Ganglioglioma differential diagnosis

Low-grade epilepsy-associated neuroepithelial tumors (LEATs) create a diagnostic challenge in daily practice and intraoperative pathological consultation (IC) in particular. Intraoperative squash smear cytology are extremely useful for accurate diagnosis; however, the knowledge on cytopathologic features of LEATs is based on individual case reports. Kurtulan et al. discuss the 3 most common and well-established entities of LEATs: ganglioglioma (GG), dysembryoplastic neuroepithelial tumor (DNT), and papillary glioneuronal tumor (PGNT).

Thirty patients who underwent surgery for GG, DNT, and PGNT between 2001 and 2021 were collected. Squash smears prepared during intraoperative consultation were reviewed by 1 cytopathologist and an experienced neuropathologist.

Among the 30 tumors, 16 (53.3%) were GG, 11 (36.6%) DNT, and 3 (10%) PGNT. Cytomorphologically, all of the 3 tumor types share 2 common features such as dual cell population and vasculocentric pattern. GG smears were characteristically composed of dysplastic ganglion cells and piloid-like astrocytes on a complex architectural background of thin- to thick-walled vessels. DNT, on the other hand, showed oligodendroglial-like cells in a myxoid thin fibrillary background associated with a delicate capillary network. Common cytological features of PGNT were hyperchromatic cells with narrow cytoplasm surrounding hyalinized vessels forming a pseudopapillary pattern and bland cells with neuroendocrine nuclei dispersed in a neuropil background.

A higher diagnostic accuracy can be obtained when squash smears are applied with frozen sections. However, it is important to integrate clinical and radiologic features of the patient as well as to know the cytopathologic features of the LEAT spectrum in the context of differential diagnosis to prevent misinterpretation in the IC [1)].

Main differential diagnosis is that of other cortical tumours, with helpful distinguishing features including:

Dysembryoplastic neuroepithelial tumor differential diagnosis

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contrast enhancement uncommon 'bubbly appearance' common

Pleomorphic xanthoastrocytoma (PXA)

contrast enhancement prominent dural tail sign is often seen

Oligodendroglioma

calcifications common

Desmoplastic infantile astrocytoma and ganglioglioma

young children dural involvement prominent large often multiple lesions

If in the spinal cord consider:

astrocytoma

ependymoma

The aim of a study was to evaluate whether ganglioglioma (GGL), dysembryoplastic neuroepithelial tumour (DNET) and FCD (focal cortical dysplasia) are distinguishable through diffusion tensor imaging. Additionally, it was investigated whether the diffusion measures differed in the perilesional (pNAWM) and in the contralateral normal appearing white matter (cNAWM). Six GGLs, eight DNETs and seven FCDs were included in this study. Quantitative diffusion measures, that is, axial, radial and mean diffusivity and fractional anisotropy, were determined in the lesion identified on isotropic T2 or FLAIR-weighted images and in pNAWM and cNAWM, respectively. DNET differed from FCD in mean diffusivity, and GGL from FCD in radial diffusivity. Both types of glioneuronal tumours were different from pNAWM in fractional anisotropy and radial diffusivity. For identifying the tumour edges, threshold values for tumour-free tissue were investigated with receiver operating characteristic analyses: tumour could be separated from pNAWM at a threshold ≤ 0.32 (fractional anisotropy) or ≥ 0.56 (radial diffusivity) *10-3 mm2/s (area under the curve 0.995 and 0.990 respectively). While diffusion parameters of FCDs differed from cNAWM (radial diffusivity (*10-3 mm/s2): 0.74 ± 0.19 vs. 0.43 ± 0.05; corrected p-value < 0.001), the pNAWM could not be differentiated from the FCD [2)].

