Medulloblastoma

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Medulloblastoma is one of the primitive neuroectodermal tumours and the most frequent childhood brain tumour (30%). The majority occur in the first decade with a peak of incidence around five years. The overall survival has increased significantly during the last two decades with 80% of long survivors at five years regardless of the stage, but this is only 55% for high-risk patients. Medulloblastoma are divided into two groups: desmoplastic (10–20%) and non-desmoplastic (85%), including the classic form, large anaplastic cells (4%), and other rare subtypes.

Medulloblastoma patients are either classified as standard-risk patients or as high-risk patients according to the age at diagnosis (<3 years), histopathology diagnosis, extension of surgery (residual tumour $>1.5\,cm^2$) and metastatic status. Currently, treatments include surgery, standard chemotherapy or high-dose chemotherapy and risk-adapted craniospinal or localised radiotherapy according to age and risk at diagnosis. This stratification can lead to under- or over-treatment with large potential late effects. For these reasons a robust molecular classification is the key to future medulloblastoma treatment strategy. Different prognostic-molecular biomarkers are currently identified such as amplification of *MYC* associated with worse outcome or nuclear localisation of β-catenin associated with good outcome.

While almost 75% of patients achieve long-term event-free survival, surgery, high-dose chemotherapy and high-dose cranial radiotherapy are the sources of major toxicities. Most of the children who survive have significant neurocognitive sequelae. A risk-adapted radiotherapy consisting of decreasing the craniospinal irradiation dose (from 36 Gy to 24 Gy), and decreasing the gross target volume, hyperfractionation and use of adjuvant chemotherapy are the current ways of improving long-term neurocognitive outcome in average-risk patients with the same cure rate of more than 75% at five years. On line quality control in dosimetry and delivery of irradiation plays a significant role in the success of this strategy. Optimising radiation techniques such as dynamic IMRT and proton therapy are evaluated prospectively to minimise late morbidity.

Better comprehension of the biological processes and abnormal cellular signalling pathways involved in MB pathogenesis will lead to a new prognostic classification in order to adapt the therapeutic strategy, decrease late effects and give hope that the new therapeutic tools.

Conflict of interest statement

The authors have no conflict of interest to disclose.