ABSTRACT

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Medulloblastoma in adults: an analysis of clinico-pathological, molecular and treatment factors.

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BACKGROUND: Medulloblastoma is a highly malignant, embryonal tumor, which is rare in adults, and shows distinct clinical, histopathological, molecular and treatment response features.

METHODS: We retrospectively investigated 44 adults (age 17-48 years) with an histological diagnosis of medulloblastoma, and in 23 immunohistochemistry was used to identif y the molecular subgroups. We analyzed demographic, diagnostic, therapeutic and cognitive data, and correlated with PFS (progression-free-survival) and OS (overall survival).

RESULTS: We observed a male prevalence and a median age of 31 years. Symptoms at onset were related to infratentorial location, while myeloradicular and/or cranial nerve involvement was rare. Histological examination showed the classic variant in 75% of patients, the desmoplastic/nodular in 23% and the anaplastic in one. As for molecular diagnosis, 17 patients were SHH and 6 non-WNT/non-SHH (5 group 4 and 1 group 3), while no WNT subgroup was found. The SHH subgroup had a prevalence of high-risk patients and leptomeningeal involvement. Patients underwent grosstotal or subtotal/partial resection, and craniospinal irradiation, followed in 20 cases by adjuvant chemotherapy. Median OS and PFS were 16.9 and 12 years, respectively. Metastatic disease at presentation and subtotal/partial resection were associated with worse prognosis, while the addition of chemotherapy did not yield a significant advantage over radiotherapy alone. Cognitive impairment in long-term survivors was limited and late relapses occurred in 15% of patients.

CONCLUSIONS: Future studies with adequate sample size and long-term follow-up should prospectively investigate the role of surgery and adjuvant therapies across the different molecular subgroups to see whether a personalized approach is feasible.

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