The chemotherapy and radiation in low-grade myofibroblastic sarcoma: is there a role?

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Research

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Abstract

Background: Low-grade myofibroblastic sarcoma (LGMS) is a rare entity with a predilection in the head and neck. There are still no optimal treatment strategies for LGMS. We aimed to investigate the role of chemotherapy and radiation treatment for LGMS. Survival estimate was performed and prognostic factors were identified.

Methods: Based on the Surveillance, Epidemiology, and End Result (SEER) database, LGMS patients diagnosed between 2001 and 2015 were involved in our study. Kaplan-Meier curves and log-rank tests were used to estimate overall survival. Cox proportional hazard regression model was performed to identify prognostic factors.

Results: A total of 96 eligible patients with LGMS were included, among which 86 (89.6%) received surgical treatment. Twenty-eight (29.2%) patients received radiation treatment while chemotherapy was offered to 20 (10.4%) patients. The median age was 55.0 years old with 22 cases occurred in head and neck region. The mean OS was 125.2 (95%CI 106.3-144.2) months while 1-, 3-, 5- and 10-year OS rates were 88%, 77%, 70% and 59%, respectively. Age older than 60 years, positive nodal status and no surgical treatment were independent prognostic factors for patients with LGMS. Chemotherapy and radiation were not independent prognostic factors for LGMS.

Conclusions: Several prognostic factors for LGMS were revealed in this study. Surgical resection is the main therapy while chemotherapy and radiation showed limited effects on survival improvement. Thus, chemotherapy and/or radiation should not be routinely performed in LGMS.

Background

Low-grade myofibroblastic sarcoma (LGMS), firstly reported by Mentzel in 1998[1], is an extremely rare entity originated from mesenchymal with a predilection in the head and neck[2]. This tumor has been characterized by myofibroblastic proliferation with fibromatosis-like features[3]. Recent literatures suggested various sites of LGMS such as skin[4], larynx[5], tongue[6], orbit[7], breast[3], femur[2] and posterior chest wall[8]. LGMS was reported to be with local recurrence and with low probability to develop distant metastases[9]. Heart and lung can be the distant metastatic sites [10].

Owing to the rarity of LGMS, the optimal treatment is still unclear. As well as other soft tissue sarcomas (STS), surgical excision with negative margin is the primary modality for LGMS[11]. For patients without clear margin, radiation may be an effective treatment[6]. A previous population-based cohort study, based on 49 LGMS patients from the Surveillance, Epidemiology, and End Results (SEER) database, described the demographic and clinical characteristics of LGMS and investigated prognostic factors[12]. However, since the information of chemotherapy was not available and no patient had radiation as the single treatment, the role of adjuvant treatment was hardly investigated. In another two large cohorts, including 18 and 15 LGMS patients, there were only four and three patients received chemotherapy or radiation[1,

13]. Therefore, due to the limited sample size, the role of radiation and chemotherapy in LGMS is still controversial.

Based on the SEER database, the purpose of the present study was to investigate the role of chemotherapy and radiation treatment for LGMS. Survival estimate was performed and prognostic factors were identified.

Methods

Data source and patient population

We included LGMS patients from the SEER database. The database, named as Incidence - SEER 18 Regs Custom Data (with additional treatment fields), Nov 2018 Sub (1975–2016 varying), which newly included radiation and chemotherapy information, was released on April 2019 based on the November 2018 submission.

The patients were selected according to the third edition of the ICD-O (ICD-O-3) histological code of 8825/3: Myofibroblastoma, malignant, which represents Low-Grade Myofibroblastic Sarcoma. Patients diagnosed between 2001 to 2015 were included and the patients who were diagnosed at autopsy or via death certificate were excluded from our study .

The variables were classified as following: age at diagnosis (< 60 and ≥ 60 years), gender (male and female), marital status (married, unmarried and unknown), race (white and nonwhite), insurance recode (insured, uninsured and unknown), tumor grade (grade I-II, grade III-IV and unknown), tumor size (< 4 cm, ≥ 4 cm and unknown), nodal status (negative, positive and unknown), SEER historic stage (localized, regional, distant and unknown), surgery treatment (surgery and no surgery), the treatment of radiation and chemotherapy (yes and no/ unknown). Primary site was classified into “Head and neck” or “Non–head and neck” according to Site recode ICD-O-3/WHO 2008.

Statistical analysis

Quantitative data were described as mean ± standard deviation (SD) and categorical data were presented as the number and percentage (N, %). Overall survival (OS) was defined from the time of LGMS diagnosis to all causes of death or last known follow-up. Disease-specific survival (DSS) was defined from the time of LGMS diagnosis to death from cancer or last known event (death or follow-up). Survival curves were generated using the Kaplan–Meier method while log-rank test was used to determine the significance of difference in survival curves. In order to investigate the role of treatment on survival, the treatment of surgery, chemotherapy, radiation and variables with $P < 0.05$ in the log-rank test were further analyzed in Multivariate Cox proportional hazard regression model. All statistical analyses were performed using SPSS 22.0 (IBM Corporation, Armonk, NY) and all charts on survival were prepared by MedCalc 18.11.3. Two-sided $P < 0.05$ was considered as statistically significant.

