The Many Faces of CNS Lymphoma

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Outline

- Definition of PCNSL
- Discuss the epidemiology of PCNSL
- How does PCNSL present
- Characteristics on CT
- Characteristics on MRI
- Images of PCNSL

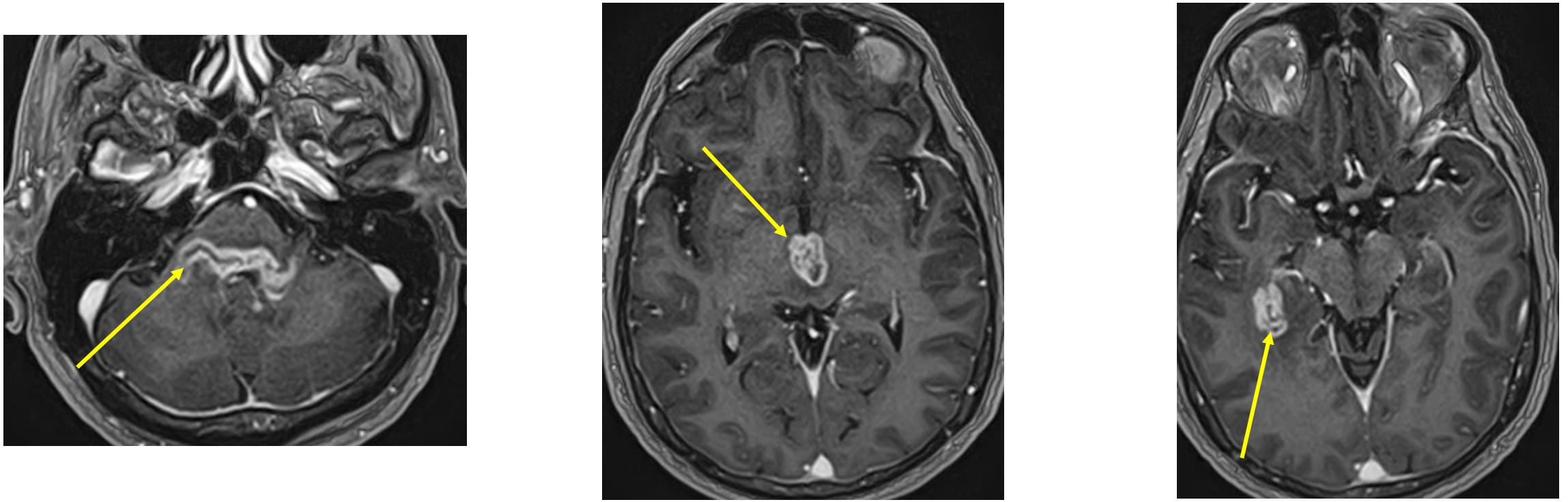
Teaching Points

- Define PCNSL
  - Primary central nervous system lymphoma (PCNSL) is commonly defined as a rare and highly aggressive extra nodal lymphoma that originates in the central nervous system.
- Discuss epidemiology of PCNSL
  - The incidence of PCNSL is estimated to be 0.4/100,000 for the general population, with a higher incidence in the elderly and immunocompromised populations [1, 2].
- How does PCNSL present
  - The clinical manifestations of PCNSL vary drastically depending on the anatomical location of lesions and range from focal neurological deficits to nonspecific cognitive and behavioral changes[2-4]. PCNSL primarily presents early as singular lesions and manifests later as multiple lesions that appear in the deep white matter and basal ganglia structures[3, 4, 6]. Joshi et al. found that a majority of lesions were found supratentorial (71%), in the periventricular region 9/22[7]. Cheng et al. found that 79.6% of lesions occur in the cerebral hemispheres, with 28.8% occurring in the basal ganglia region and 22.0% occurring in the thalamus[3].

