widespread multifocal brown violaceous or dark red colour patches, papules, plaques and/or deep skin nodules. Typically, the lesions are bilateral, symmetrically distributed along the lines of skin cleavage, involving the extremities. These lesions may involve the skin, mucosal membranes, lymph nodes and visceral organs such as the gastrointestinal tract, lungs, liver and spleen.

If left untreated, the median survival is 18 months[1]. Currently, antiretroviral therapy (ART) is the first-line therapy for Kaposi's sarcoma in patients with low CD4 counts and/or high viral loads. First-line treatment for Kaposi's sarcoma in patients with CD4 counts greater than 350 cells / µL is still unclear[2-3]. Treatment to date has been primarily palliative. Treatment goals in AIDS-KS include symptom palliation, prevention of KS progression, improvement of cosmesis and abatement of associated oedema, organ compromise and psychological stress[4].

Indications for systemic chemotherapy include widespread skin involvement (> 25 lesions), extensive oral KS, marked

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Corresponding Author: Tan Wooi Chiang, Hospital Pulau Pinang, Department of Dermatology, Jalan Residensi, 10990, Georgetown, Penang, Malaysia
Email: tanwooichiang@yahoo.com

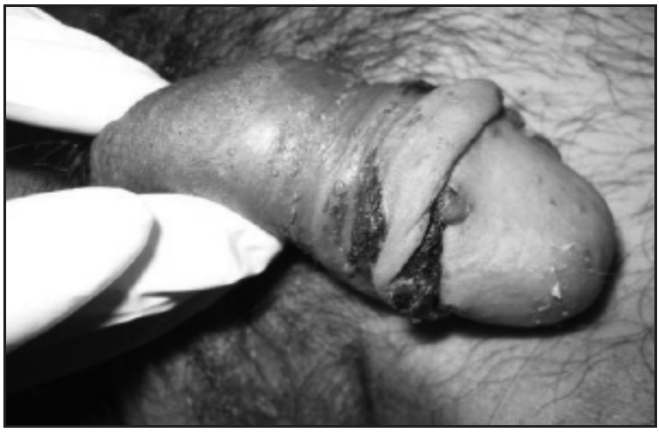

Fig. 1: Pre-treatment.

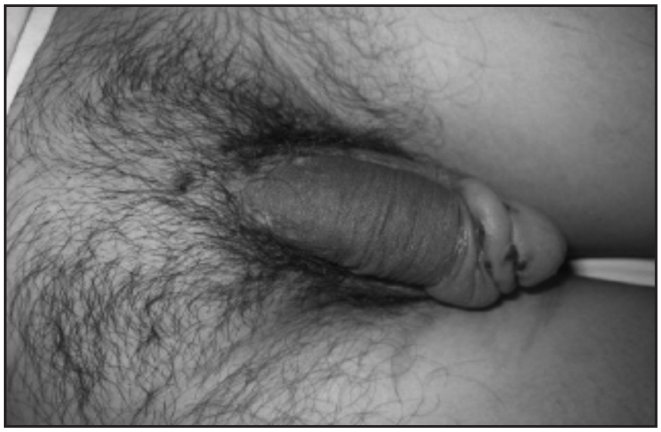

Fig. 2: After 2nd treatment.

symptomatic oedema, rapidly progressive disease, symptomatic visceral KS and KS flare. The decision to treat a patient with KS using HAART alone or HAART in combination with chemotherapy is a clinical one. Some authors suggest that patients with a poor risk (eg, age 50 years or older, occurrence of KS at or after AIDS onset, presence of co-morbid conditions) should be treated with combination of HAART and systemic chemotherapy right from the beginning, because without treatment they have a poorer outcome. Systemic chemotherapy that have been used are bleomycin and vinca alkaloids or a three drug regimen containing adriamycin (liposomal anthracycline). The taxol derivatives has been shown to be effective and with acceptable tolerance for long term administration.

There are many options for local treatment which include radiotherapy, electron beam therapy, photodynamic therapy, liquid nitrogen, intralesional vinca alkaloids, intralesional interferon alpha, topical imiquimod and others. Liquid nitrogen was selected in this case because cryotherapy is easily applied and the patient's ill condition precludes the use of systemic chemotherapy at this juncture. Tappero JW et al5 showed a favourable response to cryotherapy. 20 subjects with biopsy proven KS received 3 treatments per lesion with a mean follow-up time of 11 weeks. A complete response was observed in 80% of treated KS lesions. Tumor recurrence with cryotherapy is frequent and patients may experience pain and develop hypopigmentation[5]. Our patient shows no sign of recurrence of KS. He is in clinical clearance up to 6 months post cryotherapy.

CONCLUSION

Cryotherapy is a useful adjuvant therapy for the treatment of small and localized cutaneous KS. It may also be useful in the treatment of widespread cutaneous KS especially in patient who is unfit for systemic chemotherapy or in those with lesions that showed slow or incomplete cosmetic improvement with systemic therapies.

REFERENCES

1. Jones JL, Hanson DL, Dworkin MS, Jaffe HW. Incidence and trends in Kaposi's sarcoma in the era of effective antiretroviral therapy. J AIDS 2000: 24: 270-4.
2. Chan J, Kravcik S, Angel JB. Development of Kaposis's sarcoma despite sustained suppression of HIV plasma viremia. J AIDS 1999: 22: 209-210.
3. Cattelan AM, Calabro ML, De Rossi A, Aversa SM, *et al*. Long-term clinical outcome of AIDS-related Kaposi's sarcoma during highly active antiretroviral therapy. Int J Oncol. 2005 Sep; 27(3): 779-85.
4. Vanni T, Sprinz E, Machado MW, Santana Rde C, Fonseca BA, Schwartsmann G. Systemic treatment of AIDS-related Kaposi sarcoma: current status and perspectives. Cancer Treat Rev. 2006 Oct; 32(6): 445-55.
5. Tappero JW, Berger TG, Kaplan LD, Volberding PA, Kahn JO. Cryotherapy for cutaneous Kaposi's sarcoma (KS) associated with acquired immune deficiency syndrome (AIDS): a phase II trial. J Acquir Immune Defic Syndr. 1991; 4(9): 839-46.