Ethics Statement

The SEER database is an open database, and the data released from the SEER database do not require informed patient consent because cancer is a reportable disease in every state of the USA. The present study complied with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Results

Characteristics of the patients

A total of 96 patients were diagnosed as LGMS and detailed characteristics were shown in Table 1. In the entire population, there was a slightly female preponderance (53.1% vs 46.9%). The median age was 55.0 years old (interquartile range, 36.5–66.8 years). The majority of cohort were married (52.1%), white race (79.2%) and insured (71.9%). The tumor in seventy-four patients located at non–head and neck region such as extremities, abdomen, pelvis region and thoracic region while other 22 cases located at head and neck region. The percentage of tumor grade at I-II and III-IV was 52.1% and 22.9%, respectively. Most of the cases were diagnosed with the tumor size being larger than 4 cm (52.1%) and with negative nodal status (84.4%). As to the SEER historic stage, 49 (51.0%) cases were listed as ‘localized’, 24 (25.0%) cases as ‘regional’, and only 8 (8.3%) cases as ‘distant’. Besides, 89.6% of the patients (n = 86) received surgical treatment, 28 (29.2%) patients received radiation treatment while chemotherapy was offered to only 20 (10.4%) patients

Table 1

Characteristics of the Low-Grade Myofibroblastic Sarcoma patients in the SEER database and the results of Log-Rank Test for the Kaplan-Meier Method in subject characteristics.

Subject Characteristics	No. (%)	OS		DSS	
		Mean Survival (mon)	P (Log-Rank)	Mean Survival (mon)	P (Log-Rank)
Age					
< 60	57 (59.4)	147.2	0.002*	167.1	0.025*
≥ 60	39 (40.6)	74.9		92.5	
Gender					
Male	45 (46.9)	106.8	0.616	124.4	0.523
Female	51 (53.1)	119.3		147.7	
Marital status					
Married	50 (52.1)	116.0	0.577	136.6	0.143
Unmarried	41 (42.7)	120.0		157.0	
Unknown	5 (5.2)	90.2		90.2	
Race					
White	76 (79.2)	130.0	0.548	152.4	0.878
Nonwhite	20 (20.8)	97.1		119.9	
Insurance					
Insured	69 (71.9)	79.3	0.440	89.6	0.660
Uninsured	2 (2.1)	-		-	
Unknown	25 (26)	133.7		156.4	

Abbreviations: SEER: Surveillance, Epidemiology, and End Result; OS: overall survival; DSS: disease-specific survival.

Subject Characteristics	No. (%)	OS		DSS	
		Mean Survival (mon)	P (Log-Rank)	Mean Survival (mon)	P (Log-Rank)
Primary site					
Head and neck	22 (22.9)	116.6	0.134	121.6	0.421
Non–head and neck	74 (77.1)	117.5		149.2	
Laterality					
Unilateral	54 (56.3)	119.1	0.151	135.0	0.423
Not a paired site	42 (43.8)	114.7		146.9	
Grade					
Grade I-II	50 (52.1)	130.2	0.028*	150.7	0.016*
Grade III- IV	22 (22.9)	87.6		115.5	
Unknown	24 (25.0)	115.5		133.3	
Tumor size					
< 4 cm	20 (20.8)	132.9	0.088	-	0.014*
≥ 4 cm	50 (52.1)	86.7		98.3	
Unknown	26 (27.1)	128.7		142.9	
Nodal status					
Negative	81 (84.4)	109.8	0.036*	122.9	0.012*
Positive	3 (3.1)	15.3		15.3	
Unknown	12 (12.5)	104.3		155.9	

Abbreviations: SEER: Surveillance, Epidemiology, and End Result; OS: overall survival; DSS: disease-specific survival.

Subject Characteristics	No. (%)	OS		DSS	
		Mean Survival (mon)	P (Log-Rank)	Mean Survival (mon)	P (Log-Rank)
SEER stage					
Localized	49 (51)	132.6	0.004*	156.5	0.017*
Regional	24 (25)	123.5		128.8	
Distant	8 (8.3)	58.3		90.2	
Unknown	15 (15.6)	68.3		79.6	
Surgery					
Yes	86 (89.6)	131.2	0.011*	153.3	0.450
No	10 (10.4)	71.1		119.1	
Radiation					
Yes	28 (29.2)	105.9	0.326	121.9	0.163
No/ Unknown	68 (70.8)	129.8		159.6	
Chemotherapy					
Yes	10 (10.4)	80.7	0.021*	112.6	0.016*
No/ Unknown	86 (89.6)	119.5		140.8	

Abbreviations: SEER: Surveillance, Epidemiology, and End Result; OS: overall survival; DSS: disease-specific survival.

