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ORIGINAL ARTICLE

Survival analysis of malignant peripheral nerve sheath tumor: Experience of a tertiary center in Taiwan

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Abstract

Background: This study aimed to analyze the demographic characteristics and prognostic factors of malignant peripheral nerve sheath tumor (MPNST) in a Taiwanese population. Single-center treatment outcomes were also presented.

Methods: This retrospective cohort study analyzed the medical records of 54 patients with pathological diagnoses of MPNSTs from 2005 to 2021 at a single institution. The primary endpoint was the 5-year overall survival rate of MPNST, and the secondary endpoint was recurrence-free 5-year survival. Variables including patient characteristics, metastasis status at initial diagnosis, and surgical outcomes were analyzed with competing risk analysis.

Results: Among all 41 eligible patients diagnosed with MPNST, female predominance was noted, and the median age at diagnosis was 44 years. The most common site of lesion was found at the trunk (46.34%), and eight patients were diagnosed with notable metastasis. Twelve patients were diagnosed with type 1 neurofibromatosis (NF1). The 5-year overall survival rate was 36.84% and the 5-year recurrence-free survival was 28.95%. Metastasis diagnosed at presentation, large lesion sizes, and recurrence were identified as significant poor prognostic factors of survival. Metastasis diagnosed at presentation was identified as the only significant risk factor of recurrence.

Conclusion: In our series, metastasis diagnosed at presentation, large lesion sizes, and recurrence were identified as significant poor prognostic factors of survival. Metastasis was also identified as the only significant risk factor of recurrence. NF1-associated MPNSTs presented with significantly larger tumor sizes and additional treatment postoperatively did not significantly improve survival. The limitations of this study include its retrospective nature and sample size.

Keywords: Malignant peripheral nerve sheath tumor (MPNST); Neurofibromatoses; Survival analysis

1. INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) is a rare but aggressive soft-tissue sarcoma. Most MPNSTs arise in association with a peripheral nerve and are hypothesized to be of neural crest origin.[1] These tumors usually present as a slowly enlarging mass, accompanied by pain, paresthesia, and neurologic deficits. It most commonly occurs in the proximal portions of the upper and lower extremities, near nerve roots and bundles in the extremities and the pelvis, including the sciatic nerve, brachial plexus, and sacral plexus.[2] Early metastasis is not uncommon, and metastasis at diagnosis is reported in 11% to 16% of the patients, and the most common site is the lungs.[3,4]

MPNSTs may be sporadic and observed mostly in the middle-aged population, equal among men and women,[5] and a higher incidence in Blacks was identified.[6] However, 50% of the cases are observed in patients with neurofibromatosis type 1 (NF1), occurred in a younger population, and are more common in men.[2,7] The incidence of MPNSTs among patients with NF1 is 1:3500 in comparison with the incidence of 1:100 000 among the general population.[8] Another main risk factor for MPNST development is radiation exposure. Approximately 10% of all patients with MPNST have a clinical history of prior radiation exposure,[2] and the latency period for radiation-associated MPNSTs can be more than 10 years.[9]

The treatment for localized high-grade MPNSTs is surgical resection and adjuvant radiation, and chemotherapy is administered in those with metastatic MPNSTs. The 5-year local recurrence rate ranges from 27.3% to 85.7%.[3,7,10,11] However, MPNST carries a poor prognosis. The 5-year overall survival rate ranges from 21.4% to 52%.[3,5,10]

Given the relative rarity of MPNSTs in the Asian population, the literature on MPNSTs in the Taiwanese population includes mostly case reports, and comprehensive survival analysis in Taiwan is still lacking.[12,13]

In this study, we present our 16-year experience in treating MPNST cases in a single center in Taiwan. This study aimed to analyze the demographic characteristics of the patients and

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identify potential prognostic factors of overall survival and recurrence-free survival in a Taiwanese population.

