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MRI of Long-Term Epilepsy-Associated Tumors

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In 20 to 30% of patients with long-term drug-resistant epilepsy neuroepithelial tumors, usually glioneuronal tumors are found. Gangliogliomas and dysembryoplastic neuroepithelial tumors (DNTs) are well characterized, both clinically and on MRI. Both tumor types are located in the cortex or in the cortex and subcortical white matter, gangliogliomas most commonly in the mesial temporal lobe ("around the collateral sulcus"). Both tumor types have typical imaging features, and from both, location and imaging features, they can be usually distinguished from glial tumors. This distinction is important since more than 70% of patients with drug resistant temporal lobe epilepsy caused by gangliogliomas and DNTs get seizure free following extended lesionectomy.
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In 20 to 30% of patients with long-term drug-resistant epilepsy neuroepithelial tumors are found.[1] Clinically, two different groups exist in this cohort. The first contains typical epilepsy-associated tumors such as gangliogliomas, dysembryoplastic neuroepithelial tumors (DNTs), pleomorphic astrocytomas (PXAs), and supratentorial pilocytic astrocytomas, WHO grade I, with usually benign behaviour. The second group consists of diffuse astrocytomas, WHO grade II, oligodendrogliomas, WHO grade II, with a 5-year-survival rate of 50 to 65%, and a few anaplastic cases, classified as WHO grade III, with a median survival time of 2–3 years. Histopathologically, glioneuronal and glial tumors can be distinguished. Among the glioneuronal tumors, gangliogliomas and DNTs are the best known, whilst a new entity designed as angiocentric neuroepithelial tumor (ANET) has been described recently.[2,3] Within the spectrum of glial tumors a so-called isomorphic astrocytoma is associated with a clinically more benign behaviour.[4,5]

Here, we describe the typical imaging features of the long-term epilepsy-associated tumors and discuss important differential diagnoses related to temporal lobe seizures.

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Glioneuronal Tumors

Gangliogliomas

Gangliogliomas are usually benign intraaxial neoplasms that were first described by Perkins in 1926.[6] They are composed of dysplastic neurons and neoplastic glial cells. Both cell populations may show heterogeneity, with the morphological spectrum ranging from a predominantly neuronal phenotype to a predominant glial population. Some gangliogliomas may also exhibit clear cell morphology which makes the differential diagnosis of oligodendrogliomas or DNT difficult. However, the immunohistochemical profile of gangliogliomas (e.g., expression of the stem cell epitope CD34) usually allows a specific diagnosis.[7,8] The vast majority of gangliogliomas correspond to WHO grade I, in a series of 326 gangliogliomas collected from German, Austrian and Swiss neuropathological departments 30 (9%) tumors were classified as WHO grade II and 17 (5%) as WHO grade III tumors, respectively. WHO grade III tumors (anaplastic) were more frequently in extratemporal locations.[7,9]

On MRI, gangliogliomas associated with temporal lobe epilepsy are consistently located in the cortex or in the cortex and subcortical white matter and have a spatial preponderance for the parahippocampal and lateral temporooccipital gyri (Fig.1A-D). The classical imaging features are the combination of intracortical cyst(s), a circumscribed area of cortical (and subcortical) signal increase on FLAIR and T2-weighted images and a contrast enhancing nodule[10] (Fig. 2A-D). Calcifications are present in 1/3 of cases. If contrast enhancement is absent (≈50% of cases), gangliogliomas may be difficult to distinguish from cortical dysplasias. Especially,

