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Pediatric meningiomas: A literature review and diagnostic update

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Abstract

Background. Meningiomas have always represented the most frequently observed primary central nervous system (CNS) tumor in adults. Multiple advances concerning the genetic and epigenetic characterizations of adult meningiomas have been made over the last few years, and a new proposition for integrated histo-molecular grading has recently been offered in the literature. Pediatric meningiomas represent a very small proportion of all diagnosed meningiomas. New literature has determined that pediatric meningiomas are clinically, histopathologically, genetically, and epigenetically distinct from their adult counterparts. Herein, we reviewed and performed a synthesis of literature investigating pediatric meningiomas. We then compared and contrasted pediatric meningiomas with their adult counterparts.

Methods. We performed an extensive review of cases from English-language literature available in Pubmed using the keywords "pediatric" and "meningioma" as well as "children" and "meningioma". We reviewed and analyzed fifty-six papers that include 498 cases.

Results. This literature review revealed that pediatric meningiomas differ from their adult counterparts clinically (location, sex ratio) and also in terms of etiology (germline mutations), histopathology (a greater incidence of clear cell subtype), molecular biology, and epigenetics.

Conclusions. Pediatric meningiomas are, like other brain tumors (such as low-grade and high-grade gliomas), clinically and biologically different from their adult counterparts. Further studies are needed to better understand the tumorigenesis of pediatric meningiomas and to optimize their stratification in terms of outcome and therapeutic strategy.

Keywords

meningioma | pediatric.

The classification of central nervous system (CNS) tumors has undergone a vast transformation in the last few years, thanks to important advances in genetic and epigenetic technologies (based on DNA-methylation profiling). Former diagnostic categories based solely on histopathological criteria are now obsolete as multiple tumor types have emerged and well-constituted, mostly molecularly defined groups have replaced them.[1] Moreover, pediatric and adult gliomas

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are now considered separately due to differential genetic drivers and molecular pathways.[1] Meningiomas have always represented the most frequently observed primary CNS tumors in adults.[1,2] Multiple advances concerning the genetic and epigenetic characterizations of adult meningiomas have been made over the last few years, and a new proposition for integrated histo-molecular grading has recently been considered in the literature.[3,4] Pediatric meningiomas represent a very small proportion of all diagnosed meningiomas (ranging from 1 to 5% according to CBTRUS data).[5,6] New literature has determined that pediatric meningiomas are clinically (sex ratio, location of tumors), histopathologically (histological grading criteria), genetically (molecular abnormalities), and epigenetically (methylation classes) distinct from their adult counterparts. These specific characteristics have yet to be confirmed by additional studies.[7,8] In this article, we perform a literature review and synthesis of pediatric meningiomas studies and compare and contrast them with their adult counterparts.

Literature Review

We performed an extensive review of cases from the English-language literature in Pubmed using the keywords "pediatric" and "meningioma" as well as "children" and "meningioma". Fifty-six papers including 498 cases as well as papers representing series without specified details were analyzed for this review. We compared and contrasted those results with results from four series of adult meningiomas.[9–12]

Localization and Clinical Features

As in adults, pediatric meningiomas can be found throughout the CNS, with the most common sites being the cerebral convexities (46% of reported cases), and the skull base (27% of reported cases)[3,7,8,13–37] (Figure 1). A greater incidence of intraventricular and spinal meningiomas tend to occur in children (10% and 7% of reported cases, respectively).[3,7,8,13–37] Other locations include the posterior fossa (7% of reported cases), and also exceptionally the orbit, the optic nerve, and olfactory grooves (all these rare locations representing less than 4% of reported cases).[3,7,8,13–37] Meningiomatosis is described in 15% of reported pediatric cases.[3,7,8,13–37] Presenting symptoms depend on the tumor's location. Due to their supratentorial predilection, pediatric meningiomas present with seizures, and intracranial hypertension syndrome.[3,7,8,13–37]