Survival Estimate And Prognostic Factors

The mean OS for each variable was listed in Table 1. For the total cohort, the mean OS was 125.2 (95%CI 106.3-144.2) months while 1-, 3-, 5- and 10-year OS rates were 88%, 77%, 70% and 59%, respectively (Fig. 1A). Survival curves for surgery, chemotherapy and radiation were shown in Fig. 1B-D. In the log-rank test, age, tumor grade, nodal status, SEER stage, surgery and chemotherapy showed different survival rates. However, the OS was not different between patients with or without treatment of radiation. In

Multivariate Cox regression analysis, age older than 60 years, positive nodal status HR 16.31 (95%CI 2.15-123.76) and no surgical treatment HR 4.84 (95%CI 1.15–20.33) were independent prognostic factors for patients with LGMS. The treatment of chemotherapy and radiation showed no significant influence on OS. The results of Multivariate Cox regression analysis were shown in Table 2.

Table 2

Multivariable Cox regression for analyzing the prognosis factors for Low-Grade Myofibroblastic Sarcoma patients in the SEER database.

Subject Characteristics	OS		DSS	
	P value	HR (95% CI)	P value	HR (95% CI)
Age				
< 60	1.0	1	1.0	1
≥ 60	0.003	4.34 (1.67–11.31)	0.089	2.98 (0.85–10.52)
Grade				
Grade I-II	1.0	1	1.0	1
Grade III-IV	0.083	2.30 (0.90–5.89)	0.070	3.86 (0.90–16.60)
Unknown	0.484	0.66 (0.20–2.13)	0.852	1.16 (0.25–5.42)
Tumor size				
< 4 cm	1.0	1	1.0	1
≥ 4 cm	-	-	0.932	NA
Unknown	-	-	0.936	NA
Nodal status				
Negative	1.0	1	1.0	1
Positive	0.007	16.31 (2.15-123.76)	0.009	24.90 (2.24-276.87)
Unknown	0.596	1.38 (0.42–4.52)	0.961	1.06 (0.11–9.89)
SEER stage				
Localized	1.0	1	1.0	1
Regional	0.362	1.62 (0.58–4.56)	0.094	3.44 (0.81–14.61)
Distant	0.136	2.95 (0.71–12.24)	0.989	0.98 (0.07–13.93)
Unknown	0.086	2.66 (0.87–8.12)	0.020	7.70 (1.38–42.96)
Surgery				
Yes	1.0	1	1.0	1
No	0.031	4.84 (1.15–20.33)	0.344	2.80 (0.33–23.55)

Abbreviations: SEER: Surveillance, Epidemiology, and End Result; OS: overall survival; DSS: disease-specific survival.

Subject Characteristics	OS		DSS	
	P value	HR (95% CI)	P value	HR (95% CI)
Radiation				
Yes	1.0	1	1.0	1
No/Unknown	0.053	0.39 (0.15–1.01)	0.140	0.34 (0.08–1.43)
Chemotherapy				
Yes	1.0	1	1.0	1
No/Unknown	0.805	0.85 (0.22–3.21)	0.832	0.83 (0.14–4.92)
Abbreviations: SEER: Surveillance, Epidemiology, and End Result; OS: overall survival; DSS: disease-specific survival.				

As to the DSS, the mean DSS was 152.4 (95%CI 135.9–169.0) months. The 1-, 3-, 5- and 10-year DSS rates were 93%, 85%, 79% and 76%, respectively (Fig. 2A). The mean DSS for each variable was listed in Table 1. The log-rank test showed that the following factors were significantly associated with DSS: age, tumor grade, tumor size, nodal status, SEER stage and chemotherapy. Survival curves for surgery, chemotherapy and radiation were shown in Fig. 2B-D. Except for the unknown SEER stage, the positive nodal status was the only independent prognostic factor for DSS HR 24.90 (95%CI 2.24-276.87). Neither the treatment of surgery nor the treatment of chemotherapy or radiation was the independent prognostic factor for DSS. The results of Multivariate Cox regression analysis for DSS were shown in Table 2.

Role of chemotherapy and radiation for patients treated with surgery

In the whole cohort, 10 cases did not receive surgery, among whom three cases received radiation and two underwent chemotherapy treatment. Among the other 86 patients, 54 received surgical treatment alone and the mean OS of 114.8 (95%CI 99.2-130.4) months. Seven patients received both surgery and chemotherapy treatments, four patients of these seven deceased at the last follow up while three of them died due to the tumor. The mean OS for the seven patients was 88.6 (95%CI 22.8-154.3) months. Besides, 24 patients received the treatment of surgery plus radiation with the mean OS of 115.9 (95%CI 82.9-148.9) months. One patient had both surgery and chemoradiotherapy treatments. This patient was still alive at the last follow up with 39 months of survival. The distribution of patient's survival outcome was shown according to different treatment strategies in Fig. 3. Survival curves for different treatments were generated in Fig. 4 (P = 0.051).

Discussion

LGMS was classified as a distinct type of soft-tissue tumors in the World Health Organization classification[6]. The purpose of our study was to estimate LGMS survival based on the United States

population and to investigate the role of chemotherapy and radiation treatment. In our study, a total of 96 patients were enrolled and analyzed, among whom 86 received surgical treatment. The median age was 55.0 years old with 22 cases occurred in head and neck region. The mean OS for total cohort was 125.2 (95%CI 106.3-144.2) months while the mean DSS was 152.4 (95%CI 135.9–169.0) months.

