HHS Public Access

Author manuscript

Ann Thorac Surg. Author manuscript; available in PMC 2019 August 29.

Published in final edited form as:

Ann Thorac Surg. 2018 August ; 106(2): 333–339. doi:10.1016/j.athoracsur.2018.03.068.

Treatment Approaches and Outcomes for Primary Mediastinal Sarcoma: Analysis of 976 Patients

Kathryn E. Engelhardt, MD, **Malcolm M. DeCamp, MD**, **Anthony D. Yang, MD, MS**, **Karl Y. Bilimoria, MD, MS**, **David D. Odell, MD, MMSc**

Surgical Outcomes and Quality Improvement Center, Northwestern University, Chicago, Illinois; Department of Surgery, Medical University of South Carolina, Charleston, South Carolina; Division of Thoracic Surgery, Northwestern University, Chicago, Illinois; Division of Surgical Oncology, Northwestern University, Chicago, Illinois; Center for Healthcare Studies, Northwestern University, Chicago, Illinois; and Robert H. Lurie Comprehensive Cancer Center, Northwestern University, Chicago, Illinois

Abstract

Background—Primary mediastinal sarcomas are rare and deadly. Our objective was to describe the clinicopathological features, treatment strategies, and overall survival outcomes for a contemporary cohort of patients diagnosed with primary mediastinal sarcoma in the United States.

Methods—We queried the National Cancer Database for cases of mediastinal sarcoma diagnosed from 2004 to 2012. Five-year overall survival (OS) was examined using the Kaplan-Meier method. Differences in OS were assessed using log-rank analysis and Cox proportional hazards regression.

Results—The mean age of diagnosis was 53 years (range, 0 to 90) with a male predominance (59.2%). The most common histological subtype was hemangiosarcoma (27.1%). Fewer than half of patients underwent surgery (48.9%), and 19.7% of patients had no treatment. For all patients, OS was 14.8%. The best unadjusted OS was seen in patients treated with surgery and radiation (40.1%); untreated patients had the worst unadjusted OS (4.2%). Of those who underwent surgery (n = 477, 48.9%), OS was significantly better for those who achieved an R0 resection (30.1% versus 18.9%; $p = 0.002$). In multivariable analysis, surgery combined with radiation therapy was again associated with the best survival (HR, 0.24; 95% CI, 0.16 to 0.36). Other factors associated with improved OS included younger age, fewer comorbidities, and leiomyosarcoma histology. Worse OS was associated with poorly differentiated or undifferentiated grade, metastases, treatment in the New England region, and having Medicaid or no insurance. Sex and tumor size had no effect on OS.

Conclusions—The 5-year OS for primary mediastinal sarcoma is poor. Surgical resection can be successful and should be considered whenever possible.

Soft tissue sarcomas are malignant tumors of mesenchymal origin most frequently found in the extremities or within the abdomen. Primary mediastinal sarcomas, by contrast, are rare,

Address correspondence to Dr Odell, 633 N Saint Clair St, 20th Flr, Chicago, IL 60611; dodell@nm.org.

The Supplemental Table can be viewed in the online version of this article [https://doi.org/10.1016/j.athoracsur.2018.03.068] on http://www.annalsthoracicsurgery.org.

representing ~1% of all soft tissue sarcomas [1]. The variety of tissue types within the mediastinum results in great heterogeneity of potential tumors in this location [2, 3]. Because the behavior and prognosis of sarcoma can be heavily dependent on the underlying histology, clinicians have struggled to define the optimal treatment strategy for this disease. The resulting variability in treatment of mediastinal sarcomas has resulted in a failure to meaningfully improve survival rates with this disease.

The rarity and variety of mediastinal sarcoma has also limited comprehensive investigation and comparative assessment of treatment strategies and outcomes. The current literature is limited to single-institution studies and small case series of mediastinal sarcoma; or larger studies of sarcomas from any location, mediastinal tumors of any histology, and specific histologic subsets of sarcoma. Because mediastinal sarcoma is such a small subset of all sarcomas, however, the findings of these large studies may not be accurate for mediastinal tumors, which are unique in terms of their histologic subtype and anatomic association with major vessels and vital organs.