2. METHODS

2.1. Patient selection and inclusion criteria

This single-center, retrospective cohort study was conducted by the Plastic Surgery Department of Taipei Veterans General Hospital, Taiwan. The study was approved by the institutional review board of this hospital. Patients with pathological diagnoses of MPNSTs were included through the electronic patient record system on January 2005 to January 2021. The 5-year survival status was confirmed through electronic patient records. Patients with follow-up time <1 year and incomplete data were excluded. Patients who received nonsurgical treatment or subtotal resection (R2 resection) were also excluded.

2.2. Data extraction and selection

The demographic characteristics of the patients, including sex, age of diagnosis, anatomic lesion site, maximal diameter of the lesion, metastasis status at initial diagnosis, and history of NF1, were extracted and recorded. The depth of lesion invasion was divided into lesions above the fascia and below the fascia. The treatment type was classified into surgical excision alone or surgical excision with neoadjuvant therapy or adjuvant therapy (either radiotherapy or chemotherapy). Surgical outcomes, including the status of the excision margin, recurrence, and recurrence-free interval, were also recorded. Microscopically negative margins were defined as R0 margin status while macroscopically negative margins with microscopically positive margins was defined as R1 margin status.

2.3. Primary and secondary endpoints

The primary endpoint was the 5-year overall survival rate of MPNSTs, which was defined as the interval between the date of pathological diagnosis to the date of death of any cause. The secondary endpoint was the recurrence-free 5-year survival, which was defined as the interval between the date of primary surgery to the date of recurrence or death of any cause. The risk factors of mortality and recurrence were analyzed.

2.4. Statistical analysis

Continuous variables were presented as mean and standard deviation, and the discrete variables were presented in percentages. The univariate competing risk analysis with a cause-specific hazard model was applied to evaluate the variables individually to identify the potential factors of poor prognosis in both 5-year overall survival and recurrence. The significant prognostic factors were further analyzed in the multivariate competing risk analysis with the cause-specific hazard model. In the subgroup analysis, the univariate competing risk analysis with the cause-specific hazard model would be applied. Significance was set at $p \leq 0.05$ for each test. All data were analyzed using the SAS® 9.4 software, SAS Institute Inc., Cary, NC, USA.

3. RESULTS

Between January 2005 and January 2021, 54 patients were diagnosed with MPNSTs in our hospital. Among these patients with MPNST, eight were excluded, including two patients who received nonsurgical treatment, two patients who underwent primary surgery at other hospitals, three patients who underwent subtotal resection, and one patient with incomplete data. Nevertheless, five patients with a follow-up interval of less than 1 year were also excluded (Fig. 1).

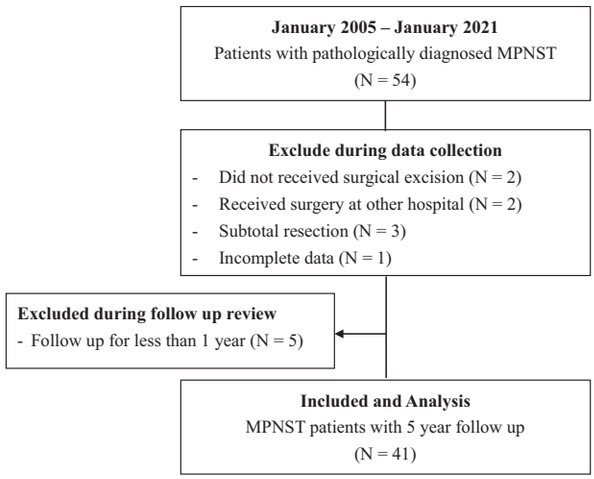

Fig. 1 Data extraction, exclusion and inclusion criteria. MPNST = malignant peripheral nerve sheath tumor.

