48-year-old male with malignancy with new-onset seizures.

Ryan P. Joyce, MD
Leo Wolansky, MD

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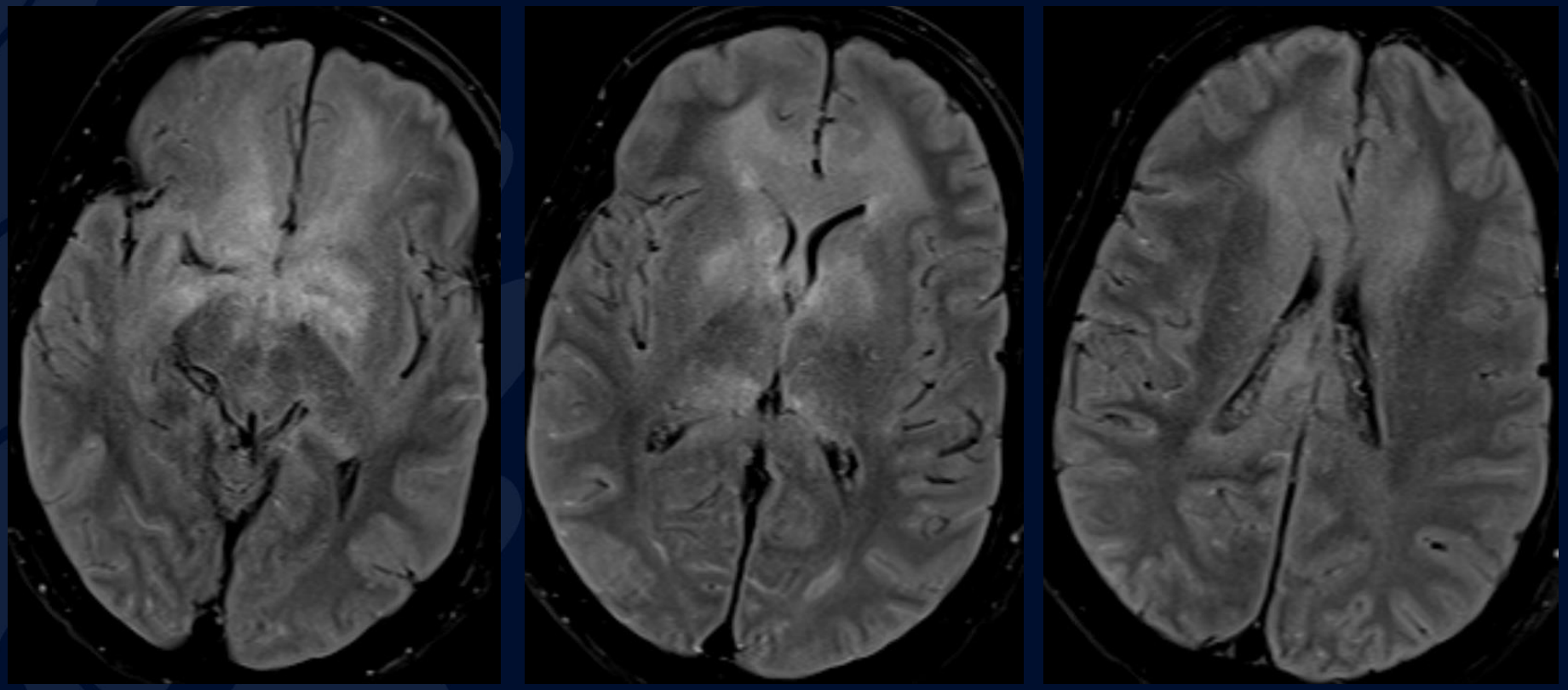