Epidemiology

Epidemiological data for pediatric meningiomas vary according to the studies and countries concerned, the age of patients, and the expertise of the neurosurgery departments. The *Surveillance, Epidemiology, and End Results* database reports that meningiomas occurring between 0 and 21 years of age account for only 0.64% of all meningiomas.[16] Among pediatric CNS tumors, meningiomas represent 1.42 to 5% of cases.[5,6,17]

Worldwide incidence data for pediatric meningiomas is very limited. In the Netherlands, pediatric meningiomas occur at an average annual rate of 1 case per 1 million in the pediatric population.[36] Contrary to their adult counterparts, pediatric meningiomas affect males and females equally (52 *vs.* 48% of reported cases).[3,7,8,13–37] For childhood meningiomas, the median age of at presentation is 13 (ranging from 0 to 18 years), with infantile cases (less than 1 year of age) representing less than 4% of cases.[3,7,8,13–37]

Etiology

Similar to adult tumors, ionizing radiation may constitute an established environmental risk factor for pediatric meningiomas (representing 8% of all reported cases), particularly after radiation therapy for a medulloblastoma, ependymoma, glioma, or lymphoma.[3,7,8,13–37] Neurofibromatosis type 2 is encountered in 21% of reported pediatric meningiomas, with multiple locations in the majority of cases.[3,7,8,13–37] Pediatric meningiomas have also been described in families with germline mutations in *NF1*,[26] *SMARCE1*,[38] *BAP1*,[39] and *SUFU*[40] genes. The identification of clear cell meningioma may help detect germline loss of function mutations of the *SMARCE1* gene and – through genetic counseling – also diagnose, parents with a family history of meningiomas/meningiomatosis.[41]

Imaging

Using magnetic resonance imaging (MRI), pediatric meningiomas (similar to their adult counterparts) are isohypointense on T1-weighted imaging and hyperintense on T2-weighted imaging.[42] Intense gadolinium enhancement is frequent.[42] Neoplastic dural infiltration is determined by the presence of the «dural tail» which is inconstant.[42] Computer tomography (CT) scan demonstrates a hyperdense mass with an intense and homogeneous enhancement after contrast injection, calcifications, edema, and hyperostosis.[42] Pediatric meningiomas are more likely to be cystic and larger than their adult counterparts.[42,43] The absence of dural attachment and calcifications is more frequently seen in children than adults.[42]

Histopathology

Meningiomas are histopathologically classified and graded according to the World Health Organization (WHO) classification.[1] Pediatric meningiomas may present as any of the 15 histopathological subtypes, with, as in adults, a majority of classical morphologies (meningothelial, transitional, and fibroblastic). Our meta-analysis of 56 studies (which included 571 pediatric meningiomas) shows that they can be broken into the following subtypes present as: 28% meningothelial subtype, 21% transitional subtype, 15.5% atypical subtype, 10.5% fibroblastic subtype, 6% clear