Table 1 lists the demographic characteristics of the remaining 41 eligible patients. Slightly female predominance was noted (n = 24, 58.54%), and the median age at diagnosis was 44 (mean, 47.37 years; range, 15–-0) years, with 11 (26.83%) patients diagnosed at age >65 years. As for the anatomical distribution of lesions, 19 (46.34%) lesions were found at the trunk, followed by 17 lesions (41.46%) found in the extremities, and five lesions (12.20%) found in the head and neck region. Most of the patients had lesion ≤10 cm in size (n = 31, 75.61%). At the first presentation, eight (19.51%) patients were diagnosed with notable metastasis, and lung metastasis was noted in all of these

Table 1

Demographics of 41 study patients with MPNST

Variable	Patients (n = 41)
Gender	
Male	17 (41.46%)
Female	24 (58.54%)
Age (y)	(Median 44 y, range 15–90)
Age = 65 y old or less	30 (73.17%)
Age elder than 65 y old	11 (26.83%)
Anatomic site of lesion	
Trunk	19 (46.34%)
Extremity	17 (41.46%)
Head and neck region	5 (12.20%)
History of radiation exposure	1 (2.43%)
Underlying diseases	
Without any underlying diseases	15 (36.59%)
NF1	12 (29.27%)
CV diseases	8 (19.51%)
Metabolic or endocrine diseases	7 (17.07%)
GI disease	5 (12.20%)
Autoimmune diseases	1 (2.44%)
History of any malignancy	3 (7.32%)
Size of lesion (maximal diameter)	(Mean 9.24 cm, range 2–26.1 cm)
<5 cm	7 (17.07%)
5–10 cm	24 (58.54%)
>10 cm	10 (24.39%)
Metastasis at presentation	
No	33 (80.49%)
Yes	8 (19.51%)

CV = cardiovascular diseases; GI = gastrointestinal disease; MPNST = malignant peripheral nerve sheath tumor; NF1 = neurofibromatosis type 1.

eight patients. Among the 41 patients, 26 (63.41%) were diagnosed with at least one underlying disease, and the most common underlying disease is NF1 (n = 12, 29.27%), including one patient (2.43%) with a history of radiation exposure (Table 1).

3.1. Pathological results and surgical outcomes

All patients received surgical excision, including one patient who received neoadjuvant chemotherapy. Based on the pathological results of the resected tumor, most lesions (36, 87.8%) were located deeper than the fascia. R0 resection was reported in 23 (56.01%) lesions, whereas 18 (43.90%) lesions were reported as R1 resection, or the margin cannot be accessed because of fragmented specimens. During the 5-year follow-up interval, 15 (36.59%) patients received radiotherapy, five (12.20%) received chemotherapy, and five (12.20%) received both (Table 2).

3.2. Overall survival rate and risk factors

The 1-year overall survival rate was 82.93%, and among the 38 patients who completed the 5-year follow-up, the 5-year overall survival rate was 36.84% (Table 3). The 1- and 5-year overall survival rates were analyzed by subgroups and listed in Table 3. The univariate competing risk analysis of the 5-year all-cause mortality was conducted (Table 4). Patients with lesion sizes of >10 cm have a high risk of 5-year mortality than those with smaller lesions ($p = 0.0004$). Nevertheless, metastasis diagnosed at presentation was also a significant risk factor for 5-year mortality ($p < 0.0001$). During the 5-year follow-up, a poor prognosis was also observed in a patient who experienced recurrence ($p = 0.024$) and who received chemotherapy ($p = 0.0128$). These four variables were further analyzed in the multivariate competing risk analysis, whereas only three of them were identified as significant risk factors for the 5-year overall mortality (Table 5). Metastasis diagnosed at presentation revealed the highest hazard ratio ($p = 0.0003$), followed by lesion sizes of >10 cm ($p < 0.0001$) and recurrence ($p = 0.0108$). No other significant prognostic factor was observed in 5-year mortality, including age, anatomic sites of lesions, treatment combined with chemotherapy or radiotherapy, or history of NF1.