Axial FLAIR

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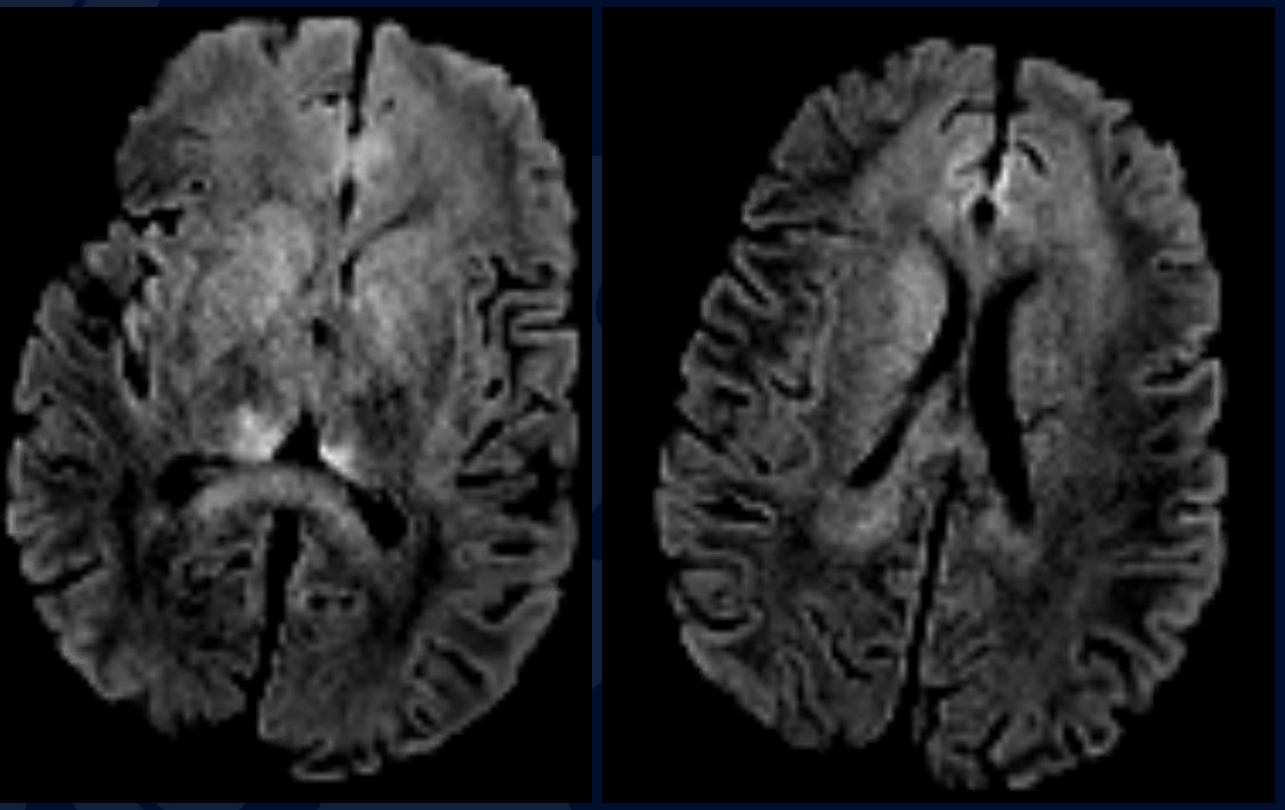

