We are IntechOpen, the world’s leading publisher of Open Access books
Built by scientists, for scientists

6,600 Open access books available
177,000 International authors and editors
195M Downloads

154 Countries delivered to
TOP 1% Our authors are among the most cited scientists
12.2% Contributors from top 500 universities

WEB OF SCIENCE™
Selection of our books indexed in the Book Citation Index in Web of Science™ Core Collection (BKCI)

Interested in publishing with us?
Contact book.department@intechopen.com

Numbers displayed above are based on latest data collected.
For more information visit www.intechopen.com
1. Introduction

The management of patients with malignancies of the central nervous system (CNS) has presented a challenge for many years. For benign tumors such as pituitary adenomas or low-grade meningiomas, surgery is often adequate to ensure a good outcome, whereas for high-grade gliomas, maximal surgery followed by radiation and chemotherapy is the standard of care. Newer modalities such as immunotherapy and tumor-treating fields have shown benefit, and have been added to the armamentarium that neuro-oncologists employ to care for patients with challenging high-grade tumors.

2. WHO classification

In 2016, the World Health Organization (WHO) revised its classification scheme for tumors of the central nervous system. The 2000 [1] and 2007 [2] classification schemes relied on histologic findings, including immunohistochemistry, to determine tumor type based on similarities to the cell of origin, as well as tumor grade. The 2016 [3] changes combined the histologic findings with genetic information in order to better characterize these tumors and to establish data on prognosis. An updated classification scheme is coming this year [4] and builds on the 2016 version, incorporating updated molecular and genetic biomarkers.

3. Pediatric CNS tumors

CNS tumors are the most common solid tumors in children. The ones most commonly found in the posterior fossa are medulloblastoma, juvenile pilocytic astrocytoma, ependymoma, diffuse pontine glioma, and atypical teratoid-rhabdoid tumor [5]. Medulloblastomas [6] are characterized by molecular features and histopathology, and there are four subtypes: WNT-activated, SHH-activated, and Group 3 and Group 4 non-WNT/non-SHH. Each subtype has its own molecular and histologic characteristics, and all are considered WHO Grade 4. Ependymomas [7] occur in both adults and children and can occur in the brain and spinal cord. The majority occur in the posterior fossa and can be WHO Grade 2 or 3. A supratentorial RELA-fusion-positive variant was first described in 2016. Astrocytomas, oligodendrogliomas, craniopharyngiomas, gangliogliomas, primitive neuroectodermal tumors, and meningiomas are most often found supratentorially. Often these are low-grade, slow-growing tumors that can be treated with surgery alone.
4. Adult CNS tumors

Meningiomas [8] arise from the meninges and are most often benign, Grade I tumors, though there are rare higher-grade meningiomas that can invade into the brain tissue. As mentioned above, ependymomas can occur in both children and adults. A myxopapillary variant is commonly found at the filum/cauda equina and is more common in adults. The new 2021 WHO classification has changed this subtype from a Grade 1 tumor to Grade 2 due to its more aggressive clinical course. The two glands within the brain also can develop tumors: Pituitary adenomas [9] arise from the pituitary gland within the sella turcica and can cause endocrinopathies and visual field loss. Tumors of the pineal gland [10] can lead to symptoms from hydrocephalus, among others. However, the most common primary CNS tumor is also the most aggressive, glioblastoma multiforme, which are classified as IDH mutant or wildtype with very different survival outcomes. Maximal surgical resection correlates with better survival [11], and surgical advances have led to improved outcomes. Following surgery, post-operative radiation and chemotherapy are the standard of care. Only temozolomide chemotherapy [12] and Optune tumor-treating fields [13] have been shown to improve overall survival in these patients, though new therapies are being investigated.

Neuro-oncology remains a very exciting field with new therapies being developed based on our better understanding of the genetics and molecular characteristics of these tumors. Targeted therapies [14] and immunotherapy [15], including vaccines, are being developed. This is truly a very exciting time for the field, and I am sure that, as our understanding of the most aggressive types of primary brain tumors grows, we will make even greater strides toward successfully treating these deadly malignancies.
References


[11] Lacroix M et al. Extent of surgery correlates with survival and surgical advances have led to improved survival. We include a discussion of awake craniotomies and new neuro-navigation and brain mapping techniques for surgical planning. Journal of Neurosurgery. 2001;92(2):190-198


