Giant cell glioblastoma with unique bilateral cerebellopontine angle localization considered as extraaxial tumor growth in a patient with neurofibromatosis Type 1.

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Abstract
Giant cell glioblastoma multiforme (GCGBM) is a rare variant of glioblastoma, occurring predominantly in the cerebral hemispheres. Its infratentorial localization has been documented occasionally, while GCGBM in the cerebellopontine angle (CPA) region has not been described so far. We report a case of GCGBM presenting primarily as an extraaxial bilateral CPA tumor in a 29-year-old woman with neurofibromatosis Type 1 (NF1). The patient died shortly after surgery of the right CPA tumor. Postmortem study of the brain revealed large tumor masses, located in the CPA bilaterally, encasing the brainstem base and cisternal portions of the cranial nerves. Tumor masses were demarcated from the brainstem and cerebellum and covered by leptomeninges. Microscopically, a slight subpial tumor seeding from the leptomeninges into the brain parenchyma was observed in the right CPA region. The tumor showed highly pleomorphic, giant and multinucleated cells, densely cellular sheets of poorly differentiated cells and pseudopalisading necroses. Tumor cells were positive for GFAP, S-100 protein, and p53 and negative for neuronal antigens. The MIB-1 labeling index was very high in densely cellular areas. To our knowledge this is the second report of GCGBM in an NF1 patient and the first reported case of GCGBM presenting as an extraaxial leptomeningeal lesion with bilateral CPA localization, which might be considered as primary leptomeningeal gliomatosis.

PMID: 22943956 [PubMed - as supplied by publisher]