Although the utility of complete resection is clear from prior studies, the role of adjuvant therapy is still debated. Our objective was to describe the clinicopathologic features, treatment strategies, and overall survival outcomes for a contemporary, multiinstitution cohort of patients diagnosed with primary mediastinal sarcoma in the United States.

Patients and Methods

Data Source

Data were obtained from the National Cancer Database (NCDB), a joint program of the American College of Surgeons and American Cancer Society. The largest cancer registry in the world, the NCDB is estimated to capture approximately 70% of all new cancer diagnoses in the United States and Puerto Rico [4]. Data are collected by certified tumor registrars who undergo extensive training and are audited to ensure accuracy of the database. Hospital and patient identity are protected and not included in the participant use file (PUF). Data released in the PUF are in compliance with the privacy requirements of the Health Information Portability and Accountability Act. The institutional review board at Northwestern University determined that this study was exempt as it uses publicly available deidentified data.

Population

The NCDB was queried to identify all cases of mediastinal sarcoma diagnosed from January 1, 2004, to December 31, 2012 in the United States. We included patients of all ages. We included first-occurrence, primary malignant sarcoma as codified by the International Classification of Disease for Oncology, Third Edition (ICD-O-3) terms. Malignant tumors were identified by an ICD-O-3 Invasive Behavior code in conjunction with the histology code. Location was limited to heart and mediastinum (ICD-O-3 codes C38.0 to C38.8). Thymic tumors were excluded. Patients who were diagnosed at death were excluded (n = 15). Patients with missing data were included in the unadjusted survival analysis but were excluded from the multivariable analysis (n = 80).

Outcomes and Variables

We evaluated overall survival as our primary outcome measure. Overall survival (OS) was defined as the period from the date of diagnosis until death or last follow-up. Our primary predictors of interest were treatment modality (grouped into 6 categories: no treatment, radiation and/or chemotherapy, surgery only, surgery and radiation, surgery and chemotherapy, and surgery and chemoradiation) and, if surgery was performed, the extent of resection (dichotomized into R0 versus R1, R2, or unknown). Patients were coded as receiving radiation or chemotherapy regardless of the timing of these therapies in relation to surgery, if performed. Unfortunately, the NCDB PUF does not provide information on the surgical approach or indication for radiation therapy (adjuvant, prophylactic, for recurrent, etc). Patient, tumor, and hospital-level characteristics were assessed. These factors included tumor characteristics (size dichotomized into up to 10 cm or larger than 10 cm, histology, grade, and presence of metastases), patient-level factors (sex, age, race/ethnicity, modified Charlson-Deyo comorbidity score [5], median income, median education level, and insurance status), and hospital-level factors (facility location and academic status). Similar to previous literature, we included histology as a categorical variable in 6 groups: hemangiosarcoma, leiomyosarcoma, synovial sarcoma, malignant peripheral nerve sheath tumor, sarcoma not otherwise specified, and other. Race and ethnicity were grouped into 4 categories, as defined by the Commission on Cancer (CoC): non-Hispanic white, non-Hispanic African American, Hispanic, and other. Patients were grouped into quartiles for income and education derived from census data for their zip code, as defined by the CoC. Insurance status was dichotomized into 2 groups presumed to represent lower and higher socioeconomic status: Medicaid and uninsured versus all others [6]. A modified Charlson-Deyo comorbidity score was included as a categorical variable as defined by the CoC: 0, 1, and 2 or more. Facility characteristics were accounted for in the model with geographic location, urban versus rural location, and academic status. Academic hospitals were defined by the CoC as having 500 or more newly diagnosed cancer types per year and offering graduate medical education programs in greater than 4 disciplines.

Statistical Analyses

Survival was examined using the Kaplan-Meier method. Point estimates for 5-year OS were obtained from the Kaplan-Meier survival functions. Differences in survival were assessed using unadjusted log-rank analysis. Subsequently, we adjusted for the available covariates using Cox proportional hazards regression. We ran 2 sensitivity analyses of our regression model. In the first, we included distance traveled to treating facility to assess for a possible referral bias. Distance traveled was grouped into 3 categories, according to previously published categories important for cancer care (<12.5 miles, 12.5 to 50 miles, and >50 miles) [7]. In the second, we assessed for an interaction between grade and treatment strategy. In this model, we limited the analysis to those for whom grade was known and dichotomized grade into low (well and moderately differentiated) and high (poorly differentiated and undifferentiated) grade. We then limited the analysis to surgical patients only and dichotomized treatment into surgery only versus surgery plus any adjuvant therapy. We then created an interaction term and included it in our regression model. Analyses were performed using the STATA v14 (College Station, TX) and SAS v9.4 (Cary, NC) statistical

software packages. Two-sided p values of less than 0.05 were considered statistically significant.