Based on the largest population with LGMS, a series of prognostic factors were revealed. Although nodal status was an independent prognostic factor for both OS and DSS in statistical analysis, its clinical value should not be overestimated because there were only three cases with positive nodal status (3.1%). In a previous SEER cohort including LGMS patients from 2001 to 2012, since there was one patient with lymph metastasis, the nodal status was not significantly associated with the survival [12]. Age order than 60 years was reported to be significantly associated with worse OS and DSS (HR = 11.3, P = 0.01; HR = 15.5, P = 0.02 for OS and DSS, respectively) [12]. In our study, age at diagnosis was further proved to be an independent prognostic factor for OS. Due to the rarity of LGMS, more factors can be found with the accumulation of cases.

Surgical resection with a negative margin is the primary therapy to prevent the local recurrence in soft tissue tumors[14]. Keller et al emphasized the importance of surgery on the long-term survival in two children diagnosed with stage I myofibrosarcoma [11]. In our cohort, patients with surgery showed longer OS than those without surgical treatment. No surgery treatment was a poor prognostic factor for OS although it was not an independent prognostic factor for DSS.

There has been no guideline recommending chemotherapy for patients with LGMS and the role of adjuvant chemotherapy still remains unclear[12]. The application of chemotherapy for LGMS was reported in some case reports[5, 8, 15]. The previous study recommended adjuvant chemotherapy as a potential therapy, particularly when the tumor is difficult to be excised completely. Chemotherapy should also be considered if the tumor showed the invasion to the adjacent tissues, or the evidence of the lymphatic and/or hematological metastasis [16]. Chemotherapy was previously reported in a LGMS patient with significant clinical improvement[8]. However, due to the absence of sufficient clinical evidence on efficiency and side effects, chemotherapy was not recommended[7]. Previous reports have suggested limited role of chemotherapy on LGMS[11, 17]. An 8-year-old girl diagnosed with LGMS received three courses of neoadjuvant chemotherapy and presented a 0.5 cm tumor diameter increase. Finally, the girl underwent surgical treatment and there was no imaging evidence of recurrence at 6 years after surgery[11]. In our study, patients without chemotherapy treatment or with unknown information about chemotherapy showed longer survival than those who received chemotherapy. Chemotherapy was not an independent prognostic factor for LGMS patients. The inverse survival outcome may be explained by small sample size of patients with chemotherapy or treatment bias by the oncologist.

A case report suggested that radiation might be a curative treatment for intermediate-grade myofibroblastic sarcoma[6]. However, LGMS was thought to be poorly responsive to radiation therapy in the literature[18]. The role of radiation could not be assessed due to the fact that no patient had radiation as a single modality therapy[12]. In our study, 28 patients received radiation treatment while only two of

them had single radiation treatment. Radiation was not an independent prognostic factor for LGMS. Besides, in order to access the adjuvant effects of chemotherapy and radiation after/before surgery, we analyzed the survival for patients who underwent surgery. The result showed chemotherapy and radiation therapy had a limited role when added to surgery in the treatment of LGMS.

This work had some limitations. First, some variables were not available in SEER database such as extent of surgical resection, lymph node dissection, margin status, which were reported to be commonly associated with STS patient's survival. Second, previous studies have documented that oral cavity is the preferred location for LGMS[7, 17]. However, only four cases occurred in the oral cavity in the present cohort. The difference may attribute to the inaccurate coding within the SEER database[12]. Third, the specific medical regimen of the adjuvant chemotherapy was not available in SEER database. Thus, we cannot analyze the effect of certain chemotherapy regimens on LGMS.

Conclusions

LGMS is an extremely rare sarcoma with the mean overall survival of 125.2 months. Nodal status was an independent prognostic factor for both OS and DSS while age older than 60 years and no surgical treatment were poor independent prognostic factors for OS. Surgical resection is the primary modality for LGMS while chemotherapy and radiation showed limited effects. The adjuvant application of chemotherapy and/or radiation in LGMS was not correlated with the improved survival, thus, chemotherapy and/or radiation should not be routinely performed in LGMS.

Abbreviations

LGMS:Low-grade myofibroblastic sarcoma; SEER:Surveillance, Epidemiology, and End Result; STS:Soft tissue sarcomas; SD:standard deviation; OS:Overall survival; DSS:Disease-specific survival.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

CZ and GW designed the study. YX collected the data. YX, GX and XW analyzed the data. YX, GX, MM and HW organized the manuscript. VP.B, VP.C and KP reviewed the papers and revised the manuscript. All the authors (YX, GX, XW, MM, HW, VP.B, VP.C, KP, GW and CZ) have read and approved the final manuscript. All authors contributed toward data analysis, drafting and revising the paper and agree to be accountable for all aspects of the work.

