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Topical Review

Pediatric Neuroglial Tumors: A Review of Ependymoma and Dysembryoplastic Neuroepithelial Tumor

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Ependymoma

Ependymomas are the third most common type of malignant pediatric intracranial neoplasm, constituting on average about 6% to 10% of CNS tumor diagnoses in children.[1,2]

These neuroepithelial tumors develop from ependymal cells, which line the ventricular system, with some research showing that supraventricular ependymoma originates from radial glial cells.[3] These tumors predominantly occur in males and are most prevalent in early childhood. Pediatric ependymomas are primarily located…

Posterior fossa ependymoma

Posterior fossa ependymomas account for about 65% to 75% of pediatric ependymomas.[17] These tumors primarily develop near the fourth ventricle and can cause symptoms of obstructive hydrocephalus, such as ataxia, vomiting, early morning headaches, and occasionally cranial nerve palsies, depending on the tumor's size and location. These tumors are characterized by location (midline versus lateral) as this impacts their resectability and thus prognosis.[8]

Posterior fossa ependymomas are divided into…

Supratentorial ependymoma

Supratentorial ependymomas are less common than posterior fossa ependymomas and often present with headaches, seizures, or focal deficits, depending on their location. These tumors are divided into two main groups based on genetic characteristics: ZFTA and YAP1 fusion types.

ZFTA fusion tumors, which account for about 70% of supratentorial ependymomas, have a median age of onset of 8 years and may be seen in adults.[7] Tumors of this group are more aggressive and have a poorer prognosis. These…

Myxopapillary ependymoma

Myxopapillary ependymomas predominantly occur in the caudal cord region and account for 25% of all spinal cord tumors. These tumors typically present with symptoms such as lower back pain, lumbar radiculopathy (including tingling and weakness in the lower extremities), and bowel or bladder dysfunction. The median age at diagnosis is 39 to 41 years, and in the pediatric population these tumors are usually found in adolescents or teens.[4,5] These tumors generally have a favorable prognosis, with…

Metastatic disease

Metastasis is not an uncommon phenomenon for ependymomas due to their location in the CSF.[24] Metastasis is most often found in disease recurrence, with the majority of metastases found to be arising from the posterior fossa.[25] Isolated distant metastases account for 3% to 7% of relapses, whereas coexisting local and distant metastases account for 10% to 20% of relapses.[26,27] Metastasis is most commonly within the CNS, either near the primary site or in the spinal cord.[28] Metastasis outside of…

Treatment

Treatment of ependymoma requires a multidisciplinary approach across teams compromising a neurosurgeon, neuro-oncologist, radiation oncologist, neuro-radiologist, and neuropathologist. In an effort to reduce treatment-related morbidities and improve outcomes, treatment paradigms and approaches have evolved over the past few decades. Surgery with or without radiation therapy (RT) are the two cornerstones of treatment modalities for all types of ependymoma. The majority of the ependymomas in…

Future considerations

Understanding the molecular landscape of ependymomas has shed light on the intratumoral and spatial heterogeneity and has revealed that molecular subgroups have distinct clinical outcomes. Current clinical trials are moving away from utilizing histologic features like anaplasia toward decision making in the context of molecular characteristics, except for supratentorial ependymomas where histology still appears to be important. Recent clinical trials are incorporating genomic profiling to…

Recurrent intracranial ependymoma

Despite multimodal treatment, approximately 30% to 35% of patients with ependymoma relapse. Around 80% of relapses are local, but distant recurrence is seen in around 10% of patients.[53] Although there is no standard of care for recurrent or relapsed ependymomas, surgical resection or reirradiation, wherever feasible, has been shown to improve survival time. Unfortunately, many relapses follow an indolent chronic course and 90% of relapsed patients die from their disease.53, 54, 55 Craniospinal…

Dysembryoplastic neuroepithelial tumors

DNETs are a type of low-grade gliomas mostly found within the pediatric population. DNETs are World Health Organization grade I tumors classified within glioneuronal and neuronal tumors. Although they typically follow a benign course characterized by slow growth, they do have a very low chance of becoming malignant.