Results

We identified 976 patients for analysis (Table 1). The mean age of diagnosis was 53 years (range, 0 to 90) with the majority of patients being male. The most common histological subtype was hemangiosarcoma. For those patients whose grade was known, the majority of tumors were poorly differentiated or undifferentiated. Fewer than half of patients underwent surgery, and 19.7% (n = 192) of patients had no treatment (Table 2). An R0 resection was accomplished in only 33.8% (n = 161) of patients undergoing surgical resection. Patients who had an R0 resection received radiation at the same rate as patients with positive margins (n = 45 of 161 [28%] versus n = 95 of 316 [30%]; $p = 0.794$). Additionally, the majority of patients who received radiation in addition to surgery received radiation postoperatively (Supplemental Table 1; n = 114 of 140, 81.4%).

Five-year OS was 14.8% for the entire cohort. The patients who received a combination of surgical resection and radiation therapy had the best 5-year survival; patients who had no treatment had the worst OS (Table 2; Fig 1). Of those who underwent surgery (n = 540, 48.4%), 5-year survival was significantly better for those who achieved an R0 resection compared with those who did not ($p = 0.002$; Fig 2). Patients who had an incomplete resection (R1 or R2) had a better 5-year survival rate when compared with those who had nonsurgical (chemotherapy and/or radiation therapy) or when compared with no therapy (both $p < 0.001$; Table 2).

Next, we built a Cox proportional hazards model to assess survival after adjusting for tumor-, patient-, and hospital-level characteristics (Table 3). In this model, the combination of surgery and radiation therapy was again associated with the best survival (HR, 0.24; 95% CI, 0.16 to 0.36). This strategy had a significantly better OS than treatment approaches that did not include surgical resection, such as no treatment, radiation alone, chemotherapy alone, or chemoradiation. There was no significant difference among the 4 surgery-based strategies. Besides treatment modality, factors associated with significantly better survival included younger age, fewer comorbidities, and leiomyosarcoma histology. Worse survival was associated with poorly or undifferentiated grade, metastases, treatment in the New England region, and having either Medicaid or no health insurance.

We then performed two sensitivity analyses of note. First, we assessed for an association between distance traveled and survival. We were particularly interested to see if including this term in our model would affect the significance of the urban/rural designation. In this model, the construct of distance traveled was not significantly associated with OS; additionally, including this construct in our model did not qualitatively change the results of our primary model. Second, we estimated a model with dichotomized grade and treatment modality, limited to surgical patients for whom grade was known. Although there were significantly more high-grade patients who received adjuvant therapy (χ^2 $p < 0.001$); there was no significant interaction between grade and treatment in our regression analysis.

Comment

Mediastinal sarcoma encompasses a range of histologic tumor types, all of which are rare cancers. The rarity of the diagnosis precludes randomized controlled trials to inform management decisions in these patients. Consequently, the literature on this disease is limited and the ideal treatment strategy is poorly defined. This retrospective cohort analysis of data from the NCDB again illustrates the poor prognosis associated with mediastinal sarcoma. In both unadjusted and adjusted analysis, the best survival was seen in those patients who underwent a combination of surgery and radiation.

To our knowledge, this is the largest nationwide series of patients with mediastinal sarcoma. We describe the clinicopathologic features, treatment strategies, and OS outcomes for 976 patients diagnosed in the United States from 2004 to 2012. Utilizing the NCDB has, for the first time, provided a patient cohort large enough to support a multivariable analysis of factors independently associated with survival in mediastinal sarcoma. As has been previously reported, there was a male predominance in our study sample [8, 9]. The most common histologic subtype in our analysis was hemangiosarcoma, consistent with prior reviews of uncommon primary mediastinal tumors [10]. Smaller studies were frequently dominated by another subtype less common in our nationwide sample. [1, 9, 11, 12]. A review article found that nerve sheath tumors represented the most common histology, but that review included both benign and malignant soft tissue tumors, whereas we limited our analysis to invasive tumors. Some studies have excluded malignant peripheral nerve sheath tumors as they are of neurogenic rather than mesenchymal origin [12]. However, Burt and colleagues included these tumors in their analysis of the largest single-center series in the literature [1] and the ICD-O-3 codes classify these tumors as sarcomas; as such, we included this histology in our analysis. In contrast to Burt and associates, neurogenic tumors were a minority of our overall patient sample and were not associated with survival in adjusted analysis. We were not able to determine the presenting clinical complaint or the precise intramediastinal location because of the limitations in the NCDB data set. Smaller studies, however, have found the primary presenting complaint to be pain (chest or back) and the most common location to be the posterior mediastinum [1, 12].

