Temporal Encephaloceles: Evidence of Epileptogenicity
Zeke Campbell, M.D.
Assistant Professor

Disclosures
• There are no relevant disclosures for this presentation

Objectives
• To understand the meaning and characteristics of encephaloceles
• To understand the relationship between temporal encephaloceles and epilepsy
• To understand the potential reasons for increased prevalence of encephaloceles in patients with epilepsy

Case

Case #1
• 37-year-old right-handed woman with a history of migraines, sickle cell trait
• Seizures began at age 32
• Epilepsy risk factors: normal birth and development; no major CNS injury/infections, no febrile seizures
• Current meds: zonisamide 300 mg qhs, oxcarbazepine 1200 mg bid
• Prior AEDs: levetiracetam
• Comorbid conditions: Depression, following with a psychiatrist

Semiology
Typical events:
• Aura: “rush” sensation over her face, sweating, lightheadedness
• Ictus: impairment of awareness with oral and manual automatisms. Events sometimes progress to increased tonicity in all extremities and bilateral convulsions that last approximately one minute
• Post-ictal: fatigued and confused for hours
• 2-3 times per month

Other occasional events during which she will hear people talking but cannot respond
Exam

- Obese (BMI 38)
- Vital signs within normal limits
- Unremarkable general exam
- Neuro exam non-focal

Initial Seizure

Initial Seizure Work-up

- Three separate EMU admissions:
  - Interictal: left anterior temporal sharp waves
  - Three left temporal seizures (see figure)
  - Two nonepileptic events
  - SPECT injection performed during a nonepileptic event (increased perfusion in the right frontal cortex)
- PET: no definitive seizure focus

EEG

EEG in bipolar montage: sensitivity 10 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

Patient feels "off"  EEG seizure begins  "Don't, don't, don't..."

EEG

EEG in bipolar montage: sensitivity 10 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

Oral automatisms
EEG

**EEG in bipolar montage:**
- Sensitivity: 10 µV/mm
- Low frequency filter at 1 Hz
- High frequency filter at 70 Hz

**Oral automatisms continue**

**EEG seizure ends**

MRI

**T2 SPACE (A) axial and (B) sagittal sections prior to LiTT**

Images courtesy Milad Yazdani, MD

**Treatment Recommendations**

- **Wada:**
  - Left hemisphere representation of language
  - Bilateral representation of memory
- **Multidisciplinary refractory epilepsy conference (REC)** → candidate for LiTT

MRI

- Initially interpreted as normal
- Later reviewed and determined to have subtle increased T2 signal in the left hippocampus

MRIs courtesy Milad Yazdani, MD
Post-ablation Course

- Patient continued to have 4-8 seizures per month for 6 months following LiTT
- Re-evaluated in the EMU:
  - 6 left temporal seizures
  - 2 nonepileptic events
  - 5 events that were poorly-visualized or were not associated with impairment of awareness

EEG in bipolar montage: sensitivity 10 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

"No," when asked if OK
EEG

EEG in bipolar montage: sensitivity 10 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

Moaning intensifies

EEG

EEG in bipolar montage: sensitivity 20 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

Oral automatisms

Grunting

EEG

EEG seizure ends

Treatment

Recommendations

- Case re-discussed → Proceed with left ATL

EEG

EEG in bipolar montage: sensitivity 10 µV/mm; low frequency filter at 1 Hz; high frequency filter 70 Hz.

EEG seizure ends

MRI – post-ATL

Postoperative Course

- Following the operation: right upper quadrantanopsia, memory loss, and occasional HAs
- Patient continues to be seizure-free for the last 18 months since her ATL
- She continues to follow with psychiatry
Temporal Encephaloceles

What are encephaloceles?

- Herniations of the brain through dura mater and skull
  - Acquired or congenital
  - Asymptomatic or result in varying pathologies based on anatomic location

Spontaneous encephaloceles

- Result in the absence of trauma, iatrogenesis, neoplasms, or inflammatory conditions
- Noted association with IIH
- 67% with TE and CSF leak → elevated ICP
- Higher average BMIs

Temporal lobe encephaloceles

- Involve the middle cranial fossa
- Associations:
  - Recurrent meningitis
  - CSF fistulas
  - Otitis
  - Hearing loss
  - Drug-resistant epilepsy

How common are encephaloceles?

- 6% to 34% of temporal bones show dehiscence involving the mastoid tegmen
  - 1% to 6% revealing multiple dehiscences
- Clinically relevant encephaloceles – ~1 in 35,000
- TEs represent an even more uncommon subset of an already rare disease

Improved detection of TEs

- 23 patients with TEs
  - 0.3% of MRIs in newly-diagnosed patients with epilepsy
  - 1.9% in drug-resistant patients
- Epilepsy surgery because of encephaloceles accounted for 10% of temporal lobe resections
Discrepancy in prevalence

- Obesity in the developing world → IHH → skull defects → small encephaloceles
- Other possible factors include referral patterns and surgical selection process
- Improvements in imaging technology

Small TEs in epilepsy

- 22 patients with STPEs
- 9.6% of patients with temporal lobe epilepsy (TLE)
- 0.5% of those with extra-TLE
- STPEs in patients with TLE:
  - Initial MRI study reported as normal – 23.3%
  - MRI-visible lesions – 1.4%