Teaching Points

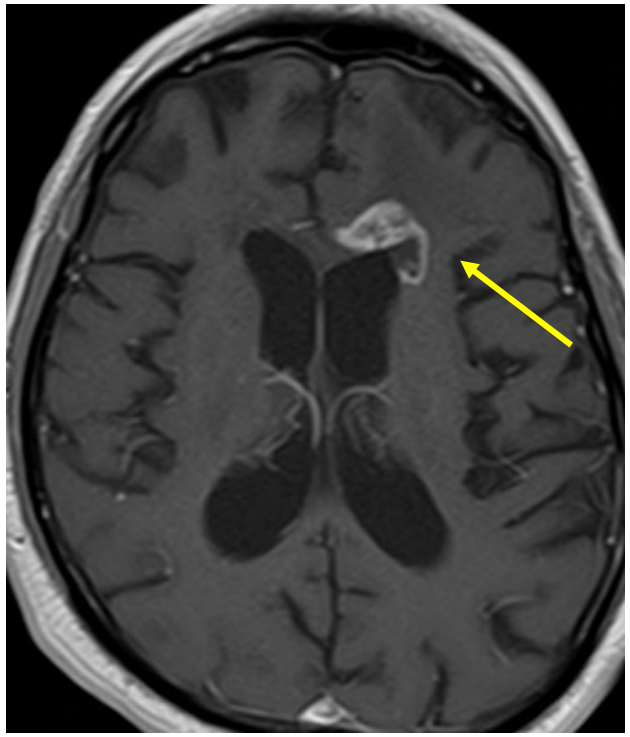
- Characteristics on CT
  - On CT, PCNSL commonly appears as hyperdense to isodense lesions with high density shadowing that are nonspecific and undistinguishable from other neoplasms and intracranial pathologies[3, 6].
- Characteristics on MRI
  - MRI remains the gold standard for PCNSL imaging [8]. PCNSL appears hyperintense on T1 and iso-hypointense on T2 and FLAIR, with most lesions exhibiting patchy enhancement [7]. Cheng et al. concluded that some PCNSL exhibit the characteristic “butterfly sign” when lesions cross the corpus callosum; however, in Wang et al. only 25% of primary lesions exhibited the characteristic butterfly sign of PCNSL. Linear enhancements along the periventricular area with “angiotropic” invasive tumor growth, arranged centripetally around the Virchow-robin space, were commonly seen on enhanced MRI[3]. On DWI, PCNSL lesions show restricted diffusions that correlates with lower apparent diffusion coefficient values in comparison to other primary brain tumors, thus having utility in differentiating PCSNL on imaging[7, 9, 10].

Intraventricular Lymphoma

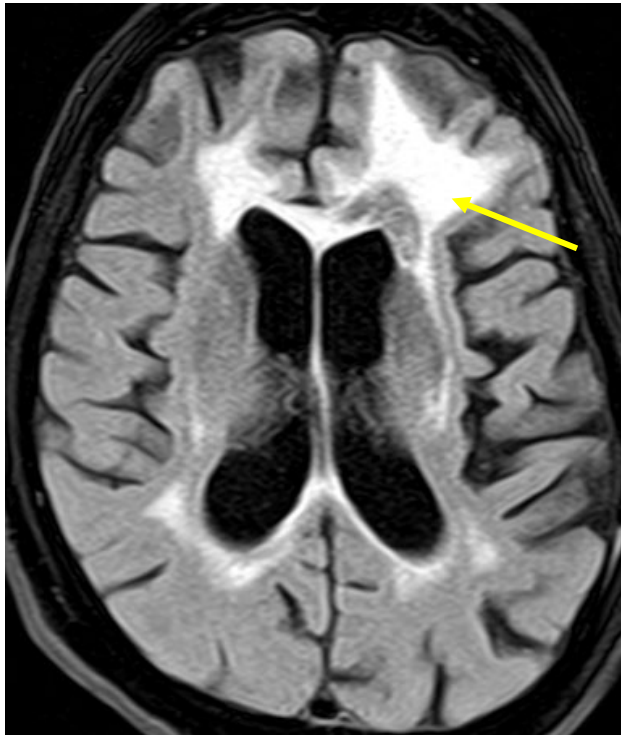


Axial T1 postcontrast images show multiple intraventricular nodular enhancing lesions.