Data on the role of radiation and chemotherapy in the treatment of mediastinal sarcoma has previously been somewhat limited. The most common treatment modality we observed was radiation and/or chemotherapy without surgical resection. This is likely because mediastinal sarcomas are frequently large and abutting or invading vital structures when they are diagnosed, precluding resection [13]. In their analysis of patients treated at a major academic medical center specializing in cancer care, Burt and coworkers found that the most frequent treatment strategy was surgery combined with radiation therapy [1]. The higher rate of nonoperative therapy in our analysis is most likely due to a difference in treating facilities. As the NCDB includes both academic and community cancer programs and captures 70% of all cancer diagnoses in the United States, these data likely more accurately represent treatment trends nationally.

In our analysis, OS was best in those patients treated with a combination of surgery and radiation (40.1%). Additionally, we found that OS for those patients who had an incomplete

resection was superior to those patients treatment with nonsurgical modalities or no treatment at all, though this unadjusted comparison was likely subject to selection bias to some extent. Similarly, Burt and colleagues found that OS for patients who underwent complete resection was 49%, compared with 30.1% in our cohort [1]. This difference may be due to improved patient selection or improved surgical technique at a specialized center when compared with care at other centers. A population-based analysis in Canada also found that, although OS was poor, surgical resection resulted in the best OS [9]. Similarly, an article from Egypt using Survival, Epidemiology, and End Results data found that surgical treatment offered the best survival [14]. In a published report of 21 cases in a single province in China, the authors reported improved survival after complete resection with no additional benefit gained from chemotherapy or radiation [12]. Similarly, although there are dramatic differences in unadjusted OS between the 4 surgery-based treatment groups in our study (surgery alone, surgery and radiation, surgery and chemotherapy, or surgery and chemoradiation), after adjusting for patient and tumor characteristics no significant difference was seen among these 4 strategies. On the contrary, in a 2008 update to a metaanalysis to determine the utility of chemotherapy in localized, resectable soft-tissue sarcomas, the authors concluded that there is a slight survival advantage to combination doxorubicin and ifosfamide chemotherapy postoperatively [15]. This meta-analysis included sarcomas in any anatomic location, however, and did not provide discrete information on the subgroup of mediastinal tumors.

In addition to treatment strategy, we found several additional factors were independently associated with OS. While most previous studies have not found better OS with younger age [1, 9, 12], our study, similar to one previous [14], did find that increasing age was associated with worse OS. Despite the observed male predominance in the incidence of mediastinal sarcoma, sex does not appear to be correlated with survival [9, 12]. In our analysis, increased comorbidity score was associated with worse OS; previous studies have not used the Charlson Comorbidity Score but one study did find an association between poor performance status and worse OS [12]. Although prior studies of mediastinal sarcomas have not found an association between histologic subtype and OS, we found that leiomyosarcoma was associated with improved OS and that hemangiosarcoma had the worst OS. However, in a study of soft tissue sarcomas of any site, hemangiosarcoma had the worst OS of the relevant histologic subtypes [16]. Even though most previous studies have failed to show an association between grade and OS [9, 12], like our cohort, one previous study [14] showed improved survival in lower grade tumors. Whereas a single study showed that tumor size larger than 10 cm was a significant risk factor for metastatic recurrence risk, 2 other studies found, like ours, that there was no association between tumor size and OS [9, 12, 13]. The study with findings dissimilar to ours was not limited to mediastinal sarcoma, and, because the authors grouped mediastinal and abdominal wall tumors together, it is unclear what percentage of their patients are comparable to ours [13].

