Primary CNS Lymphoma Masquerading as Autoimmune CNS Vasculitis on MRI

Case Exhibit

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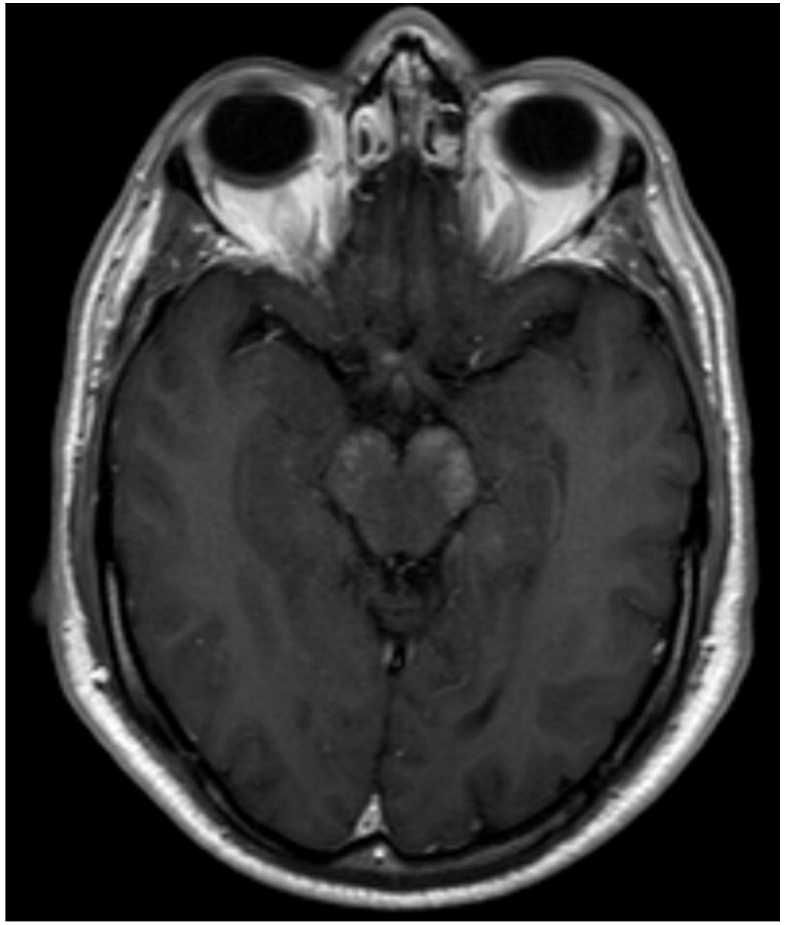
Initial Clinical Presentation

- 61 yo Iranian M w/ history of migraines and HLD presenting w/ 4 weeks of L facial numbness, diplopia, tinnitus and R handgrip weakness
- Physical exam
  - CN
    - CN2: diplopia looking to left
    - CN3, 4, 6: left lateral gaze palsy
    - CN5: left sided upper facial numbness
  - Motor: R handgrip weakenss
  - Sensation: SILT b/l UE
- Admitted to neurology for LP w/ full workup including opening pressure, cell count, protein, glucose, IgG index, oligoclonal bands, gram stain…
- CSF: 6 nucleated cells, protein 30, absent oligoconal bands, negative meningitis/encephalitis panel, negative paraneoplastic panel, normal CSF ACE 1.7

