Chapter 11
Sarcomas

Lynn A. Gloeckler Ries, Kevin C. Ward, and John L. Young, Jr.

INTRODUCTION

Sarcomas are tumors of diverse cell types which are mostly of mesodermal origin. They primarily arise in the soft tissues of the body including the retroperitoneum and peritoneum, pleura, heart, mediastinum and spleen but can also arise in the structural cells or parenchyma of specialized organs such as the stomach and kidney. The classification of tumors according to both the anatomic site in which they arose and the morphology of the tumor itself using the International Classification of Disease for Oncology (1) allows these tumors to be analyzed together as a group. Consequently, authors of other chapters in this monograph may have elected to exclude the sarcomas from their analyses knowing that they would be included here.

MATERIALS AND METHODS

The NCI contracts with medically-oriented, nonprofit institutions located in specific geographic areas to obtain data on all cancers diagnosed in residents of the SEER geographic areas. SEER collects data on all invasive and in situ cancers except basal cell and squamous cell carcinomas of the skin (of non-genital anatomic sites) and in situ carcinomas of the uterine cervix. SEER actively follows all previously diagnosed patients on an annual basis to obtain vital status allowing the calculation of observed and relative survival rates.

This analysis is based on data from 12 geographic areas which collectively cover about 14% of the total US population. The areas are the States of Connecticut, Iowa, New Mexico, Utah, and Hawaii; the metropolitan areas of Detroit, Michigan; Atlanta, Georgia; San Francisco, San Jose, and Los Angeles, California; Seattle, Washington; and 10 counties in rural Georgia. Los Angeles contributed data for diagnosis years 1992 to 2001, all other areas for 1988-2001.

Cases diagnosed in children and adolescents aged 0-19 have been excluded. Some patients have more than one diagnosis of cancer, but only the first diagnosis of cancer has been included. Death certificate only cases, autopsy only cases, and all other cases with no survival time have been excluded. Further, cases with no microscopic confirmation have been excluded. Finally, sarcomas arising in bone (osteosarcomas) have also been excluded from this analysis but are included in the bone chapter (2). Table 11.1 shows the numbers of cases excluded by category.

Survival analysis is based on relative survival rates calculated by the life-table (actuarial) method. Relative survival, defined as observed survival in the cohort divided by expected survival in the cohort, adjusts for the expected mortality that the cohort would experience from other causes of death. Expected survival is based on unabridged life tables for the United States in 1990. Although the American Joint Committee on Cancer's Staging Manual

Table 11.1: Sarcomas: Number of Cases and Exclusions by Reason, 12 SEER Areas, 1988-2001

Number selected/ remaining	Number excluded	Reason for exclusion/selection
41,408	0	Select 1988-2001 diagnosis (Los Angeles for 1992-2001 only)
37,314	4,094	Select first primary only
36,979	335	Exclude death certificate only or at autopsy
36,425	554	Exclude unknown race
36,337	88	Active follow-up and exclude alive with no survival time
33,820	2,517	Exclude children (000-019)
33,820	0	Exclude in situ cancers
30,183	3,637	Exclude no or unknown microscopic confirmation
28,758	1,425	Exclude cancer of the bone (C40.0-C41.9)

Table 11.2: Sarcomas: Number and Distributions by Sex, Race, SEER Summary Stage 1977 and Histology Group, Ages 20+, 12 SEER Areas, 1988-2001

	Sex			Race		Stage Percent			
Histology Group	Total	Male	Female	White	Black	Localized	Regional	Distant	Unstaged
Total	28,758	18,226	10,532	23,195	3,588	37.5	12.6	10.5	39.4
Perivascular sarcomas	174	95	79	130	31	28.2	28.2	28.2	15.5
Liposarcomas	2,368	1,413	955	1,986	190	65.9	21.3	5.7	7.1
Dermatofibrosarcomas	2,142	996	1,146	1,612	366	84.8	5.2	0.5	9.5
Other fibrosarcomas	511	250	261	391	71	57.1	23.9	9.2	9.8
Fibrohistiocytic sarcoma	3,063	1,823	1,240	2,626	218	61.2	21.1	9.9	7.8
Leiomyosarcomas	4,486	1,625	2,861	3,516	544	51.0	18.6	21.9	8.5
Rhabdosarcomas	299	178	121	223	49	27.8	28.8	29.8	13.7
Kaposi sarcoma	9,513	9,208	305	7,849	1,374	0.0	0.0	0.0	100.0
Vascular sarcomas excluding Kaposi	614	332	282	513	44	41.5	18.9	24.1	15.5
Chondro-oseous sarcomas	93	42	51	76	11	45.2	32.3	15.1	7.5
Sarcomas of uncertain differentiation	5,495	2,264	3,231	4,273	690	45.9	20.3	22.5	11.3

