ABSTRACT

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Glioblastoma with primitive neuronal component: An immunohistochemical study and review of literature.

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Glioblastoma with primitive neuronal component, a rare neoplasm, is recognized as a distinct histological pattern of glioblastoma. In this study we report the morphological and immunohistochemical features of three cases of glioblastoma with primitive neuronal component diagnosed at the Institute along with a comprehensive literature review. The cases include: (1) 11-year-old girl with right fronto-parietal lesion, (2) 48-year-old male with right parietal lesion, and (3) 36-year-old male with left fronto-parietal lesion. Case 1 had prior history of glioblastoma. All the cases had classic morphology of glioblastoma along with GFAP-negative and synaptophysin-positive primitive neuronal component. The latter was poorly demarcated from the glial component in case 1, while well-defined in the remaining two. All the three cases exhibited diffuse p53 positivity and a higher MIB-1 labelling index in the neuronal component compared to the glial component. One of them (case 3) was IDH1 R132H-mutant with loss of ATRX expression. None were positive for K27M-mutant H3 or G34R-mutant H3.3. Literature review of 50 published cases of glioblastoma with primitive neuronal component was performed. The age of onset ranged from 3 months to 82 years (mean: 50 years) with M:F of 1.5:1. 18.8% of tumors were IDH-mutant, 87.5% were p53 positive and three cases showed H3F3A gene mutations. There was a greater propensity for neuraxial dissemination, noted in 20% of cases. Overall survival of glioblastoma with primitive neuronal component was similar to that of IDH-wildtype glioblastoma (13 months) which was significantly shorter compared to the overall survival of IDH-mutant glioblastoma (33.6 months).

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