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Understanding diffuse leptomeningeal glioneuronal tumors

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Abstract

Introduction: Diffuse leptomeningeal glioneuronal tumors (DLGNTs) pose a rare and challenging entity within pediatric central nervous system neoplasms. Despite their rarity, DLGNTs exhibit complex clinical presentations and unique molecular characteristics, necessitating a deeper understanding of their diagnostic and therapeutic nuances.

Methods: This review synthesizes contemporary literature on DLGNT, encompassing epidemiology, clinical manifestations, pathological features, treatment strategies, prognostic markers, and future research directions. To compile the existing body of knowledge on DLGNT, a comprehensive search of relevant databases was conducted.

Results: DLGNT primarily affects pediatric populations but can manifest across all age groups. Its diagnosis is confounded by nonspecific clinical presentations and overlapping radiological features with other CNS neoplasms. Magnetic resonance imaging (MRI) serves as a cornerstone for DLGNT diagnosis, revealing characteristic leptomeningeal enhancement and intraparenchymal involvement. Histologically, DLGNT presents with low to moderate cellularity and exhibits molecular alterations in the MAPK/ERK signalling pathway. Optimal management of DLGNT necessitates a multidisciplinary approach encompassing surgical resection, chemotherapy, radiotherapy, and emerging targeted therapies directed against specific genetic alterations. Prognostication remains challenging, with factors such as age at diagnosis, histological subtypes, and genetic alterations influencing disease progression and treatment response. Long-term survival data are limited, underscoring the need for collaborative research efforts.

Conclusion: Advancements in molecular profiling, targeted therapies, and international collaborations hold promise for improving DLGNT outcomes. Harnessing the collective expertise of clinicians, researchers, and patient advocates, can advance the field of DLGNT research and optimize patient care paradigms.

Keywords: Brain tumor; Central nervous system neoplasm; Diffuse leptomeningeal glioneuronal tumor (DLGNT); Leptomeningeal dissemination; Review.

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