

(Malignant peripheral nerve sheath tumor, MPNST)

1986 2 1996 11 MPNST 34
 가 17 , 가 17 41 (18 74)
 가 17 , 11 , 4 , 2 . AJC(American Joint Com-
 mittee on Cancer) stage IA가 2 , stage IIA 2 , stage
 IIB 6 , stage III 16 , stage IV가 8 26
 3 , 3
 33.5 (5.6 141.1)
 Kaplan-Meier , log rank test
 : 14 CDF(continuous disease free) , 2 가
 NED(no evidence of disease), 2 AWD(alive with disease), 14 가 DOD
 (died of disease) . (actuarial) 5 10 53.5% , 35.7%
 . 24.1% . 5 stage I 100%,
 stage II 85.7%, stage III 55.9% stage IV 2 14.3%
 (p=0.04). Stage II, III 21 , 가
 15 5 76.0% 6 40.0%
 (p=0.26). 4 (8) 5 71.4%
 3 (6) 3 83.3%
 (p=0.96). Stage II, III 19
 5 3 가 (60.0%) 14 4 가
 (28.6%). 8

(surgical margin)

10 cm 가 16 (47.1%) .

가 2

(Malignant Peripheral Nerve Sheath Tumor, MPNST) 32 .
 Malignant Schwannoma (staging) AJC (American Joint Committee on Cancer)
 Schwann cell (neurilema) Stage IA 2 , Stage IIA 2 , Stage IIB 5
 ma) 가 , Stage III 16 (47.1%), Stage IV가
 , MPNST 7 .

가

가

가 ^{1,2,4,6,7,10,11,12} (excisional biopsy)

(multimodality therapy) (re-excision) .
 MPNST 32 23 (71.9%)
 , 가 , 3
 , 3 , 3

, 3

1986 2 1996 11 (cycle) (9)

34 MPNST (epidemiology) 4 (14)

가 1 가 .

가 32 29 6 ,

23 (79.3%) . Enneking

가 17 가 17 8) 20 (69.0%)가

18 , 74 41 , 7 (24.1%) ,

30 , 40 8 , 16 (47.1%) 2 (6.9%)가

protocol ifos-

가 11 (32.3%), 17 famide + cisplatin 62 , CYVADIC (cyclo-
 (50.0%), 4 (11.8%), 2 phosphamide, vincristine, adriamycin,
 (5.9%) . 가 12 (35.3%) dacarbazine) 34 , ifosfamide + adriamycin

9 (26.5%) . 11 , cyclophosphamide + adriamycin 6 ,

MRI, MAID(mesna, adriamycin, ifosfamide, dacar-
 -bazine) 2 .

2.0 cm 20.0 cm 9.1

cm . 5 cm 가 7 (20.5%), 5 4000 6000

5 cm 10 cm 11 (32.4%), cGy . 3 3000 cGy

10

(

33.5 (6

1 5000 cGy 146)

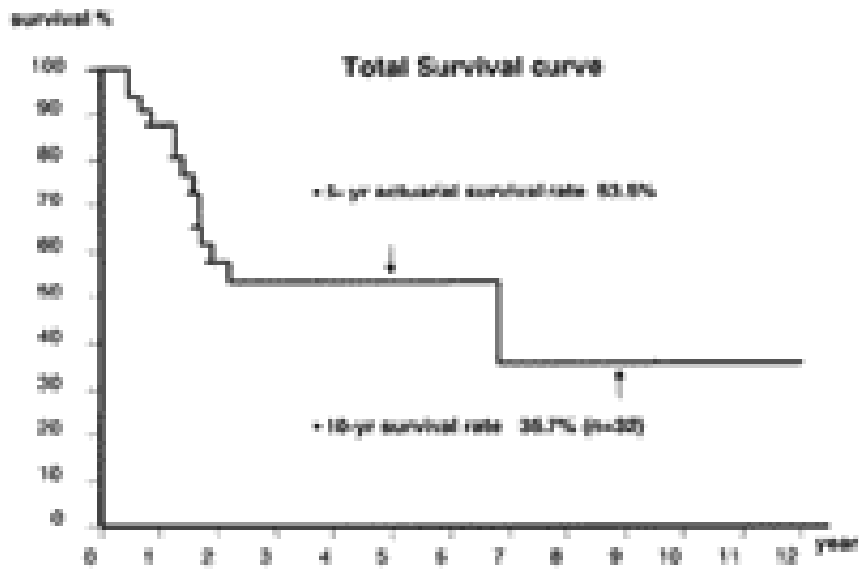


Fig. 1. Total survival curve. This graph shows actual survival proportion of MPNST patients in the study.