To our knowledge, this was the first study to examine the association between OS and treating facility characteristics or patient-specific social determinants of health. We found that patients treated in New England had worse OS when compared with all regions except the east south central and west north central regions. Though this finding results from a multivariable analysis adjusting for available tumor, patient, and facility characteristics, this

result should be interpreted with caution because of the small number of patients treated in this region during our study time period. Nevertheless, this finding should be explored further to determine whether this is a result of specialized centers in this geographic area or regional differences in epidemiology, disease state, or socioeconomic factors. Similarly, we found that patients who are uninsured or on public insurance for low-income families had slightly worse OS. This finding is consistent with the large body of literature on social determinants of health [17, 18].

Our study does have important limitations. First, data extracted from any database are subject to coding error. The NCDB data are collected by trained and audited abstractors, however, improving reliability. Additionally, the data definitions are standardized. Thus, the effect of these differences is likely minimal. Second, disease-specific survival and local or distant recurrence were not available in the NCDB for analysis, thus we limited our outcomes metric to OS. Third, there may be selection bias for which we cannot correct based on the limited comorbidity information and lack of performance status data provided in the NCDB. Additionally, we grouped all chemotherapy together and all radiation together, although there may be variation in treatment protocols. Nevertheless, this is the largest cohort of mediastinal sarcomas reported on in the literature. This cohort of patients with primary mediastinal sarcoma is heterogeneous and thus our results may not be equally generalizable to all patients with this diagnosis.

In summary, the 5-year OS for primary mediastinal sarcoma is poor, but surgical resection can be successful and should be considered whenever possible. Radiation may offer an important survival advantage but the contribution of chemotherapy remains unclear. Further research is needed to evaluate the effect of specialized centers and social determinants of health on oncologic outcomes, but our results suggest that these factors may play a role in mediastinal sarcoma, as they do in other oncologic conditions. We recommend that this rare and deadly condition be treated by multidisciplinary teams.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

Dr Odell receives support from the National Cancer Institute (award number K07CA216330), The AATS Graham Foundation Oz Lemole Scholarship, The American College of Surgeons Faculty Research Fellowship, and the Thoracic Surgery Foundation Research Scholarship. The National Cancer Data Base (NCDB) is a joint project of the Commission on Cancer (CoC) of the American College of Surgeons and the American Cancer Society. The CoC's NCDB and the hospitals participating in the CoC NCDB are the source of the deidentified data used herein; they have not verified and are not responsible for the statistical validity of the data analysis or the conclusions derived by the authors.

References

1. Burt M, Ihde JK, Hajdu SI, et al. Primary sarcomas of the mediastinum: results of therapy. J Thorac Cardiovasc Surg 1998;115:671–80. [PubMed: 9535456]
2. den Bakker MA, Marx A, Mukai K, Strobel P. Mesenchymal tumours of the mediastinum—part I. Virchows Arch 2015;467:487–500. [PubMed: 26358059]

3. den Bakker MA, Marx A, Mukai K, Ströbel P. Mesenchymal tumours of the mediastinum—part II. Virchows Arch 2015;467:501–17. [PubMed: 26358060]

4. Bilimoria KY, Stewart AK, Winchester DP, Ko CY. The National Cancer Data Base: a powerful initiative to improve cancer care in the United States. Ann Surg Oncol 2008;15: 683–90. [PubMed: 18183467]

5. Deyo RA, Cherkin DC, Ciol MA. Adapting a clinical comorbidity index for use with ICD-9-CM administrative databases. J Clin Epidemiol 1992;45:613–9. [PubMed: 1607900]

6. Jemal A, Fedewa SA. Lung cancer screening with low-dose computed tomography in the United States—2010 to 2015. JAMA Oncol 2017;3:1278–81. [PubMed: 28152136]

7. Vetterlein MW, Loppenberg B, Karabon P, et al. Impact of travel distance to the treatment facility on overall mortality in US patients with prostate cancer. Cancer 2017;123: 3241–52. [PubMed: 28472547]

8. Ortega P, Suster D, Falconieri G, et al. Liposarcomas of the posterior mediastinum: clinicopathologic study of 18 cases. Mod Pathol 2015;28:721–31. [PubMed: 25475695]

9. Paquette M, Truong PT, Hart J, et al. Primary sarcoma of the mediastinum: a report of 16 cases referred to the British Columbia Cancer Agency. J Thorac Oncol 2010;5: 898–906. [PubMed: 20521357]

