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Original Paper

Descriptive Epidemiology of Soft Tissue Sarcomas in Vaud, Switzerland

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Trends in incidence and survival from soft tissue sarcomas (STS) were analysed for the period 1974–1994 using data from the Cancer Registry of the Swiss canton of Vaud. A total of 645 cases were registered. The most common histotypes were fibrosarcoma (0.82/100 000 males, 0.86/100 000 females, world standard, 1990–1994), leiomyosarcoma (0.90/100 000 males, 1.28/100 000 females, 1990–1994), and Kaposi's sarcoma (3.10/100 000 males in 1990–1994). Overall incidence rates for STS increased from 2.68/100 000 males in 1974–1979 to 6.86 in 1990–1994, and from 3.61 to 4.27 in females. However, after excluding Kaposi's sarcoma, no consistent trend over time was observed, peak rates (approximately 4.40/100 000) being registered in the late 1980s for both sexes, with some levelling off thereafter. Five-year relative survival was 17% for Kaposi's sarcomas, and 51% for other STS (all STSs, 45%). These data indicate that there has been no major new risk factor for STS of such a relevance to modify appreciably the overall rates on a population level, except from the impact of the AIDS epidemic for Kaposi's sarcoma. © **1999 Elsevier Science Ltd. All rights reserved.**

Key words: soft tissue sarcoma, Kaposi's sarcoma, morphology, trends, incidence rates, survival, descriptive epidemiology, registry, Switzerland
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INTRODUCTION

SOFT TISSUE sarcomas (STS), or cancers of connective tissues, are rare neoplasms arising from mesenchymal tissue, including muscle, fat, blood vessels, and fibrous tissues.

Most information on descriptive epidemiology of STS comes from data collected between 1975 and 1989 by the US Surveillance, Epidemiology and End Results (SEER) Programme [1–3]. These indicate that fibrosarcoma, leiomyosarcoma, blood vessel (including Kaposi's) sarcoma were the most common histological types. Incidence rates increased from the early 1980s onwards, this being essentially explained by the AIDS-related Kaposi's sarcoma epidemic, whilst no consistent pattern of trends was observed for other histologies.

Other population-based series from Denmark [4], New York State [5], Iceland [6] and North West England [7], showed overall stable rates between 1950 and 1980 and suggested that the incidence of STS is somewhat higher in the black population than caucasians, and in females than in males, due to tumours of the female genital tract.

International variation in STS incidence shows high rates in New Zealand Maoris and low ones in Japanese, but registered rates may be influenced by differences in case ascertainment and registration [8]. Kaposi's sarcoma was extremely rare in western populations, and was observed mainly in elderly Italian or Jewish men, often with an indolent clinical course. This pattern changed following the AIDS-related Kaposi's sarcoma epidemic, mainly spreading in young males from North America and Northern Europe [9]. Trends in incidence of other histological types have been related to consumption of phenoxy-herbicides, but this issue remains open to discussion [4].

Five-year relative survival rates were approximately 55% in the US National Cancer Database [10], and in a study from Italy [11], but only 36% in the SEER database, including

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AIDS-related Kaposi's sarcomas [2], and even lower in a study of 129 cases from Iceland over the period 1955–1988 [6]. Five-year relative survival was 59% for both sexes in the EUROCARE dataset, and tended to be lower in eastern Europe [12]. Overall survival was highest for liposarcomas and fibrosarcomas, and lowest for blood vessel (Kaposi's) sarcomas [2].

Since additional studies that examine other populations or more recent time periods have been recommended for a better understanding of the epidemiology of STS [1], we systematically analysed the data collected by the Vaud Cancer Registry over the period 1974–1994.

MATERIALS AND METHODS

The information was extracted from the Vaud Cancer Registry file which includes data on cases of malignant neoplasms in the canton (the population, according to the 1980 census was 529 000 and according to the 1990 one, was 602 000) [13]. Information collected by the register includes general demographic characteristics of the patients (age, sex, municipality of residence), site and histological type of the tumour according to the standard International Classification of Diseases for Oncology (ICD-O) [14], and time of diagnostic confirmation.

