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Neuroradiology / Neuropathology Conference

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Case 1

- 3 month old baby girl
- Presented with new onset of seizures
Newborn. Questionable blurring of the gray-white junction within the right occipital lobe. “Findings suspicious for heterotopic gray matter adjacent to the posterior aspect of the temporal horn of the right lateral ventricle.”
4 year-old. Unchanged from prior exam.
• **Diagnosis:**
  - Focal cortical right occipital lobe dysplasia

• **EEG** – seizures originating electrographically from the right parieto-occipital region.

• **Surgery (intractable complex seizures):** right partial occipital lobectomy including the most inferior occipital and mesial region plus lesionectomy.
Molecular layer neurons

Neuronal microcolumns (Radial and tangential)
Cortical Dysplasia - MRI

- Focal cortical thickening or thinning, focal brain atrophy, blurring of the gray-white matter junction, increased T2/FLAIR signal in the gray and subcortical white matter, decreased T1 signal in the subcortical white matter, and abnormal sulcal or gyral pattern.

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<th>Type I – Temporal lobe; adults; mild symptoms</th>
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<td>▪ Significant segmental or lobar hypoplasia/atrophy often coexistent with reduced volume of subcortical white matter. Hippocampal atrophy is frequently seen.</td>
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<td>▪ There are no essential differences between type Ia and Ib. Type Ib is more often located outside of the temporal lobe and in this cases difficult to differentiate it from type II.</td>
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<th>Type II – Extra-temporal (frontal lobe); children; more severe symptoms</th>
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<td>▪ Cortical thickening, marked blurring of GM/WM junction, and abnormal WM signal is often extended towards the ventricle (transmantle sign), which is observed exclusively in FCD type II. PVS may also be enlarged.</td>
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Cortical Dysplasia

- MRI may be normal, particularly in type I and the lesion may be smaller than the seizure-generating region seen in the EEG.
  - MRI- PET (reduced FDG metabolism); Ictal SPECT (may localize the dysplastic focus), Magnetoencephalography (is able to differentiate type I and II and also localizes anatomically the functional areas: sensory, motor and speech).
  - DTI may be helpful in determining the microstructure of affected brain and fiber tract.
  - Intracranial EEG can be performed if methods described above are insufficient to determine the epileptogenic focus.
Case 2

• 4 year old boy
• Presented with:
  – Lesion in the pons seen on outside brain CT
  – History of left esotropia for 2 weeks
Heterogeneous hypo T1 /hyper T2 SI mass expanding the pons extending to the left midbrain. No hydrocephalus (IV ventricle remains patent). No tonsillar or transtentorial herniation. Normal MRA with mild mass effect on the inferior basilar artery and V4 segments of both vertebral arteries.
Some areas of low ADC. No hemorrhage or calcifications are seen on SWI. Heterogeneous enhancement along the inferior (more exophytic) portion of the lesion.
• Differential diagnosis:
  – Diffuse Intrinsic  Pontine Glioma
  – ?
  – Infection, ADEM, Osmotic demyelination

• Surgery (11/14/2014): posterior fossa decompression

• Radiation therapy
Suboccipital craniectomy. Decreased size. Increased degree of enhancement that could represent radiation necrosis or tumor progression.
Stable size. A focus of enhancement along the floor of the IV ventricle slightly increased. Otherwise the heterogeneous enhancement is unchanged.
Diffuse Intrinsic Pontine Glioma
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Diffuse Intrinsic Pontine Glioma

• Brainstem gliomas are divided into 2 categories:
  – 20% are considered to be low grade, arising in the midbrain, medulla, or dorsal pons, and are often focal, dorsally exophytic, and amenable to resection with a relatively good prognosis.
  – The remaining 80% arise and occupy the majority of the pons and are diffuse in nature with poor prognosis - Diffuse intrinsic pontine glioma

• DIPG typically present in childhood (3 to 10 years of age)

• MRI findings:
  – Pontine enlargement with tumor centered in and involving more than 50-70% of the cross sectional area of the pons.
  – Hypointense on T1- and hyperintense on T2-WI with slight or absence contrast enhancement. It can enhance post radiotherapy.
  – DWI: usually normal, occasionally mildly restricted.
  – Anterior exophytic extension with “embracement” of the basilar artery and extension into the midbrain and middle cerebellar peduncles are common.
Diffuse Intrinsic Pontine Glioma

• The prognosis is generally poor; a subset of these tumors is histologically low grade at initial clinical presentation and rapidly evolves into high-grade.

• MRI prognostic findings:
  – Areas of low T2 signal, enhancement and restricted DWI represent focal anaplasia. rCBV values of these areas were notably higher (increased vascularity).
  – The T2 hypointensity itself, rCBV values higher than those of GM, and high FA and low ADC values correlated with a worse clinical outcome.
  – FA, ADC and Perfusion may also be a useful for the assessment of treatment response and tumor progression.

Case 3

• 8 year old girl
• Presented with:
  – Fell at school and hit her head followed by vomiting, without loss of consciousness.
  – Frontal headache and intermittent vomiting for the last couple of weeks.
Large heavily calcified right frontal mass extending into the anterior lateral ventricles with marked hydrocephalus (obstruction at the foramen of Monro) with transependymal flow of CSF. Erosion and mass extension through the adjacent right frontal skull into the subcutaneous soft tissues. Severe right to left midline shift.
Large calcified right frontal lobe mass extending into the right lateral and III ventricles. Hydrocephalus and midline shift. Restricted DWI.
Minimal enhancement on Post contrast T1-WI. Normal MRA and MRV.
• **Differential diagnosis:**
  – Aggressive primary brain neoplasm: PNET, ATRT, sarcoma, GB, undifferentiated.

• **Surgery (03/28/2015) and RT**
  – Operative findings: Large left frontal mass with areas of soft easily suctioned tumor, and areas of white caseating-type necrosis and areas of some hypervascularity; the tumor appeared to be invasive into the surrounding pia at multiple areas; likely gross total resection.
Malignant Glio or glioneuronal tumor?

- Three glioneuronal tumors with slightly different clinical, radiologic, and pathologic findings:

1- Papillary glioneuronal tumor:
   - Low-grade tumor (variant of ganglioglioma).
   - Well circumscribed, may be located in any lobe (with a slight predilection for the temporal), and frequently lies near a lateral ventricle.
   - Peritumoral edema and mass effect are generally mild, and some cases are focally calcified.

2- Rossetted glioneuronal tumor (glioneuronal tumor with neuropil-like islands):
   - One or more cerebral lobes involved by a T1-hypointense, T2-hyperintense mass with variable mass effect and edema (findings all common for diffuse glioma).
   - Contrast enhancement usually minimal at diagnosis, may become conspicuous with tumor progression.
Malignant Glio or glioneuronal tumor?

3- Rosette-forming glioneuronal tumor of the fourth ventricle
   – Midline masses in the fourth ventricle or cerebral aqueduct, commonly involving the cerebellum (especially vermis).
   – Relatively circumscribed and often partially cystic with heterogeneous contrast enhancement.
   – There is often involvement of surrounding periventricular and periaqueductal tissue such as dorsal pons, midbrain, thalamus, or pineal region, and multinodularity or multicentricity may be observed.

• Some anaplastic tumors with histologic mixtures of neoplastic neuronal, neurocytic, and astrocytic forms in various proportions are named “malignant mixed glioneuronal tumor”.
Case 4

• 22 year old female
• Presented with:
  – Outside CT performed after a motor vehicle accident.
  – CT: No acute findings. Possible IV ventricle lesion.
Heterogeneously enhancing mass within the fourth ventricle. No hydrocephalus.
• **Differential diagnosis:**
  – Ependymoma
  – Subependymoma
  – Choroid plexus tumor

• **Surgery (05/21/2015)**
  – Soft tannish tumor with good plane between it and the fourth ventricular surface; likely gross total resection.
Ependymoma WHO II

Smear

Frozen
Ependymoma

- Ependymomas are generally well-circumscribed lesions that occur supra-tentorially (40%) or in the posterior fossa (60%). When supra-tentorial most are intra-parenchymal.
- Young patients / children: posterior fossa (IV ventricle).
- Ependymomas involving the fourth ventricle tend to fill the ventricle and may extend through the foramina of Luschka, foramen of Magendie, or foramen magnum.
- Frequently demonstrate cystic components and calcification. Occasionally, intratumoral hemorrhage.
- Restricted diffusion in the solid components may be seen, especially in anaplastic tumors. DWI should be interpreted with caution (hemorrhage or calcification).
- Intra-ventricular ependymomas are associated with a small risk of spread throughout the CSF and imaging of the entire neuroaxis should be performed to exclude dissemination.
- Surgical resection is often difficult due to adherence and the infiltrating nature of the tumor.
- Poor prognostic factors include location in the 4th ventricle, anaplastic variant and incomplete resection.