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References

1. Mentzel T, Dry S, Katenkamp D, Fletcher CD: **Low-grade myofibroblastic sarcoma: analysis of 18 cases in the spectrum of myofibroblastic tumors**. *AM J SURG PATHOL* 1998, **22**(10):1228-1238.
2. Arora R, Gupta R, Sharma A, Dinda AK: **A rare case of low-grade myofibroblastic sarcoma of the femur in a 38-year-old woman: a case report**. *J Med Case Rep* 2010, **4**:121.
3. Myong NH, Min JW: **Low-grade myofibroblastic sarcoma arising in fibroadenoma of the breast-A case report**. *DIAGN PATHOL* 2016, **11**:33.
4. Diaz-Cascajo C, Borghi S, Weyers W, Metze D: **Fibroblastic/myofibroblastic sarcoma of the skin: a report of five cases**. *J CUTAN PATHOL* 2003, **30**(2):128-134.
5. Covello R, Licci S, Pichi B, Spriano G, Vidiri A, Morelli L, Rosenberg AE: **Low-Grade Myofibroblastic Sarcoma of the Larynx**. *INT J SURG PATHOL* 2011, **19**(6):822-826.
6. Takacsi-Nagy Z, Murakozy G, Pogany P, Fodor J, Orosz Z: **Myofibroblastic sarcoma of the base of tongue. Case report and review of the literature**. *STRAHLENTHER ONKOL* 2009, **185**(3):198-201.
7. Zhang S, Ma Y, Ma T, Wang Z: **Low-grade myofibroblastic sarcoma of the orbit**. *MEDICINE* 2017, **96**(51):e9172.
8. Katalinic D, Santek F: **Giant low-grade primary myofibroblastic sarcoma of the posterior chest wall**. *WORLD J SURG ONCOL* 2017, **15**(1).
9. Agaimy A, Wunsch PH, Schroeder J, Gaumann A, Dietmaier W, Hartmann A, Hofstaedter F, Mentzel T: **Low-grade abdominopelvic sarcoma with myofibroblastic features (low-grade myofibroblastic sarcoma): clinicopathological, immunohistochemical, molecular genetic and ultrastructural study of two cases with literature review**. *J CLIN PATHOL* 2007, **61**(3):301-306.
10. Oylumlu M, Yildiz A, Ercan S, Oylumlu M, Davutoglu V: **Cardiac metastasis of a low-grade myofibroblastic sarcoma**. *Echocardiography* 2014, **31**(1):E1-E4.
11. Keller C, Gibbs CN, Kelly SM, Haller JR, White KS, Coffin CM, Lemons RS: **Low-grade myofibrosarcoma of the head and neck: importance of surgical therapy**. *J Pediatr Hematol Oncol* 2004, **26**(2):119-120.
12. Chan JY, Gooi Z, Wong EW, Ng SK, Tong MC, Vlantis AC: **Low-grade myofibroblastic sarcoma: A population-based study**. *LARYNGOSCOPE* 2017, **127**(1):116-121.
13. Montgomery E, Goldblum JR, Fisher C: **Myofibrosarcoma: a clinicopathologic study**. *AM J SURG PATHOL* 2001, **25**(2):219-228.
14. Eyden B, Banerjee SS, Shenjere P, Fisher C: **The myofibroblast and its tumours**. *J CLIN PATHOL* 2009, **62**(3):236-249.
15. Peng L, Tu Y, Li Y, Xiao W: **Low-grade myofibroblastic sarcoma of the pancreas: A case report and literature review**. *J CANCER RES THER* 2018, **14**(Supplement):S796-S799.
16. Meng GZ, Zhang HY, Bu H, Yang GH, Zhang XL, Yang G: **Myofibroblastic sarcoma of the nasal cavity and paranasal sinus: a clinicopathologic study of 6 cases and review of the literature**. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2007, **104**(4):530-539.

17. Tomohiro YTYN: **Low-grade myofibroblastic sarcoma of the palate**. *International Journal of Oral Science* 2012, **4**(3):170-173.
18. Smith DM, Mahmoud HH, Jenkins JR, Rao B, Hopkins KP, Parham DM: **Myofibrosarcoma of the head and neck in children**. *Pediatr Pathol Lab Med* 1995, **15**(3):403-418.