10. Macchiarini P, Ostertag H. Uncommon primary mediastinal tumours. Lancet Oncol 2004;5:107–18. [PubMed: 14761815]

11. Zehani A, Ayadi-Kaddour A, Daghfous H, et al. Primary mediastinal sarcomas. Rev Mal Respir 2011;28:14–24. [PubMed: 21277470]

12. Luo DX, Huang MJ, Xiong B, et al. Primary mediastinal sarcoma: surgical outcomes of 21 cases. Interact Cardiovasc Thorac Surg 2013;17:982–6. [PubMed: 24027167]

13. Coindre JM, Terrier P, Bui NB, et al. Prognostic factors in adult patients with locally controlled soft tissue sarcoma. A study of 546 patients from the French Federation of Cancer Centers Sarcoma Group. J Clin Oncol 1996;14:869–77. [PubMed: 8622035]

14. Abdel-Rahman O An analysis of clinical characteristics and patient outcomes in primary mediastinal sarcomas. Expert Rev Anticancer Ther 2017;17:1071–6. [PubMed: 28889760]

15. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. Cancer 2008;113:573–81. [PubMed: 18521899]

16. Corey RM, Swett K, Ward WG. Epidemiology and survivorship of soft tissue sarcomas in adults: a National Cancer Database report. Cancer Med 2014;3:1404–15. [PubMed: 25044961]

17. Link BG, Phelan JC. Understanding sociodemographic differences in health—the role of fundamental social causes. Am J Public Health 1996;86:471–3. [PubMed: 8604773]

18. Monfared ED, Mohseni M, Amanpour F, Jarrahi AM, Joo MM, Heidarnia MA. Relationship of social determinants of health with the three-year survival rate of breast cancer. Asian Pac J Cancer Prev 2017;18:1121–6. [PubMed: 28547951]

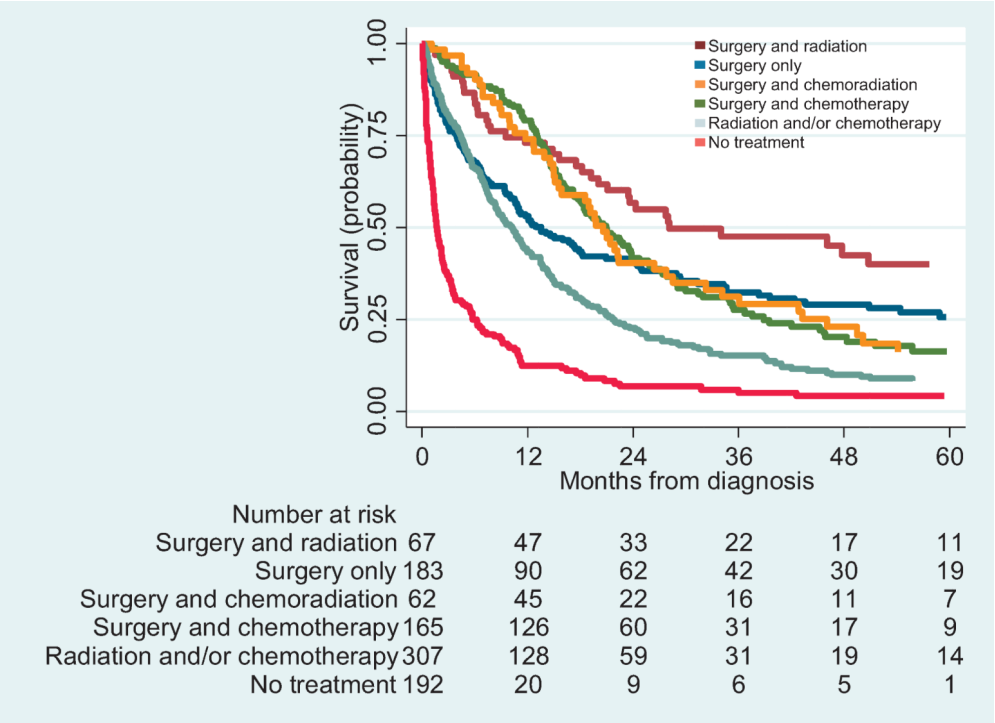

Fig 1.
Kaplan-Meier by treatment, $p < 0.0001$.

