AMSER Case of the Month February 2022

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?

Select the applicable ACR Appropriateness Criteria

Acute Mental Status Change, Delirium, and New Onset Psychosis

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.**

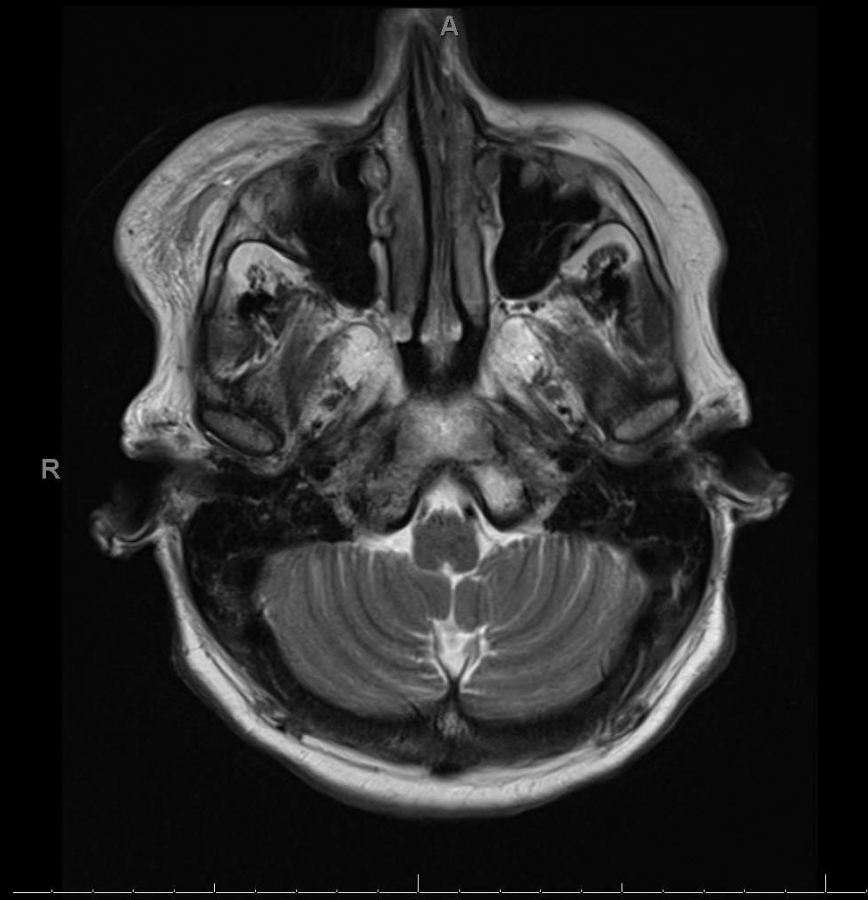
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢ ☢ ☢
CT head without and with IV contrast	May Be Appropriate	☢ ☢ ☢
CT head with IV contrast	Usually Not Appropriate	☢ ☢ ☢

This imaging modality was ordered

Findings (unlabeled)

Axial T2 MRI

Our Patient



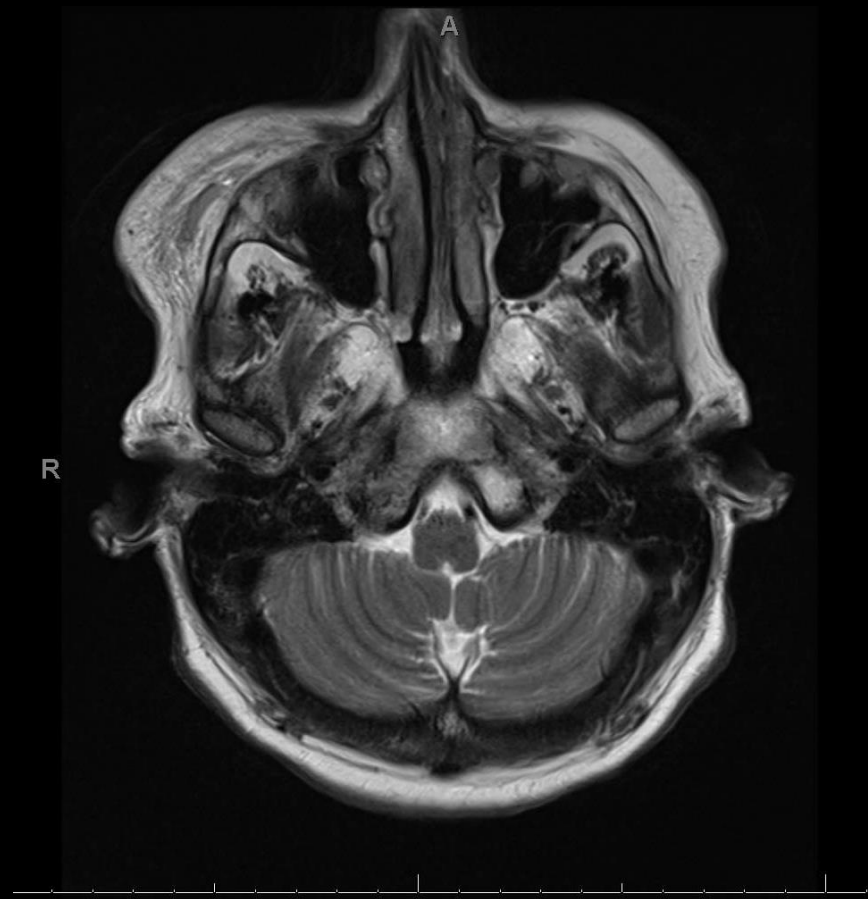
Normal Comparison

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.

Axial T2 MRI

Our Patient



Normal Comparison

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

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