A unique case of aggressive Large B Cell Lymphoma with unique presentation

Anand Bhagat D.O.; Shruti Kumar M.B.B.S.

University of Connecticut School of Medicine, Primary Care Internal Medicine

Introduction

- Primary diffuse large B cell lymphoma (PDLBcL) of the central nervous system (CNS) is a rare B-cell non-Hodgkin's lymphoma that accounts for less than 1% of all lymphomas.
- The disease is primarily present in immunocompromised individuals, however increased prevalence in competent individuals is reported.
- Initial presentation is typically due to signs of high intracranial pressure, neuropsychiatric signs, or focal neurologic deficits.

Case Presentation

- 73-year-old male with a history of dementia, transient ischemic attack presented to the emergency department with fever and vomiting.
- Patient reported right ear pain that radiated to his jaw and eye burning.
- Patient was also noted to have worsening auditory hallucinations, become more aggressive and have increased profanity over one week period.

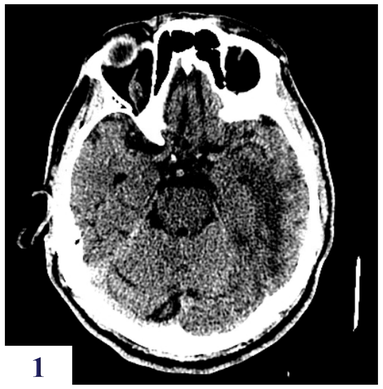

Figure 1: CT scan shows regional white matter edema in left anterior temporal lobe concerning for occult parenchymal lesion vs herpes encephalitis

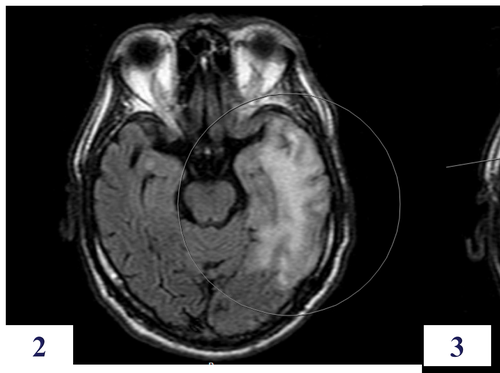

Figure 2: Axial Flair MRI shows diffuse signal abnormalities in left temporal lobe extending to the left inspiratory region

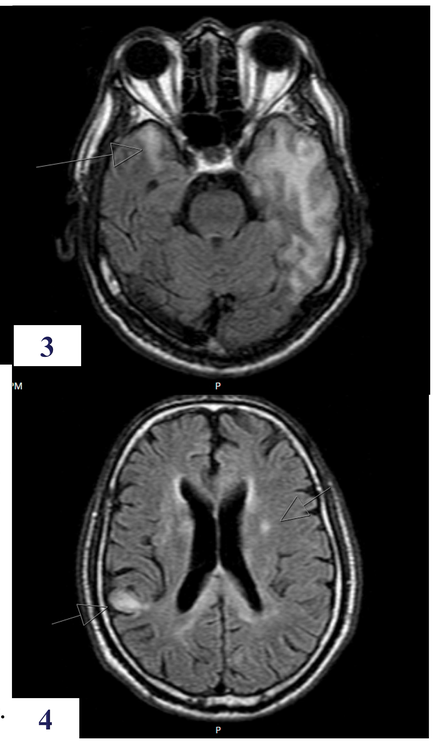

Figure 3: Axial Flair MRI shows abnormal signal in the right temporal lobe

Figure 4: Axial Flair MRI shows small area of abnormal signal in right posterior inferior frontal lobe.

Hospital Course

- Computer tomography (CT) scan and Magnetic resonance imaging (MRI) of the head showed fluid opacity in the right mastoid complex and right middle ear and enhancing mass in left in portal lobe with extensive vasogenic edema concerning for herpes encephalitis.
- Electroencephalogram consistent with mild encephalitis.
- Cerebrospinal fluid analysis showed non-conclusive cytology with mild lymphocytic pleocytosis.
- Patient continued to have fluctuating mental status and repeat MRI brain showed extensive vasogenic edema in left temporal region with enhancing mass.
- Patient underwent left temporal biopsy. Pathology confirmed concerns for diffuse aggressive B- Cell lymphoma.
- Patient discharged with outpatient hematology oncology follow-up.

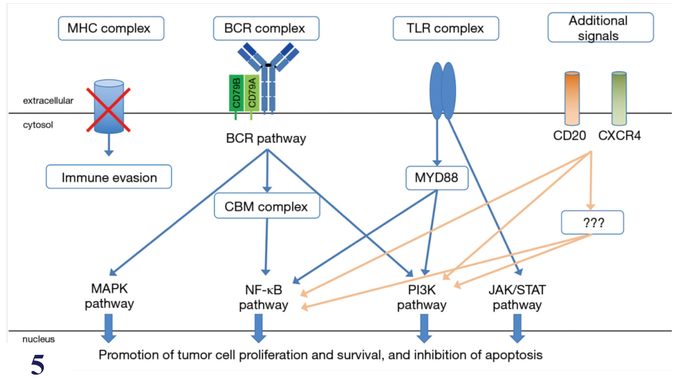

Figure 5: Most common altered pathways for the pathogenesis of CNS-PDLBcL.

Discussion

- While primary diffuse large BcL of the CNS is primarily seen in immunocompromised individuals, increase prevalence is noted in immunocompetent individuals.
- Patients typically have a variety of presentations, different neurological imaging appearances, heterogeneity of morphology and no specific lab examination making it difficult to identify and diagnose.
- Our patient's unique presentation of CNS-PDLBcL in an immunocompetent individual shows the difficulty diagnosing PDLBcL. Our patient initially presented with signs concerning for meningitis with imaging suggesting herpes encephalitis.
- Further workup including lumbar puncture revealed signs concerning for malignancy and biopsy confirmed PDLBcL.
- Internist should consider primary diffuse large BcL of the CNS as prompt identification and treatment may help prolong survival.

References

[1] Chen D, Gu W, Li W, Liu X, Yang X. Primary diffuse large B-cell lymphoma of the central nervous system: A case report and literature review. Oncol Lett. 2016 May;11(5):3085-3090. doi: 10.3892/ol.2016.4365. Epub 2016 Mar 22. PMID: 27123068; PMCID: PMC4840863.

[2] Newell, M.E., Hoy, J.F., Cooper, S.G., DeGraaff, B., Grulich, A.E., Bryant, M., Millar, J.L., Brew, B.J. and Quinn, D.I. (2004), Human immunodeficiency virus–related primary central nervous system lymphoma. Cancer, 100: 2627-2636. https://doi.org/10.1002/cncr.20300

[3] Bertoni F, Montesinos-Rongen M. Primary diffuse large B-cell lymphoma of the central nervous system: molecular and biological features of neoplastic cells. Ann Lymphoma 2022;6:1.