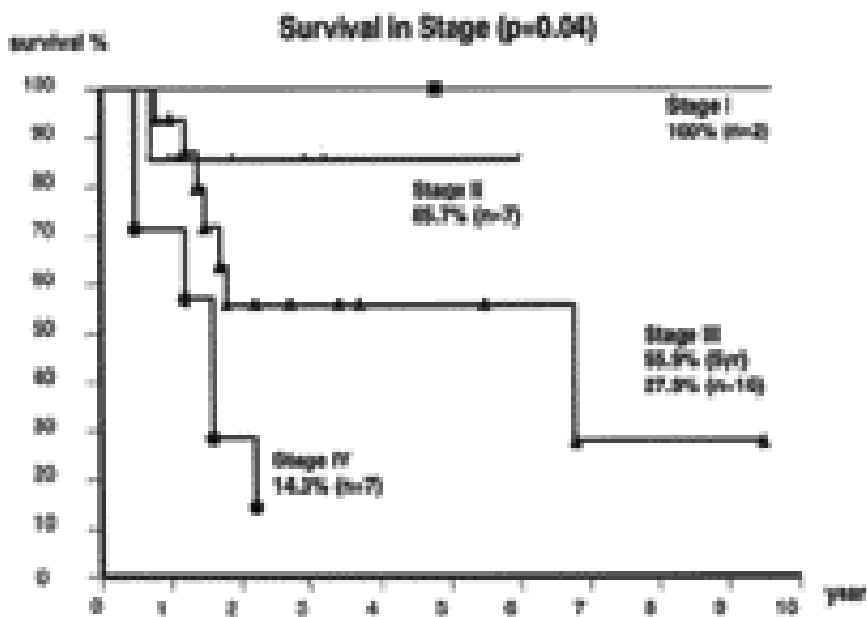


Fig. 2. Survival curve according to stages. This graph shows actual survival proportion according to stages. In log rank test, the p-value was 0.04.

Kaplan-Meier 5
 10 stage II, III
 log-rank 14 . 4
 (8) 5 71.4% 3
 (6)
 (actuarial) 5 10 3 83.3% , 2
 53.5%, 35.7% (continu
 ous disease free survival) 44.1%,
 22.1% (Fig. 1).
 (CDF) 14 , (NED) 2 , (p=0.96, Fig. 4).
 (AWD) 2 , (DOD) 14 . 29 7
 AJC stage 5 24.1% .
 stage I 100%, stage II 85.7%, stage III 3 2 2
 55.9% stage IV 2 12.6 .
 14.3% (p=0.04, Fig. 2). 가
 stage II, III stage II, III
 21 , 19
 가 15 5 5 3 가
 76.0% 60.0%
 6 40.0% . 14 4 가 28.6%
 가
 (p=0.26, Fig. 3).