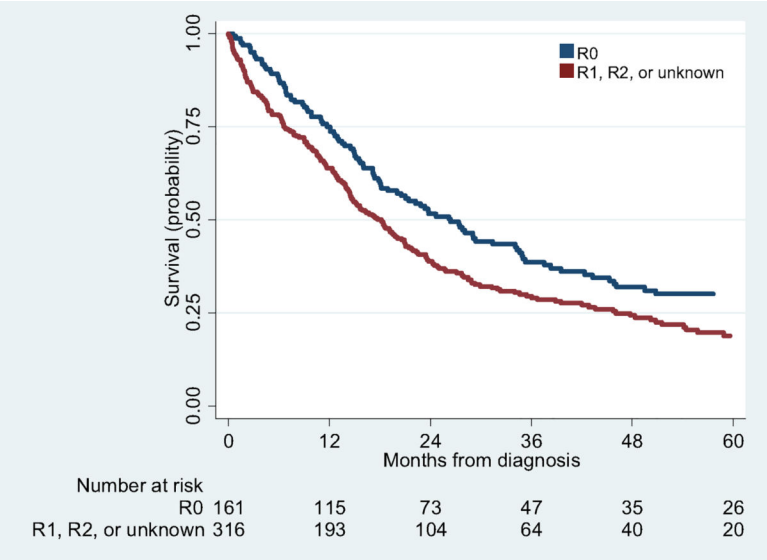

Fig 2.
Kaplan-Meier by R status, p = 0.0019.

Table 1.

Characteristics of Study Sample, n = 976

Variable	Number (n)	Percent (%)
Sex		
Male	578	59.2
Female	398	40.8
Age[a], years		
<20	52	5.3
20–39	219	22.4
40–59	309	31.7
60–79	301	30.8
80+	93	9.5
Race[a]		
Hispanic	79	8.1
Non-Hispanic white	719	73.4
Non-Hispanic black	125	12.8
Other	49	5.0
Charleson-Deyo score		
0	709	72.6
1	209	21.4
2+	58	5.9
Histology		
Hemangiosarcoma	264	27.1
Sarcoma, NOS	214	21.9
Leiomyosarcoma	101	10.4
Synovial sarcoma	101	10.4
Malignant peripheral nerve sheath tumor	51	5.2
Other	245	25.1
Grade		
Well-differentiated	37	3.8
Moderately differentiated	45	4.6
Poorly differentiated	257	26.3
Undifferentiated	169	17.3
Unknown	468	47.9
Size		
≤10 cm	490	50.2
>10 cm	206	21.1
Unknown	280	28.7
Medicaid or uninsured[a]	145	14.9
Income[a]		
<$38,000	166	17.0

Variable	Number (n)	Percent (%)
$38,000-$47,999	219	22.4
$48,000-$62,999	250	25.6
≥$63,000	315	32.3
Education quartile		
Lowest	179	18.3
Second	220	22.5
Third	296	30.3
Highest	257	26.3
Urban[a]	932	95.5
Academic	340	34.8
Facility location		
New England (CT, MA, ME, NH, RI, VT)	24	2.5
Middle Atlantic (NJ, NY, PA)	128	13.1
South Atlantic (DC, DE, FL, GA, MD, NC, SC, VA, WV)	132	13.5
East North Central (IL, IN, MI, OH, WI)	128	13.1
East South Central (AL, KY, MS, TN)	34	3.5
West North Central (IA, KS, MN, MO, ND, NE, SD)	66	6.8
West South Central (AR, LA, OK, TX)	66	6.8
Mountain (AZ, CO, ID, MT, NM, NV, UT, WY)	37	3.8
Pacific (AK, CA, HI, OR, WA)	88	9.0
Unknown	273	28.0

[a]Age was missing in 2 patients (0.2%), race was missing in 7 patients (0.7%), insurance status was missing in 34 patients (3.5%), income status was missing in 26 patients (2.7%), education was missing in 24 patients (2.5%), urban status missing in 36 patients (3.7%).

NOS = not otherwise specified.

Table 2.