Axial B1000 DWI

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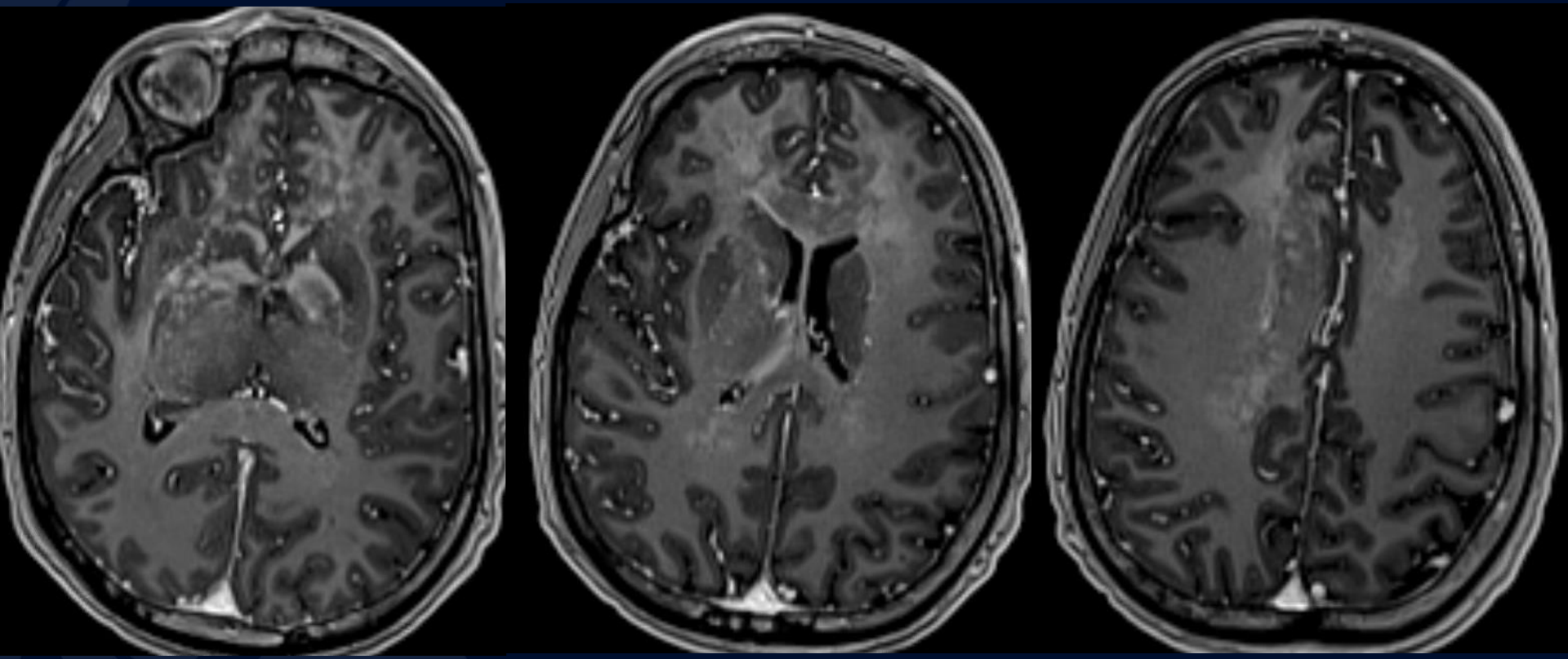
Axial T1 Gd-enhanced Volumetric Gradient-Echo



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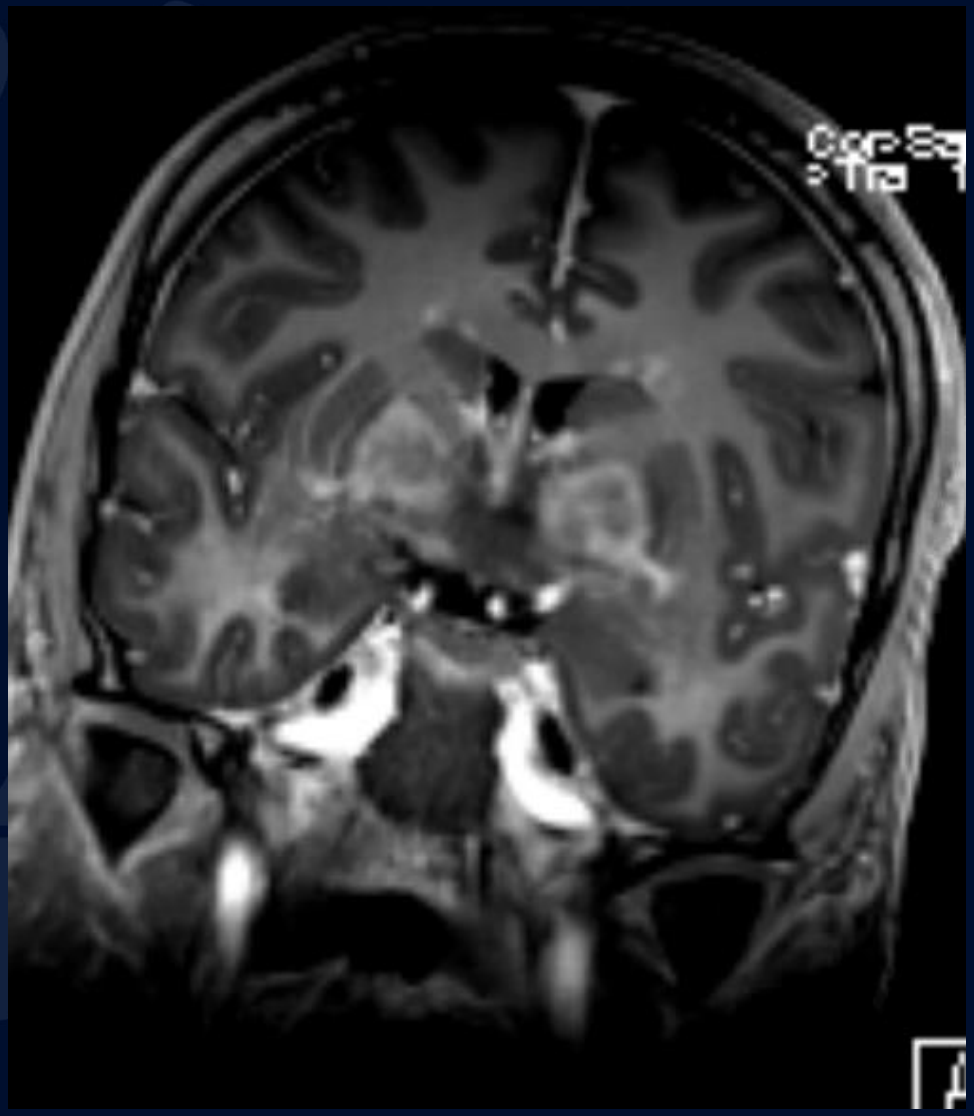
Axial T1 Gd-enhanced Volumetric Gradient-Echo



