

occurs more commonly in the vicinity of the aortic arch or the neck remains largely unresolved.

In conclusion, this preliminary report suggests that thoracoscopic oesophagectomy is associated with a high

morbidity and this must be taken into consideration by those who are likely to undertake such a procedure. The role of this procedure in the treatment of oesophageal cancer certainly needs further evaluation.

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# A Report of Two Non-AIDS associated Kaposi's Sarcoma in Malaysia

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## Summary

Kaposi's sarcoma is an uncommon cutaneous neoplasm seen classically in elderly males of East European or Jewish extract. It has been known to be endemic in sub-Saharan Africa for many years. Numerous cases had been described in patients on long-term immunosuppressive therapy and in patients living with acquired immunodeficiency syndrome (AIDS). In spite of the increasing number of organ transplant recipients and people living with AIDS. Kaposi's sarcoma remains rare in Asia. We report two cases seen in Johor, Malaysia.

**Key Words:** Kaposi's sarcoma, Non-AIDS associated, Malaysia

## Introduction

Kaposi's sarcoma (KS) was first described in 1872 by a Hungarian dermatologist, Moritz Kaposi. During the

next century, this uncommon tumour was seen in four different clinical settings<sup>1</sup>. Classic KS occurs in elderly men (male/female ratio, 10-15:1) of Mediterranean, East European or Jewish heritage. The lesions are

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usually confined to the legs and the disease runs an indolent course with patients dying from unrelated causes after 10-15 years. Secondary malignancies develop in about 35% of cases.

KS has been known to be endemic in sub-Saharan Africa since early 1960s. African-endemic KS affects two distinct age groups. Fulminant lymphadenopathic disease presents with multiple enlarged lymph nodes in young African children (mean age, 3 years; male/female ratio, 3:1) and it is invariably fatal within 2 to 3 years. The other affected age group are young African adults (mean age, 35 years; male/female ratio, 13-17:1) with a generally benign nodular disease. But, sometimes the disease can be aggressive and fatal within 5 to 8 years.

KS has been described in a wide spectrum of patients receiving chronic immunosuppressive therapy. Iatrogenic immunosuppressive drug-associated KS is seen predominantly in organ transplant recipients. Although the disease may be aggressive or chronic, spontaneous remission occurs after cessation of immunosuppressive therapy.

It is the AIDS epidemic that thrust this relatively unknown tumour into the medical limelight in 1981 when Centre for Disease Control, Atlanta, reported 26 cases in homosexual men with AIDS. Subsequently, KS was described in homosexual men without Human immunodeficiency virus (HIV) infection. However, KS whether AIDS or non-AIDS associated is rare in Asia. Hence, two cases seen in Malaysia are described here.

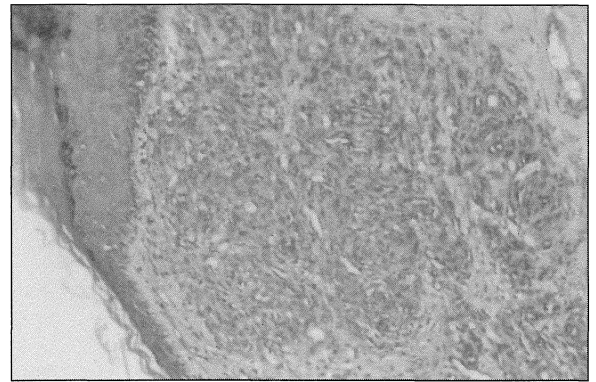
### Case reports

#### Patient 1

A 29-years-old Malay man, a bank officer, was admitted to our orthopaedic ward for septic arthritis of his right hip in September 1993. He was referred to us two weeks later for a slowly enlarging non-irritating skin lesion on his right iliac fossa that has been present for two years. He has been married for three years and denied homosexuality, multiple sexual partners and intravenous drug use. However, he stayed with 3 men who had urethral discharge before his marriage.

Physical examination revealed a 4cm by 2cm non-tender, purplish oval plaque on his right iliac fossa. There was no lymphadenopathy or visceral involvement. Histopathological examination of a skin biopsy specimen (Fig. 1) was compatible with the diagnosis of KS. Patient rejected genital examination and only permitted serological screening for syphilis and HIV infection. His HIV antibody was repeatedly negative (ELISA and western blot). Both VDRL (dilution, 1:2) and TPHA were positive. His full blood count was normal. The absolute number of CD4 T lymphocytes was also normal but there was a reversal of CD4 to CD8 T cell ratio.

Both patient and wife had late latent syphilis and were treated with intramuscular penicillin. Patient chose to leave the KS alone and defaulted 6 months later.



**Fig. 1:** Shows a dermal nodule consisting of a mass of spindle cells and multiple slit-like vascular spaces filled with red blood cells and some haemosiderin.

#### Patient 2

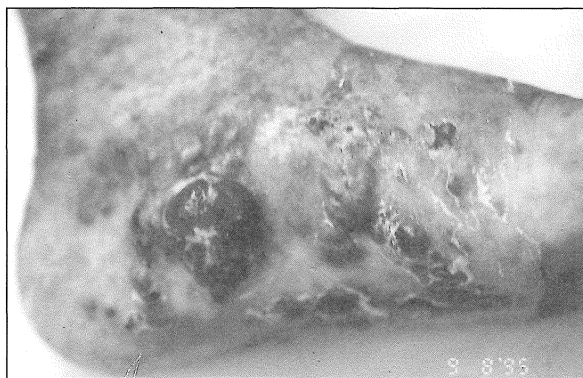
A 51-years-old Chinese lorry driver, presented in August 1995, with a 2-month history of multiple painful nodules on his left foot. He first noted painful nodules on the dorsum of his left foot which subsequently spread up his left lower limb. He had been divorced for 10 years and denied homosexuality, multiple sexual partners and intravenous drug use. He was on glibenclamide for diabetes mellitus and perindopril for hypertension.

