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Gliomatosis cerebri in young patients' report of three cases and review of the literature.

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Abstract

Gliomatosis cerebri (GC) is a rare disease, defined as a diffuse neoplastic glial cell infiltration of the brain. Diagnosis and management of GC are difficult. **METHOD:** The authors report a literature review and their experience based on three patients, two male, and one female, all younger than 10 years, who were treated for GC. **RESULTS:** Our series of three patients were combined for the purposes of survival assessment together with the 22 patients from the literature review yielding 25 evaluable patients with diagnosis of GC. We compared the patients treated (16) with chemo, RT, or both combined, with untreated patients (7) to evaluate the median survival. Even though, as expected, the number is too small to show a statistically significant increase of survival ($p = 0.08$ log rank test), we still demonstrated a slight increase in survival in the group of patients treated (26.6 vs 14.8 months). We also compared the overall survival according to treatment. The comparison between the group of five patients treated with radiotherapy only, ten treated with chemotherapy with TMZ and ten with chemo and RT combined, showed a slight increase in mean survival, although not statistically significant, in the second and third groups ($p = 0.6$ log rank test). **CONCLUSION:** The optimal treatment in children under 10 years with GC is still obscure and absolutely not clear because total surgical resection is impossible to perform for the diffuse nature of the disease; CHT with TMZ seems to be the best treatment for children because it demonstrates a little reduction of the extension tumoral mass, but the responsiveness of this treatment is extremely variable from case to case.

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