The present series comprises histologically confirmed malignant STS registered between 1974 and 1994 in the population of the canton. After reclassification of imprecisely classified tumours, and exclusion of 29 cases (whose ICD-O M [14] is listed in Table 1), 645 cases of STS were available for the present report and grouped into the following 12 morphological categories: (1) sarcomas not otherwise specified (NOS) (ICD-O M: 8800-4); (2) fibrosarcomas (ICD-O M: 8810-32); (3) liposarcomas (ICD-O M: 8850-70); (4) leiomyosarcomas (ICD-O M: 8890-1); (5) blood vessel sarcomas, including myosarcomas (ICD-O M: 8894, 8895, 9120, 9130, 9150); (6) rhabdomyosarcomas (ICD-O M: 8900-20); (7) stromal sarcomas (ICD-O M: 8930); (8) Kaposi's sarcomas (ICD-O M: 9140); (9) synovial sarcomas (ICD-O M: 9040-4); (10) meningiosarcomas (ICD-O M: 9530); (11) mesenchymoma (ICD-O M: 8990); (12) other morphological types (ICD-O M: 9240 = mesenchymal chondrosarcoma; 9370 = chordoma; 9490 = ganglioneuroblastoma; 9500 = neuroblastoma; 9540 = neurofibrosarcoma; 9560 = malignant schwannoma; 9581 = alveolar soft part sarcoma). Six separate groups of sites of origin were considered (stomach, intestines (incl. small intestine), (retro)-peritoneum, soft tissue, skin, corpus uteri) plus a heterogeneous group of others and undefined.

Table 1. *Distribution of* 29 *excluded cases according to morphological type*

Morphological type (ICD-O M; 14)	*n*
8000: unclassified	6
8010: carcinoma not otherwise specified	2
8033: pseudo-sarcomatous carcinoma	3
8200: adenoid cystic carcinoma	1
8743: superficial spreading melanoma	1
8951: mesodermal mixed tumour	1
9071: endodermal sinus tumour	2
9180: osteosarcoma	1
9580: granular cell myoblastoma	1
9591-9696: malignant lymphoma	4
9700: mycosis fungoides	1
9720: malignant histiocytosis	1
9731: plasma cell sarcoma	1
9990: tumour not microscopically confirmed	4

Overall age-standardised incidence rates (by the direct method, world standard population) were computed for the calendar periods 1974–1979, 1980–1984, 1985–1989 and 1990–1994 [15].

Information on survival is regularly integrated from mortality statistics into the registry database and, for patients who are apparently alive, through an active follow-up based on verification of vital status from registries of current residence. The vital status of each patient was verified up to 31 December 1996. On the basis of these data, 5-year survival rates were computed according to the product-limit method [15]. Relative survival was derived after allowance for the general lifetable of the canton [16].

RESULTS

Table 2 gives the distribution of the 645 registered cases of STS according to histological type, sex and calendar year, and the corresponding age-standardised (world population) incidence rates. The most common histotypes were fibrosarcoma (0.82/100 000 males, 0.86/100 000 females, 1990–1994), leiomyosarcoma (0.90/100 000 males, 1.28/100 000 females, 1990–1994), and Kaposi's sarcoma (3.10/100 000 males in 1990–1994). Overall age-standardised incidence rates for all STS increased from 2.68/100 000 males in 1974–1979 to 6.86/100 000 in 1990–1994, and from 3.61 to 4.27/100 000 females. However, after excluding Kaposi's sarcoma no consistent pattern of trends over time was observed, peak rates being registered in the late 1980s for both sexes, approximately 4.40/100 000, and with some levelling off thereafter.

The pattern of trends for all STS, except Kaposi's sarcoma, and Kaposi's sarcoma only in both sexes combined is also given in Figure 1 according to seven 3-year calendar periods. Incidence rates for all STS, as well as for Kaposi's sarcoma only rose up to the early 1990s, and stabilised thereafter. No consistent trend was observed for STS other than Kaposi's sarcoma.

The age distribution of Kaposi's and other STS in males and females over the period 1985–1994 is given in Figure 2. Kaposi's sarcoma, which is essentially related to the AIDS epidemic, peaked in males between 30 and 40 years of age. Other STS showed a peak in the first decade (essentially due to childhood rhabdomyosarcoma), and then rose again from the age of 30 years onwards in both sexes, to reach a peak in the sixth and seventh decades.