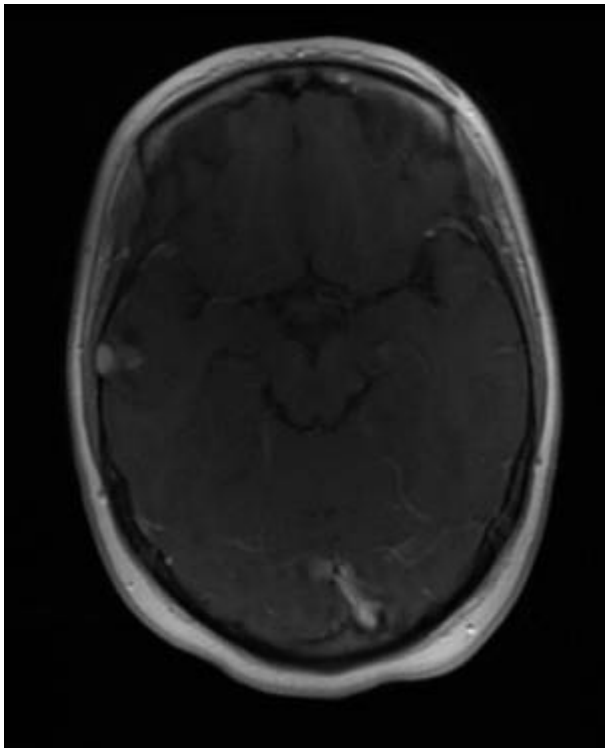
Case report

14-year-old woman admitted due to a right temporal lobe tumor.

She was transferred from other Hospital after finding a right temporal lesion on MRI in the context of seizures.

Unprovoked focal seizure. Paroxysmal episodes of blank stare, unresponsiveness, Orofacial Dyskinesia, Guttural sounds, and hypersalivation lasting approximately 30 seconds. Transient global amnesia. He refers to a similar episode a month ago.

Cranial magnetic resonance imaging without and with intravenous contrast (8ml gadovist) was performed with the usual protocol: sagittal T1 TSE, axial T2 TSE, coronal T2 TSE, axial T2 FLAIR, axial T2 EG and axial diffusion.



A signal alteration centered on the anterior pole of the right temporal lobe of approx. 2.2×2.7×1.7cm (TxAPxCC) associates diffuse cortical thickening and the presence of a heterogeneous lesion with a solid and microcystic component that is hypointense in the T1 sequences and hyperintense in the T2 sequences, it also presents a hyperintensity of the peritumoral signal and an increase in diffusion in DWI sequences without presenting signal drop in the ADC. The perfusion sequences did not show an increase in cerebral perfusion at this level with ADC: 1.3. This lesion presents a heterogeneous contrast uptake, drawing attention to the presence of a solid pole adjacent to the dura that presents intense enhancement, but does not present dural enhancement. These findings may be related to a dysembryogenic neuroepithelial tumor (DNET) or to a Ganglioglioma as the main differential diagnoses. No microbleeds were seen in the gradient echo T2 sequence or calcifications. The rest of the cerebral, cerebellar and brainstem parenchyma show no morphological or signal alterations. Middle line centered. Free basal and perimesencephalic cisterns. Centered ventricular system with preserved ventricular size. The main arterial and intracranial venous vessels show a caliber and signal void within normality. Unoccupied paranasal sinuses and mastoid cells. Slight descent of the cerebellar tonsils not significant (2 mm).

Diagnostic impression:

Heterogeneous lesion centered on the anterior temporal pole of the right temporal lobe with a solid / cystic component and enhancement after contrast administration, with tumor characteristics suggesting a Dysembryoplastic neuroepithelial tumor (DNET) or a ganglioglioma as the main differential diagnoses.

1)
Kurtulan O, Bilginer B, Soylemezoglu F. Challenges in the Intraoperative Consultation of Low-Grade Epilepsy-Associated Neuroepithelial Tumors by Cytomorphology in Squash Preparations. Acta Cytol. 2022 Jan 11:1-7. doi: 10.1159/000521249. Epub ahead of print. PMID: 35016169.
2)
Rau A, Kellner E, Foit NA, Lützen N, Heiland DH, Schulze-Bonhage A, Reisert M, Kiselev VG, Prinz M, Urbach H, Mader I. Discrimination of epileptogenic lesions and perilesional white matter using diffusion tensor magnetic resonance imaging. Neuroradiol J. 2018 Nov 21:1971400918813991. doi: 10.1177/1971400918813991. [Epub ahead of print] PubMed PMID: 30461353.