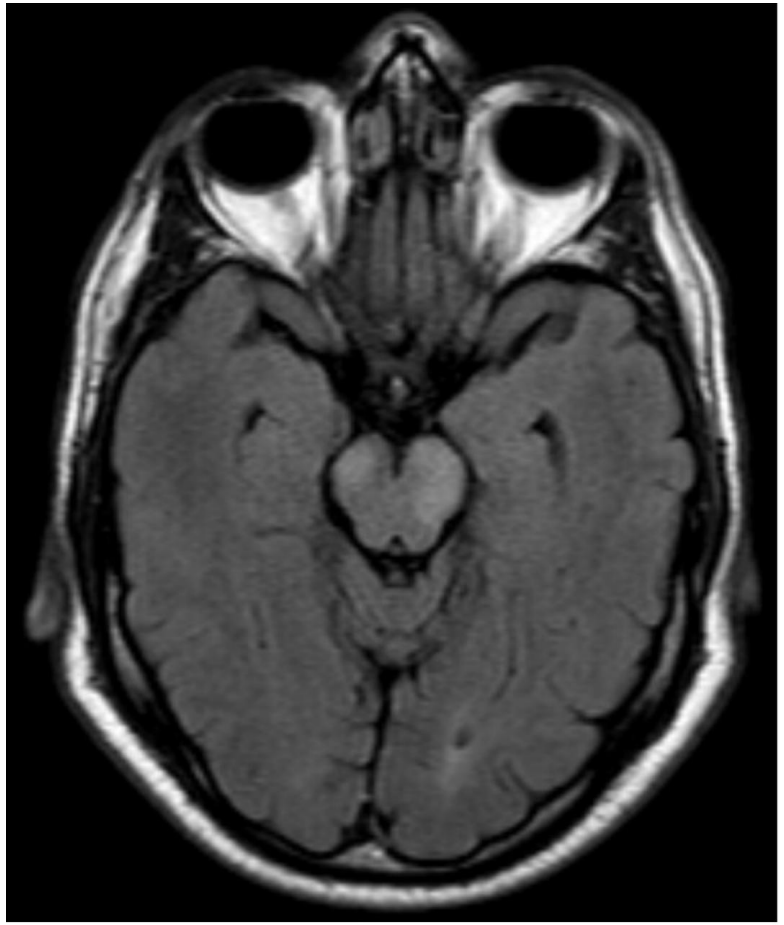
Initial Clinical Presentation

- DDx: CLIPPERS, Behcet's, paraneoplastic syndrome, neuroimmunological lesions, or infectious/metabolic etiologies
- Given MRI findings and of Iranian descent, concern for neuroinflammatory process (Behcet's) and treated with 3d IV Solu-Medrol
- Ocular symptoms resolved, other symptoms persisted

Initial Presentation



Axial T1 Post-contrast



T2 FLAIR

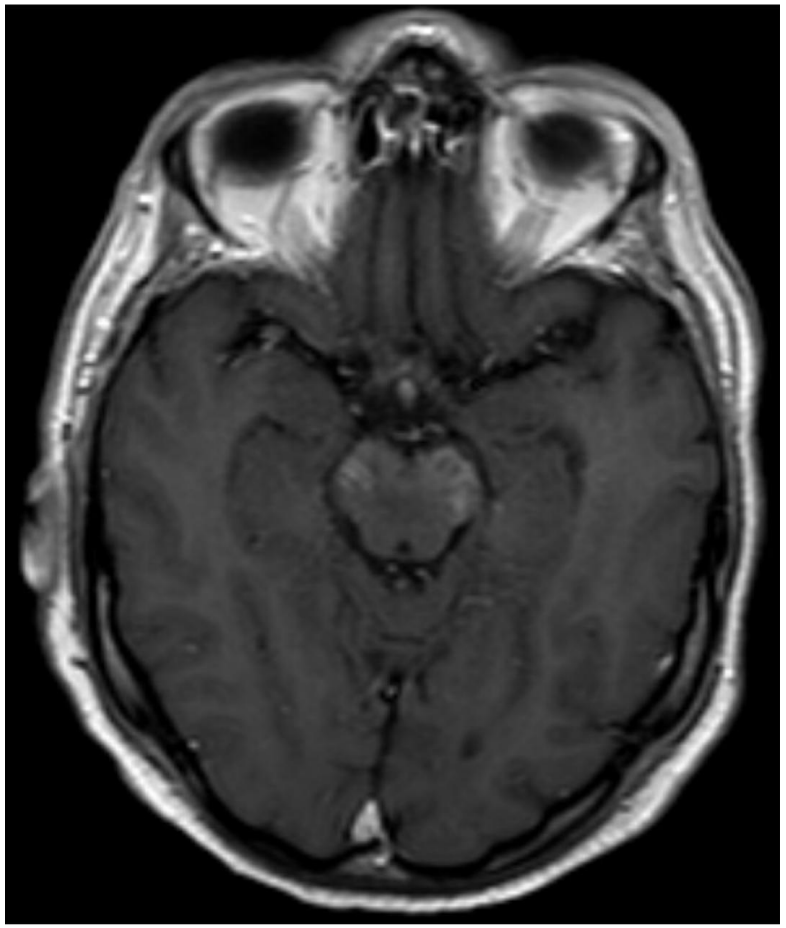
Follow-up and further testing

- Seen by rheumatology and neurology outpatient
  - Could not definitively diagnose Behcet's (eg. patient had no history of ulcers)
- Symptoms persisted and without improvement on repeat imaging
- NSGY consulted for possible biopsy, but no lesion amenable to biopsy
- Admitted for 5 days of PLEX with slight improvement