Table 3 gives the distribution of the 645 registered cases according to histological type and site. Overall, the most frequent sites were soft tissues, unspecified (37.1%), skin (20.8%), (retro)peritoneum (6.7%), uterus (6.4%, mainly leiomyosarcoma), intestines (4.3%), and stomach (3.3%).

Overall 5-year relative survival after STS was 45% (41% in males, 50% in females). Excluding Kaposi's sarcoma, overall survival was 51% (52% in males, 50% in females). With reference to morphological groups, the highest survival rates were observed for patients with fibrosarcoma and liposarcoma (67%); rates were intermediate for stromal sarcoma (53%), synovial sarcoma (52%), rhabdomyosarcoma (45%), leiomyosarcoma (39%), and blood vessel sarcoma (excluding Kaposi's sarcoma) (41%); the lowest rates were for sarcomas not otherwise specified (37%), meningiosarcoma (29%) and

Table 2. Age-standardized incidence rates (world population; rates per 100 000) of soft tissue sarcomas by sex, morphological group and calendar period, Vaud, Switzerland, 1974–1994

	Males					Females					Males and females				
Morphological type	1974–1979	1980–1984	1985–1989	1990–1994	(*n*) (Total)	1974–1979	1980–1984	1985–1989	1990–1994	(*n*) (Total)	1974–1979	1980–1984	1985–1989	1990–1994	(*n*) (Total)
Sarcoma NOS	0.21	0.21	0.34	0.19	(16)	0.23	0.23	0.59	0.63	(33)	0.21	0.22	0.47	0.42	(49)
Fibrosarcoma	0.52	0.84	1.21	0.82	(61)	0.48	0.55	0.77	0.86	(63)	0.50	0.68	0.98	0.84	(124)
Liposarcoma	0.31	0.40	0.52	0.51	(32)	0.29	0.20	0.23	0.16	(23)	0.30	0.30	0.37	0.32	(55)
Leiomyosarcoma	0.51	0.64	0.39	0.90	(46)	1.31	1.24	1.49	1.28	(118)	0.93	0.96	0.96	1.10	(164)
Blood vessel sarcoma	0.17	0.15	0.26	0.15	(14)	0.31	0.34	0.44	0.24	(26)	0.24	0.23	0.36	0.19	(40)
Rhabdomyosarcoma	0.67	0.30	0.52	0.36	(23)	0.21	0.38	0.02	0.09	(12)	0.42	0.34	0.26	0.23	(35)
Stromal sarcoma	–	–	–	–		0.22	0.07	0.14	0.19	(13)	0.12	0.04	0.08	0.10	(13)
Kaposi's sarcoma	–	0.42	1.94	3.10	(92)	–	–	0.12	0.40	(9)	–	0.21	1.03	1.74	(101)
Synovial sarcoma	–	0.06	0.16	0.15	(5)	0.05	0.13	0.33	0.18	(10)	0.02	0.09	0.25	0.17	(15)
Meningiosarcoma	0.08	–	0.06	0.04	(4)	–	0.10	–	0.04	(3)	0.03	0.05	0.03	0.04	(7)
Mesenchymoma	0.04	0.07	–	0.03	(3)	–	–	–	–	–	0.02	0.03	–	0.01	(3)
Other and NOS	0.18	0.17	0.94	0.61	(21)	0.52	0.29	0.36	0.19	(18)	0.35	0.23	0.67	0.38	(39)
Total	2.68	3.25	6.33	6.86		3.61	3.53	4.50	4.27		3.13	3.37	5.45	5.55	
(*n*)	(52)	(51)	(95)	(119)	(317)	(83)	(63)	(89)	(93)	(328)	(135)	(114)	(184)	(212)	(645)
Total minus Kaposi	2.68	2.83	4.39	3.76		3.61	3.53	4.38	3.87		3.13	3.16	4.42	3.81	
(*n*)	(52)	(45)	(64)	(64)	(225)	(83)	(63)	(87)	(86)	(319)	(135)	(108)	(151)	(150)	(544)

NOS, not otherwise specified.

for Kaposi's sarcoma (17%). Survival curves are given in Figure 3 for Kaposi's sarcomas and STS other than Kaposi's sarcomas for both sexes combined. Ten-year relative survival was 13% for Kaposi's sarcoma and 48% for all other STS. Five-year relative survival for non-Kaposi's STS was 43% for cases diagnosed in 1974–1984 and 58% for cases diagnosed in 1985–1994.

