Dysembryoplastic neuroepithelial tumor (DNT) in childhood

Nolan et al. examined the long-term outcome and prognostic features of DNT-associated epilepsy in 26 children managed surgically. Although 85% of children were seizure-free at 1 year, at longer follow-up this fell to 62%. Residual tumor on postoperative MRI was the most important factor in predicting long-term seizure outcome.

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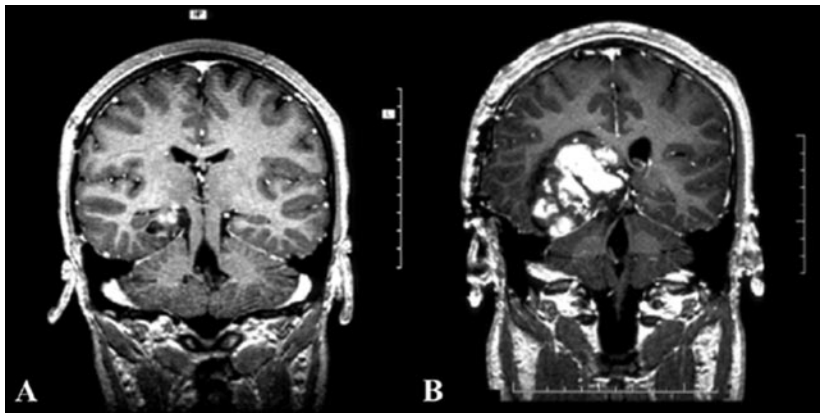
Commentary by Scott Pomeroy, MD

Dysembryoplastic neuroepithelial tumors (DNT) are glial-neuronal neoplasms that occur in children or young adults, typically in association with long-standing intractable partial seizures.[1] They localize in cortex and inconsistently enhance on MRI, and are characterized by columns of axon/oligodendroglial bundles that surround neurons floating in a pale, eosinophilic matrix. Frequently they are associated with cortical dysplasias. DNTs are thought to be dysembryoplastic in origin and without potential for recurrence once surgically removed.[2]

Nolan et al. extend our understanding of their natural history by following 26 children with DNT for up to 11 years after initial diagnosis and resection. They report an initial favorable prognosis for seizure control, with 85% of children free of seizures 1 year after diagnosis. Longer follow-up, however, demonstrates seizure recurrence in many other children. At the end of the study, only 65% remained free of seizures with mean follow-up of 4.3 years. Moreover, three of the children had tumor recurrence. This adds to a growing literature indicating that DNTs can recur after resection, and may even undergo malignant transformation.[3]



Recurrence of DNT, gadolinium enhanced coronal T1-weighted images. A. Postoperatively, there is residual enhancing DNT visible in medial right temporal lobe. B. Same patient, 3 years later. DNT recurrence with increase in size of mass.

Collectively, these results support a need for close follow-up of children and young adults who have been diagnosed and treated for DNT, monitoring for recurrence of seizures and for potential of regrowth of the tumor.

References

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