Kaposi's sarcoma in patients with the acquired immunodeficiency syndrome: the Hong Kong experience

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Acquired immunodeficiency syndrome-related Kaposi's sarcoma has become an important disease entity, both in the field of human immunodeficiency virus/acquired immunodeficiency syndrome as well as neoplastic disorders. In the 10-year period from February 1985 through December 1994, 17 (13%) of the 130 acquired immunodeficiency syndrome patients who reported to the Department of Health, Hong Kong, had Kaposi's sarcoma as primary (11) or subsequent (6) acquired immunodeficiency syndrome-defining illnesses. All of these patients had acquired the virus through sexual transmission, with 88% via homo- or bi-sexual contacts. Nine patients were Chinese, and seven were diagnosed in or after 1992. Solitary or multifocal cutaneous lesions were the commonest presentations. Only one patient had visceral involvement. The median CD4$^+$ T-lymphocyte count at diagnosis was 91 cells/μL (normal range, >290 cells/μL). Eleven patients had died, but no deaths were directly related to the Kaposi's sarcoma. The mean survival after diagnosis of Kaposi's sarcoma was 14.4 months.

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Introduction

Kaposi's sarcoma (KS), an angioproliferative disease, was first described more than 100 years ago by a dermatologist in Vienna.[1] Before the acquired immunodeficiency syndrome (AIDS) era, two main forms of KS existed. These were the endemic form in Central Africa and the rare form that affected only elderly men of Mediterranean origin. With the discovery of its association with the human immunodeficiency virus (HIV) and AIDS, the epidemiology of KS has changed considerably. Compared with the risk of the general population, the risk of KS developing in men with AIDS is a few hundred times higher. Acquired immunodeficiency syndrome-associated KS has become the most important subgroup of the disease in the past decade.

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It is known that KS is more common among patients who acquire HIV through sexual contacts, particularly via homosexual activity. Hence, the geographical distribution of KS also reflects the pattern of sexually-acquired HIV infection throughout the world. Currently, research on KS is focused on the American and European nations, and on Africa to a lesser extent. This study explores the dimension and features of KS in Hong Kong, a predominantly Chinese community. The epidemiology of the HIV/AIDS pandemic in Hong Kong is closely following that seen in the United States and other western countries. The main difference is that the infection started later, in the mid- to late- 1980s. An in-depth study of AIDS-related clinical conditions in Hong Kong would be useful for the development of clinical services in Asian communities.

Materials and methods

Both HIV infection and AIDS are not notifiable diseases in Hong Kong. Instead, a voluntary reporting system has been adopted for the collection of epidemiological data on the epidemic. Medical practitioners are urged to report cases of HIV infection and AIDS to the Department of Health, and to notify the department whenever new AIDS-defining conditions are di-

agnosed. This reporting system has been in operation in Hong Kong since 1984.

In Hong Kong, AIDS is diagnosed in accordance with the 1987 surveillance definition of the Centers for Disease Control (CDC) in the United States.[2] A patient is diagnosed as having AIDS if they have laboratory evidence of HIV infection and an AIDS-defining clinical condition is present. Infection with *Penicillium marneffei*, a fungus that is endemic in Southeast Asia, is also included as an AIDS-defining condition.[3] The first AIDS patient in Hong Kong was reported in early 1985, and this study looks at patients reported during the 10-year period between February 1985 and the end of December 1994.

The records of AIDS patients whose presenting clinical condition was KS, or who developed KS after AIDS diagnosis, were retrieved. The year of diagnosis, demography, clinical presentation, treatment given, and outcome were noted.

Results

As of 31 December 1994, 520 patients with HIV infection had been reported to the Department of Health, of whom 130 were known to have deveoloped AIDS. All had HIV-1 infection and no HIV-2 infection had reported during the study period. The majority (77.3%) of patients acquired the infection through sexual intercourse, and only 8% were women (Table 1). Seventeen (13%) of the 130 AIDS patients had KS—11 had it as a primary AIDS-defining illness. The clinical records of all patients were available for analysis.