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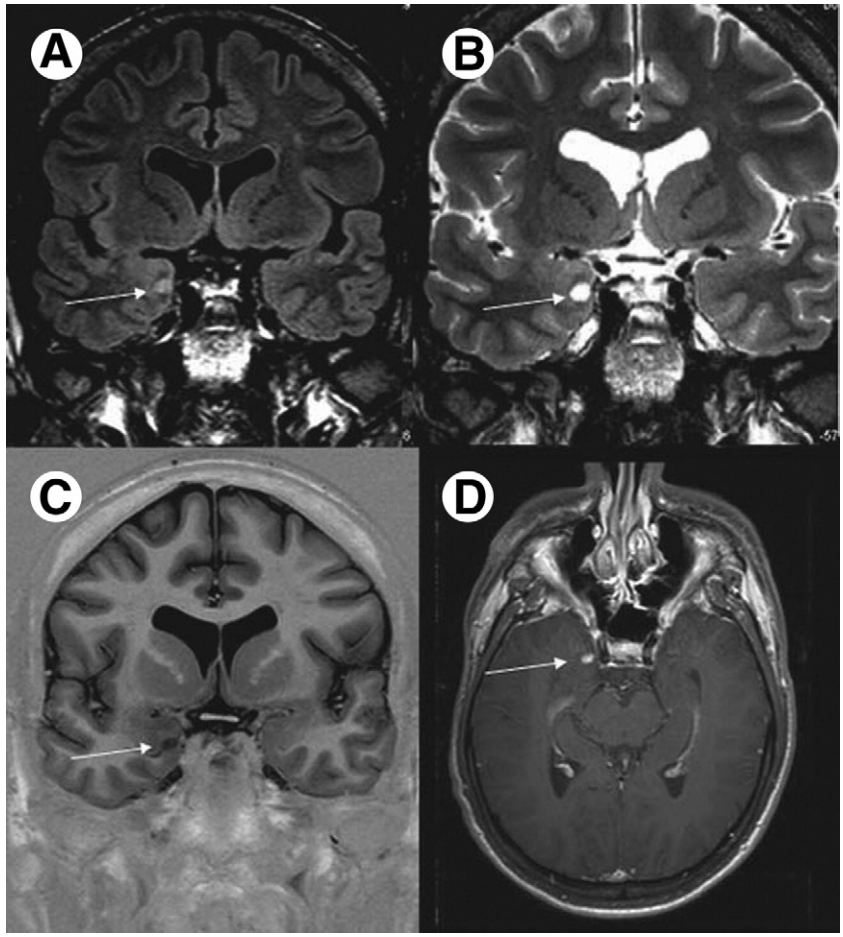

Figure 1 (A-D) Ganglioglioma WHO grade I of the right temporo-polar cortex just anterior to the amygdala (A-D: arrow). The tumor appears cystic on FLAIR TSE (A), T2-weighted TSE (B) and T1-weighted IR (C) images and enhances contrast material (D). Diagnostic clues are the (pure) cortical location, the cystic appearance and the contrast enhancement, by which this lesion can be distinguished from a focal cortical dysplasia, for example. Contrast enhancement, however, is absent in half of the cases.

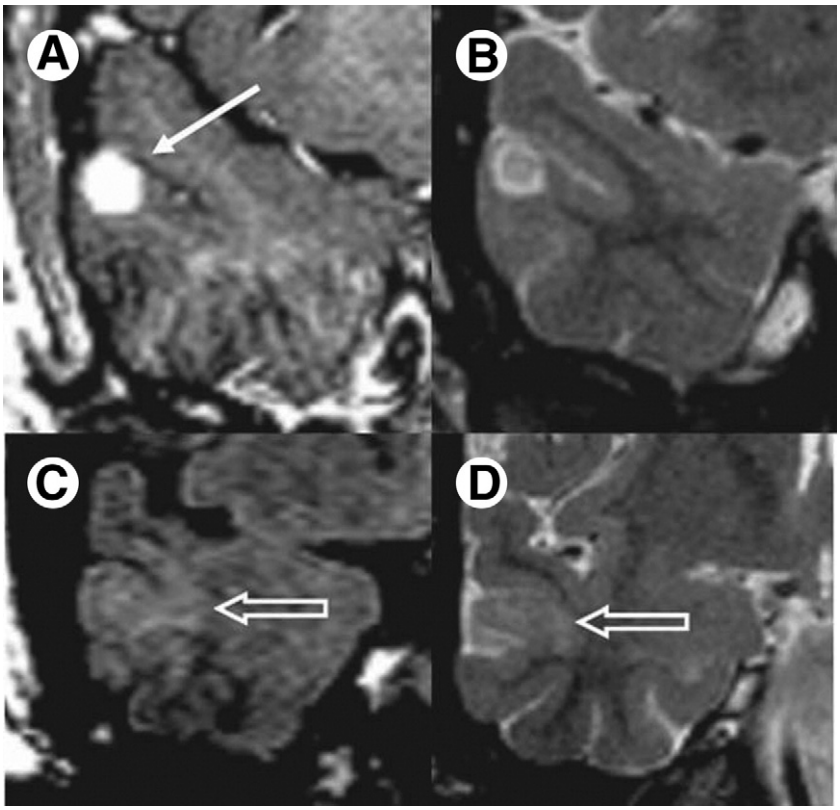

Figure 2 (A-D) Ganglioglioma WHO grade I of the right middle temporal gyrus. (A and B) are T1-weighted contrast enhanced and T2-weighted images at the same position, respectively. (C and D) are FLAIR and T2-weighted images 8 mm posteriorly. Note the coexistence of a contrast enhancing nodule (A: arrow) and FLAIR and T2-hyperintense segments of the cortex and subcortical white matter (C, D: hollow arrow).