Physical examination revealed on oedematous lower limb with multiple purplish papules and nodules,

ranging in size from 2mm by 2mm to 2cm by 2cm, which were located mainly on the dorsum and medial aspect of left foot, and on the medial aspect of the left lower limb. A 3cm by 3cm ulcerated nodule was also present on the medial aspect of his left foot (Fig. 2). There was no significant lymphadenopathy or organomegaly.

Histopathological examination of a skin specimen was compatible with a diagnosis of KS. His full blood count was normal except for a low total lymphocytes of 800/ $\mu$ l and less than 1% of this was CD4 cells. Repeated blood tests were negative for HIV antibody. VDRL and TPHA were repeatedly positive.

The patient was treated for late latent syphilis and was referred to Hospital Kuala Lumpur for further management of the KS. He was deemed unfit for radiotherapy because of development of cardiac failure secondary to ischaemic heart disease and hypertension. Intravenous vinblastine was started but patient rejected further treatment because he attributed his cardiac failure to vinblastine and was transferred back to us. He could not tolerate cryotherapy and refused simple excision. Subcutaneous interferon  $\alpha$ -2b, 5 million IU thrice a week was started. After 2 months of treatment, pain subsided with flattening of skin lesions and patient permitted excision of the much shrunken ulcerated nodule. After 6 months of treatment, there were only a few tiny papules left. Subsequently, the patient died suddenly at home.



**Fig. 2: Kaposi's sarcoma; multiple purplish nodules**

## Discussion

Although the AIDS epidemic prompted zealous research on all aspects of KS, the cell of origin of KS and its pathogenesis remain obscure<sup>1</sup>. KS most probably arises from mesenchymal cells of lymphatic or vascular endothelium. Epidemiologic studies suggest that KS is caused by a sexually transmitted agent which is more readily expressed in HIV infection and other immunocompromised states. Infective agents implicated include cytomegalovirus (CMV), HIV itself, human papilloma virus (HPV) and, recently, a newly identified human herpes virus type 8 (HHV8). The risk of developing KS is associated with sexual practices that involve faecal contact such as insertive anilingus, insertive fisting and insertive anal intercourse<sup>2</sup>.

Although both our patients are HIV negative, they had some degree of immunosuppression as reflected by the reversal of CD4 to CD8 T cell ratio in the first patient and a severe CD4 lymphopaenia in the second patient. Both patients had late latent syphilis and hence, were at risk of contracting other sexually transmitted diseases including KS. KS remains rare in Malaysia in spite of increasing number of organ transplant recipients and people living with AIDS probably due to under-diagnosis, under-reporting and low prevalence of, the causative agent and sexual practices involving faecal contact.

There is no definitive cure for KS. All treatment modalities are palliative and have only temporary efficacy<sup>1</sup>. Specific indications for treatment include cosmetic control of disfiguring KS, shrinkage of problematic lesions, abatement of pain and oedema associated with lymphadenopathy or extensive cutaneous disease and alleviation of symptomatic systemic disease. Local therapies include simple excision, cryotherapy, laser treatment and intralesional therapy.

Simple excision is occasionally of benefit but frequently unsatisfactory. Cryotherapy leads to a complete or partial response rate of about 85%. Usefulness of laser therapy which has a similar response rate as cryotherapy is limited by possible risk of infectious viral particles in the vapour plume. Intralesional vinblastine is fast and inexpensive but is only suitable for isolated lesions. Side-effects include pain, skin

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irritation,<sup>2</sup> ulceration and postinflammatory hyperpigmentation. Intralesional interferon is still experimental and offers no advantage over cryotherapy or intralesional vinblastine.

Lesions of KS are highly radiosensitive. Radiotherapy is well tolerated and temporarily controls large localised lesions. Patients with non-AIDS associated KS treated with either localised or extended field radiotherapy or total electron beam therapy had response rates of more than 80%. Radiotherapy is particularly useful in controlling symptoms due to mass effects such as painful localised lymphadenopathy and localised lymphoedema of extremities and penis.

Systemic interferon- $\alpha$  has been evaluated extensively in AIDS-associated KS. However, large doses are required to obtain response rates of between 20% to 40%, depending on the immunologic status of the patients. Efficacy of low-dose interferon- $\alpha$  treatment in endemic and classic KS had been reported in several small series. Costa da Cunha *et al*<sup>3</sup> reported the results

of subcutaneous interferon- $\alpha$ , 5 million U, 3 times a week for at least 6 months in 13 patients with classic KS and 3 patients with endemic KS. One patient had a complete response while 9 patients showed major improvements. The response is less in those with advanced disease. Our patient benefited from low-dose interferon- $\alpha$  and was most relieved when the local pain and oedema subsided.

Single-agent or combined systemic chemotherapy is used mainly for patients with rapidly progressive KS or those with symptomatic visceral disease. The most widely used drugs are vinca alkaloids (vinblastine and vincristine). Other beneficial agents include bleomycin, doxorubicin, dactinomycin, dacarbazine, etoposide, teniposide and epirubicin.

### Acknowledgement

I would like to thank the Director General, Ministry of Health, Malaysia, for his permission to publish this paper.

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