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Clinical, pathological, radiological features and prognosis of epithelioid glioblastoma: a retrospective single center study

Guo-Zhong Yi [#1], Jin-Feng Lin [#1], Yan-Xia Lu [2], Zhi-Yong Li [1], Shan-Qiang Qu [1], Wen-Yan Feng [3], Guang-Long Huang [4]

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Abstract

Background: Epithelioid glioblastoma (eGBM) is a rare variant of glioblastoma (GBM). Due to the limited number of reported cases, our understanding of eGBM remains constrained. The updated definition of GBM has further highlighted the need for more comprehensive studies on this rare subtype.

Materials and methods: This study retrospectively reviewed 17 cases of eGBM diagnosed at a single center between 2016 and 2023. Detailed clinicopathological data were analyzed, including clinical presentation, pathological characteristics, radiological findings, treatment modalities, and patient outcomes.

Results: The cohort comprised 8 females and 9 males with an average age of 40.88 years. Tumors were predominantly located in the supratentorial region (64.71%, 11/17), while uncommon locations included the basal ganglia (11.76%, 2/17), sellar region (5.88%, 1/17), and pineal region (5.88%, 1/17). Multi-modal MRI scans revealed consistent features such as T1 contrast enhancement, elevated Cho/NAA ratios, increased nCBF and nCBV values, and disruption of white matter tracts. Additional radiological findings included mild peritumoral edema (64.71%, 11/17), hemorrhage (17.65%, 3/17), dural tail sign (35.29%, 6/17), and necrosis (29.41%, 5/17). Pathologically, all cases exhibited microvascular proliferation, IDH1 wild-type status, and high Ki-67 indices. Other notable findings included EMA positivity or partial positivity (47.06%, 8/17), INI-1 positivity (100%, 11/11), BRAF-V600E mutation (76.47%, 13/17), necrosis (82.35%, 14/17), and leptomeningeal dissemination (29.41%, 5/17). All patients underwent gross total resection (GTR), and 14 received adjuvant chemoradiotherapy. The median overall survival was 12.53 months. Two patients who received TTFields therapy survived for 12.53 and 16.63 months, respectively. Two patients treated with BRAF-V600E-targeted therapy had survival times of 16.93 and 22.67 months, respectively.

Conclusion: eGBM is a distinct and aggressive variant of GBM characterized by shorter survival times. The variability in MRI features and unusual tumor locations can lead to misdiagnosis, which multi-modal MRI may help mitigate. High frequencies of BRAF V600E mutations and INI-1 positive expression are observed in eGBM. Combined therapies, including TTFields and BRAF V600E-targeted treatments, appear to be associated with improved outcomes.

Keywords: Clinicopathological features; Epithelioid glioblastoma; Prognosis; Radiological features; Treatment.