Cases of TEs with Epilepsy in the Literature

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<tbody>
<tr>
<td>Cases</td>
<td>12</td>
<td>7</td>
<td>4</td>
<td>4</td>
<td>30</td>
<td>28</td>
<td></td>
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</table>

Imaging and TE detection

- TEs confirmed after “close inspection” of previously normal 3T MRI or high-res CT
- TE on 7T MRI after initial 3T MRI was interpreted as normal
- Majority of cases prior to 2014 were noted incidentally at the time of surgery

Detection and characteristics of TEs

- MRIs in patients with refractory epilepsy over a 7-year period reviewed by an expert neuroradiologist
- 418 patients with available MRI:
  - 7 had TEs reported on initial imaging
  - 52 (12.5%) had TEs on expert review

Demographics

<table>
<thead>
<tr>
<th></th>
<th>Total</th>
<th>TE (N = 32)</th>
<th>Without TE (N = 356)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>AGE</td>
<td>43.3 ± 13.5</td>
<td>43.3 ± 12.2</td>
<td>43.3 ± 13.6</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>BMI</td>
<td>28.7 (24.1, 35.4)</td>
<td>36.9 (30.0, 40.7)</td>
<td>27.7 (23.9, 33.7)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Age at Onset</td>
<td>15.0 (6.0, 29.0)</td>
<td>35.0 (29.0, 45.0)</td>
<td>14.0 (5.0, 22.0)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Years with Epilepsy</td>
<td>16.0 (7.0, 27.0)</td>
<td>4.5 (2.0, 5.1)</td>
<td>17.0 (9.0, 28.0)</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Male</td>
<td>176 (42.1%)</td>
<td>9 (17.3%)</td>
<td>167 (45.6%)</td>
<td>&lt;0.001</td>
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Radiologic Findings

<table>
<thead>
<tr>
<th>Normal</th>
<th>MRI</th>
<th>MRI Magn + 3T</th>
<th>SPACE was used</th>
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</thead>
<tbody>
<tr>
<td>Total</td>
<td>TE</td>
<td>Without TE</td>
<td>p</td>
</tr>
<tr>
<td></td>
<td>N = 52</td>
<td>N = 366</td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>157 (37.6%)</td>
<td>31 (90.6%)</td>
<td>126 (37.7%)</td>
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<tr>
<td>MRI</td>
<td>341 (81.8%)</td>
<td>50 (96.2%)</td>
<td>291 (79.7%)</td>
</tr>
<tr>
<td>MRI Magn + 3T</td>
<td>175 (41.9%)</td>
<td>48 (92.3%)</td>
<td>126 (34.7%)</td>
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<tr>
<td>SPACE was used</td>
<td>11 (2.6%)</td>
<td>5 (9.6%)</td>
<td>6 (1.8%)</td>
</tr>
<tr>
<td>Abnormal</td>
<td>98 (23.4%)</td>
<td>48 (92.3%)</td>
<td>50 (13.7%)</td>
</tr>
<tr>
<td>Abnormal sella turcica</td>
<td>9 (2.2%)</td>
<td>6 (11.5%)</td>
<td>3 (0.8%)</td>
</tr>
</tbody>
</table>


Seizure Localization

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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>N = 250</td>
<td>N = 326</td>
<td>0.06**</td>
</tr>
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<td>Total</td>
<td>250</td>
<td>326</td>
<td>0.06**</td>
</tr>
<tr>
<td>Temporal</td>
<td>223 (57.6%)</td>
<td>46 (90.2%)</td>
<td>177 (52.7%)</td>
</tr>
<tr>
<td>Non-localizable</td>
<td>109 (26.4%)</td>
<td>5 (9.8%)</td>
<td>94 (28.9%)</td>
</tr>
<tr>
<td>Non-temporal</td>
<td>62 (16.0%)</td>
<td>0 (0.0%)</td>
<td>62 (18.0%)</td>
</tr>
<tr>
<td>No-Seizures</td>
<td>1 (1.9%)</td>
<td>1 (12.5%)</td>
<td>0 (0.0%)</td>
</tr>
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<td>Temporal</td>
<td>250</td>
<td>326</td>
<td>0.06**</td>
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<td>1 (12.5%)</td>
<td>0 (0.0%)</td>
</tr>
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Conclusion

- Careful inspection of MRI with special attention to high-resolution T2 sequence in patients with RTLE by an experienced, board-certified neuroradiologist can increase the detection of subtle TEs


Intraoperative ECoG in TEs

- 9 patients with TE
- 6 underwent intraoperative ECoG and resection
  - Interictal EDs arising from the region of the TE in all cases (6/6)
  - Interictal EDs from HCP in 4/5
  - When seizures were recorded, the TE was always involved at onset but with synchronous or rapid spread to the HCP in both cases


Summary

- TEs are seen in 2 to 12.5% of patients with refractory epilepsy and more commonly seen in patients with TLE
- TE detection can be improved by careful review of high-resolution T2 MRI by an experienced neuroradiologist
- Some patients may achieve seizure freedom after lesionectomy, even when the mesial temporal structures are not resected


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References