does contain a staging scheme for sarcomas, the scheme only applies to those sarcomas arising in soft tissues. Since this analysis is based on all sarcomas including those arising in any anatomic site except bone, the staging definitions utilized in this chapter are those of the 1977 Summary Staging Guide (3) whose staging categories are generally equivalent across the spectrum of anatomic sites. For simplicity, all categories of regional disease in the summary staging scheme have been added together into a single group. Finally, the sarcomas have been categorized into 11 subgroups roughly following the recommended classification of the World Health Organization (4) as shown below based on ICD-O-3 codes:

Perivascular Sarcomas 8680-8713
Liposarcomas 8850-8858
Dermatofibrosarcomas 8832-8833
Other fibrosarcomas 8810-8811,8813-8814,8825
Fibrohistiocytic sarcoma 8830
Leiomyosarcomas 8890-8891,8894,8896
Rhabdosarcomas 8900-8902,8910,8912,8920
Kaposi sarcoma 9140
Vascular sarcomas excluding Kaposi 9120,9133,9161
Chondro-oseous sarcomas 9180-9185,9192-9193,9221, 9230,9240,9243,9250
Sarcomas of uncertain differentiation 8800-8806,8823,8930-8936,8963,8990-8991,9040-9044,9231,9260-9261, 9364,9370-9371,9560-9561,9571,9580-9581.

RESULTS

Table 11.2 shows the distribution by sex, race, and summary stage for eleven categories of sarcomas. The largest category by far is Kaposi sarcoma (33%) with the smallest being the chondro-oseous (extraosseous) sarcomas (0.3%). Sarcomas occur more commonly among males, especially Kaposi of which only 3% of the cases occurred among females. Since there was no specific summary staging scheme for Kaposi, 100% of these cases were classified as "unstaged." Almost 30% of perivascular sarcomas and rhabdosarcomas were already staged as distant at the time of diagnosis.

Table 11.3 presents 5-year relative survival rates by anatomic site, histology and sex. Patients with sarcomas arising in soft tissues had much better survival than did patients whose tumors arose in other non-parenchymous sites (pleura, mediastinum, heart, retroperitoneum, peritoneum, and spleen) or in other organs, 68% vs. 43% and 44%, respectively. For Kaposi sarcoma, survival for those tumors arising in the soft tissue vs. other sites (primarily skin) was more than double – 53% vs. 25%. The best survival, regardless of site was among patients with dermatofibrosarcoma, 99.9%.

The 1-, 3-, 5- and 10-year relative survival rates for each subgroup are shown for males and for females in Table 11.4. The relative survival rates for dermatofibrosarcomas were almost 100% across all years being 98% at 10 years for males and 99.9% for females.

The poorest 10-year relative survival rate among males was experienced by patients with Kaposi sarcoma, 18%, followed closely by patients with other vascular sarcomas, 24%. Among females, the poorest relative survival at both 5 and 10 years was among those with a diagnosis of rhabdosarcoma.

A comparison of 1-,3-,5-, and 10-year relative survival rates for whites and for blacks is shown in Table 11.5. Overall, whites had higher survival rates at each interval compared to blacks, but the advantage diminished with time since diagnosis with the advantage being small at 10 years, 44% for whites compared to 41% for blacks. For the category of "other fibrosarcomas", blacks had much

Table 11.3: Sarcomas: 5-Year Relative Survival Rates (5) by Histology Group, Site, and Sex, Ages 20 +, 12 SEER Areas, 1988-2001