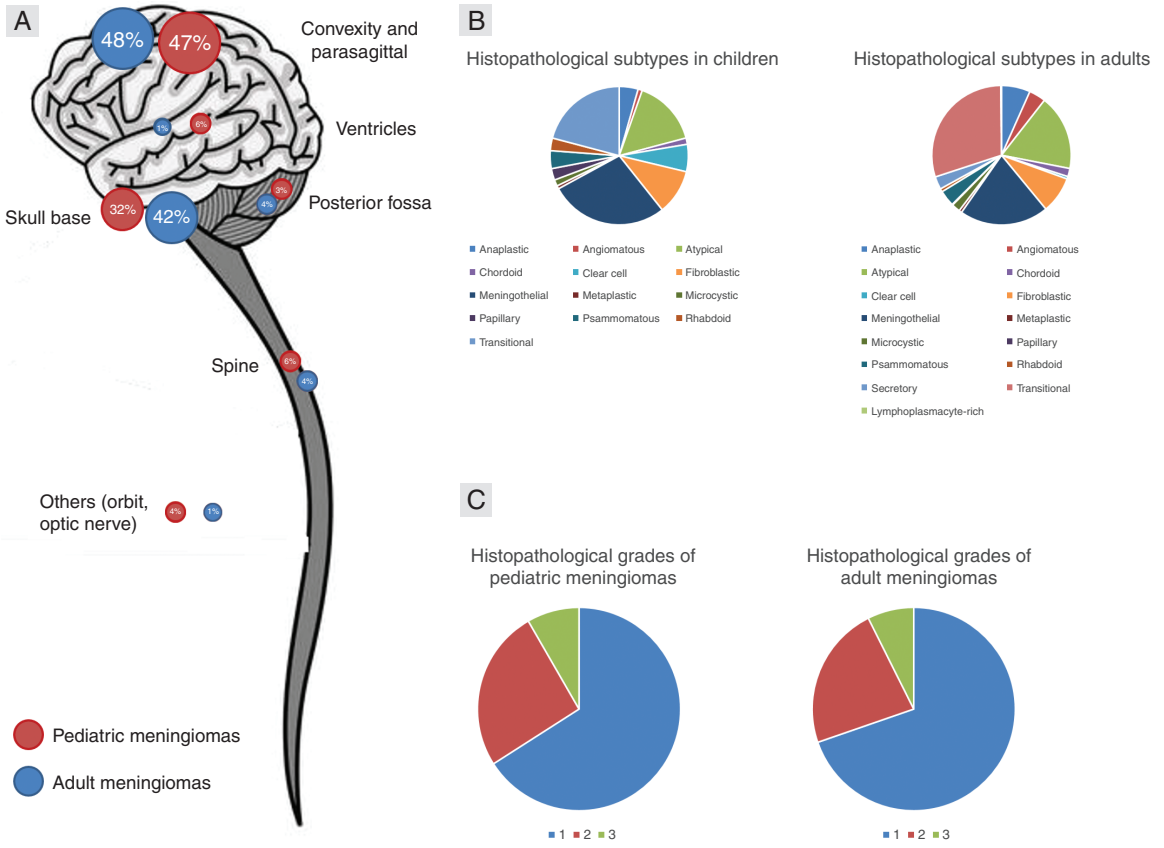

Figure 1. Locations and histopathological subtypes of pediatric meningiomas. Those results included all articles concerning pediatric meningiomas of the literature review and the four series of adult meningiomas.

cell subtype, 4% anaplastic subtype, 4% psammomatous subtype, 3% papillary subtype, 3% rhabdoid subtype, 1.5% microcystic subtype, 1.5% chordoid subtype, 1% angiomatous subtype, <1% secretory subtype, <1% metaplastic subtype, and <1% lymphoplasmacyte-rich subtype[3,7,8,13–37] (Figure 1). This meta-analysis is biased by the use of various past WHO classifications. These results provide evidence that the clear cell subtype is more prevalent in the pediatric population than in the adult population.[44] However, the distribution between the three tumor grades is quite similar to that in adults.[3,7,8,13–37] In the majority of cases, the histopathological diagnosis of a meningioma does not present difficulties for pathologists. However, recent literature has identified a potential morphologic mimicker, the intracranial mesenchymal tumor, *FET::CREB*-fused which can be found in adult and pediatric populations and which shares anatomic, radiological, and histological features with meningiomas.[45,46] However, this tumor type does not present SSTR2a immunoexpression,[46] *NF2* alterations, is defined by a fusion between a member of the *FET* (mainly *EWSR1*) and the *CREB* (*CREB1*, *CREM*, or *AFT1*) family of genes respectively,[1,45,46] and also has a distinct DNA-methylation profile.[46,47] Another potential histopathological confusion may happen with other CNS tumor with *BCOR* internal tandem duplication[1]: it has often been falsely diagnosed as a papillary meningioma in a subset of pediatric cases in the literature.[48]

