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Advances in molecular prognostication and treatments in ependymoma

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Abstract

Ependymoma is the third most common brain tumour of childhood and historically has posed a major challenge to both pediatric and adult neuro-oncologists. Ependymoma can occur anywhere in the central nervous system throughout the entire age spectrum. Treatment options have been limited to surgery and radiation, and outcomes have been widely disparate across studies. Indeed, these disparate outcomes have rendered it extraordinarily difficult to compare studies and to truly understand which patients are low and high-risk. Over the past two decades there have been tremendous advances in our understanding of the biology of ependymoma, which have changed risk stratification dramatically. Indeed, it is now well accepted that ependymoma comprises multiple distinct entities, whereby each compartment (supratentorial, posterior fossa, spinal) are distinct, and within each compartment there exist unique groups. The driver events, demographics and response to treatment vary widely across these groups and allow for a better classification of thee disease. Herein, we review the advances in the molecular stratification of ependymoma including how an improved classification and risk stratification allows for more precise therapies.

Keywords: Ependymoma; Genomics; Molecular stratification; PFA; PFB; Radiotherapy; ZFTA.