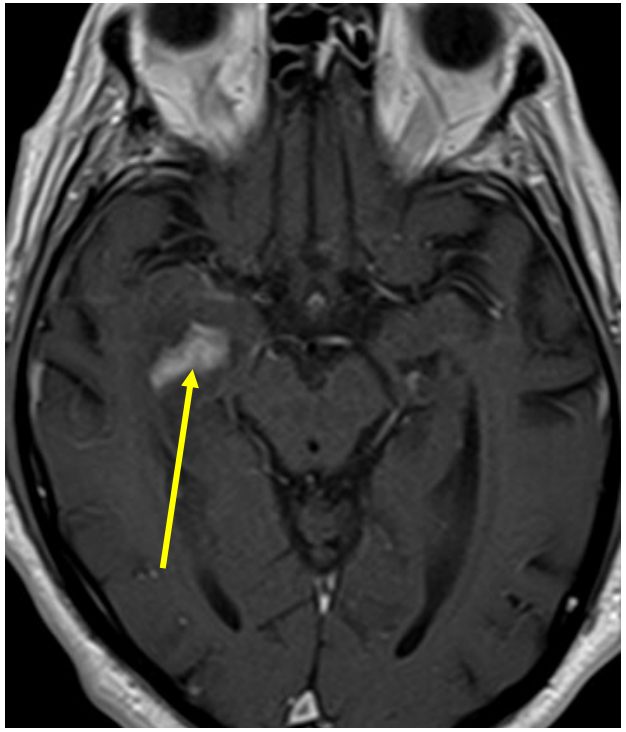
Periventricular lymphoma



Axial T1 postcontrast shows enhancing left frontal periventricular mass.

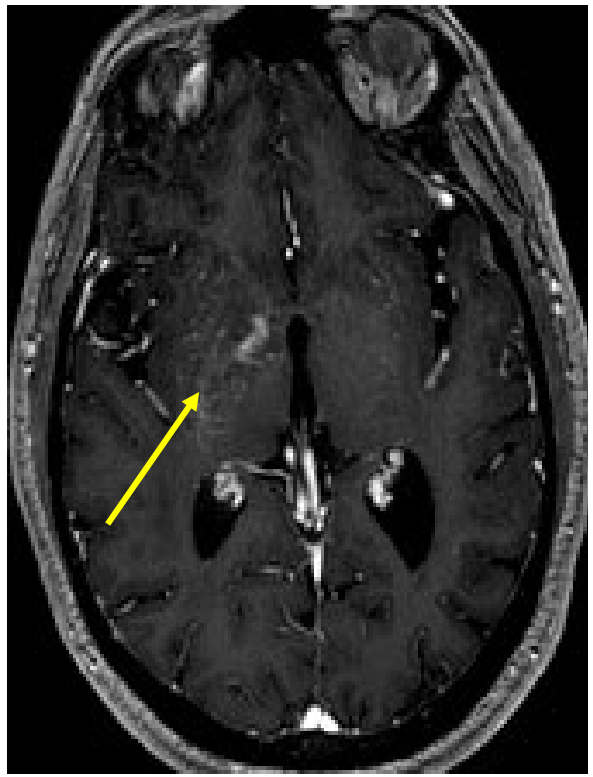


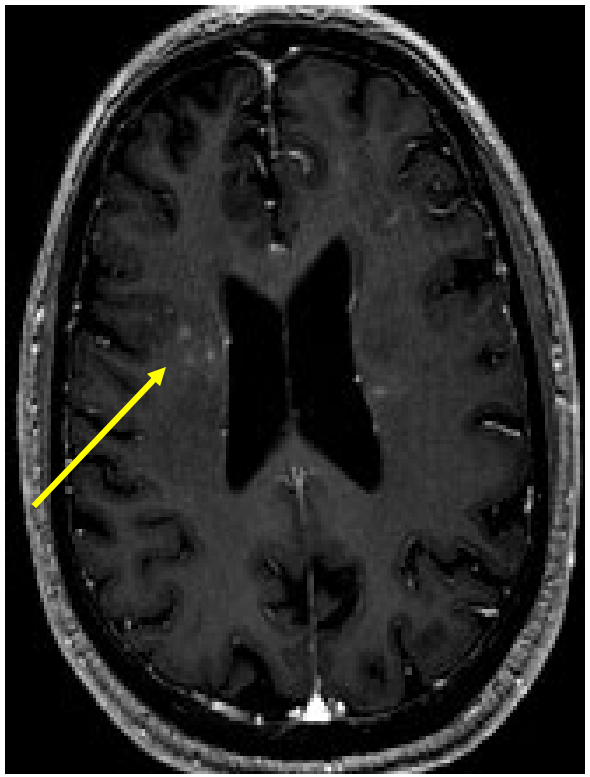
Axial FLAIR shows vasogenic edema surrounding the left frontal mass which is isointense to brain parenchyma.