Figures

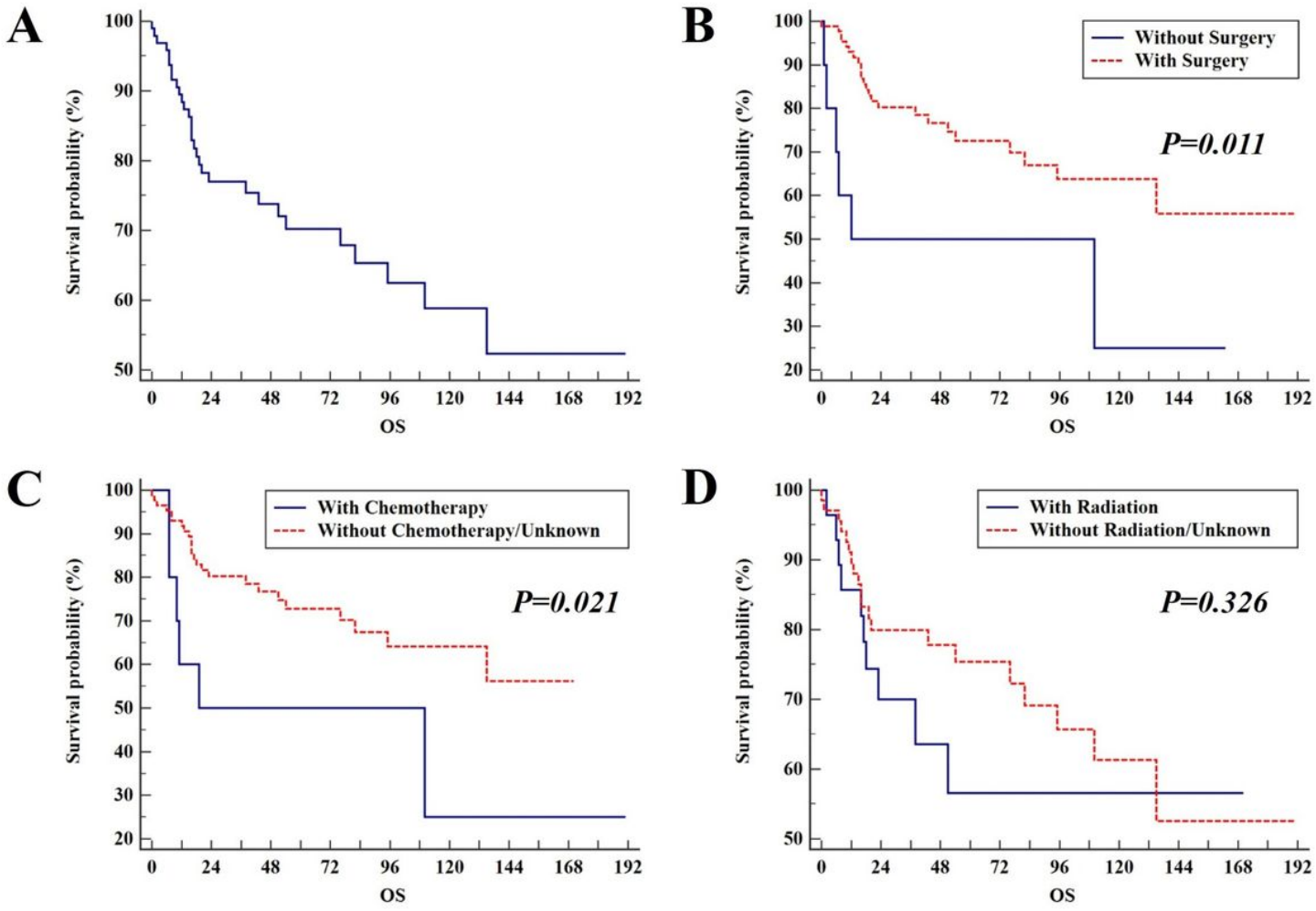

Figure 1

Overall survival for the total cohort (A) and survival curves for patients with and without surgery (B), chemotherapy (C) and radiation (D).

