Oral Oncol, Eur J Cancer, Vol. 29B, No. 3, pp. 187–189, 1993
Printed In Great Britain

0964–1955/93 $6.00 + 0.00

Papers

Epidemic Oro-facial Kaposi's Sarcoma (eKS)—Report on 124 Cases

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Oro-facial epidemic Kaposi's sarcoma (eKS) was observed in 124 patients (123 homo-/bisexual men, average age 37.9 years, and one 35-year-old transsexual patient). Average survival time was 1 year 9 months (range: 3 months–4 years 6 months). 57.3% of the patients had died. The hard and/or soft palate, gingiva and tongue were most frequently affected, with falling frequency within these locations. Oral manifestations of eKS appear to be common; treatment modalities must be optimised.
Oral Oncol, Eur J Cancer, Vol. 29B, No. 3, pp. 187–189, 1993.

INTRODUCTION

ORAL MANIFESTATIONS of epidemic Kaposi's sarcoma (eKS) have been classified as lesions strongly associated with the acquired immunodeficiency syndrome (AIDS) [1]. Besides a number of earlier case reports on oral manifestations of eKS, several larger studies have been published describing clinical, therapeutic and morphological aspects of this entity [2–6]. An extensive review of head and neck malignancies associated with HIV infection with particular emphasis on eKS was recently published [7, 8].

The purpose of this communication was to present epidemiological, clinical and therapeutic findings of eKS with oro-facial manifestations in 124 patients observed in the Department of Oral Surgery (North), Freie Universität Berlin, during the period from January 1984 to June 1992 and to compare findings with those of other reports.

PATIENTS AND METHODS

All patients with HIV infection and AIDS who were seen from January 1984 to June 1992 were registered and classified according to the CDC classification. Oral manifestations were monitored following the EEC classification which was re-evaluated in 1991 [1]. The following parameters of 124 patients were registered: gender, mean age, year of birth, risk group, number of initial diagnoses of oral eKS per year, average survival time, number of deaths, oral localisation of eKS, clinical appearance and size of lesions, patterns of complaints, and types and modalities of treatment.

Diagnosis of eKS was made on clinical and histological criteria, particularly in cases with oral lesions only. In cases with disseminated eKS oral lesions were not always proven by means of biopsy (one-third of cases). Oral eKS in patients at risk for AIDS is clinically characteristic, so that biopsy is often a formality [7].

RESULTS

From January 1984 to June 1992 a total of 873 patients with HIV infection were seen in the Department of Oral Surgery (North), Freie University of Berlin. Of these, 498 patients were classified as AIDS patients. Of these, 470 were men [412 homosexual, 8 bisexual, 24 intravenous drug users (IDU), 1 haemophiliac, 25 unknown] and 28 were women (26 IDU, 1 heterosexual, 1 transplant patient). 124 patients (24.9% of AIDS cases) were observed with extraoral and/or oral eKS as the initial diagnosis. In addition 32 patients not included in this series were seen with extraoral eKS alone. Of these, 15 revealed facial manifestations of eKS. Of 124 patients, 102 had manifestations of oral eKS and 25 patients presented with oral eKS alone. 123 patients were homosexual/bisexual with a mean age of 37.9 years (±1.2 years, range 21–64 years); one 35-year-old patient was transsexual. Table 1 shows the number of patients with oral eKS per year of birth. The majority of patients were born between 1941 and 1960 ($n=82$). Table 2 shows the number of cases with oral eKS in relation to number of AIDS cases per year of initial diagnosis. Most cases were seen in 1988 and 1989 ($n=38$). The average survival of patients with oral eKS was 1 year 9 months with a range of 3 months to 4 years 6 months. 71 patients have died (57.3%). The hard and soft palate was most often affected by oral eKS ($n=49$), followed by the gingiva ($n=39$), tongue ($n=17$), buccal mucosa ($n=11$), pharynx ($n=8$) and uvula ($n=2$) (several localisations per patient). In addition, 2 cases of intraosseous KS were seen [9]. Clinically oral eKS lesions appeared as bluish, brownish or violet in colour except 1 case which was non-pigmented [13]. 59 of the lesions were flat, 67 exophytic or nodular; of these, five were superficially ulcerated (several lesions per patient). 38 of the oral eKS lesions were less than 1 cm in diameter, 50 were between 1 and 3 cm and 38 were more than 3 cm in size (several eKS lesions per patient). There were not many complaints caused by oral eKS including local discomfort ($n=18$), irradiating pain ($n=6$),

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Received 5 Nov. 1992; revised manuscript received 12 Nov. 1992; revised manuscript accepted 7 Dec. 1992.

Table 1. Year of birth of patients with oral eKS

Year	n
Before 1930	2
1930–1940	7
1941–1950	43
1951–1960	39
1961–1970	9
After 1970	2

dysphagia ($n=5$), taste of blood ($n=4$), problems with breathing ($n=1$) and paresthesia ($n=1$).

