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Glioblastoma with a primitive neuroectodermal component: two cases with implications for glioblastoma cell-of-origin.

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BACKGROUND: Glioblastoma (GBM) is the most common primary brain malignancy, but much remains unknown about the histogenesis of these tumors. In the great majority of cases, GBM is a purely glial tumor but in rare cases the classic-appearing high-grade glioma component is admixed with regions of small round blue cells with neuronal immunophenotype, and these tumors have been defined in the WHO 2016 Classification as "glioblastoma with a primitive neuronal component."

METHODS: In this paper, we present two cases of GBM-PNC with highly divergent clinical courses, and review current theories for the GBM cell-of-origin.

RESULTS AND CONCLUSIONS: GBM-PNC likely arises from a cell type competent to give rise to glial or neuronal lineages. The thesis that GBM recapitulates to some extent normal neurodevelopmental cellular pathways is supported by molecular and clinical features of our two cases of GBM-PNC, but more work is needed to determine which cellular precursor gives rise to specific cases of GBM. GBM-PNC may have a dramatically altered clinical course compared to standard GBM and may benefit from specific lines of treatment.

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