DISCUSSION

The major value of the present study is in providing a comprehensive overview of general aspects of descriptive epidemiology of STS in a European population, covered by a cancer registration system which has maintained a uniform structure over the 21-year period considered. Incidence rates in this Swiss population were relatively high on a European scale [17], and somewhat higher than those reported for North West England [7]. However, after allowance was made for different standard populations, the rates were comparable with those of the SEER Programme [1]. After excluding Kaposi's sarcoma, which is essentially related to the AIDS epidemic [9, 18], the distribution of registered cases by site and histological type was also broadly consistent with those of the caucasian American

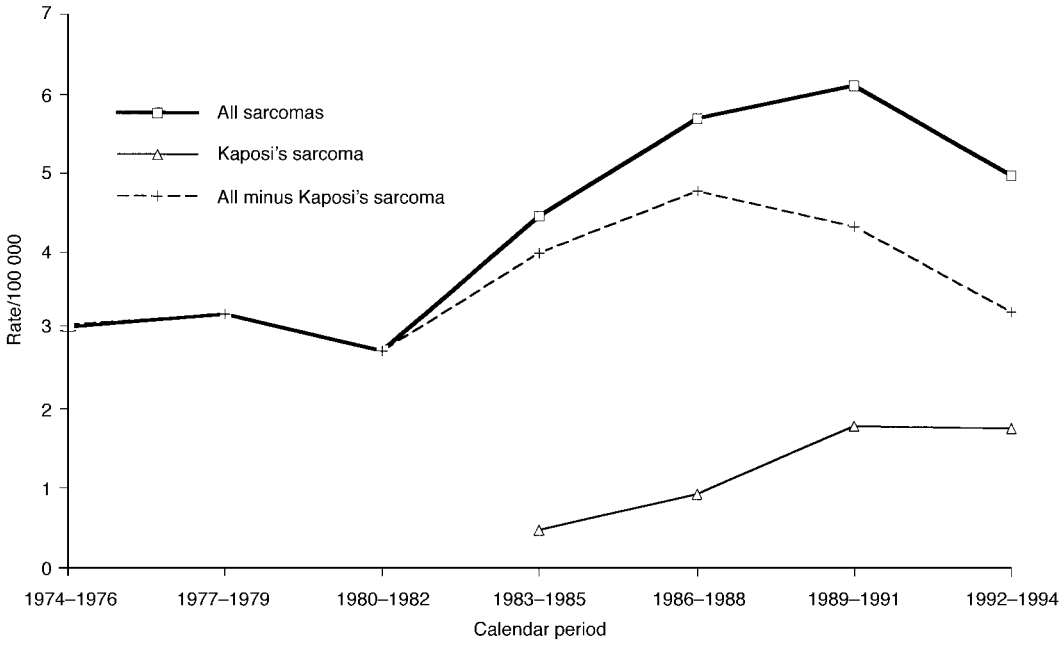

Figure 1. Trends in overall age-standardised (world population) incidence rates (per 100 000) of all soft tissue sarcomas, Kaposi's sarcomas and sarcomas other than Kaposi's in both sexes combined. Vaud, Switzerland, 1974–1994.