Treatment regimens varied during the entire period of observation which makes comparison very difficult. Patients received systemic cytostatic drugs (bleomycine, vinblastine, vincristine, actinomycin D, doxorubicin, etoposide a. o. and combinations thereof) ($n=39$); 8 patients received systemic immuno-modulatory treatment using interferon α_2. Radiation (dosage: 20 Gy) was used in 43 cases. Intralesional cytostatic drugs (vinblastine, bleomycin) were used in 12 patients. Surgical excision was performed in 5 cases (several modalities of treatment per patient). Details of dosages and outcome of different treatment modalities are presented in a further paper and are discussed here only briefly.

It was of interest to see oral hairy leukoplakia (HL) in 38 of 102 patients with oral eKS (37.3%). Oral candidiasis was observed in 60.8% (different types), HIV-associated gingivitis was seen in 13.7%, periodontitis in 11.8% and necrotising gingivitis in 7.8%. 11.8% of the patients showed atypical ulcerations. In 29.2% of the patients with oral eKS this was the only presenting symptom. 27.1% of the patients suffered from additional opportunistic infections. Of these 12.5% were pneumocystis carinii pneumonia (PCP) and 14.6% were other opportunistic infections including toxoplasmosis, herpes simplex virus and cytomegalovirus infections.

DISCUSSION

In Berlin (West) 1774 patients with AIDS had been registered by June 1992. Of these 1680 were men and 94 were women; 801 patients were registered as having died. Berlin (west) has for some time attracted people who have to be considered risk groups, particularly homosexual men; therefore the incidence of HIV-infected patients and AIDS per 100 000 is the highest in Germany. In the whole of Germany 8463 cases of AIDS had been registered at that time. Of these 1279 (15.1%) were registered with eKS. Another 436 (5.2%) were registered with opportunistic infections and eKS [15]. Compared with the percentage of cases with eKS in Germany (15.1% vs. 5.2%) the percentage of patients with oral eKS in relation to the number of AIDS cases (20.5%) is high and may be due to the specific hospital setting where most of the patients are hospitalised as full-blown AIDS cases. In about 25% ($n=25$) of the cases manifestations of eKS were restricted to oral lesions. In an earlier study [5] KS exclusive of other sites was seen in 7 of 47 AIDS patients (15%). In other studies it was 5% [4] and 4% [10]. Small numbers of patients with oral eKS were also recorded in other studies ($n=2$ of 33) [11]; 2 of 4 AIDS patients [12]. In another study [6] more than 20% of patients had the oral cavity as the initial site of manifestation. All patients except 1 were homosexual or bisexual, a finding which is consistent with other studies of oro-facial manifestations of eKS [2, 4, 6]. The average age of patients was 37.9 years which compares well with other figures [4], and 34 years [6]. A little less than 50% (50 of 102 patients) were below the age of 41. As is shown in Table 2 a decrease of patients with oral KS was observed since 1984. In another study the relative rate of eKS patients with AIDS had dropped from 34 to 20% during an observation period of 4 years [6].

Table 2. Initial diagnosis of oral eKS/year

Year	No. of eKS patients	No. of new AIDS patients/year	eKS patients/year (% of AIDS patients)
1984	5	14	35.7
1985	8	29	27.6
1986	12	43	27.9
1987	10	44	22.7
1988	18	78	23.1
1989	20	89	22.5
1990	14	83	16.9
1991	10	79	12.7
1992	5	39	12.8

The average survival time of all patients with oral eKS including those with opportunistic infections was 21 months with a range of 3 months to 4 years and 6 months. In a former study median survival time was 14 months in patients with oral eKS without opportunistic infection. It was 6–9 months in patients with opportunistic infection and oral eKS [6].

The present study has confirmed the more common localisations for oral eKS being the hard and soft palates, the gingiva and the tongue [6, 16]. The clinical appearance of oral epidemic KS is typical although variations such as non-pigmented KS or intraosseous manifestations may occasionally be seen [9, 13]. 35 of 102 patients (34.9%) had symptoms because of the presence of oral eKS, the percentage being comparable with the one published by Ficarra *et al.* [6] (26.8%). The incidence of other associated oral lesions in patients with eKS is comparable with other studies [16], although in the latter study HL was observed in 60.6% compared to 37.3% in our cohort.

The multiple types of treatment of oral eKS including systemic and/or local therapies are difficult to compare and evaluate. Although 94 patients received azidothymidine (AZT) the anti-retroviral therapy is not an eKS treatment *per se* and is supposed to have no significant effect on the course of the disease [14]. All treatments used so far are palliative, of which some have now been given up. Particularly surgical excision is not considered to be effective; only 5 patients were treated by these means. The number of patients who were treated by intra-lesional cytostatic drugs is too small to allow for a definite evaluation. In 43 of the cases intraoral radiation was used. However, side-effects such as mucositis ($n=6$) or palatal perforation ($n=1$) have been observed. Treatment modalities and results for oral eKS are still not satisfactory, and as has been stated "many patients require infinite treatment for control of lesions" [6]. Controlled therapy studies for oral eKS are necessary to improve oral health in these patients.

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