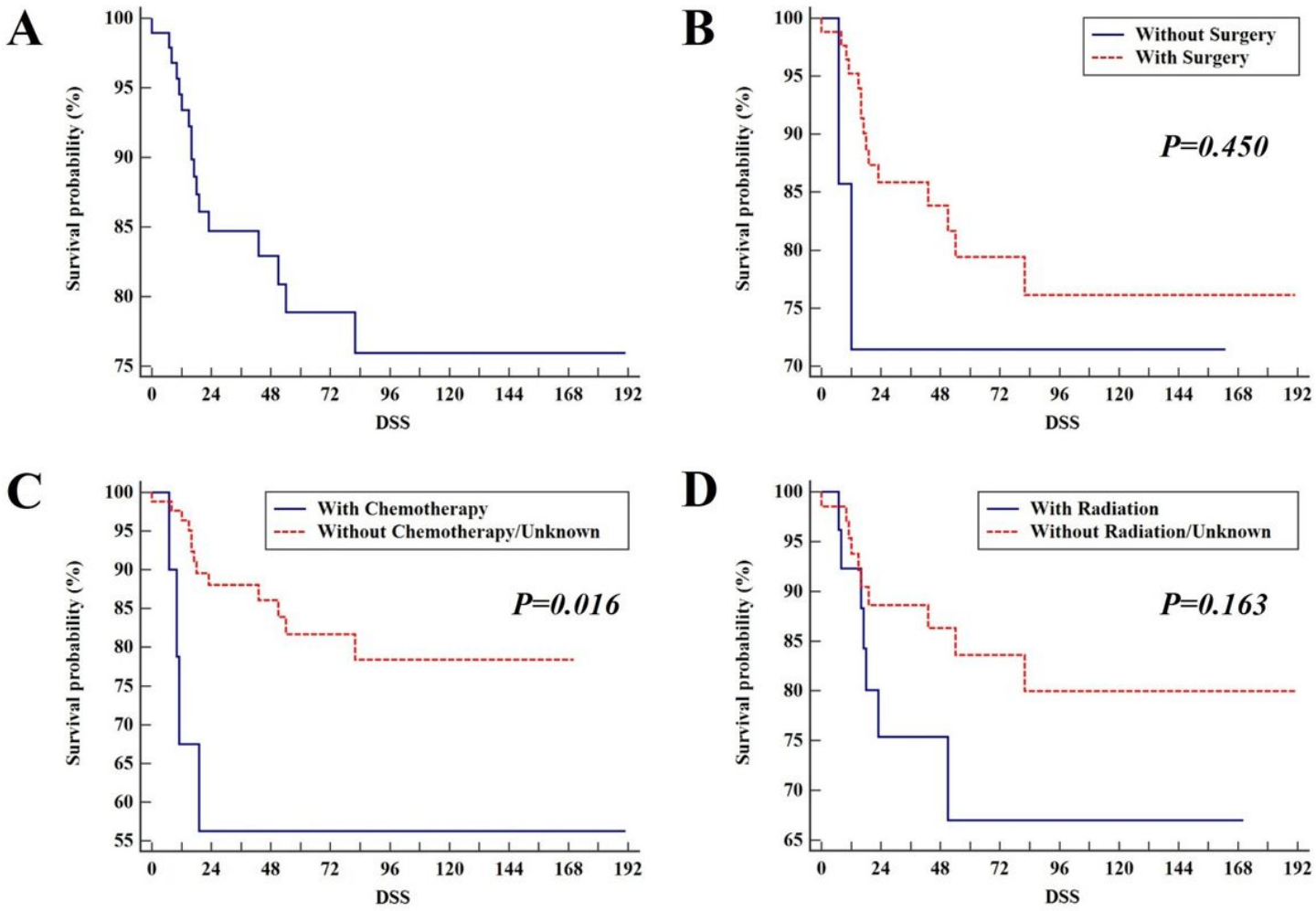

Figure 2

Disease-specific survival for the total cohort (A) and survival curves for patients with and without surgery (B), chemotherapy (C) and radiation (D).

