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The effect of surgical margins on outcomes for low grade MPNSTs and atypical neurofibroma

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Abstract

Background and objectives: While convention defines atypical neurofibroma as benign and low-grade malignant peripheral nerve sheath tumors (MPNSTs) as malignant, sparse outcomes data exist for these tumors. This study reviews clinical outcomes of surgically resected low-grade MPNST and atypical neurofibroma, focusing on the effect of surgical margins on outcome.

Methods: This study is a retrospective review of 23 patients who underwent surgical resection of a low-grade MPNST or atypical neurofibroma. Treatment characteristics of adjuvant therapy and surgical margin were noted. Endpoints of local recurrence, presence of metastatic disease, disease-specific survival, and overall survival were reviewed.

Results: Eighteen of 23 patients (78%) had microscopically positive margins on the resection. Disease-specific survival was 100% for both atypical neurofibroma patients and those with low-grade MPNST, regardless of surgical margin. Local recurrence in terms of recurrence of measureable disease occurred in 2/12 (16.7%) of LGMPNST patients and 1/11 (9.1%) of atypical NF patients, all of whom had microscopically positive surgical margins.

Conclusions: In a study dedicated exclusively to "intermediate" nerve sheath tumors, no patients developed metastatic disease nor died of disease despite a high rate of microscopically positive surgical margins (78%). While positive margins did lead to increased rates of local recurrence, these data suggest that surgeons potentially can temper their zeal for negative surgical margins in the setting of low-grade MPNST and atypical neurofibroma, as surgical morbidity may be more important than a presumed survival benefit of wide resection.

Keywords: MPNST; atypical neurofibroma; low grade MPNST; nerve sheath tumor; neurofibromatosis.