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MULTIMODALITY TREATMENT IN MALIGNANT PERIPHERAL NERVE SHEATH TUMORS

Yusuf Karakas (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)
Omer Dizdar (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)
Kemal Kosemehmetoglu (Hacettepe Üniversitesi, Patoloji Bilim Dalı, Ankara)
Gokhan Gedikoglu (Hacettepe Üniversitesi, Patoloji Bilim Dalı, Ankara)
Figen Soylemezoglu (Hacettepe Üniversitesi, Patoloji Bilim Dalı, Ankara)
Utku Burak Bozbulut (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)
Metin Demir (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)
Alev Turker (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)
Ayse Kars (Hacettepe Üniversitesi, Medikal Onkoloji Bilim Dalı, Ankara)

Introduction - Purpose : Malignant peripheral nerve sheath tumor (MPNST) is a rare malignancy with poor prognosis, which commonly diagnosed in Neurofibromatosis type 1 (NF1) patients. Data regarding treatment options and the prognosis is limited. We aimed to investigate treatment outcomes and prognosis along with PD-L1 expression in our patients with MPNST.

Methods - Tools : Twenty-seven patients diagnosed with MPNST between 2000 and 2016 at Hacettepe University were evaluated. The patient and tumor characteristics, survival data and treatment modalities were obtained from medical charts. Slides prepared from 4-mm diameter microarray tissue were stained for PD-L1 antibody (Cell Signaling, E1L3N®) using Leica Bond Autostainer. Any membranous staining over %5 of the cells was regarded as positive.

Findings : The median age was 36 years (range 19-89), and 37% of patients were male. Median tumor size was 8.7 cm, and 62% of patients had high grade tumors. Most common tumor localizations were trunk (48%), extremity (41%), and head-neck (11%). Only 2 patients (7%) had distant metastases at the time of diagnosis. Seven patients (26%) had NF1 and presence of NF-1 was associated with poorer overall survival ($p=0.056$). Majority of the patients underwent primary surgery (96.3%), and R0-R1 resection was achieved in 70%. Median follow-up was 16 months (range 1 to 178 months). During the follow-up, 16 patients (59%) had recurrence (44% local, 15% distant recurrence). 3-year disease-free survival (DFS) rate according to R0-R1 and R2 resection were 57 vs 17%, respectively ($p<0.001$). After surgery, 48% of the patients received adjuvant therapy. Two patients (7%) received only chemotherapy, 5 patients (19%) received only radiotherapy (RT), and 6 patients (22%) received both. Patients who received both CT and RT had longer DFS compared to those who received either therapy alone and to those who received no adjuvant therapy. (3-year DFS 100% vs 14% vs 44%, respectively, $p=0.003$) (Figure 1). PDL-1 expression was positive in 5 of 13 patients evaluated (38%), and was not associated with DFS.

Discussion : Multimodality treatment with surgery, CT and RT may improve DFS in patients with MPNSTs. High rate of PDL-1 expression suggests potential responsiveness to immune checkpoint inhibitors.

Keywords: MPNST, Multimodality treatment, PDL1 expression