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Polymorphous low-grade neuroepithelial tumour of young (PLNTY): the new kid on the block

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Abstract

Introduction: Polymorphous low grade neuroepithelial tumor of the young (PLNTY) is a newly described epileptogenic tumor first reported by Jason. T. Huse et al. in 2016. Only a very few cases have been reported so far and has been recently incorporated in the World Health Organization (WHO) Central Nervous System Classification of tumours, 5th edition, 2021. Here we report a rare case of PLNTY which closely resembles DNET (Dysembryoplastic neuroepithelial tumor) with plenty of interesting findings which would otherwise go unnoticed resulting in a nonspecific or misclassified diagnosis.

Case report: A 12 year old boy presented to the Neurosurgery OPD with seizures for the past five years and was given multiple antiepileptics for the same. Magnetic resonance imaging (MRI) showed a well-defined lobulated cortical mass with T1 hypo intensity and T2 hyperintensity in the left temporal lobe measuring 2.1 × 2 × 1.3 cm suggesting a DNET. Left temporal craniotomy and excision of the lesion was done. Frozen section showed features of a low grade glial neoplasm. Routine sections demonstrated polymorphous findings including oligodendroglia like features, neuronal nuclear pleomorphism, spindled astroglial elements, perivascular rosettes, calcification, and vascular mineralization. By immunohistochemistry (IHC), the tumor cells were diffusely positive for GFAP and CD34.Ki67 labelling index was low. A final diagnosis of PLNTY was made based on the above findings. The child has been epilepsy free since the past one-month post-surgery and is on follow up.

Discussion/conclusion: PLNTY is a newly discovered distinct pediatric low grade glial neoplasm which was earlier grouped into nonspecific forms of DNET. It is characterized morphologically and molecularly by the presence of oligodendroglial component, CD34 expression, BRAFV600E mutation and alterations in the MAP kinase pathway. They are known to behave in a low-grade fashion amenable to control by excision with occasional cases of recurrence reported. It is important to recognize and report similar tumors to determine the long-term risk of recurrence and create a more complete understanding on their radiology and molecular genetics.

Keywords: Epilepsy; Neuroepithelial tumour; Paediatric glioma.