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

Ryan P. Joyce, MD
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Axial B1000 DWI
Axial T1 Gd-enhanced Volumetric Gradient-Echo
Axial T1 Gd-enhanced Volumetric Gradient-Echo
Intravascular (angiocentric) lymphoma
Hyperintense FLAIR signal within the basal ganglia, thalami, periventricular and deep white matter
Hyperintense DWI signal, representing restricted diffusion, within the same regions; the basal ganglia, thalami, periventricular and deep white matter
Patchy, punctate, & linear enhancement involving same regions; basal ganglia, thalami, periventricular & deep white matter.

Axial T1 Gd-enhanced Volumetric Gradient-Echo
Patchy and confluent enhancement involving same regions; basal ganglia, thalami, periventricular & deep white matter

Axial T1 Gd-enhanced Volumetric Gradient-Echo
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
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
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.
References


4. Statdx.com