Demography

All patients with KS were men aged from 26 to 59 years (mean age, 36 years). All acquired HIV through sexual exposure—the transmission routes being homosexual (11), bisexual (4), and heterosexual (2). This gives an odds ratio of 8.5 (95% CI, 1.7-56.6) for homo-/bi-sexual transmission. To date, no known HIV-infected injecting drug users or recipients of blood or blood products have been reported as having KS. Approximately two to three patients were reported yearly. Although nearly half of all patients were Chinese, the proportion has increased in recent years, as 77.7% of those diagnosed in or after 1992 were Chinese, whereas this was only 25% prior to 1992. This difference did not, however, reach statistical significance (P = 0.056, Fisher's exact test).

Clinical manifestation

Kaposi's sarcoma was the primary AIDS-defining condition in 8% of all reported AIDS patients during the study period, ranking third overall, after *Pneumocystis carinii* pneumonia (44%), and tuberculosis (10%). The majority had significantly impaired immunological function at the time of diagnosis. Of the 16 patients whose $CD4^+$ levels were available, only two had counts above 200 cells/μL, and the median level was 91 cells/μL (range, 6-499 cells/μL).

The commonest presentation was of multiple cutaneous lesions, usually presenting as purplish macules or nodules (Table 2). Two had solitary lesions on the forearm and the calf. Dissemination to the gastrointestinal tract and bone was diagnosed in one patient. Diagnosis of KS was confirmed histologically

Table 1. Cumulative statistics on human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS) reported in Hong Kong as of 31 December 1994

		Total	
		HIV infection	AIDS
Gender	Male	478	122
	Female	42	8
Ethnicity	Chinese	346	90
	Non-Chinese	174	40
Transmission route	Homo-/bi-sexual	190	68
	Heterosexual	212	41
	Injecting drug users	11	2
	Blood/blood product recipients	66	12
	Mother-to-child	1	1
	Undetermined	40	6
	Total	520	130

in five patients and was based on macroscopic appearance in the remainder. Seven patients had other clinical problems at the time of diagnosis, these being pneumonia (3), tuberculosis (2), cytomegalovirus retinitis (1), and eosinophilic folliculitis (1).

Treatment and outcome

No specific treatment was given to 10 patients. The various treatments included α-interferon (2), radiotherapy (2), intralesional vinblastine (1) and intravenous vinblastine (2). The indication for providing the treatment was symptomatic relief in three patients and for cosmetic reasons in four. Improvement was noted in almost all patients after treatment. Eleven patients had already died but none was directly related to KS. The median survival of the deceased patients was 14.4 months (range, 2-36 months) after the diagnosis of KS.

Discussion

Kaposi's sarcoma is an important AIDS-defining condition in patients infected with HIV. In the early 1980s, some 35% to 40% of all reported AIDS patients in the United States presented first with KS.[4] The incidence has, however, been decreasing in recent years. Kaposi's sarcoma accounted for 11% of AIDS patients reported in 1989,[5] falling to only 4% in 1994.[6] In Australia, 17.2% of all adult AIDS patients present with KS as the initial AIDS-defining illness.[7] The incidence of KS in Orientals is not well-known, and only a limited number of cases have been reported in Chinese people.[8,9] The current study attempts to explore the epidemiological and clinical characteristics of AIDS-related KS in Hong Kong, which has a predominantly Chinese community.

Hong Kong has a population of approximately 6 million people. The first HIV-infected patient in Hong Kong was reported in 1984. As of 31 December 1994, a cumulative total of 520 and 130 HIV and AIDS patients, respectively, had been reported. However, the estimated numbers stood at 3000 and 250.[10] Eleven of the 130 (8%) AIDS patients presented with KS as the primary AIDS-defining condition, similar to the incidence found in the United States in the early 1990s. Because of the small number of people involved, analysis of the temporal trend cannot be made.