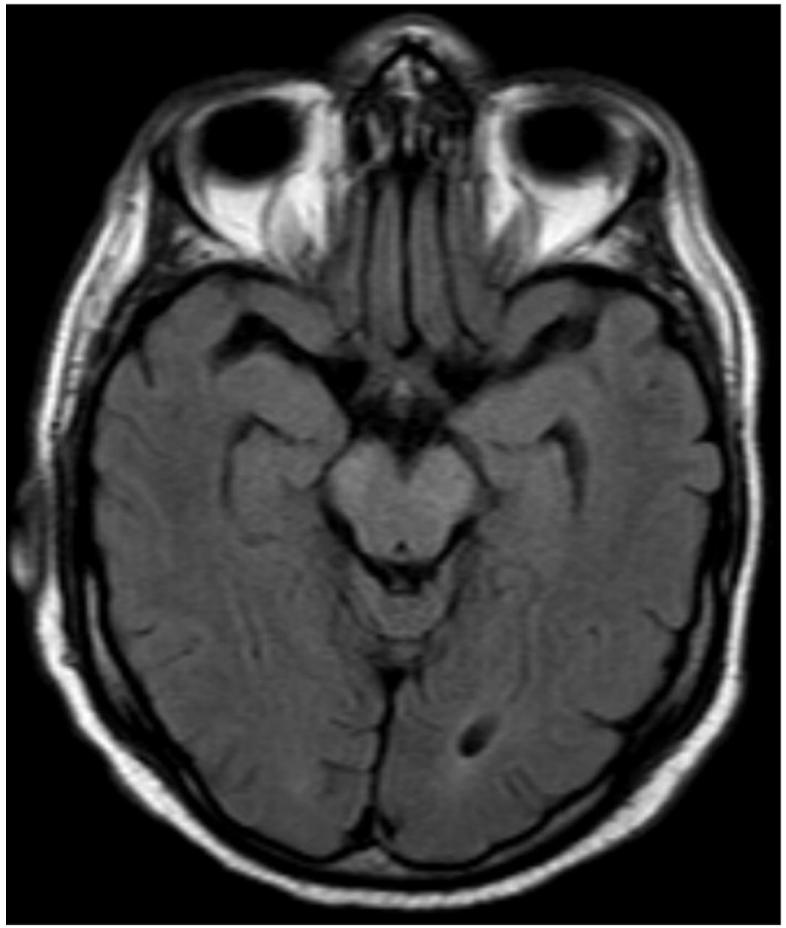
Subsequent Encounter

- 3 months since initial presentation, now with R-sided weakness, dysphagia, and unintelligible speech
- Admitted for worsening of symptoms, hospital course complicated by ventilator dependency s/p tracheostomy, septic shock 2/2 ESBL Klebsiella UTI, followed by pseudomonas and ESBL Klebsiella pneumonia
- Discussed extensively at tumor board and underwent repeat MRI for surgical biopsy imaging