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Intravascular (angiocentric) lymphoma

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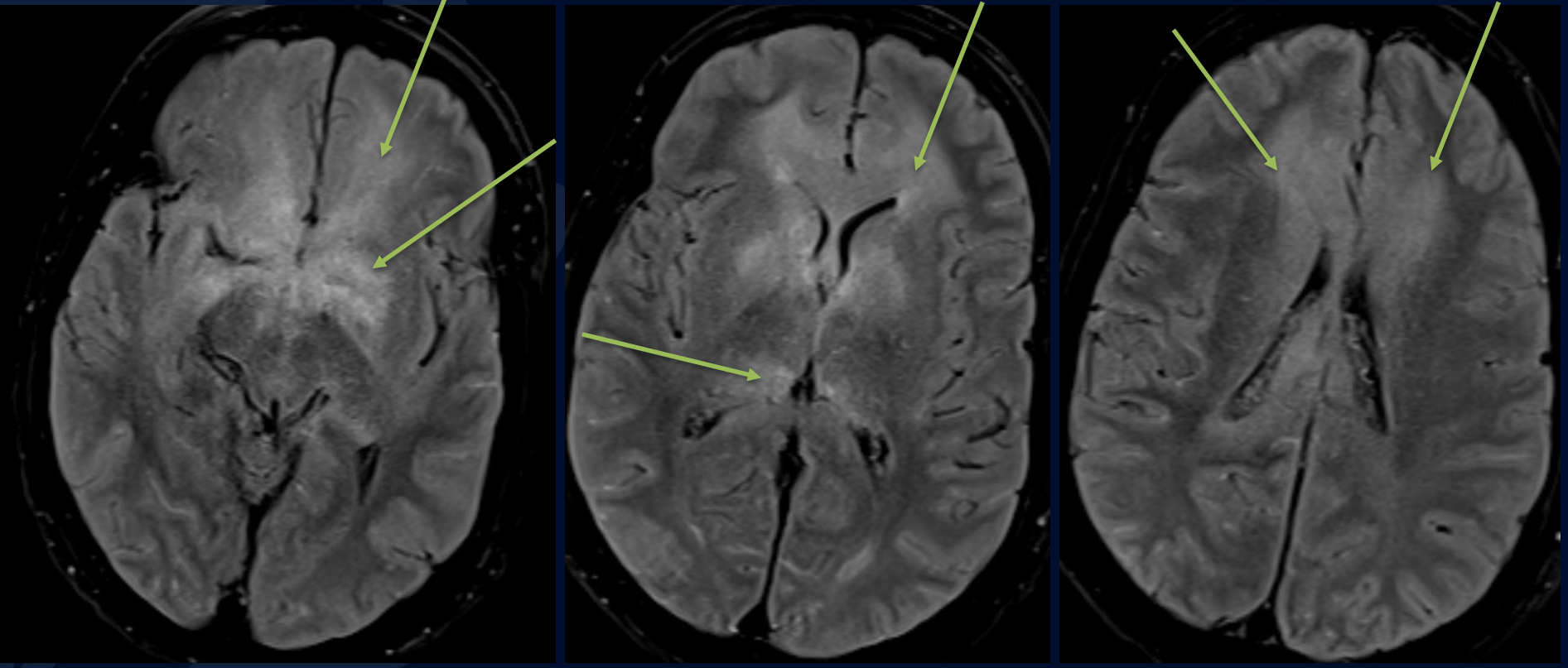
Hyperintense FLAIR signal within the basal ganglia, thalami, periventricular and deep white matter