Treatment and Overall Survival for Primary Mediastinal Sarcoma

Treatment (n = 976)	n	%	5-year OS, %
Surgery and radiation therapy	67	6.9	40.1
Surgery only	183	18.8	25.6
Surgery and chemoradiation	62	6.4	16.1
Surgery and chemotherapy	165	16.9	14.3
Radiation and/or chemotherapy	307	31.5	8.5
No treatment	192	19.7	4.2
Extent of resection (n = 477)			
R0	161	33.8	30.1
R1, R2, or unknown	316	66.2	18.9

OS = overall survival.

Table 3.

Cox Proportional Hazards Model

Variable	HR (95% CI)	*p* Value
Sex		
Male	Ref	-
Female	0.90 (0.77–1.06)	0.199
Age, years		
<20	Ref	-
20–39	1.39 (0.93–2.09)	0.112
40–59	3.29 (1.80–6.05)	<0.001
60–79	4.53 (2.48–8.29)	<0.001
80+	8.50 (4.44–16.3)	<0.001
Race		
Hispanic	0.83 (0.60–1.15)	0.276
Non-Hispanic white	Ref	-
Non-Hispanic black	1.20 (0.93–1.53)	0.160
Other	1.24 (0.86–1.78)	0.256
Charleson-Deyo score		
0	Ref	-
1	1.51 (1.24–1.84)	<0.001
2+	1.85 (1.35–2.54)	<0.001
Histology		
Hemangiosarcoma	Ref	-
Sarcoma, NOS	0.86 (0.68–1.08)	0.186
Leiomyosarcoma	0.48 (0.35–0.66)	<0.001
Synovial sarcoma	0.88 (0.67–1.16)	0.367
Malignant peripheral nerve sheath tumor	0.76 (0.51–1.13)	0.179
Other	0.59 (0.47–0.75)	<0.001
Tumor Size		
≤10 cm	Ref	-
>10 cm	1.16 (0.94–1.44)	0.164
Unknown	1.29 (1.07–1.56)	0.007
Grade		
Well-differentiated	Ref	-
Moderately differentiated	1.59 (0.81–3.1)	0.177
Poorly differentiated	2.58 (1.51–4.41)	0.001
Undifferentiated	3.24 (1.88–5.58)	<0.001
Unknown	2.63 (1.55–4.44)	<0.001
Metastases	1.60 (1.30–1.97)	<0.001
Treatment		
Surgery and radiation	0.24 (0.16–0.36)	<0.001
Surgery only	0.36 (0.27–0.47)	<0.001

Variable	HR (95% CI)	*p* Value
Surgery and chemoradiation	0.30 (0.21–0.44)	<0.001
Surgery and chemotherapy	0.33 (0.25–0.43)	<0.001
Radiation and chemotherapy	0.49 (0.39–0.61)	<0.001
No treatment	Ref	-
Facility location		
New England (CT, MA, ME, NH, RI, VT)	Ref	-
Middle Atlantic (NJ, NY, PA)	0.51 (0.32–0.82)	0.006
South Atlantic (DC, DE, FL, GA, MD, NC, SC, VA, WV)	0.49 (0.30–0.79)	0.004
East North Central (IL, IN, MI, OH, WI)	0.55 (0.34–0.90)	0.017
East South Central (AL, KY, MS, TN)	0.57 (0.32–1.03)	0.063
West North Central (IA, KS, MN, MO, ND, NE, SD)	0.60 (0.35–1.01)	0.056
West South Central (AR, LA, OK, TX)	0.41 (0.24–0.70)	0.001
Mountain (AZ, CO, ID, MT, NM, NV, UT, WY)	0.52 (0.29–0.93)	0.028
Pacific (AK, CA, HI, OR, WA)	0.56 (0.34–0.94)	0.027
Urban (versus non-urban)	0.76 (0.35–1.64)	0.480
Academic (versus non-academic)	0.89 (0.73–1.09)	0.249
Medicaid or uninsured	1.26 (1.00–1.62)	0.050
Income		
<$38,000	Ref	-
$38,000-$47,999	1.14 (0.87–1.49)	0.345
$48,000-$62,999	0.97 (0.73–1.29)	0.826
≥$63,000	0.98 (0.73–1.33)	0.917
Education quartile		
Lowest	Ref	-
Second	0.97 (0.75–1.25)	0.809
Third	1.05 (0.80–1.38)	0.740
Highest	1.01 (0.73–1.38)	0.967

CI = confidence interval; HR = hazard ratio; NOS = not otherwise specified; Ref = reference.