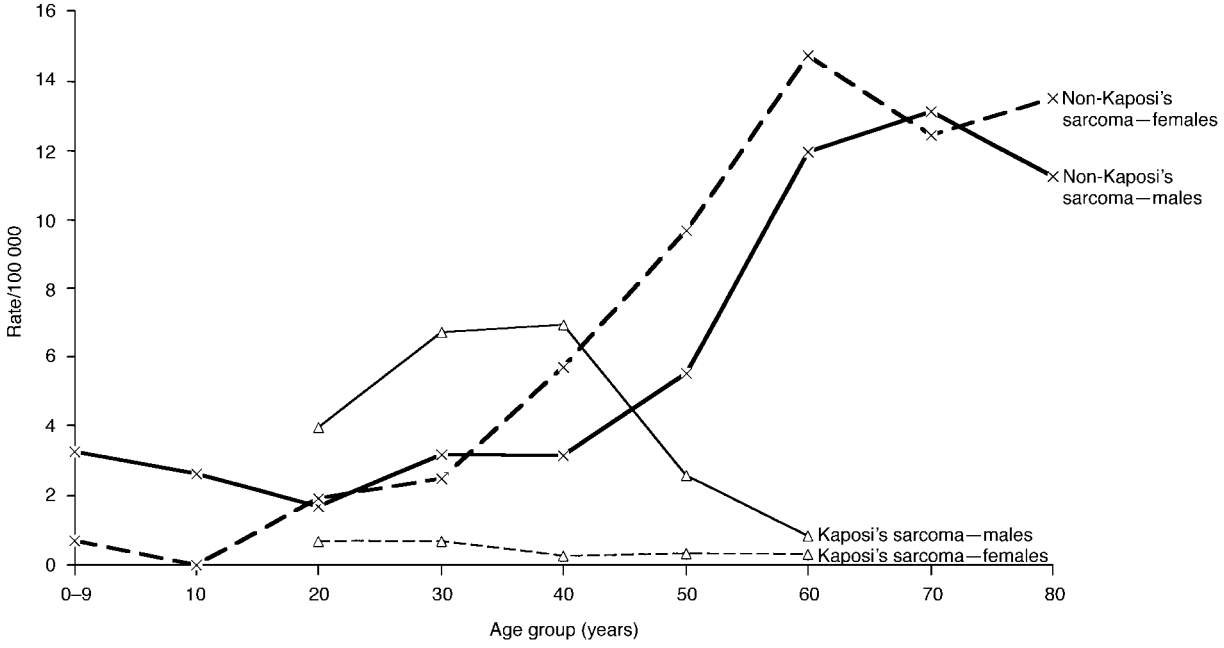

Figure 2. Age-specific incidence rates per 100 000 for Kaposi's sarcomas and soft tissue sarcomas other than Kaposi's, by gender. Vaud, Switzerland, 1985–1994.

Table 3. Site distribution of morphological groups of soft tissue sarcomas, Vaud, Switzerland, 1974–1994

Morphological group	Number of cases (%) by site							
	Stomach	Intestines	(Retro)peritoneum	Soft tissue	Skin	Uterus (corpus)	Other sites	Total
Fibrosarcoma	1 (0.8)	–	5 (4.0)	58 (46.8)	42 (33.9)	–	18 (14.5)	124 (100.0)
Liposarcoma	–	1 (1.8)	17 (30.9)	33 (60.0)	–	–	4 (7.3)	55 (100.0)
Leiomyosarcoma	19 (11.6)	22 (13.4)	17 (10.4)	45 (27.4)	6 (3.7)	25 (15.2)	30 (18.3)	164 (100.0)
Blood vessel sarcoma	–	2 (5.0)	–	17 (42.5)	3 (7.5)	–	18 (45.0)	40 (100.0)
Rhabdomyosarcoma	–	–	2 (5.7)	12 (34.3)	1 (2.9)	1 (2.9)	19 (54.3)	35 (100.0)
Kaposi's sarcoma	1 (1.0)	3 (3.0)	–	6 (5.9)	80 (79.2)	–	11 (10.9)	101 (100.0)
Other morphologies	–	–	2 (1.6)	68 (54.0)	2 (1.6)	15 (11.9)	39 (30.9)	126 (100.0)
Total, all morphologies	21 (3.3)	28 (4.3)	43 (6.7)	239 (37.1)	134 (20.8)	41 (6.4)	139 (21.5)	645 (100.0)
Rate per 100 000*	0.13	0.16	0.24	1.68	0.95	0.25	0.95	4.37

*All age-standardised rate on the world population.

population [2, 3, 10, 19], and of a smaller study from Iceland [6], although classification may be variably influenced by changes in diagnostic techniques, including immunochemistry and molecular pathology [1, 2, 10]. The major strength of this paper is the possibility of providing information on various histotypes and anatomical sites, including relative survival by histotype, together with the relatively small number of imprecisely classified or miscellaneous tumours.