3.3. Recurrence-free survival and risk factors of recurrence

At the end of the 5-year follow-up, 18 patients experienced tumor recurrence (43.90%), with a median recurrence time of 12.42 (mean, 29.48 months; range 2.4-137.3) months. The recurrence-free survival at the end of the first year was 68.29%, and among the 38 patients who completed the 5-year follow-up, the recurrence-free survival was 28.95% (Table 3). Table 3 lists the subgroup analysis of the 5-year recurrence-free survival.

A univariate competing risk analysis of recurrence in the 5-year follow-up interval was conducted (Table 6). Metastasis diagnosed at presentation was the only significant risk factor of recurrence (hazard ratio [HR] = 7.302, confidence interval [CI] = 1.471-36.234, $p = 0.015$).

3.4. Subgroup analysis

In the subgroup analysis of 33 patients with localized diseases, similar risk factors of mortality were identified. Lesion sizes of >10 cm (HR = 4.845, CI = 1.622-14.476, $p = 0.0047$), chemotherapy (HR = 2.997, CI = 1.027-8.741, $p = 0.0445$), and recurrence (HR = 5.566, CI = 1.776-17.448, $p = 0.0032$) were significant poor prognosis factors of 5-year overall survival. Nevertheless, NF1 was also identified as a significant risk factor of mortality (HR = 2.743, CI = 1.014-7.421, $p = 0.0469$) in the univariate cause-specific analysis. No significant risk factor of recurrence was noted in the subgroup analysis.

As for the subgroup analysis of 29 patients with sporadic MPNSTs, lesion sizes of >10 cm (HR = 7.027, CI = 2.044-24.158, $p = 0.002$), metastasis diagnosed at presentation (HR = 16.593, CI = 3.77-73.021, $p = 0.0002$), and recurrence (HR = 3.345, CI = 1.106-10.113, $p = 0.0324$) were significant risk

Table 2

Surgical data of 41 study patients with MPNST

Variable	Patients (n = 41)
Resection margin of tumor	
R0 resection	23 (56.10%)
R1 resection	18 (43.90%)
Depth of tumor invasion	
Above fascia	5 (12.20%)
Fascia invasion or deeper	36 (87.80%)
Types of treatment	
Excision only	15 (36.59%)
Neo-C/T + excision	1 (2.44%)
Excision + R/T	15 (36.59%)
Excision + C/T	5 (12.20%)
Excision + C/T and R/T	5 (12.20%)
Recurrence	18 (43.90%)
	(Median 12.42, range 2.4–137.3 mo)

C/T = adjuvant chemotherapy; MPNST = malignant peripheral nerve sheath tumor; Neo-C/T = neoadjuvant chemotherapy; R/T = adjuvant radiotherapy; R0 resection = microscopically negative margins; R1 resection = macroscopically negative margins with microscopically positive margins.

Table 3

5-year overall survival and recurrence free survival status

	5-y OS	5-y RFS
Entire cohort (n = 38)	36.84% (n = 14)	28.95% (n= 11)
Gender		
Female (n = 23)	30.43% (n = 7)	21.74% (n = 5)
Male (n = 15)	46.67% (n = 7)	40.00% (n = 6)
Age >65 y/o		
No (n = 29)	41.38% (n = 12)	31.03% (n = 9)
Yes (n = 9)	22.22% (n = 2)	22.22% (n = 2)
Head and neck lesion		
No (n = 33)	33.33% (n = 11)	27.27% (n = 9)
Yes (n = 5)	60.00% (n = 3)	40.00% (n = 2)
Size >10 cm		
No (n = 29)	48.28% (n = 14)	37.93% (n = 11)
Yes (n = 9)	0.00% (n = 0)	0.00% (n = 0)
Fascia invasion		
No (n = 4)	50.00% (n = 2)	0.00% (n = 0)
Yes (n = 34)	35.29% (n = 12)	32.35% (n = 11)
Metastasis		
No (n = 30)	46.67% (n = 14)	36.67% (n = 11)
Yes (n =8)	0.00% (n = 0)	0.00% (n = 0)
Additional therapy		
Excision only (n = 13)	46.15% (n = 6)	38.46% (n = 5)
Yes (n = 25)	32.00% (n = 8)	24.00% (n = 6)
Concurrent NF1		
No (n = 27)	48.15% (n = 13)	37.04% (n = 10)
Yes (n = 11)	9.09% (n = 1)	9.09% (n = 1)
Margin status		
R0 resection (n = 22)	45.45% (n = 10)	36.36% (n = 8)
R1 resection (n = 16)	25.00% (n = 4)	18.75% (n = 3)
Recurrence		
No (n = 20)	55.00% (n = 11)	N/A
Yes (n = 18)	16.67% (n = 3)	

