Dysembryoplastic neuroepithelial tumor

- Co-Occurrence of Dysembryoplastic Neuroepithelial Tumor and Hodgkin Lymphoma in a Patient with Noonan Syndrome and Mutation in the PTPN11 Gene
- Differentiation of multinodular and vacuolating neuronal tumor and dysembryoplastic neuroepithelial tumor based on MRI
- Epilepsy in patients with pediatric brain tumors: Etiology, treatment & management
- Metachronous tumour (DNET and haemorrhagic chiasmal tumour) in a patient with encephalocraniocutaneous syndrome (ECCL)
- The Mickey Mouse's hand sign in brain MRI points out multinodular and vacuolating neuronal tumors in mesial temporal lobe structures
- Ultrahigh-field imaging (7 Tesla) in DNET: Unmasking microstructural imaging characteristics - A case report
- Atypical Lesions in Virchow-Robin Spaces: A Case Report
- Are PDFGRA Dinucleotide Alterations Definitional for Myxoid Glioneuronal Tumor? Report of PDFRA p. K385L Mutation in a Neonatal High-Grade Glioma

Dysembryoplastic neuroepithelial tumors (DNTs) commonly abbreviated DNT or DNET are usually benign tumors of neuroepithelial origin arising from the cortical gray matter.

Defined as “a usually supratentorial glial-neuronal neoplasm occurring in children and young adults and characterized by a predominantly cortical location and by drug-resistant partial seizures”.

It appears similar to oligodendroglioma, but with visible neurons.

First coined by Daumas-Duport and colleagues to describe a cortical lesion presenting in childhood [1].

The World Health Organization (WHO) has categorized it under grade 1 tumors.

The vast majority are centered in cortical grey matter, arise from secondary germinal layers and are frequently associated with cortical dysplasia (up to 80% of cases).

Classically, DNETs have been described to have a benign course with cortical dysplasia rather than true neoplasias.

Rare midline neoplasms with similar histological features to those found in DNETs have been described near the septum pellucidum and termed 'DNET-like neoplasms of the septum pellucidum'. Due to their rarity, these tumors have been described in just a few reports and their genetic alterations sought only in small series [2].

Epidemiology

Dysembryoplastic neuroepithelial tumors (DNETs) are uncommon neural tumors presenting most often in children and young adults.

The temporal lobe is the most common site (62%), followed by the frontal lobe (31%)

Classification

Dysembryoplastic neuroepithelial tumor classification.

Genomic landscape

Pagès et al. used targeted methods (IHC, FISH, targeted sequencing) and large-scale genomic methodologies including DNA methylation profiling to perform an integrative analysis to better characterize a large retrospective cohort of 82 DNTs, enriched for tumours that showed progression on imaging.

They confirmed that specific DNTs are characterized by a single driver event with a high frequency of FGFR1 variants. However, a subset of DNA methylation-confirmed DNTs harbour alternative genomic alterations to FGFR1 duplication/mutation. They also demonstrated that a subset of DNTs sharing the same FGFR1 alterations can show in situ progression. In contrast to the specific forms, “non-specific/diffuse DNTs” corresponded to a heterogeneous molecular group encompassing diverse, newly-described, molecularly-distinct entities.

Specific DNT is a homogeneous group of tumours sharing characteristics of pediatric low-grade gliomas: a quiet genome with a recurrent genomic alteration in the RAS-MAPK signalling pathway, a distinct DNA methylation profile, a good prognosis but showing progression in some cases. The “non-specific/diffuse DNTs” subgroup encompasses various recently described histo-molecular entities, such as polymorphous low-grade neuroepithelial tumor of the young and Diffuse astrocytoma MYB or MYBL1 altered [3].

Immunophenotype

The stellate astrocytes within the specific glioneuronal element (SGNE) are positive for GFAP

The oligodendrocyte-like cells are typically S100 and OLIG2 positive, and may also express NOGO-A and myelin-oligodendrocyte glycoprotein.

The floating neurons are positive for NeuN.

Importantly, DNETs are negative for IDH mutations, TP53 mutations, and do not demonstrate 1p/19q co-deletion. These features are helpful in distinguishing DNETs from low-grade astrocytomas (usually IDH mutated) and oligodendrogliomas (IDH mutated and 1p19q co-deleted) [4]

Pathology

Dysembryoplastic Neuroepithelial Tumor Pathology.

Clinical Features

Dysembryoplastic neuroepithelial tumor Clinical Features

Diagnosis

Dysembryoplastic neuroepithelial tumor diagnosis.

Differential diagnosis

Dysembryoplastic neuroepithelial tumor differential diagnosis.

Treatment

Dysembryoplastic neuroepithelial tumor treatment.

Outcome

Dysembryoplastic Neuroepithelial Tumor Outcome.

Case series

Dysembryoplastic neuroepithelial tumor case series.

Case reports

Dysembryoplastic neuroepithelial tumor case reports.

1)
Daumas-Duport C, Scheithauer BW, Chodkiewicz JP, Laws ER Jr, Vedrenne C. Dysembryoplasic neuroepithelial tumor: A surgically curable tumor of young patients with intractable partial seizure. Report of thirty-nine cases. Neurosurgery 1988;23:545-56.
2)
Chiang JCH, Harreld JH, Tanaka R, Li X, Wen J, Zhang C, Boué DR, Rauch TM, Boyd JT, Chen J, Corbo JC, Bouldin TW, Elton SW, Liu LL, Schofield D, Lee SC, Bouffard JP, Georgescu MM, Dossani RH, Aguiar MA, Sances RA, Saad AG, Boop FA, Qaddoumi I, Ellison DW. Septal Dysembryoplastic Neuroepithelial Tumor: A Comprehensive Clinical, Imaging, Histopathologic and Molecular Analysis. Neuro Oncol. 2019 Feb 6. doi: 10.1093/neuonc/noz037. [Epub ahead of print] PubMed PMID: 30726976.
3)

Pagès M, Debily MA, Fina F, Jones DTW, Saffroy R, Castel D, Blauwblomme T, Métais A, Bourgeois M, Lechapt-Zalcman E, Tauziède-Espariat A, Andreiuolo F, Chrétien F, Grill J, Boddaert N, Figarella-Branger D, Beroukhim R, Varlet P. The genomic landscape of dysembryoplastic neuroepithelial tumours and a comprehensive analysis of recurrent cases. Neuropathol Appl Neurobiol. 2022 Jul 14:e12834. doi: 10.1111/nan.12834. Epub ahead of print. PMID: 35836307.

4)

Louis DN, Ohgaki H, Wiestler OD et-al. “WHO Classification of Tumours of the Central Nervous System. 4th Edition Revised” ISBN: 9789283244929