Axial FLAIR

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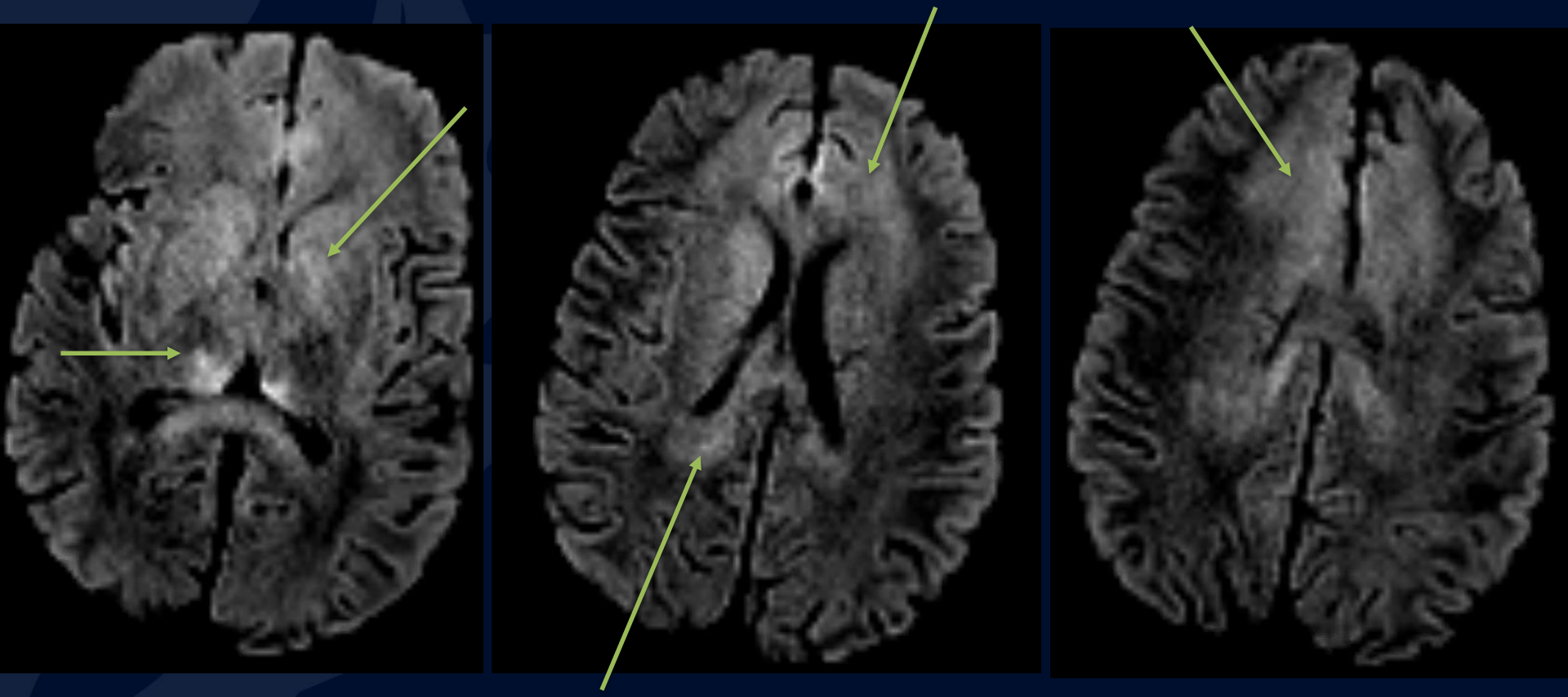
Hyperintense DWI signal, representing restricted diffusion, within the same regions; the basal ganglia, thalami, periventricular and deep white matter



Axial B1000 DWI

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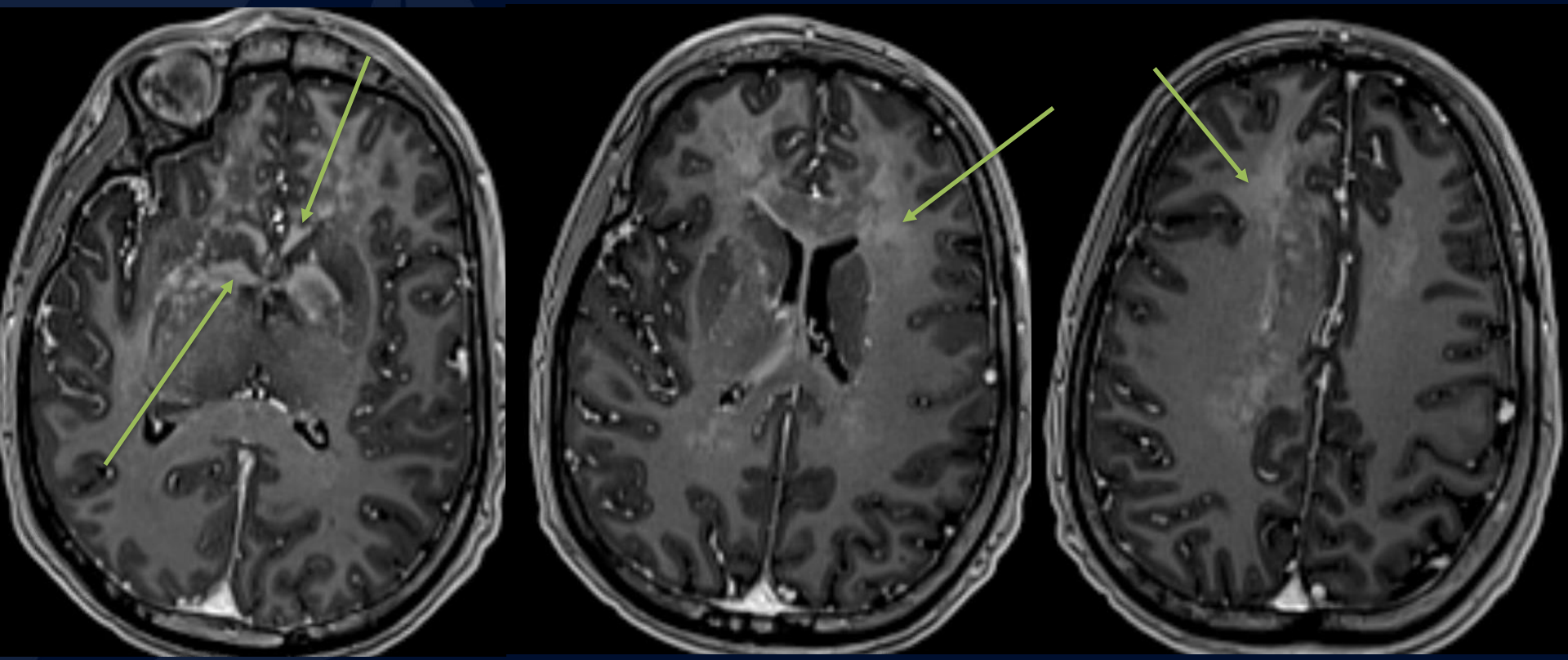
Patchy, punctate, & linear enhancement involving same regions; basal ganglia, thalami, periventricular & deep white matter