	Site											
	Total			Soft Tissue			Other Non-parenchymous			All Other		
Histology Group	All	Male	Female	All	Male	Female	All	Male	Female	All	Male	Female
Total	50.3	42.6	64.0	68.0	68.1	67.9	42.6	39.8	45.3	43.9	34.7	64.4
Perivascular sarcomas	63.3	62.0	64.9	~	~	~	69.6	~	~	61.8	60.1	63.5
Liposarcomas	82.8	82.4	83.1	85.9	84.8	87.1	65.1	60.0	70.2	94.4	97.0	81.9
Dermatofibrosarcomas	99.9	99.6	100.0	98.4	96.6	99.3	!	!	!	99.9	99.7	100.0
Other fibrosarcomas	72.4	68.5	75.5	79.8	79.0	80.3	38.6	~	~	61.1	40.4	70.7
Fibrohistiocytic sarcoma	67.0	67.9	65.7	67.6	67.6	67.6	25.4	26.4	24.2	77.7	79.5	72.9
Leiomyosarcomas	51.9	55.7	49.9	62.0	67.4	57.0	36.4	32.7	38.6	50.3	52.8	49.4
Rhabdosarcomas	35.0	35.9	33.9	40.4	40.1	39.7	~	~	~	31.0	32.6	29.4
Kaposi sarcoma	24.7	23.9	54.9	52.6	45.9	~	~	~	!	24.5	23.8	53.4
Vascular sarcomas excluding Kaposi	36.3	32.1	40.9	37.9	42.0	33.4	12.9	12.3	~	40.0	30.9	48.6
Chondro-oseous sarcomas	54.7	46.8	59.4	53.6	~	54.4	~	~	~	60.8	~	~
Sarcomas of uncertain differentiation	55.6	49.1	59.9	57.4	56.0	58.9	35.4	34.0	36.0	57.8	43.9	63.0

~ Statistic not displayed due to less than 25 cases.
! Not enough intervals to produce rate

Table 11.4: Sarcomas: 1-, 3- ,5- and 10-Year (Yr) Relative Survival Rates (%) by Histology Group and Sex, Ages 20+, 12 SEER Areas, 1988-2001

	Relative Survival Rate (%)											
	Total				Male				Female			
Histology Group	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr	1-Yr	3-Yr	5-Yr	10-Yr
Total	77.7	56.8	50.3	43.9	74.8	49.8	42.6	35.7	82.8	69.2	64.0	58.6
Perivascular sarcomas	90.2	75.1	63.3	47.5	88.5	71.1	62.0	38.4	92.2	80.0	64.9	58.8
Liposarcomas	93.4	85.8	82.8	74.4	93.9	85.8	82.4	76.4	92.8	85.9	83.1	71.0
Dermatofibrosarcomas	100.0	99.9	99.9	99.3	100.0	99.6	99.6	98.2	100.0	100.0	100.0	99.9
Other fibrosarcomas	87.2	77.8	72.4	65.4	86.3	76.8	68.5	63.1	88.1	78.6	75.5	66.5
Fibrohistiocytic sarcoma	85.0	71.2	67.0	64.0	85.4	72.5	67.9	63.5	84.3	69.4	65.7	63.2
Leiomyosarcomas	80.6	60.8	51.9	43.1	81.9	63.0	55.7	47.5	79.9	59.5	49.9	40.8
Rhabdosarcomas	65.6	42.6	35.0	30.5	70.4	43.9	35.9	32.3	58.5	40.5	33.9	27.2
Kaposi sarcoma	66.9	32.9	24.7	18.8	66.4	31.9	23.9	18.2	80.9	65.4	54.9	47.2
Vascular sarcomas excluding Kaposi	60.2	40.6	36.3	29.5	55.9	35.8	32.1	23.7	65.3	46.2	40.9	35.5
Chondro-osseous sarcomas	83.3	65.7	54.7	48.0	78.1	62.3	46.8	44.0	87.5	68.1	59.4	50.9
Sarcomas of uncertain differentiation	76.0	60.4	55.6	50.5	73.8	55.2	49.1	40.7	77.6	63.8	59.9	56.0

better survival than did whites with the 10-year relative survival rate being 79% for blacks compared to only 61% for whites. For rhabdosarcomas, the opposite was true with whites having much higher survival at each interval with the 10-year rate being 35% for whites compared to only 19% for blacks. The 10-year rate for patients with Kaposi sarcoma was almost equal among the two groups – 19% for whites and 18% for blacks.

Table 11.6 presents 5- and 10-year survival rates by summary stage. Patients with distant disease had uniformly poor survival (11% overall) at 10-years while survival for patients diagnosed at a localized stage was 78%. Among patients with localized disease, those with rhabdosarcoma had the poorest survival with rates of 59% at 5 years and 53% at 10 years.