C/T = adjuvant chemotherapy; NF1 = neurofibromatosis type 1; Neo-C/T = neoadjuvant chemotherapy; OS = overall survival; RFS = recurrence-free survival; R/T = adjuvant radiotherapy; R0 resection = microscopically negative margins; R1 resection = macroscopically negative margins with microscopically positive margins.

Table 4

Univariate competing risk analysis of 5-year mortality

Variate	Hazard ratio	95% CI	*p*
Gender (male)	0.707	0.302–1.652	0.423
Age >65 y old	1.429	0.589–3.468	0.430
Lesion of head and neck region	0.431	0.101–1.842	0.256
Size of lesion >10 cm	4.823	2.024–11.492	**<0.001**
Metastasis at presentation	17.983	5.477–59.05	**<0.001**
Excision + additional therapy	1.584	0.656–3.825	0.307
Concurrent with NF1	2.239	0.987–5.077	0.054
Not R0 resection	1.709	0.761–3.838	0.195
Recurrence	2.624	1.136–6.064	**0.024**
Fascia invasion	2.106	0.494–8.982	0.314
Received C/T	2.931	1.257–6.838	**0.013**
Received R/T	0.904	0.406–2.012	0.804

Significant *p* values ($p < 0.05$) have been emphasized with bold.
Additional therapy = neoadjuvant or adjuvant therapy; C/T = adjuvant chemotherapy; CI = confidence interval; NF1 = neurofibromatosis type 1; R/T = adjuvant radiotherapy; R0 resection = microscopically negative margins.

Table 5

Multivariate competing risk analysis of 5-year mortality

Variate	Hazard ratio	95% CI	*p*
Metastasis at presentation	11.525	3.055–43.485	**<0.001**
Size of lesion >10 cm	7.284	2.704–19.620	**<0.001**
Recurrence	3.402	1.327–8.721	**0.011**
Received C/T	2.053	0.686–6.146	0.199

Significant *p* values ($p < 0.05$) have been emphasized with bold.
C/T = adjuvant chemotherapy; CI = confidence interval.

Table 6

Univariate competing risk analysis of recurrence

Variate	Hazard ratio	95% CI	*p*
Gender (male)	0.453	0.163–1.261	0.130
Age >65 y old	0.945	0.312–2.86	0.920
Lesion of head and neck region	0.819	0.237–2.839	0.754
Size of lesion >10 cm	1.419	0.39–5.168	0.596
Metastasis at presentation	7.302	1.471–36.234	**0.015**
Excision + additional therapy	1.735	0.624–4.83	0.291
Concurrent with NF1	1.427	0.533–3.822	0.479
Not R0 resection	1.394	0.562–3.458	0.474
Fascia invasion	0.669	0.219–2.041	0.480
Received C/T	2.434	0.900–6.583	0.080
Received R/T	0.902	0.366–2.224	0.823

Significant *p* value ($p < 0.05$) has been emphasized with bold.
Additional therapy = neoadjuvant or adjuvant therapy; C/T = adjuvant chemotherapy; CI = confidence interval; NF1 = neurofibromatosis type 1; R/T = adjuvant radiotherapy; R0 resection = microscopically negative margins.

factors of survival. No significant risk factor of recurrence was identified in the subgroup analysis.