Pre-Biopsy



Axial T1 Post-contrast

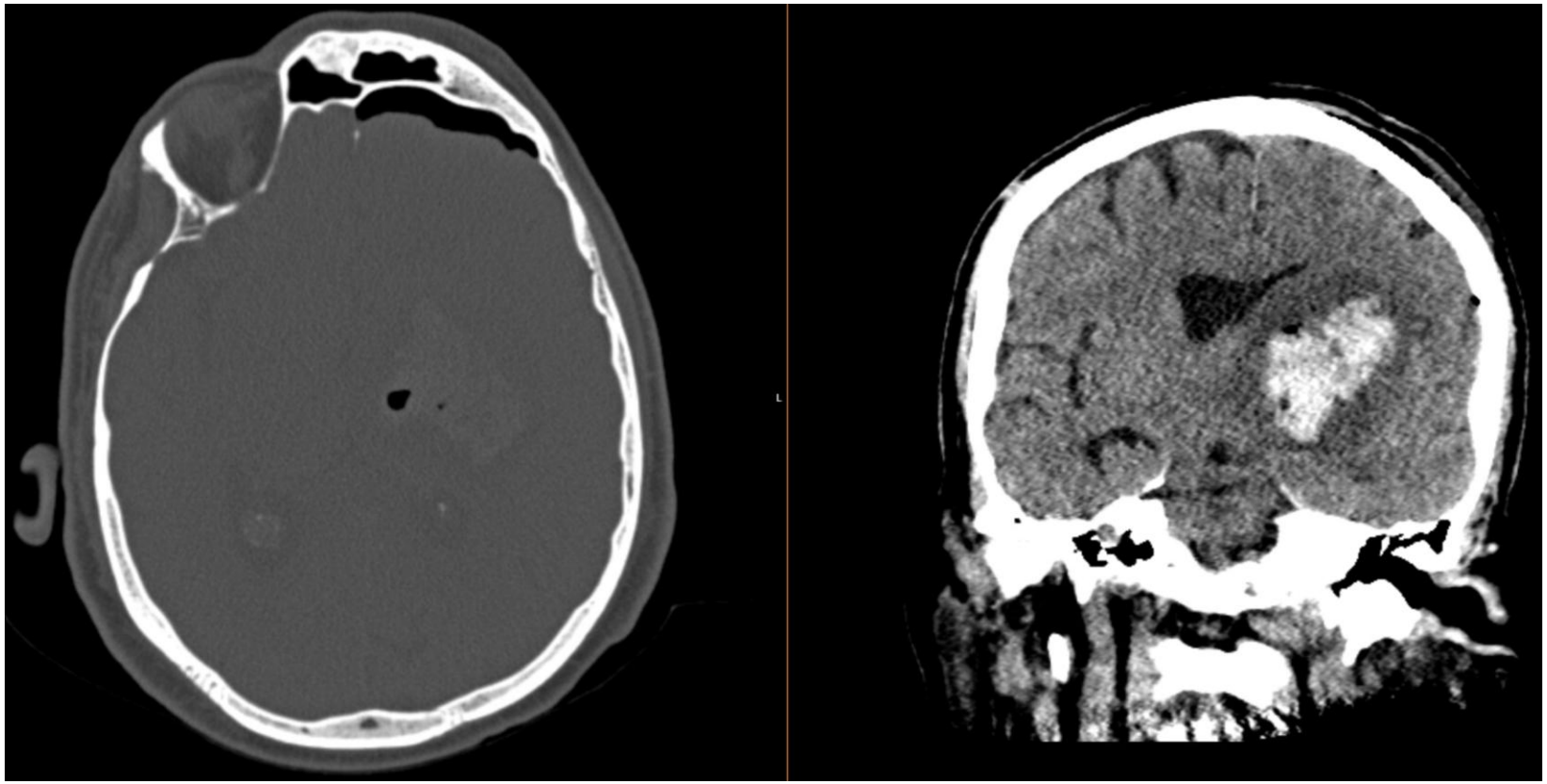


T2 FLAIR

Hospital Course

- Post-biopsy, noted to have worsened neuro exam and increased somnolence
- Head CT showed 4.6cm L basal ganglia hemorrhage
- Biopsy showed B-cell lymphoma (PCNSL – DLBCL)