Pathogenesis

As in adults, monosomy of chromosome 22 is the most frequently reported genetic abnormality in pediatric meningiomas.[7] Loss of heterozygosity of 22q with bi-allelic inactivation of the *NF2* tumor suppressor gene, located at 22q12.2, is recognized as an early tumorigenic event of meningioma.[49] The *NF2* gene encodes the protein Merlin which plays a role in regulating cell growth and cell motility as well as binding several transmembrane signaling proteins.[49] *NF2* alterations have been more frequently reported in children (49% of cases) than in adults (22% of cases).[7,8,12,50] Whereas *TRAF7*, *AKT1*, *KLF4*, *SMO*, and *PIK3CA* alterations have been reported in a large part of adult meningiomas, particularly those located in the skull base, these mutations are rare in the pediatric population.[7,13,51] Genetic mutations in these pathways (Hedgehog signaling for *SMO*- and SUFU-altered meningiomas, PIK3 pathway for *PIK3CA*- and *AKT*-altered meningiomas) seem to correlate with the anatomical distribution of meningiomas (anterior and medial skull base, convexity) and embryological origins.[51,52] This anatomic/genetic correlation suggests different molecular driver events may be present in children. In non-*NF2* driven pediatric meningiomas, *YAP1* fusions (particularly with the *MAML2* gene) as an alternative pathogenic alteration have been

reported.[35,53] *YAP1* is a transcriptional co-activator and downstream effector of the HIPPO pathway which regulates the expression of genes involved in cell proliferation and apoptosis. *YAP1* fusions have been evidenced in a series of meningiomas, mainly pediatric (one adult case was described), but no clear phenotype (different morphologies of meningiomas have been reported with these fusions, including poorly differentiated forms initially misdiagnoses as gliomas) or grade was found to be associated.[35] A group of rhabdoid and papillary meningiomas share *BAP1* (*BRCA1*-associated protein-1) and *PBRM1* alterations.[54,55] Loss of function mutations in the *BAP1* gene is easily detectable immunohistochemically by protein loss.[56] Even though pediatric rhabdoid/papillary meningiomas are exceedingly rare (only one reported case with *BAP1* alteration),[39] the clinical implications are important because a subset of meningiomas associated with *BAP1* alterations may reveal the presence of the *BAP1* tumor predisposition syndrome including tumors of different organs and lineage (mesotheliomas, uveal, and cutaneous melanomas, clear cell carcinoma of the kidney).[56,57] Moreover, loss of BAP1 staining was associated with an aggressive clinical behavior in these subtypes of meningioma.[56] However, only adult cases of these subtypes of meningioma have been reported to date. Bi-allelic alterations in *SMARCE1* constitute the sole specific abnormality of a histopathological subtype the clear cell meningioma, which is mostly encountered in children, adolescents, and young adults.[38,44,58–60] The *SMARCE1* gene, located on chromosome 17q21, encodes a component of the SWI/SNF (BAF57) protein complex which is a modulator for chromatin remodeling and gene transcription.[58]This loss of function alteration may be easily and specifically evidenced by the loss of SMARCE1 protein expression using immunohistochemistry.[44] Finally, the DNA-methylation profiling of pediatric meningiomas revealed that they represent three different epigenetic subgroups (named as 1, 2a, and 2b), distinct from those described in adults (distributed throughout 6 prognostic subgroups: benign-1, benign-2, benign-3, intermediate-A, intermediate-B, and malignant).[7,12] Even pediatric meningiomas presenting *NF2* alterations did not fall into the cluster of *NF2*-altered adult meningiomas.[7] These methylation classes include different histopathological subtypes and grades. However, group 1 seems to be largely encompassed by clear cell meningiomas and *NF2* alterations seem to be excluded from group 2B.[7] These results (obtained via DNA-methylation profiling) seem to suggest the presence of a *NF2* alteration in a pediatric meningioma. However, germline mutation may be difficult to diagnose by classical molecular analyses.[7]