DNETs were first discovered in 1988, where Daumas-Duport et al. identified a unique glioneuronal tumor in 39 patients found to be causing partial complex seizures refractory to…

Conclusion

Ependymomas and DNETs are common pediatric neuroglial tumors, each with distinct biology, molecular characteristics, classification systems, and clinical behaviors. These unique features underscore the importance of a tailored approach to diagnosis, treatment, and management of these tumors. Advances in molecular diagnostics have revolutionized our understanding of ependymomas, allowing for more precise categorization and treatment strategies that aim to improve patient outcomes. Similarly, the …

CRediT authorship contribution statement

Melissa Arfuso: Writing – review & editing, Writing – original draft. **Sandeepkumar Kuril:** Writing – review & editing, Writing – original draft. **Harshal Shah:** Writing – review & editing, Writing – original draft. **Derek Hanson:** Writing – review & editing, Writing – original draft, Supervision, Conceptualization.…

Declaration of competing interest

None.…

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Pediatric brain tumors are the most common solid malignancy in children. Although categorized by their location in the central nervous system (CNS), this group of tumors is highly heterogeneous, composed of numerous different tumor types, each with a unique cell of origin, clinical behavior, and prognosis. Advances in molecular diagnostics have pushed the field of pediatric neuro-oncology forward in recent years. Tumors that were once considered a singular diagnosis are now known to be multiple entities with a unique genetic signature; this has led to more tailored and complex treatment and management approaches for patients. Given the wide range of neurological issues that children with CNS tumors face, pediatric neurologists play a crucial role in their care. In this review, we focus on two specific pediatric neuroglial tumors: ependymoma and dysembryoplastic neuroepithelial tumor (DNET). These tumors often exhibit symptoms and complications that necessitate the expertise of pediatric neurologists. A thorough understanding of the clinical presentation, diagnosis, and treatment of these tumors is essential for physicians to provide the best

Ependymoma

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These neuroepithelial tumors develop from ependymal cells, which line the ventricular system, with some research showing that supraventricular ependymoma originates from radial glial cells.[3] These tumors predominantly occur in males and are most prevalent in early childhood. Pediatric ependymomas are primarily located intracranially, most commonly in the posterior fossa, followed by supratentorial regions, but can occur infrequently in the spine.[1,4,5]

Symptoms at initial diagnosis of ependymomas can vary but are often related to increased intracranial pressure, particularly in tumors located in the posterior fossa. These tumors can interrupt cerebrospinal fluid flow in the fourth ventricle, leading to obstructive hydrocephalus. Typical presenting symptoms include

possible neurological care for these children.

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nausea, vomiting, early morning headaches, balance and coordination issues, gait abnormalities, and, in infants, increased head circumference. Ependymomas can also cause seizures or focal neurological deficits, most often caused by supratentorial tumors due to mass effect. Spinal cord ependymomas may present with focal deficits, myelopathies, or radiculopathies, depending on the spinal level affected.

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References (83)

M.D. Taylor *et al.*
Radial glial cells are candidate stem cells of ependymoma
Cancer Cell (2005)

E.I. Hwang *et al.*
Why haven't we solved intracranial pediatric ependymoma? Current questions and barriers to treatment advances
Neoplasia (2023)

A.E. Gillen *et al.*
Single-cell RNA sequencing of childhood ependymoma reveals neoplastic cell subpopulations that impact molecular classification and etiology
Cell Rep (2020)

K.W. Pajtler *et al.*
Molecular classification of ependymal tumors across all CNS compartments, histopathological grades, and age groups
Cancer Cell (2015)

D.S. Tsang *et al.*
Outcomes after reirradiation for recurrent pediatric intracranial ependymoma
Int J Radiat Oncol Biol Phys (2018)

S. Khatua *et al.*
Current therapy and the evolving molecular landscape of paediatric ependymoma
Eur J Cancer (2017)

T. Tihan *et al.*
The prognostic value of histological grading of posterior fossa ependymomas in children: a Children's Oncology Group study and a review of prognostic factors
Mod Pathol (2008)

R.G. Grundy *et al.*
Primary postoperative chemotherapy without radiotherapy for intracranial ependymoma in children: the UKCCSG/SIOP prospective study
Lancet Oncol (2007)

E. Bouffet *et al.*
Survival benefit for pediatric patients with recurrent ependymoma treated with reirradiation
Int J Radiat Oncol Biol Phys (2012)

B.R. Eaton *et al.*
Use of proton therapy for re-irradiation in pediatric intracranial ependymoma
Radiother Oncol (2015)

View more references

Cited by (0)

1 Drs. Arfuso and Kuril were equally responsible for the work described in the paper and will be joint first authors.

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