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High-grade gliomas with autoimmune encephalitis-like presentation: case report and systematic review of the literature

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Abstract

Background: Autoimmune encephalitis (AIE) consists of a heterogeneous group of inflammatory disorders affecting the central nervous system. Since several conditions, including primary brain tumors, can mimic AIE, diagnosis may be challenging.

Methods: We report the case of a 69-year-old woman initially diagnosed and treated for a suspected AIE, who later received a histological diagnosis of glioblastoma. Based on this case, we performed a systematic review of the literature to identify cases of high-grade gliomas (HGG) with an AIE-like presentation. Data were collected from each article to characterize patient demographics, clinical manifestations, cerebrospinal fluid (CSF) results, antibody profiling, neuroradiological and other findings, treatment options and outcome.

Results: Overall, 15 studies that described 21 patients were included in the literature revision. Median age was 59 years (range 32-86). Seizures were present in 17 patients. The first brain MRI did not reveal typical HGG features in any case. CSF analysis showed pleocytosis in 6 patients, increased protein content in 4, CSF-restricted oligoclonal bands (OCBs) in 1 patient. Autoantibodies were detected in 8 patients. Four patients retrospectively met the 2016 criteria for a definite AIE diagnosis by Graus et al. The median diagnostic delay to the later diagnosis of glioma was 3 months (range 1-24). One patient was diagnosed post-mortem.

Conclusions: HGG may uncommonly have an AIE-like presentation, showing potentially initial overlapping clinical and radiological features. Moreover, HGG may present with CSF pleocytosis and elevated protein count, CSF-restricted OCBs, positive autoantibodies and transitory response to immunotherapy. Patients who initially meet AIE diagnostic criteria must be followed closely over time, as distinctive oncologic features may emerge later during the disease course.

Keywords: Anti-neural antibodies; Autoimmune encephalitis; Glioblastoma; High-grade glioma.