Survival appeared to improve over more recent calendar periods, and was also comparable with most available information [10–12], that is approximately 55% (5-year relative survival) for all STS other than Kaposi's, and 20% for Kaposi's sarcoma. This includes essentially AIDS-related Kaposi's sarcoma, since this was not an endemic area for this neoplasm [18]. These data also confirm that liposarcomas and fibrosarcomas have more favourable survival rates than other histological types.

An important indication emerging from this dataset is that, after exclusion of Kaposi's sarcoma, STS incidence has not appreciably nor systematically changed between the 1970s and the 1990s in this European population. This confirms the observation made in Denmark until 1982 [4], as well as in the SEER dataset until 1987 [1]. This also provides strong evidence that, in populations with reliable and constant registration schemes over time, there has been no major new risk factor for STS of such relevance to modify materially overall incidence rates, apart from the AIDS epidemic for Kaposi's sarcoma. Among these potential risk factors are phenoxyherbicides and chlorophenols, whose consumption has substantially increased in several countries between 1950 and 1970 [4]. The potential role of these chemicals in STS risk has been widely debated [2, 20–22], but appears now unlikely to have substantially influenced STS incidence on a general population level.

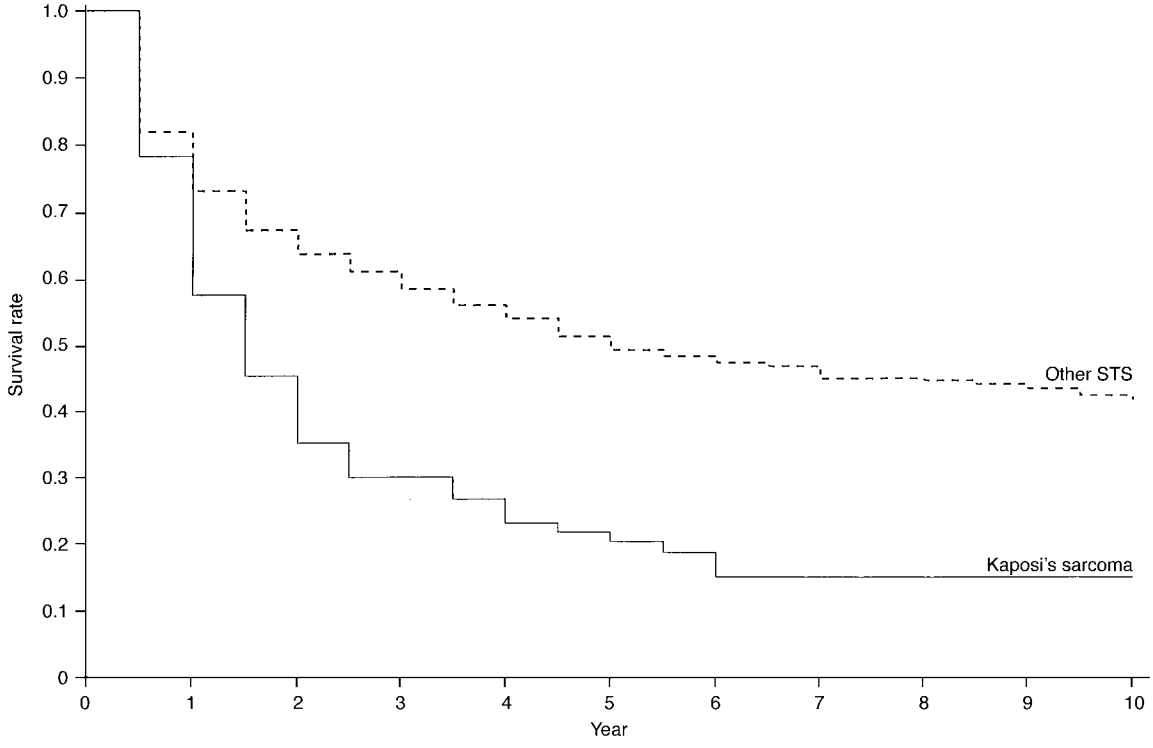

Figure 3. 10-year overall survival curves for Kaposi's sarcoma and soft tissue sarcomas other than Kaposi's. Vaud, Switzerland, 1974–1994.

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