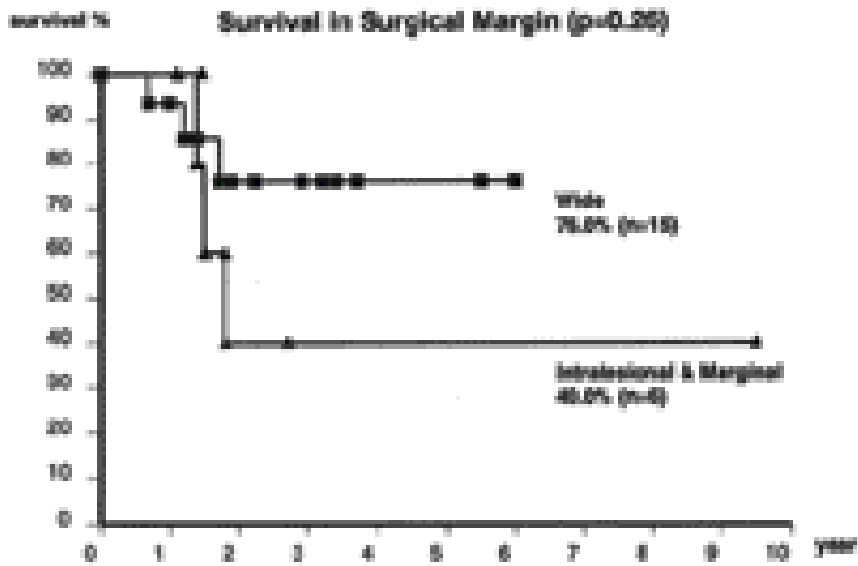


Fig. 3. Survival curve according to surgical margins. This graph shows actual survival proportion according to surgical margins. In log rank test, the p-value was 0.26.

MPNST가 1 (peripheral form neurofibromatosis) 가 가
 1/2 가
 4% MPNST가 가
 10 20 5) 34 protocol
 2

MPNST 20 50 MPNST 가
 Mayo Clinic 가
 29 40 가
 Memorial Sloan Kettering Memorial Sloan Kettering
 36 , 44 40% 65% 가
 24 14 18
 42 가 24.1%
 12.6

Wanebo 5 43.7%
 가 53.7%
 11) 가
 5 10 가
 53.5%, 35.7%
 44.1%, 22.1% 5 21.7

, 5 가
 가 9 (69.2%) 가
 Stage IV가 가
 6 가
 22.5 가 5 5
 24.1%
 14.0 가
 MPNST가 Stage II, III
 28.6%
 Stage III 가
 1997 AJC (stage) 가 가
 MPNST 가
 Stage I
 Stage
 III
 가

가
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 가
 3
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 AJC
 MPNST
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Abstract**Treatment and Survival Rate of Malignant Peripheral Nerve Sheath Tumors**

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Purpose: We analyzed our malignant peripheral nerve sheath tumor (MPNST) cases to find out their oncologic results following by each treatment modalities.

Materials and Methods: Thirty four patients with MPNST were registered in Korea Cancer Center Hospital from Feb. 1986 to Nov. 1996. Seventeen cases were male and 17, female. Average age was 41 years (range 18 to 74). Location of the tumor was as follows; 17 in lower extremity, 11 upper extremity, 4 trunk, and 2 retroperitoneum. Following the AJC classification, stage IA were 2 cases, stage IIA 2, stage IIB 6, stage III 16 and stage IV 8. Twenty six patients took operations and adjuvant chemotherapy and/or radiation therapy, 3 operation only and 3 adjuvant chemotherapy or radiation therapy. Average follow up period was 33.5 months (5.6 to 146.1). Kaplan-Meiyer method was done for survival curve, and log rank test for comparison analysis.

Results: Fourteen cases were continuous disease free, 2 no evidence of disease, 2 alive with disease and 14 dead of disease states at final follow up. Actual 5-year and 10-year survival rates were 53.5%, 35.7%. Local recurrence rate after operation was 24.1%. 5-year survival rates of stage I/II/III were 100/85.7/55.9% and 2-year survival rate of stage IV was 14.3% (p=0.04). In 21 cases operated with stage II-III, wide margin (15cases) had 76.0% 5-year survival rate, and marginal or intralesional margin (6cases) had 40.0%. The actual 5-year survival rate of the group which were done 4 or more cycles chemotherapy (8cases) was 71.4% and the actual 3-year survival rate less than 4cycles chemotherapy (6cases) was 83.3% (p=0.96). In 19 cases operated with stage II-III and which had no radiotherapy, marginal or intralesional margin (5cases) had 3 cases of local recurrences (60.0%), though wide margin (14cases) had 4 cases recurrences (28.6%). There was no local recurrence in 8cases which had pre- or post-operative radiotherapy.

Conclusions: Surgical margin is an important factor in local recurrence. Resection margin has a tendency to influence the survival despite insufficient statistical significance. Conventional chemotherapy has no definite statistical significance in the effect on local control and survival. Preoperative and postoperative radiotherapy has some positive effect on local control.

Key Words: Malignant peripheral nerve sheath tumor, Survival, Treatment

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