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Dysembryoplastic neuroepithelial tumors and cognitive outcome: cure at a price?

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Abstract

BACKGROUND: Dysembryoplastic neuroepithelial tumors (DNETs) are benign glioneuronal tumors that occur in children. These tumors are characterized by seizures, lack of neurologic deficits, and a seemingly benign course after resection.

METHODS: A retrospective review was conducted of data relating to 11 children diagnosed with DNETs between January 1988 and December 2007 at St. Jude Children's Research Hospital. This report documented the clinical features, neurocognitive function, and treatment outcomes in this institutional series.

RESULTS: The patient cohort included 8 boys and 3 girls (median age at diagnosis, 10 years); all patients presented with seizures: 4 complex partial, 3 generalized tonic-clonic, 2 absence, 1 partial simple, and 1 not classified. Of the 11 patients, 1 died of cardiac fibrosis, and tumors recurred or progressed in 4 (36%) patients. Seizure control was achieved in all patients but 1. Of the 9 patients who completed neuropsychologic testing, only 3 (33%) functioned at or above the expected level of same-age peers.

CONCLUSIONS: The high recurrence and progression rates of DNETs and the high rate of abnormal neurocognitive test results noted in the current study highlight the need for regular follow-up and appropriate academic counseling of children with these tumors.

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