Treatment outcome of Neurofibromatosis type 1 associated and Sporadic Malignant Peripheral Nerve Sheath Tumor

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Introduction

Malignant peripehral nerve sheath tumor (MPNST) is one of soft tissue sarcoma. MPNST occur spontaneously or associated with neurofibromatosis type 1(NF1). There are some disputes about prognostic differences between sporadic MPNST and NF1 associated MPNST.

Methods

95 patient charts were reviewed for retrospective study for NF1 associated MPNST. In addition to comparison NF1 associated and sporadic MPNST, patient characteristics, resection margin, pathology and stage were investigated for indentifying prognostic factors.

Results

Median age of patients was 40.7 years and patients with NF 1 showed median age of 35 years. Frequent occurent location was extremity(48.4%), then trunk(34.7%), and head&neck(16.8%) were less frequent sites. Tumor size in NF1 associated patients(9.3cm) were greater than in the other patients(5.74cm). About half of pateints underwent wide resctions.(49.5%) Resection margin free(R0) sugery was taken on the 35 patients(36.5%) and the other patients(59 patients,63.5%) was taken margin positive surgery.16 patients(16.8%) had taken isolated chemotherapy, 34 patients(35.8%) had taken radothrapy only and 11 patients(11.6%) had both. 10 year overall survival(OS) of patients in MPNST was 51.1± 6.1%. 10 year OS was 56.2 ±

7.3% in sporadic MPNST patients and 40.1± 10.8 % in NF1 MPNST patients, the data reveled p-value = 0.045 and significant difference between 2 groups. In addition, resection margin and metatstasis had effet on overall survival significantly(P-value=0.037 in 10yr OS and <0.001 in 5yr OS). On multivariate analysis, only metastasis was poor prognostic factor on OS[Odd ratio = 3.11, 95% confidence interval (CI) 1.22–7.95]

Conclusion

NF1 had effect on overall survival in MPNST and there are many characters which affect on life style and survival, such like multiple neurofibroma, tendency for malignant progression. For better management NF1, we need to have general consensus follow up periods, methods and study genetic factors to influence on malignant progression.