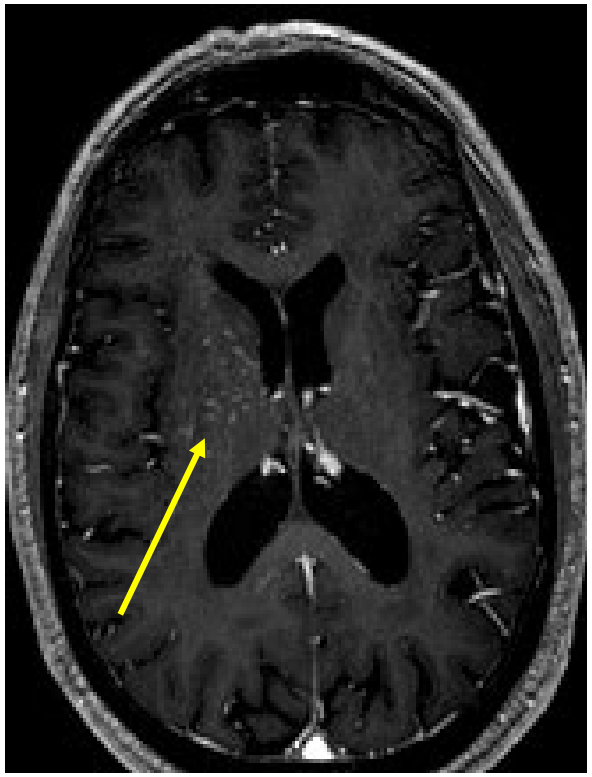


Axial T1 postcontrast shows enhancing right temporal periventricular mass.

