Supratentorial primitive neuroectodermal tumors of the central nervous system in adults: molecular and histopathologic analysis of 12 cases.


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Abstract
Advances in understanding the molecular basis of primitive neuroectodermal tumors of the central nervous system (CNS-PNET) biology are critical to improve patient outcome. Recently, new data on their molecular features have been reported, suggesting that supratentorial PNET (s-PNET) in adult patients may represent a specific tumor entity among CNS-PNETs. In this study, we analyzed the clinicopathologic and molecular features of 12 cases of s-PNET in adult patients. The follow-up analysis showed that these tumors have an aggressive clinical behavior. At the histopathologic level, they resembled their pediatric counterpart, showing a variable spectrum of neuronal differentiation. These cases did not show astrocytic differentiation; therefore, they did not qualify for the differential diagnosis of glioblastoma variants. The tumors were also screened for mutation of TP53, IDH1, IDH2, and β-catenin, using single strand conformation polymorphism-based and sequencing assays, and were analyzed for c-myc/N-myc gene copy numbers with a quantitative polymerase chain reaction-based method. The strand conformation polymorphism-based mutational analysis showed that 5 tumors harbored TP53 mutations. In 2 cases, a mutation at codon 132 of the IDH1 gene was also found. No mutations of the β-catenin or IDH2 genes were identified. No cases presented c-myc or N-myc amplifications. Only 1 case presented overexpression of epidermal growth factor receptor. In conclusion, our data show a high incidence of TP53 mutations in this group of tumors and show, in comparison with pediatric s-PNET, the absence of amplification of the c-myc/N-myc genes, indicating that s-PNET in adult patients may represent a specific subset of tumors among CNS-PNETs.

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