4. DISCUSSION

In this study, we analyzed the characteristics of patients with MPNSTs in our institution. A mild female predominance was noted in our study, which was also seen in a previous study of the Japanese population.[14] In the present study, the median and mean age at diagnosis was 44 years old, which was slightly older than in other studies on Asian populations (median, 40-41.5).[3,14–16] Nearly half of the lesions were found at the trunk, followed by the extremities and head and neck region. Compared with a previous study, a high rate of sporadic MPNSTs (70.73%) was noted in the present study. The majority of the lesions were larger than 5 cm (82.93%), and a higher rate of notable metastasis at first diagnosis was also detected (19.51%).

The characteristics of patients with NF1-associated MPNSTs and sporadic MPNSTs were also analyzed. Patients with NF1-associated MPNSTs presented with larger average tumors at the initial diagnosis (mean sizes, 7.6 vs 13.23 cm, $p = 0.021$), corresponding with previous studies.[3,4,15,17] Patients with NF1-associated MPNSTs were on average diagnosed at a younger age than those with sporadic MPNSTs, although not significantly different (mean, 39.33 vs 50.69 years, $p = 0.123$). However, no other significant difference was found between these two groups, including sex, lesion site, age at diagnosis, or lesion depth. However, in patients with localized MPNSTs, NF1 was found to be a significantly poor prognostic factor of survival, corresponding with the finding of a previous study.[15]

In the presentation, the 1-, 3-, and 5-year overall survival rates in our study were 82.93%, 56.41%, and 36.84%, respectively (Table 3), which were in the compatible range of previous reviews based on other populations (21.4%-48%).[3–5,10,14,16,18] Several poor prognostic factors have been proposed, including distant metastasis, association with NF1, location of the tumor in the head and neck, high tumor grade, and positive margin status, large tumor size, and deep to fascia.[2,3,5,7] In our study, similar risk factors were identified. Large lesions (>10 cm),[5,17] metastasis at initial diagnosis,[15,19] and recurrence[17] were significantly poor prognostic factors of 5-year overall survival (Table 5).

At the end of the 5-year follow-up, 18 patients experienced tumor recurrence (43.90%), which was compatible with previous reports (39.6%-45%).[3,14,16] A longer median recurrence time was noted, compared with a Chinese population-based study (12.42 vs 6 months).[16] In the present study, the 5-year recurrence-free survival was 28.95% (Table 3), corresponding to existing studies (24%-41.5%).[3,4,16,20] A previous review identified the risk factors of local recurrence, including anatomic site, tumor size, recurrence, and positive margins.[1,21] Nevertheless, metastasis diagnosed at presentation was the only significant risk factor for recurrence in the present study.

This study has several limitations. The retrospective collected data may inevitably lead to potential misinterpretation. Besides, inadequate description of surgical records, outpatient follow-up records, and pathological analysis may limit the precise evaluation of the tumor sizes and time to recurrence.

Considering the rarity of MPNSTs, our study included 41 illegible patients. A multicenter with a larger sample size study of the Taiwanese population is needed for further evaluation.

To the best of our knowledge, this study is the first to report a comprehensive survival analysis of MPNSTs in the Taiwanese population in the English language literature. In this study, we presented single-center treatment outcomes, which were compatible with a previous review based on other populations worldwide.

In conclusion, MPNST is a rare but aggressive soft-tissue malignant tumor. Large lesion sizes, metastasis at initial diagnosis, and postoperative recurrence were significant harmful factors of the overall 5-year overall survival. Metastasis at presentation predicted a higher risk of recurrence. Although NF1-associated MPNSTs presented with significantly larger tumor sizes, no significant poor survival was found among patients with NF1-associated MPNSTs. Additional treatment postoperatively did not significantly improve survival, and early diagnosis is recommended.

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