Intravascular Lymphoma

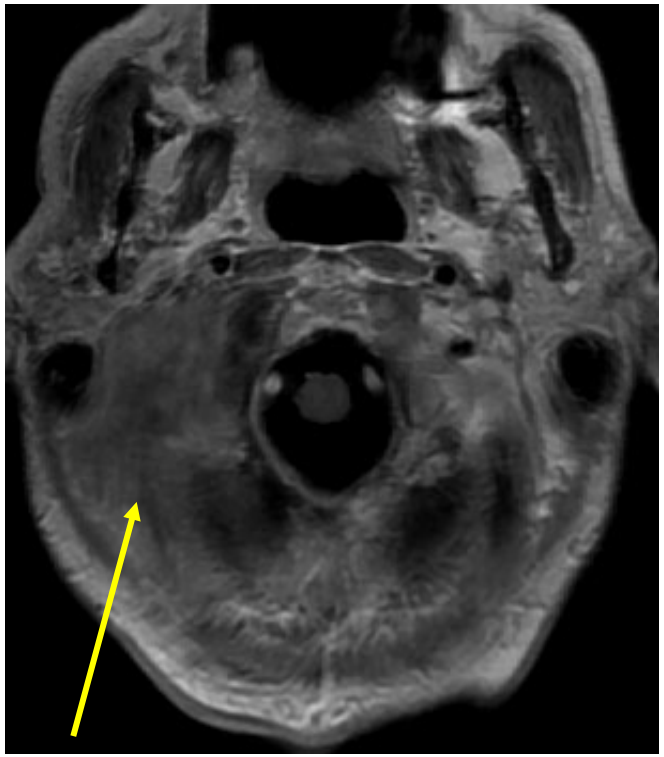




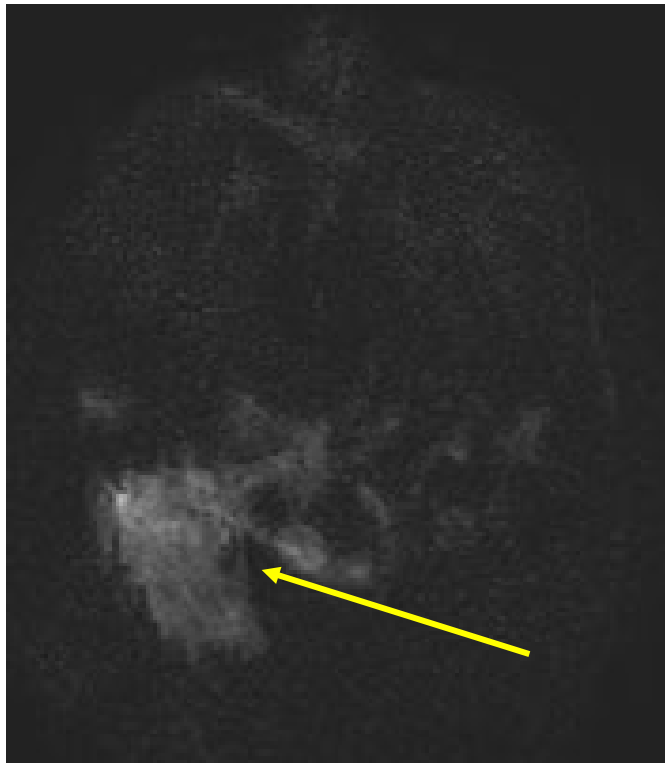


Axial T1 postcontrast images show nodular enhancement of the perivascular spaces.

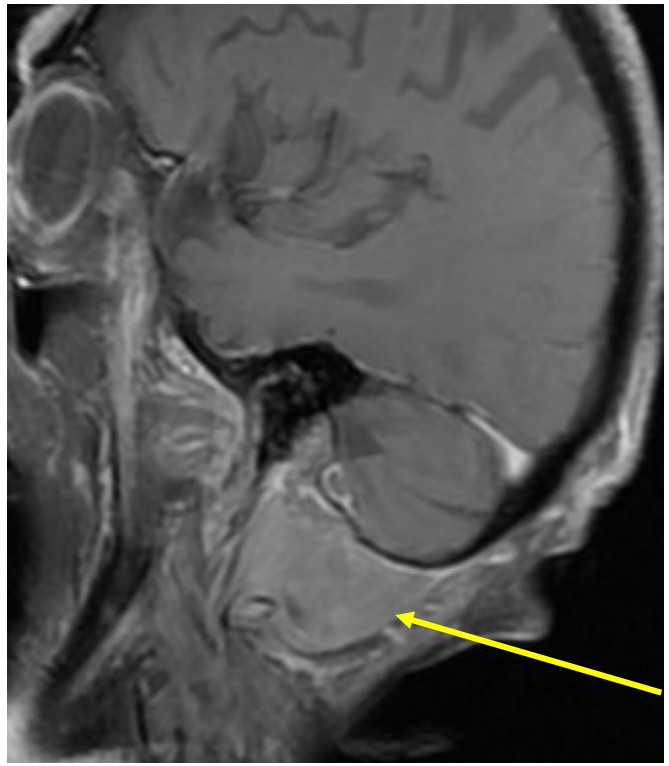
Skull base lymphoma



Axial T1 postcontrast image shows right skull base mass with mild enhancement.

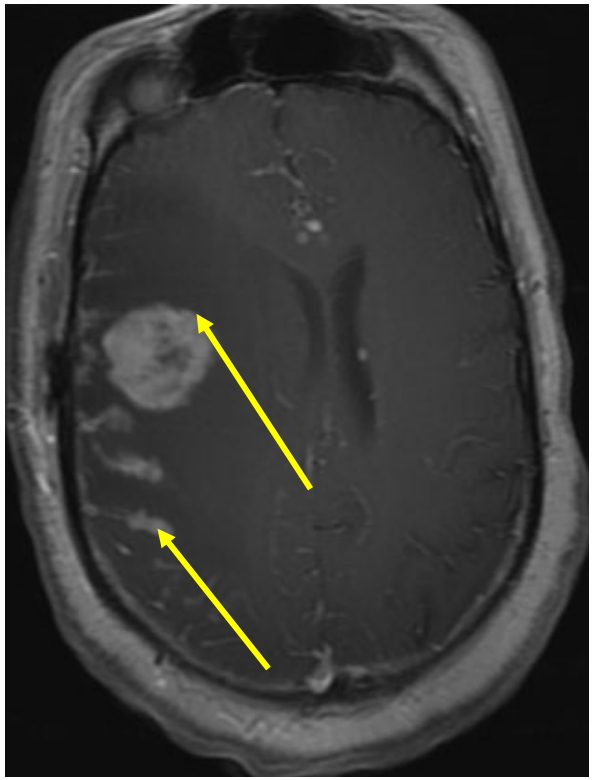


Axial DWI shows restricted diffusion in the right skull base mass.

