2017 Medulloblastoma Report

Medulloblastoma - Introduction

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Historically, medulloblastoma (MB) was first described in 1925 by Percival Bailey and Harvey Cushing; Bailey wrote: “In 1925, I isolated a type of glioma which occurs chiefly in the center of the cerebellum of children for which I suggested the name *medulloblastoma*. A preliminary study was published in collaboration with Cushing (1), who has recently made a much more complete survey of the material (2)” (3). Interestingly, despite a very precise histological description, there was some confusion with gliomas.

Our current knowledge of MB is emblematic of what medicine can, at least in my opinion, achieve at its best: necessary and fruitful collaboration between different specialists (oncologists, pediatric oncologists, neurosurgeons, pathologists, geneticists, neuropsychologists, radiotherapists, etc.) and a strong connection with research, either clinical or more fundamental. This connection is usually easy to make, as the care providers on both sides are usually.... the same. A strong network has now developed (in France and worldwide) between colleagues involved in the fight against MB.

Although MB is a highly malignant tumor (grade IV on the WHO classification), many improvements in both survival and the quality of life of survivors have been made thanks to work in each of these subspecialties. For example, the relationship between the vermis and cognition (4, 5) alerted neurosurgeons to the need to avoid any injury to it (by using, for example, a telo-velar approach or by limiting the use of retractors liable to injure the dentate nuclei); or treatment (chemotherapy, radiotherapy, neurosurgery) is adapted to the aggressiveness of the tumor (stratification and strategy). The recently revised WHO classification of tumors provides additional details of MB classification (6). The current pathological classification of MB is now very precise and includes a wide range of information, from biological to histological. This classification better summarizes the important amount of information needed to characterize the different types and subtypes of MB. Knowledge of molecular biology, oncogene amplification, pathways involved in the development of MB, etc. has enabled treatment strategy to be adapted to the aggressiveness of the lesion, providing promising targets for treatment.

Some problems, however, may be clarified in the future: in terms of age, the youngest affected children remain a challenge for both pediatric neurosurgeons and pediatric oncologists. Likewise, MB recurrence is still associated with disappointing results and poor survival. Moreover, there is still high mortality in long-term survivors of childhood medulloblastoma (7).

This work about such a fascinating tumor and such a collaborative model of care will include the following items: anatomy of the posterior fossa, approaches to the posterior fossa, clinical considerations and associated syndromes, pathology and classification, high-risk MB, low and very-low risk MB, the problem of recurrence and its dismal prognosis, the particularities of adult and young adult MB, radiotherapy, treatment side-effects, neuropsychological issues, chromosomal abnormalities and signaling pathways, perspectives for research, etc.

And finally, I want to thank and very sincerely congratulate all those who agreed to participate in this review and who gave much of their time to provide papers of high quality, documenting the state of the art about this tumor. There has been no focus upon the question since the

remarkable work of Choux and Lena published in *Neurochirurgie* in 1982 (8) and I therefore hope that this special issue of *Neurochirugie* will help the reader to have a better understanding of the many facets of medulloblastoma.

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