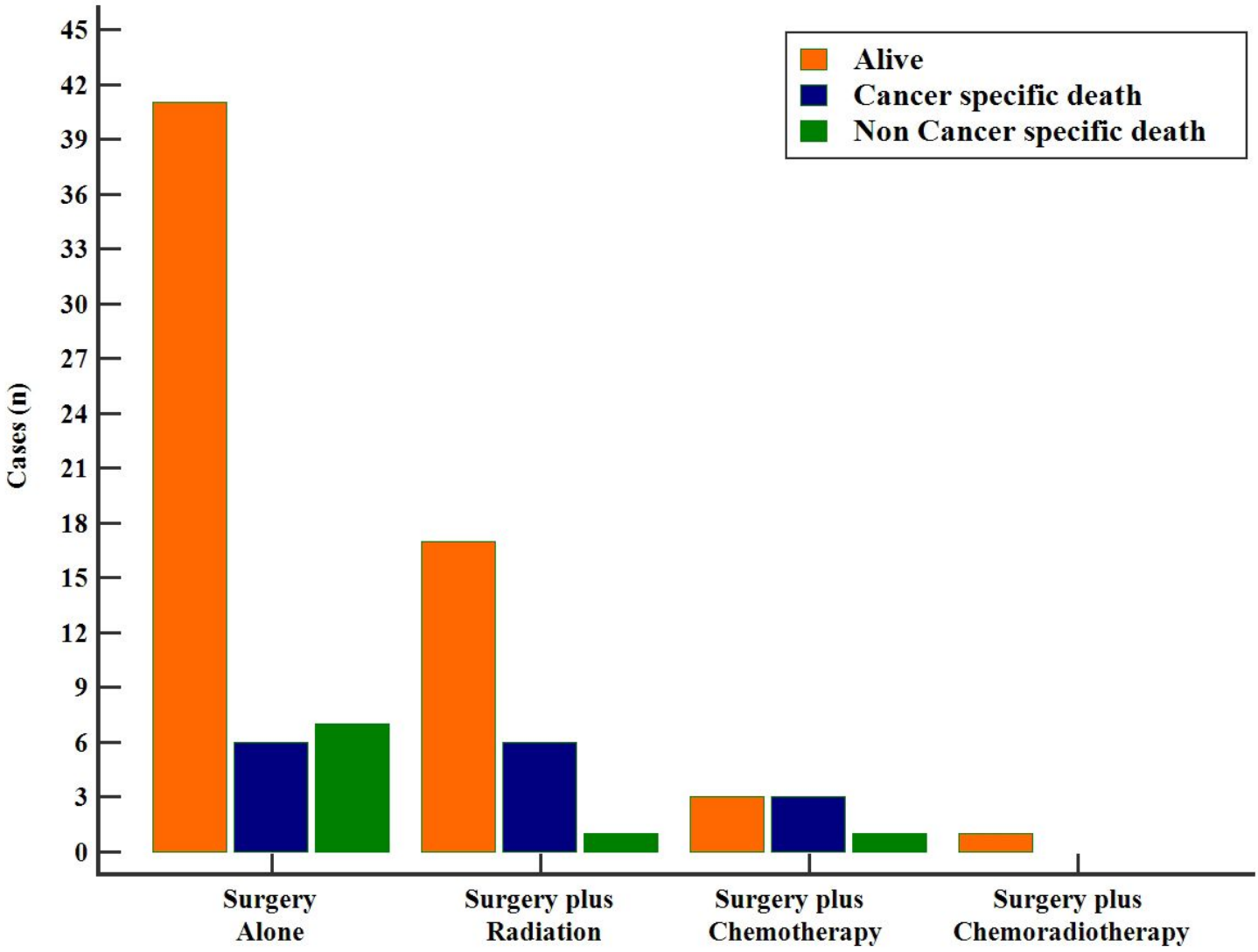

Figure 3

The distribution of patient's survival outcome according to different treatment strategies.

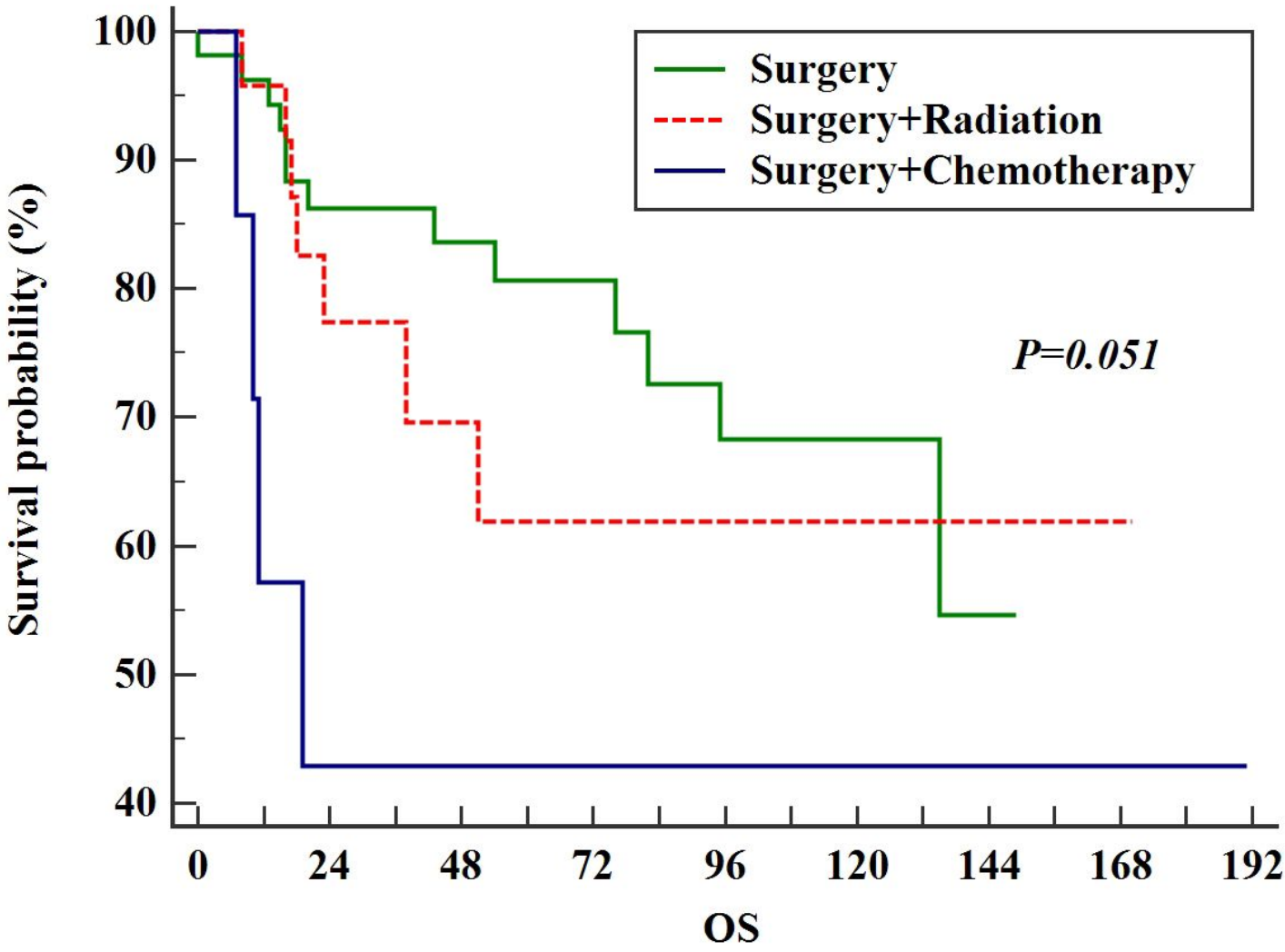

Figure 4

Survival curves for different treatments in 86 patients treated with surgery.