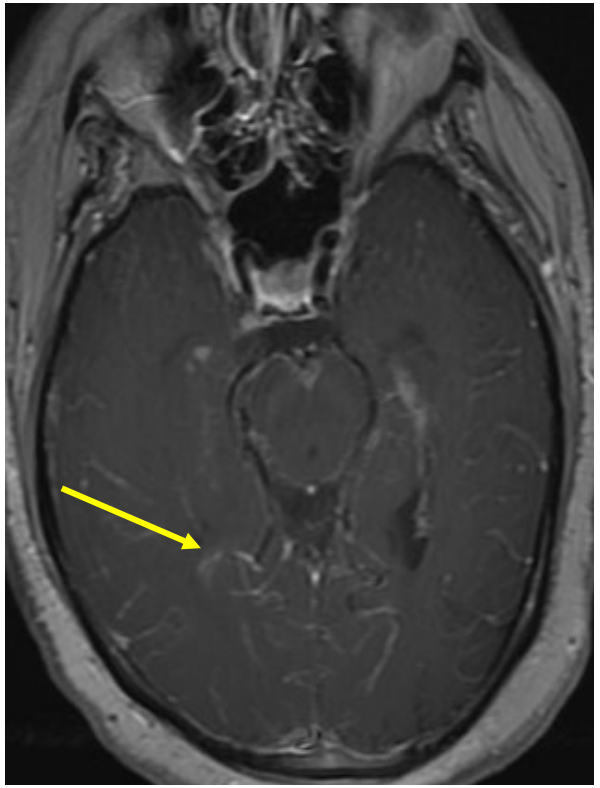


Sagittal T1 postcontrast image shows right skull base mass with mild enhancement.

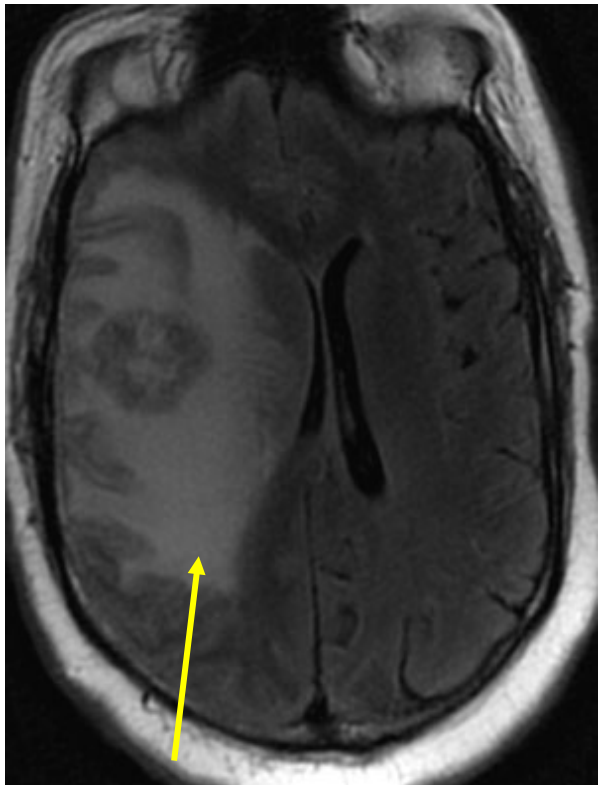
Intraparenchymal and leptomeningeal lymphoma



Axial T1 postcontrast shows right frontal enhancing mass with leptomeningeal enhancement.



Axial T1 postcontrast shows diffuse nodular leptomeningeal enhancement.



Axial FLAIR demonstrates vasogenic edema surrounding the right frontal mass.

Summary

- **Primary Central Nervous System Lymphoma (PCNSL)**: A rare, highly aggressive extranodal lymphoma originating in the central nervous system.

- **Lesion Distribution**: Initially single lesions, later multiple lesions. A majority are supratentorial, with significant presence in the periventricular region and basal ganglia.

- **Appearance on CT**: Hyperdense to isodense lesions with high-density shadowing; nonspecific and difficult to differentiate from other intracranial pathologies.

- **Appearance on MRI:**
  - **T1**: Hyperintense lesions.
  - **T2 & FLAIR**: Iso- to hypointense lesions.
  - **DWI**: Restricted diffusion with lower apparent diffusion coefficient (ADC) values, helpful for distinguishing PCNSL from other brain tumors.

References

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