AMSER Case of the Month
December 2023

67-year-old female presenting with complex partial seizure and subsequent word finding difficulty, memory difficulties, generalized weakness, and falls

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Patient Presentation

- **Chief Complaint:** Initially presented to ED with seizure and neck jerking lasting one minute in duration. Non-contrast CT head obtained, Keppra was started, and patient lost to outpatient follow-up. Over the course of two years had begun to develop generalized weakness, falls, and word finding difficulties.

- **Past medical history:** Breast cancer (IDC), squamous cell carcinoma of the nose, obstructive sleep apnea, hypertension, hyperlipidemia, transient ischemic attack,

- **Past surgical history:** Left breast lumpectomy with axillary lymph node biopsy

- **Medications:** Tamoxifen, Keppra, Lisinopril, pantoprazole, atorvastatin
Pertinent Labs

- Basic Metabolic Panel (BMP) had no abnormal findings
- Complete Blood Count (CBC) had no abnormal findings
- Vitamin B1/B12/E are within normal limits
- Microbiological workup was negative
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria [1]

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>CT head without IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI head without IV contrast</td>
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<td>O</td>
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<td>MRI head without and with IV contrast</td>
<td>May Be Appropriate</td>
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<tr>
<td>FDG-PET/CT brain</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>MEG</td>
<td>Usually Not Appropriate</td>
<td>O</td>
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<tr>
<td>MRI functional (fMRI) head without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
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<tr>
<td>HMPAO SPECT or SPECT/CT brain ictal and interictal</td>
<td>Usually Not Appropriate</td>
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This imaging modality was ordered by the ED physician.
Select the applicable ACR Appropriateness Criteria [2]

**Variant 4:** Persistent or worsening mental status change despite clinical management of the suspected underlying cause (intoxication, medication-related, hypoglycemia, sepsis, etc) or acute change in mental status of unknown cause. Initial imaging.

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This imaging modality was ordered by the neurologist.
Findings (unlabeled)

Head CT Without IV Contrast
Findings (unlabeled)
Findings (unlabeled)

T1 Pre-contrast

T1 Post-contrast
Findings (unlabeled)
Findings: (labeled)

Head CT Without IV Contrast

Low density, extra-axial cystic mass present in the left superior frontal region with mild mass effect on underlying brain parenchyma and sulcal effacement. No IV contrast was used so cannot comment on degree of enhancement.
Findings: (labeled)

No diffusion restriction seen on DWI (b1000) and ADC Map.
Findings: (labeled)

Multi-septate cystic extra-axial mass with incomplete intra-cystic FLAIR suppression.
Multi-septate cystic mass with no evidence of intra-cystic or rim-enhancement on T1 contrast image.

Findings: (labeled)
Findings: (labeled)

T2 Coronal again demonstrates an extra-axial, multi-septate cystic mass displacing the gray-white matter junction and with midline shift.
Final Dx:

Neurenteric (Endodermal) Cyst
Lesion was biopsied!

**Final Diagnosis and Attending Signature**

A. Brain, "Cyst Wall", Excision: *Endodermal Cyst.* (See comment.)

B. Brain, "Flare Change", Biopsy: *Endodermal Cyst.*

C. Brain, Thickened Tissue Over Cortex, Biopsy: *Endodermal Cyst.*

D. Brain, Thickened Tissue Over Cortex, Biopsy: *Endodermal Cyst.*

**Final Diagnosis Comment**

Histological examination of the biopsies received in this case reveal cyst walls lined by a single layer of low cuboidal cells, best visualized in block A1. Immunohistochemical staining is performed on this block (A1) for cytokeratin 7 (CK7) and epithelial membrane antigen (EMA), demonstrating strong immunoreactivity in the cells lining the cyst (positive) for both stains. In the setting of a differential diagnosis of endodermal cyst versus subarachnoid meningotheelial cyst, this staining pattern confirms the diagnosis of an endodermal cyst.
Neurenteric cysts are rare congenital lesions with endodermal origin\(^1\)
- Typically occur in the subdural space of the spinal cord
- Very rarely can occur in an intra-parenchymal location

**Supratentorial cystic lesion differential diagnosis\(^2\)**

**Intra-axial**
- Dysembryoplastic neuroepithelial tumor (DNET)
- Ganglioglioma
- Multinodular and vacuolating neuronal tumor
- Abscess
- Pleomorphic xanthoastrocytoma

**Extra-axial**
- Epidermoid cyst
- Arachnoid Cyst
- Schwannoma (cystic)
Case Discussion (3)

- Imaging findings\(^2\)
  - **CT Scan:** will demonstrate hypo to hyperdense mass with no calcium or hemorrhage
  - **MRI**
    - T1 Weighted Imaging will demonstrate a cystic, extra-axial mass iso or hyperintense to CSF
    - T2 Weighted Imaging and T2 Flair will typically show a cystic mass with hyperintensity to CSF
    - DWI: Demonstrates no diffusion!
    - T1 Weighted Imaging with Contrast may show rim enhancement but usually does not enhance
References:
