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Medulloblastoma Variants: Age-Dependent Occurrence and Relation to Gorlin Syndrome--A New Clinical Perspective.

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PURPOSE: We aimed to test the hypothesis that medulloblastoma (MB) variants show a different age distribution and clinical behavior reflecting their specific biology, and that MB occurring at very young age is associated with cancer predisposition syndromes such as Gorlin syndrome (GS). **EXPERIMENTAL DESIGN:** We investigated the frequency, age distribution, location, response to treatment, outcome, and association with familial cancer predisposition syndromes in a series of 82 cases of MB in patients ages <14 years diagnosed at the Giannina Gaslini Children's Hospital, Genoa, between 1987 and 2004. **RESULTS:** Desmoplastic MB and MB with extensive nodularity (MBEN), were present in 22 of 82 cases (27%) and were more frequent in children ages ≤ 3 years (13 of 25; 52%). In this age group, MBEN was significantly more frequent than desmoplastic MB and classic MB ($P < 0.001$) and had a good prognosis. MBEN was associated with GS in 5 of 12 cases. Overall, 8 cases occurred in the context of familial tumor predisposition syndromes (5 GS, 1 each NF1, Li-Fraumeni, and Fragile X) and 7 of these patients were ages ≤ 3 years at diagnosis. Desmoplastic histology and a more intensive treatment represented independent favorable prognostic factors in multivariate analysis ($P = 0.003$ and $P = 0.0139$, respectively). Metastasis was a predictor of bad outcome ($P = 0.0001$). **CONCLUSIONS:** Our data indicate that biologically different MB entities warrant risk-adapted treatment and that MBEN is strongly associated with GS. Patients, ages ≤ 3 years, with MB and their families should be investigated for tumor predisposition syndromes such as GS.

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