Epidemiological evidence suggests that the homo-/bi-sexual transmission of HIV is a major risk factor for the development of KS.[11] Kaposi's sarcoma could be caused by the combination of immunosuppression and a sexually transmitted agent, one possible candidate being the human herpesvirus type 6 (HHV-6).[12] In Hong Kong, all reported cases of AIDS-related KS occurred in men who contracted the virus through sexual contact—15 homo-/bi-sexual and two heterosexual men. This gives an odds ratio of 8.5 (95% CI, 1.7-56.6) for homo-/bi-sexual transmission, which is compatible with the findings of other studies. Nine patients were Chinese. With the increasing incidence of HIV infection in the local community, more KS cases can be expected to be seen in the Chinese population in the future. Kaposi's sarcoma is rare in individuals not infected with HIV. Only three cases have been diagnosed in the government's dermatology service in recent years (YM Tang, personal communication).

The commonest clinical presentation of KS is that of solitary or multifocal cutaneous nodules.[4] Fifteen of the 17 reported patients in this study had such presentations. Systemic presentation was diagnosed in only one patient. Although KS could occur in the absence of immune impairment, all of the patients here had a low $CD4^+$ count at diagnosis. This could possibly be explained by the late presentation of patients with HIV/AIDS in the local setting, especially those diagnosed in the early years. In view of the variability in the timing of presentation, there may not be a direct relationship between the $CD4^+$ count and the development of KS.[13] Nevertheless, staging systems incorporating $CD4^+$ counts have been developed for KS.[4]

The information collected from this study may have been limited by sampling bias as it relied on the reporting of cases. Nevertheless, findings of the prevalence and presentation of KS among the AIDS patients in our study are similar to those found in overseas countries. In conclusion, KS accounts for a small but significant proportion of all AIDS-defining illnesses in patients in Hong Kong. The clinical presentation is similar to that reported in other countries. Patients with KS often presented late when their $CD4^+$ count was low. An ongoing epidemiological study is warranted to detect changes in incidence and clinical presentation.

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Table 2. Summary of acquired immunodeficiency syndrome (AIDS) patients with Kaposi's sarcoma (KS) in Hong Kong

No.	Ethnicity	Risk	Age	Year of KS diagnosis	KS as 1°or 2° diagnosis	CD4 + count at diagnosis (cells/μL)	Extent of lesions	Treatment	Survival after KS diagnosis (months)	Cause of death
1	CA*	BI§	28	1985	1°	139	c	Interferon	4	*Pneumocystis carinii* pneumonia
2	CA	HO	52	1988	2°	37	a	Nil	3	*Pneumocystis carinii* pneumonia
3	CH†	HO	30	1988	1°	77	b	Nil	15	Cryptococcosis
4	CA	HO	39	1989	2°	104	c	Intralesional Vinblastine	17	Unknown
5	CA	HO	46	1989	1°	499	c	Interferon	Lost to follow up	-
6	CA	HO	30	1989	1°	199	c	Nil	22	Unknown
7	AS‡	BI	26	1990	1°	-	c	Nil	14	Cryptococcosis
8	CH	HE¶	43	1992	2°	6	c	Nil	36	Fungal infection
9	CA	HE	59	1992	1°	109	c	Intravenous Vinblastine	15	*Penicillium marneffei* infection
10	CH	HO	40	1990	1°	120	c	Intravenous Vinblastine	23	Septicaemia
11	CH	HO	35	1992	2°	216	?	Nil	2	*Pneumocystis carinii* pneumonia
12	CH	HO	34	1993	2°	91	d	Radiotherapy	Lost to follow up	
13	CA	BI	52	1993	2°	39	c	Nil	Alive	
14	CH	BI	49	1994	1°	50	a	Nil	Alive	
15	CH	HO	42	1994	1°	21	c	Nil	8	Gastrointestinal bleeding
16	CH	HO	34	1994	1°	90	b	Radiotherapy	Alive	
17	CH	HO	33	1992	1°	60	c	Nil	Alive	

*CA caucasian;†CH Chinese;‡AS Asian,non-Chinese;§BI bisexual;‖HO homosexual;¶HE heterosexual
a) Solitary b) One region c) Diffuse d) Internal organ involvement