HPI: 38 y/o M with chronic brain hemorrhage, persistent fevers, and non-improving neurological examination

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Patient Presentation

- **HPI:** 38 y/o M with previous severe traumatic brain injury from assault ~1 year ago and subsequent left-sided acquired skull defect, now with chronic brain hemorrhage s/p cranioplasty and post-op epidural hematoma, herniation, and duret hemorrhage requiring second operation, s/p septic shock post-op now resolved, now with no improvement in neurological status post stabilization
- **ROS:** persistent fevers, unresponsive, unable to obtain further ROS
- **PMHx:** Hemophilia B
- **PSHx:** emergent hemicraniectomy and drainage following traumatic brain injury, cranioplasty, emergent evacuation of epidural hematoma and replacement of PEEK cranioplasty
- **Family Hx:** positive for hemophilia
- **SocHx:** prior hx of smoking (1 ppd), illicit drug use, and mild alcohol use
- **Pertinent Physical Exam Findings:**
  - Neuro: tracheostomy in place, does not open eyes spontaneously or to pain, no vocalization, pupils sluggish bilaterally but equal, minimal movement to noxious stimuli, not improving
Pertinent Labs

• CBC
  • WBC 4.63, Hgb 8.2 (L), Hct 28.4 (L), Plt 74 (L)
  • MCV 89.6, RDW 21.4 (H)
• BMP
  • Na 136, K 4.2, Cl 101, HCO3 22, BUN 19, Cr 0.65 (L), Glu 116 (H)
  • Anion gap 13
  • Ca 8.7
  • Mg 2.1
  • PO4 3.2
• PT 14.9 (H)
What Imaging Should We Order?
# Acute Mental Status Change, Delirium, and New Onset Psychosis

Select the applicable ACR Appropriateness Criteria

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<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tr>
<td>MRI head without and with IV contrast</td>
<td>Usually Appropriate</td>
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<td>CT head with IV contrast</td>
<td>Usually Not Appropriate</td>
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This imaging modality was ordered
Findings (unlabeled)

Axial T2 MRI

Our Patient

Normal Comparison
Findings (labeled)

Explanation: T2 hyperintensity found in bilateral inferior olivary nuclei of the medulla (bright white signal within red circles). Normal comparison shows homogenous grey.
Final Dx:

Bilateral Hypertrophic Olivary Degeneration
Background

- Hypertrophic Olivary Degeneration (HOD) is a rare condition that is characterized by a unique pattern of trans-synaptic degeneration.
- Lesion occurs in the triangle of Guillain and Mollaret.
  - Corners of the triangle include: Red nucleus, Inferior olivary nucleus, Contralateral dentate nucleus.
- Interruption of connections within the triangle of Guillain and Mollaret results in hypertrophy of inferior olivary nucleus.
- Etiologies vary: posterior fossa surgery, tumor, hemorrhage, traumatic brain injury, etc.
Clinical Presentation & Treatment

- Patients are frequently asymptomatic when diagnosed
- Diagnosis commonly made incidentally with routine imaging surveillance
- If symptoms develop, they are commonly related to cerebellar dysfunction:
  - palatal tremor (most common ~20-45% of cases)
  - dentato-rubral tremor
  - ocular myoclonus
- Usually self-limited, but treatment and prognosis ultimately related to etiology
- Medical treatment can be considered for symptoms like palatal tremor (anti-seizure meds), but usually deferred due to limited effectiveness/side effects
Radiographic Features

- Radiographic features typically do not present until several months after initial insult
- MRI with T2-weighted and T2-FLAIR sequences are the preferred method of visualization
- T2 signal increases several months after insult and can be seen for 3-4 years on imaging
- Hypertrophy occurs about 1 year after insult and can be seen for 3-4 years on imaging
- Three stages:
  - T2 hyperintensity without olivary swelling
  - T2 hyperintensity and olivary swelling
  - Olivary swelling subsides but T2 hyperintensity persists
References:

