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Cortical ependymoma: an unusual epileptogenic lesion.

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Abstract

OBJECT: Supratentorial cortical ependymomas (CE) are rare, with 7 cases reported. The lesions, typically occurring in the superficial cortex in young adults and associated with a history of seizures, are not fully characterized. Furthermore, their relationship with the recently described angiocentric glioma (AG) is still being debated. This study was undertaken to summarize the authors' experience with CEs.

METHODS: Between 1997 and 2009, 202 cases of ependymoma were surgically treated at the Mayo Clinic, 49 of which were supratentorial. Among these, 9 CE cases were retrospectively identified. Clinical, imaging, and pathological features of each case were reviewed.

RESULTS: Tumors arose from the frontal (5 cases), parietal (3), and occipital (1) lobes. No tumor occurred in the temporal lobe, despite its reported association with seizures. The mean age at presentation was 27 ± 19 years (\pm SD) and age at resection was 36 ± 16 years. The mean size of the lesion was 16 ± 14 cm³. Seizures were the presenting symptom in 78%. Cross-sectional imaging in 8 cases was characterized by a heterogeneous mass with multiple cystlike areas and enhancement of the soft-tissue component. Gross-total resection was achieved in 8 of 9 tumors. Pathologically, 6 were low-grade (WHO Grade II) and 3 were anaplastic (WHO Grade III) ependymomas. All tumors exhibited the focal presence of perivascular pseudorosettes, but only 1 (11%) exhibited the focal presence of a true rosette. A bipolar spindle cell component resembling AG was present in 3 (33%) and "Schwannian-like" nodules in 2 (22%). Subpial aggregation and peripheral infiltration were present in 4 cases (44%). With a mean postsurgery follow-up of 62 ± 38 months, only 2 lesions recurred locally after imaging-confirmed gross-total resection, both being Grade III. In 5 (71%) of 7 patients presenting with seizures an Engel Class I outcome was achieved.

CONCLUSIONS: Cortical ependymomas represent a rare type of ependymoma occurring superficially in the cortex. Morphologically, these tumors are protean, varying from classic to epithelioid, clear cell, and tancytic. Some also exhibited features typical of AG. Most tumors were low grade and cured with resection. Anaplastic tumors occur and may recur locally despite provision of radiation therapy. Cortical ependymomas frequently, but not always, present with seizures, but despite their high association with epilepsy, none occurred in the temporal lobe in any of the authors' 9 patients. Overall, CEs appear to have a relatively favorable prognosis compared with other supratentorial ependymomas.

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