Meningioangiomatosis

Meningioangiomatosis is a poorly studied, rare, benign, and epileptogenic brain meningovascular lesion. Only about two-hundred cases have been reported to date.[61,62] The majority of meningioangiomatosis are sporadic, affecting predominantly male patients, younger than 20 years of age.[62] Epileptic seizures constitute the main symptom, with more than 80% of patients having uncontrolled seizures at the time of surgery.[62] Meningioangiomatosis are mainly located in the frontal and temporal lobes.[62] Imaging presentation is heterogeneous, and the diagnosis is often missed pre-operatively.[62] Histopathologically, meningioangiomatosis is characterized by an intracortical meningovascular proliferation with a perivascular spread of spindle-shaped cells along the Virchow-Robin spaces. It often encompasses psammoma bodies, fibrosis, and white matter infiltration.[62] The malformative or neoplastic origin of meningioangiomatosis is still not well-understood. However, the fact that 30% of cases are reported in association with an adjacent tumor, predominantly a meningioma[62] – and also that cytogenetic analyses of isolated meningioangiomatoses or those associated with meningiomas have been found to present alterations, including 22q12 deletions[61,62] – seems to support the idea that meningioangiomatosis represents a potential specific tumoral pattern of infiltration. The outcome of meningioangiomatosis is favorable. Indeed, the majority of patients are treated by a complete resection and have epileptic seizure control at the end of follow-up; recurrences are rare.[62]

Prognosis and Prediction

As in adults, clinical, histopathological, and biological features have been defined as prognostic factors in pediatric meningiomas. Among these factors, a major clinical predictor of recurrence and overall survival in childhood meningiomas is based on the extent of surgical resection.[26,30,43] The feasibility/quality of resection may, however, be limited by the tumor location (particularly in the skull base) and the extent of invasion. The WHO CNS classification grade has been determined as the most prevalent histopathological predictor of recurrence. However, because this grading was based on a series of 581 meningiomas, of which less than 5% of cases were pediatric,[63,64] and because some pediatric meningiomas are histopathologically classified as atypical (determined as 4 per 1.6 mm^2 for grade 2), a novel proposition of mitotic count index (6 per 1.6 mm^2 for grade 2) has been shown to correlate with recurrence-free survival.[8] The loss of H3K27me3 immunoexpression, which is associated with malignant morphology and a shorter overall survival in adult meningiomas,[65,66] seems to be very rare in the pediatric group and to date, no prognostic impact has been attributed in this age group.[8] More than the histological grade, the histopathological subtype of clear cell meningioma, *SMARCE1*-altered is considered to have an aggressive clinical behavior with a higher rate of recurrences and death compared to other histopathological subtypes of meningiomas.[3,7,8,13–37,44] Cytogenetic (1p and 14q deletions) and molecular (*CDKN2A* homozygous deletion) abnormalities have been reported to be associated with high-grade histologies, elevated risk of recurrence, and a shorter time to progression in adult and pediatric populations.[4,7,67–69] Even though *TERT* promoter mutations have been associated with a poor prognosis in adults, this mutation seems to be very rare in the pediatric population despite a high rate

of grades 2 and 3 meningiomas in this age group.[7] In adults, DNA-methylation profiling of meningiomas defines six clusters of meningiomas split into three prognostic subgroups (benign, intermediate, and malignant) which in turn correlates to prognosis.[12,70] However, this classification has not been demonstrated in childhood meningiomas, which are separated into three subgroups distinct from adult meningiomas.[7] Finally, an integrated molecular-morphologic risk stratification has been suggested in adult meningiomas and has yet to be tested in pediatric population.[7]

Conclusion

Taken together, pediatric meningiomas are, like other brain tumors (such as low-grade and high-grade gliomas), clinically and biologically different from their adult counterparts. Further studies of their histopathological, genetic, and epigenetic characteristics are needed to better understand the tumorigenesis of pediatric meningiomas and to optimize their stratification in terms of outcome and therapeutic strategy.

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