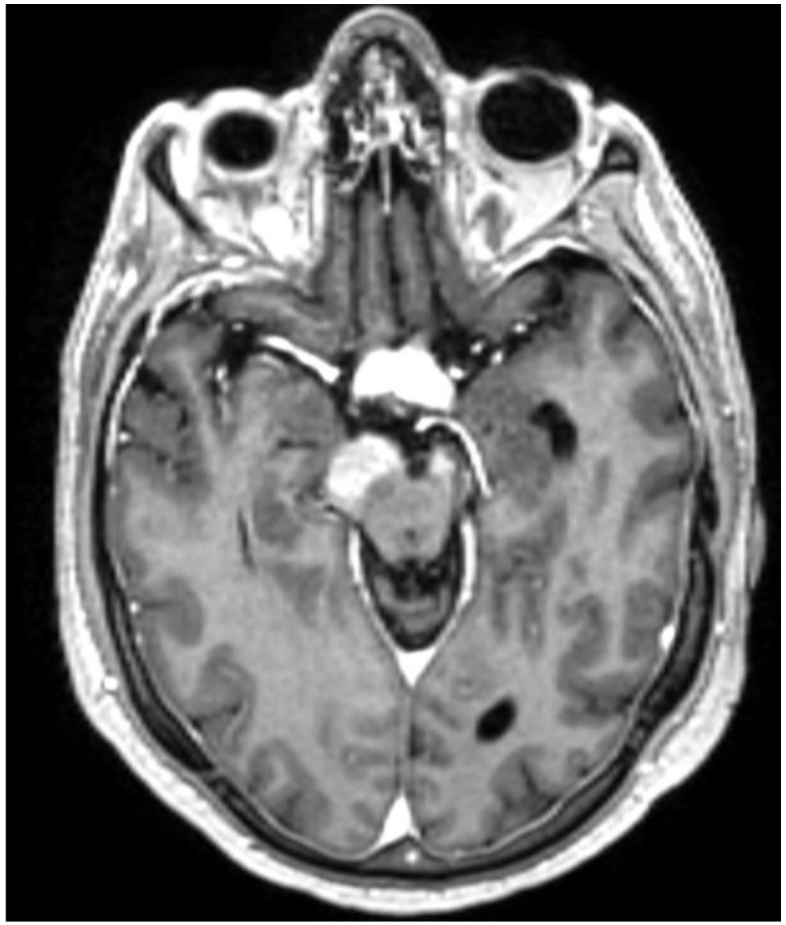
Post-Biopsy



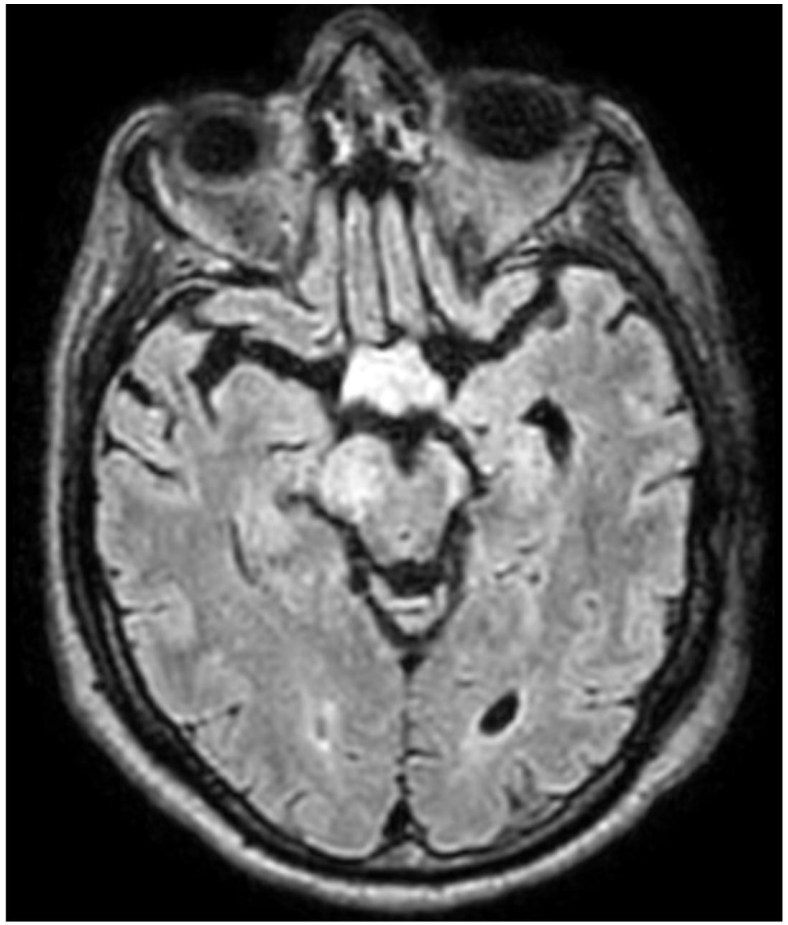
Management

- Underwent 5 cycles of rituximab, high-dose methotrexate (R-HDMTX, 21d cycles)
- Discharged to SNF and then re-admitted for septic shock 2/2 Enteroccus UTI
- Symptoms continued to progress, repeat MRI as shown
- Transitioned to palliative care

Post-Treatment



Axial T1 Post-contrast



T2 FLAIR

Outcome

- Radiation Oncology evaluated patient but determined that radiation therapy would neither improve quality of life nor prolong survival and posed a high risk of neurotoxicity in the context of intrathecal methotrexate
- Transitioned to palliative care

Take Home Points

- Radiological Mimicry
  - Progressive neurological symptoms with MRI findings such as enhancement and T2 abnormalities in the brainstem, cranial nerves, and white matter can mimic inflammatory conditions (e.g., CLIPPERS, vasculitis) but may also indicate neoplastic or infectious processes like CNS lymphoma.
- Steroid Response
  - While PCNSL often responds dramatically to steroids within days and autoimmune CNS diseases improve more gradually, a partial or slow response does not rule out PCNSL.
- Biopsy as the Gold Standard
  - Stereotactic biopsy is essential for diagnosing CNS lymphoma when imaging and other studies (e.g., CSF) are inconclusive. Risks, especially in deep brain structures, must be carefully managed.

Acknowledgements

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