EPILEPSY: neoplasms what we should know

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Learning objectives

There are various types of epileptogenic brain tumors, different type has different biological behavior and prognosis. The purpose of this exhibit is:

1. To review the MR findings of epilepsy associated tumors that are typically seen in children and young adults

2. To assess the common and uncommon radiologic imaging findings of each of them

3. To improve the accuracy of the preoperative diagnosis

4. To differentiate between these lesions and high grade cortical astrocytomas

Background

Seizures are a relatively common problem in patients with brain tumors.

Seizures may be initial manifestation of a brain tumor or may occur during the course of disease.

Factors that influence the incidence of seizures include tumor-type and location, for example, among patients with primary brain tumors, seizures are less common with high grade compared with low grade gliomas.

There are typically epilepsy associated tumors, such as, low grade astrocytoma, oligodendroglioma, pleomorphic xanthoastrocytoma and specially glioneural tumors (gangliocytoma, ganglioglioma, dysembryoplastic neuroepithelial...)

Findings and procedure details
Most intraaxial tumors are located in the white matter.

Some tumors, however, spread to or are located in the grey matter. Patients with cortical based tumors includes ganglioglioma, dysembryoplastic neuroepithelial tumor (DNET), pleomorphic xanthoastrocytoma and oligodendroglioma.

It is important to differentiate between these brain tumors and cortical dysplasia. Furthermore, high grade astrocytomas should be included in the differential diagnosis of a cortical lesion in a patient with a new onset epileptic seizure.

**Cortical dysplasia**

Focal cortical dysplasia of taylor (FCDT) is a developmental disorder associated with medically intractable epilepsy. The MR imaging findings (a solitary lesion with sign abnormality and mass effect) can easily confused with a neoplastic process. However, it is important to differentiate both entities, because the decision to operate may depend on the presence or absence of neoplastic tissue. Several MR features are associated with balloon cell FCDT rather than neoplasms, and can help distinguish balloon cell FCDT from neoplasms. These MR characteristic are:

- Grey matter thickening (Fig. 1)
- Homogeneous hyperintense signal in the subcortical white matter (Fig. 2 and Fig. 3)
- Hyperintense signal extension to the lateral ventricle (Fig. 2)
- Frontal lobe (Fig. 1, Fig. 2 and Fig. 3)

**Neuronal tumors**

Tumors of the central nervous system that contain abnormal neuronal elements, termed neuronal tumors, make up 1% of all brain tumors.

Distinguishing neuronal tumors from the more common glial tumors is important because these tumors have favorable clinical outcomes and are generally cured with surgery alone. Neuronal tumors are usually classified as pure neuronal cell tumors and mixed neuronal tumors. We describe the neuronal tumors that are associated with epilepsy: ganglioglioma-gangliocytoma and dysembryoplastic neuroepithelial tumor

**Gangliocytoma-Ganglioglioma** (Fig. 4, Fig. 5, Fig 6 and Fig. 7)
Gangliocytomas are composed of mature neurons, gangliogliomas are composed of a mixed population of ganglion cells and glial cells.

These tumors may occur in all age groups but they are frequently seen in the young with a peak between the age of 10 and 30 years.

Gangliogliomas are the most common of the neuronal-glial neoplasms arising within the central nervous system.

Although any site within the central nervous system may be affected, most tumors develop in the temporal lobe. They are commonly associated with the clinical presentation of medically refractory seizures.

At MR imaging, a well-defined cystic mass with a solid mural nodule is typically seen. However, a solid mass showing nonspecific low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images is also common. Enhanced of the solid portion is variable, ranging from nonenhancing to ringlike to intense homogeneity. There is little associated mass effect or surrounding vasogenic edema.

**Dysembryoplastic neuroepithelial tumor** (Fig. 8, Fig. 9, Fig 10, Fig. 11, Fig. 12, Fig. 13 and Fig. 14)

Dysembryoplastic neuroepithelial tumor is a benign, usually supratentorial, mixed neuronal-glial neoplasm characterized by multinodular architecture and a predominantly intracortical location. The histopathology appearance and the benign clinical course suggest that it may be a malformation rather than neoplastic lesion, furthermore, cortical dysplasia is commonly seen with this tumor.

These tumor virtually always manifest in patients with medically refractory partial seizures. The vast majority of patients are younger than 20 years.

The imaging appearance is similar to other low grade glial tumors, and in some cases it may be impossible to distinguish this tumor from diffuse astrocytoma, ganglioglioma, oligodendrogliaoma, or other low grade neoplasms.

At MR imaging, these tumors manifest as cortical masses that are hipointense on T1-weighted images and hiperintense on T2-weighted images without surrounding vasogenic edema. Some lesions may appear as an enlarged gyrus, producing a soap bubble appearance at the cortical margin. About one-third of dysembryoplastic neuroepithelial tumors enhance following intravenous administration of contrast material.

**Pleomorphic xanthoastrocytoma**
They are rare tumors, accounting for only about 1% of all brain neoplasms, but they are important because they have a characteristic imaging appearance and are highly amenable to surgical extirpation, which may be curative.

Pleomorphic xanthoastrocytoma usually occur in adolescents or young adults. A long history of seizures is very common. An over-whelming majority of tumors occur in the supratentorial brain (most commonly in the temporal lobe).

A cystic supratentorial mass containing a mural nodule that is adjacent to the peripheral leptomeninges is the classic although nonspecific imaging appearance. However, slightly more than half of the lesions reported in the literature occurred in the absence of cystic change.

At MR-imaging, they are usually hypo-to isointense relative to grey matter on T1-weighted images. Involvement of the leptomeninges is highly specific. The solid portion of the tumor enhance intensely following intravenous administration of contrast material.

Despite some variance in the imaging appearance reported in the literature, the peripheral location of these tumors is the single most consistent imaging feature.

**Low grade astrocytoma**

Diffuse low grade gliomas of the cerebral hemispheres are typically diagnosed in young adults.

The most common presenting feature is seizures. Headaches are often also present.

Best diagnostic clue: Focal ot diffuse nonenhancing white matter mass. (Fig.15).

**Oligodendroglioma** (Fig. 16)

Best diagnostic Clue: partially calcified subcortical/cortical mass in middle-aged adult. The most common site is frontal lobe, but, oligodendroglioma may involve temporal, parietal or occipital lobes.

**High grade cortical astrocytomas (HGAs)**

MR findings in HGA are well known, and they include the presence of focal areas, usually solitary, of T1 iso or hypointensity and T2 hyperintensity, with irregular an ill defined margins, mass effect, perilesional vasogenic edema, and marked enhancement with intravenous contrast administration, as well as central necrosis in glioblastomas.
Nonetheless, in the early stages, the radiologic presentation of HGAs can be atypical (fig.17 and fig.18). Due to this, HGA must be considered in the diagnosis of patients with focal lesions of mild /moderate enhancement, especially in the setting of new-onset epileptic seizures (less likely in the case of children and young adults).

The differential diagnosis in these patients must include low grade astrocytoma, cortical dysplasia and neuronal tumors. The presence of contrast enhancement is an unusual finding in LGAs and in focal cortical dysplasias (fig. 1, 2 and 3), but the latter may show enhancement when associated with certain tumors such as DNET or ganglioglioma.

Images for this section:

Fig. 1
Fig. 2
Fig. 4: Ganglioglioma in a 25-year-old woman with a 4-year-history of partial complex seizures. MRI shows a well circumscribed mass appearing within the cortex of the right temporal lobe. T1 C+ MR (fig 5) shows intense enhancement of the mass. A solid nodule vividly enhances. Ganglioglioma was found at resection. Imaging appearance mimics pleomorphic xanthoastrocytoma and pilocytic astrocytoma.
Fig. 5: Ganglioglioma in a 25-year-old woman with a 4-year-history of partial complex seizures. MRI shows a well circumscribed mass appearing within the cortex of the right temporal lobe. T1 C+ MR (fig 5) shows intense enhancement of the mass. A solid nodule vividly enhances. Ganglioglioma was found at resection. Imaging appearance mimics pleomorphic xanthoastrocytoma and pilocytic astrocytoma.
Fig. 6: Ganglioglioma in a 26-year-old girl with seizures. MR images show a mass in the pulvinar nuclei of the thalamus. It demonstrates homogeneous contrast enhancement with only as small amount of surrounding edema.
Fig. 7: Ganglioglioma in a 26-year-old girl with seizures. MR images shows a mass in the pulvinar nuclei of the thalamus. It demonstrates homogeneus contrast enhancement with only as small amount of surrounding edema.
**Fig. 8:** Dysembryoplastic neuroepithelial tumor in a 13-year-old girl with a 8-year history of seizures. (Fig.8) Axial FLAIR MR image shows characteristic bright rim along the borders of a cortically-based wedge shaped DNET.(Fig. 9) Coronal T2-weigheed MR image shows marked high signal intensity of the mass, which extends beyond the normal cortical margin. (Fig.10). Contrast-enhanced axial T1-weighted image shows no evidence of enhancement within the mass.
**Fig. 9:** Dysembryoplastic neuroepithelial tumor in a 13-year-old girl with a 8-year history of seizures. (Fig.8) Axial FLAIR MR image shows characteristic bright rim along the borders of a cortically-based wedge shaped DNET.(Fig. 9) Coronal T2-weigheed MR image show marked high signal intensity of the mass, which extends beyond the normal cortical margin. (Fig.10). Contrast-enhanced axial T1-weighted image shows no evidence of enhancement within the mass.
Fig. 10: Dysembryoplastic neuroepithelial tumor in a 13-year-old girl with a 8-year history of seizures. (Fig.8) Axial FLAIR MR image shows characteristic bright rim along the borders of a cortically-based wedge shaped DNET.(Fig. 9) Coronal T2-weigheed MR image show marked high signal intensity of the mass, which extends beyond the normal cortical margin. (Fig.10). Contrast-enhanced axial T1-weighted image shows no evidence of enhancement within the mass.
**Fig. 11:** DNET in a 45-year-old man with long-standing progressive seizures. FLAIR and T2 weighted images shows wedge shaped, multicystic (bubbly) lesion extending from cortex to ventricle.
**Fig. 12:** DNET in a 45-year-old man with long-standing progressive seizures. FLAIR and T2 weighted images shows wedge shaped, multicystic (bubbly) lesion extending from cortex to ventricle.
**Fig. 13:** DNET in a 45-year-old man with long-standing progressive seizures. FLAIR and T2 weighted images shows wedge shaped, multicystic (bubbly) lesion extending from cortex to ventricle.
**Fig. 14:** Dysembryoplastic neuroepithelial tumor involving the right temporal lobe in a 7-year-old boy with seizures.
**Fig. 15:** A 29-year-old boy with first seizure. Axial T2WI MR shows a diffuse hyperintense white matter frontal lobe mass. Axial T1 C+ MR shows no enhancement of the mass, characteristic of a low grade astrocytoma.
Fig. 16
Fig. 17: A 45-year-old man presents a first complex partial seizure. Axial T2-weighted and contrast enhanced T1-weighted images show a focal lesion located in the right occipital cortex with enhancement after intravenous contrast administration. The histologic diagnosis was glioblastoma.
Fig. 18: A 50-year-old man presents to the emergency room because of a partial seizure with secondary generalization. Fig.17. The MR sequences (FLAIR, T2-weighted and contrast enhanced T1) performed show a focal lesion in the left parieto-temporal lobe with enhancement after intravenous administration of contrast. The perfusion sequence showed increased cerebral blood flow. Fig 18. Follow-up MRI study performed 3 months later, shows marked tumor growth and characteristics sign of high grade glioma.
**Fig. 19:** A 50-year-old man presents to the emergency room because of a partial seizure with secondary generalization. Fig. 17. The MR sequences (FLAIR, T2-weighted and contrast enhanced T1) performed show a focal lesion in the left parieto-temporal lobe with enhancement after intravenous administration of contrast. The perfusion sequence showed increased cerebral blood flow. Fig 18. Follow-up MRI study performed 3 months later, shows marked tumor growth and characteristics sign of high grade glioma.
Conclusion

The superficial gliomas that are peripherally located and involve the cortical gray matter are remarkable because of their generally favorable prognosis following surgical resection. Many of these special cases of neuroepithelial tumors are associated with medically refractory seizures.

They tend to manifest in younger patients with long history of seizures.

Due to this, detection and correct interpretation of the imaging appearance of these neoplasms of central nervous system assume prime importance.

However, high grade gliomas must be included in the differential diagnosis of any cortical lesion with mild or moderate enhancement on MRI, in elder patients with new-onset seizure.

Personal information

References


