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Pathologic Response to Neoadjuvant Therapy is Associated With Improved Long-term Survival in High-risk Primary Localized Malignant Peripheral Nerve Sheath Tumors

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Abstract

Background: Malignant peripheral nerve sheath tumors (MPNSTs) comprise a rare, aggressive subtype of soft tissue sarcoma. While surgery is the mainstay of therapy for this disease, the role of neoadjuvant therapy remains undefined.

Methods: This study reviewed patients 16 years of age and older who underwent surgical treatment for MPNST between 1974 and 2012 at the authors' institution. Univariate and multivariate analyses were performed of clinicopathologic and treatment variables predictive of disease-specific survival (DSS) and disease-free survival.

Results: Eighty-eight patients with primary localized MPNST underwent surgical treatment between 1974 and 2012 at our institution. Of these, 38 (43%) underwent neoadjuvant chemotherapy and had tissue available for analysis. Neoadjuvant radiation was given to 25 patients (68%). The median follow-up time for survivors was 12.5 years (range, 4 to 27 y). Nine patients (23%) had underlying MPNST. With a cutoff of ≥90% pathologic necrosis and/or fibrosis defining response, we identified 14 responders (36%). On univariate analysis, patient age, tumor size, and pathologic response were significantly associated with DSS (P=0.015, 0.011, and 0.030, respectively).

Conclusions: Although the impact of neoadjuvant chemotherapy on the outcome of primary localized MPNST patients continues to be debated, this study shows that a pathologic response to therapy is associated with a significant improvement in DSS. The challenge moving forward is to determine upfront which patients will be "responders" to standard systemic therapy and which patients should be considered for newer investigational agents as part of a clinical trial.