DISCUSSION

In order to have sufficient numbers of cases for analyses the classification of sarcomas suggested by the World Health Organization (4) was utilized. Even so, several of the categories contained very few cases. Whenever possible, major subgroups were examined separately, namely, dermatofibrosarcomas from the fibrosarcoma group and Kaposi sarcoma from the vascular sarcoma group. In other categories, some interesting differences may have been obscured by the grouping of categories. For example, among the rhabdosarcomas, there were enough cases of alveolar rhabdosarcoma to examine separately.

Mack (5) has described in detail the heterogeneity of sarcomas and the various ways of classifying them over

Table 11.5: Sarcomas: 1-, 3- ,5- and 10-Year Relative Survival Rates (%) by Histology Group and Race, Ages 20+, 12 SEER Areas, 1988-2001

Histology Group	Relative Survival Rate (%)							
	White				Black			
	1-Year	3-Year	5-Year	10-Year	1-Year	3-Year	5-Year	10-Year
Total	78.5	57.1	50.4	43.9	71.5	51.4	46.4	40.8
Perivascular Sarcomas	88.9	74.7	62.8	47.2	94.3	75.0	66.6	49.0
Liposarcomas	93.1	85.1	82.5	73.4	94.8	88.2	83.3	79.7
Dermatofibrosarcomas	100.0	99.8	99.8	99.8	100.0	100.0	100.0	97.7
Other fibrosarcomas	85.5	75.4	69.4	61.3	98.3	90.9	87.2	79.4
Fibrohistiocytic sarcoma	85.6	73.1	69.0	65.6	78.5	55.1	53.7	52.4
Leiomyosarcomas	81.8	62.0	53.2	44.4	73.2	52.1	45.6	37.9
Rhabdosarcomas	68.1	46.1	39.1	35.0	51.5	21.4	21.4	18.6
Kaposi sarcoma	68.5	33.2	24.6	18.9	58.5	31.2	24.9	17.9
Vascular sarcomas excluding Kaposi	61.0	42.2	37.4	29.0	58.5	43.3	39.5	37.0
Chondro-osseous sarcomas	84.6	66.8	56.5	49.4	~	~	~	~
Sarcomas of uncertain differentiation	76.5	61.5	56.7	51.6	70.3	51.5	47.4	41.3

~ Statistic not displayed due to less than 25 cases.

Table 11.6: Sarcomas: 5- & 10-Year (Yr) Relative Survival Rates (%) by Histology Group and SEER Summary Stage, 12 SEER Areas, 1988-2001

Histology Group	Stage									
	Total		Local		Regional		Distant		Unstaged	
	Relative Survival		Relative Survival		Relative Survival		Relative Survival		Relative Survival	
	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr	5-Yr	10-Yr
	%	%	%	%	%	%	%	%	%	%
Total	50.3	43.9	83.1	78.1	54.0	46.1	16.2	11.1	28.8	22.2
Perivascular Sarcomas	63.3	47.5	82.1	59.3	82.6	73.5	43.0	21.0	37.3	21.0
Liposarcomas	82.8	74.4	90.9	81.5	74.4	65.4	30.8	15.4	70.1	66.9
Dermatofibrosarcomas	99.9	99.3	99.9	99.3	100.0	100.0	~	~	99.4	97.3
Other fibrosarcomas	72.4	65.4	88.3	83.0	54.3	44.4	24.2	19.5	67.3	56.0
Fibrohistiocytic sarcoma	67.0	64.0	81.4	79.1	55.2	49.8	11.8	9.3	53.8	43.7
Leiomyosarcomas	51.9	43.1	71.8	61.4	44.4	35.3	13.6	8.9	45.4	32.4
Rhabdosarcomas	35.0	30.5	58.6	52.6	40.2	34.5	6.3	6.3	34.8	27.1
Kaposi sarcoma	24.7	18.8	!	!	!	!	!	!	24.7	18.8
Vascular sarcomas excluding Kaposi	36.3	29.5	57.6	52.2	31.7	24.2	12.9	7.3	25.4	15.5
Chondro-osseous sarcomas	54.7	48.0	62.3	53.5	65.5	57.1	~	~	~	~
Sarcomas of uncertain differentiation	55.6	50.5	80.3	76.0	48.5	39.5	17.1	13.4	41.0	31.8

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! Not enough intervals to produce rate

time. He has also described the genetic determinants as well as the environmental agents believed to play a role in the etiology of sarcomas. It is not unreasonable that these factors might influence survival as well, particularly among patients with certain chromosomal abnormalities.

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