Axial T1 Gd-enhanced Volumetric Gradient-Echo

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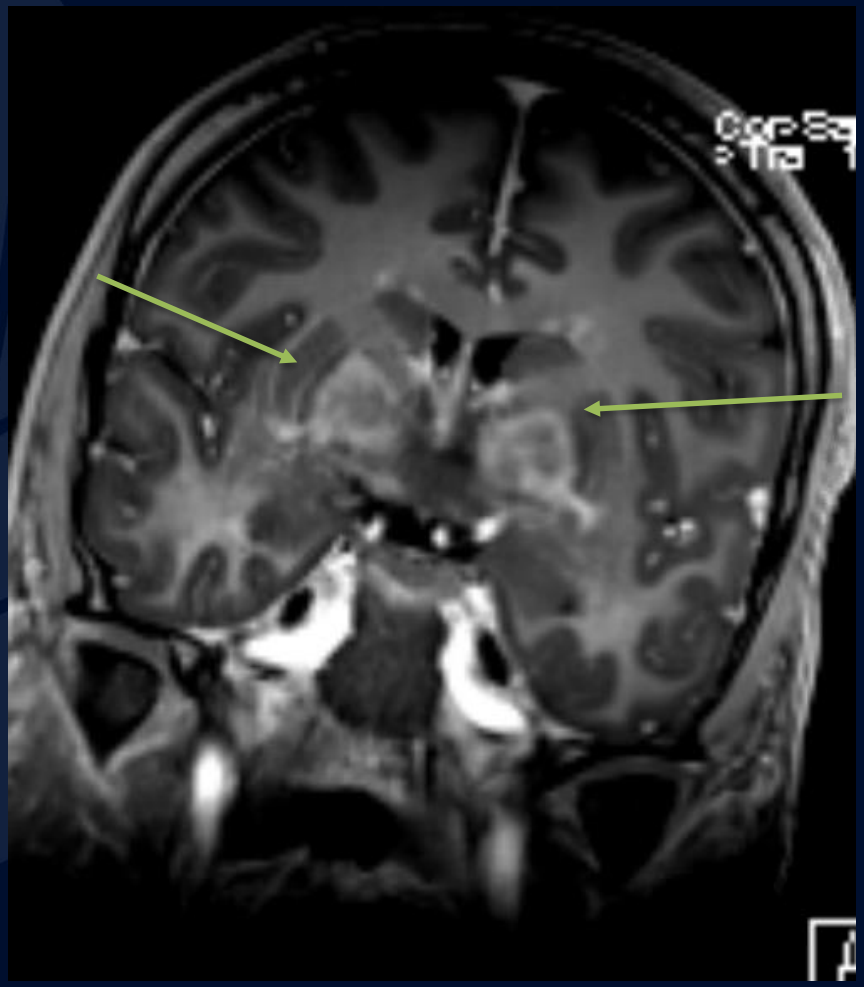
Patchy and confluent enhancement involving same regions; basal ganglia, thalami, periventricular & deep white matter



Axial T1 Gd-enhanced Volumetric Gradient-Echo

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Intravascular (angiocentric) lymphoma

- Rare, fatal form of non-Hodgkin lymphoma with angiotropic growth – proliferation of malignant lymphoid cells within vessel lumen.
  - Results in distension & occlusion of small arteries, veins, & capillaries.
  - Originally thought to be a neoplastic vascular proliferation, later shown to be lymphoma (usually B-cell).
- Preferentially involves small-to-medium sized vessels.
  - Cells primarily found in capillaries in the CNS & skin.
    - Western variant found in the CNS and skin, Asian variant characterized by hemophagocytosis.

Intravascular (angiocentric) lymphoma

- *Can involve any organ*, but predilection for CNS & skin
- Tends to not be seen on peripheral blood smears, bone marrow biopsies.
- Typically *no lymphadenopathy* on imaging.
- Highly variable clinical presentation.
- New-onset dementia most common presenting feature.
- Rapidly progressive disease with high mortality rate.
  - Mean survival 7-13 months

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Intravascular (angiocentric) lymphoma

There are no pathognomonic criteria for IVL.

Imaging findings are nonspecific & include:

- Multifocal abnormal T2/FLAIR hyperintensities in white matter and/or cortex and/or basal ganglia, with abnormal, typically linear or nodular enhancement on post-contrast imaging.
- Enhancement may be absent in early/acute phase. Seen in subacute phase.
- Frequently associated with hemorrhage or evidence of prior hemorrhage (blooming artifact).
- May mimic acute multifocal embolic stroke or vasculitis.
  - May see “beading” morphology of vessels on MRA, CTA

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Intravascular (angiocentric) lymphoma

-- >1/2 of patients are diagnosed at autopsy.

-- Recent emphasis on the importance of a skin biopsy for diagnosis.

--Treatment with chemotherapy & immunotherapy has shown better response than chemotherapy alone.

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References

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2. Awad, Amer, Bachir Estephan, and Olaf Stüve. "CNS intravascular lymphoma: A case report." *Case reports in neurological medicine* 2011 (2011).
3. Matsue, Kosei, et al. "A clinicopathological study of 13 cases of intravascular lymphoma: experience in a single institution over a 9-yr period." *European journal of haematology* 80.3 (2008): 236-244.
4. Statdx.com

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