65-year-old female with history of multiple sclerosis presenting with subacute intermittent thunderclap headaches

Ricky Paramo, MS4

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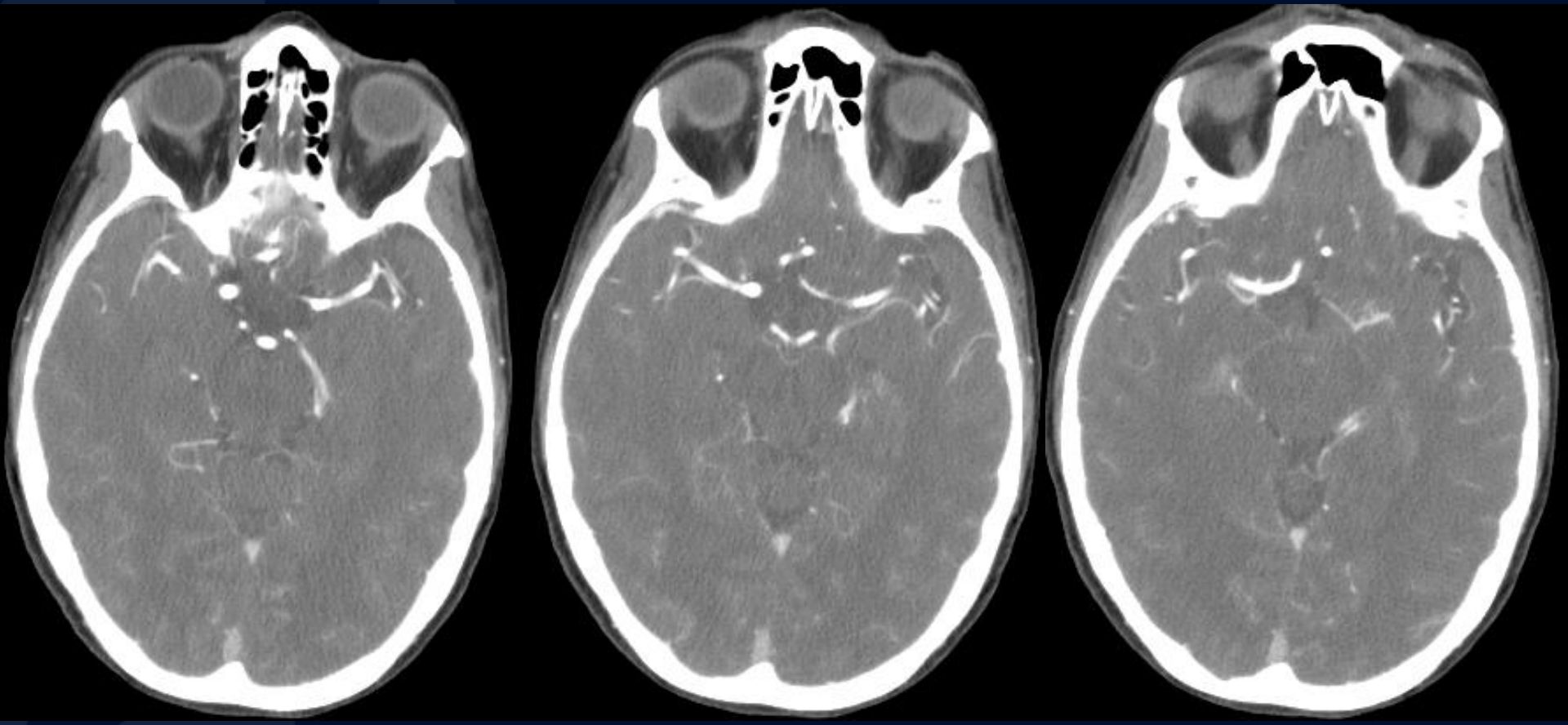
CT Angiogram



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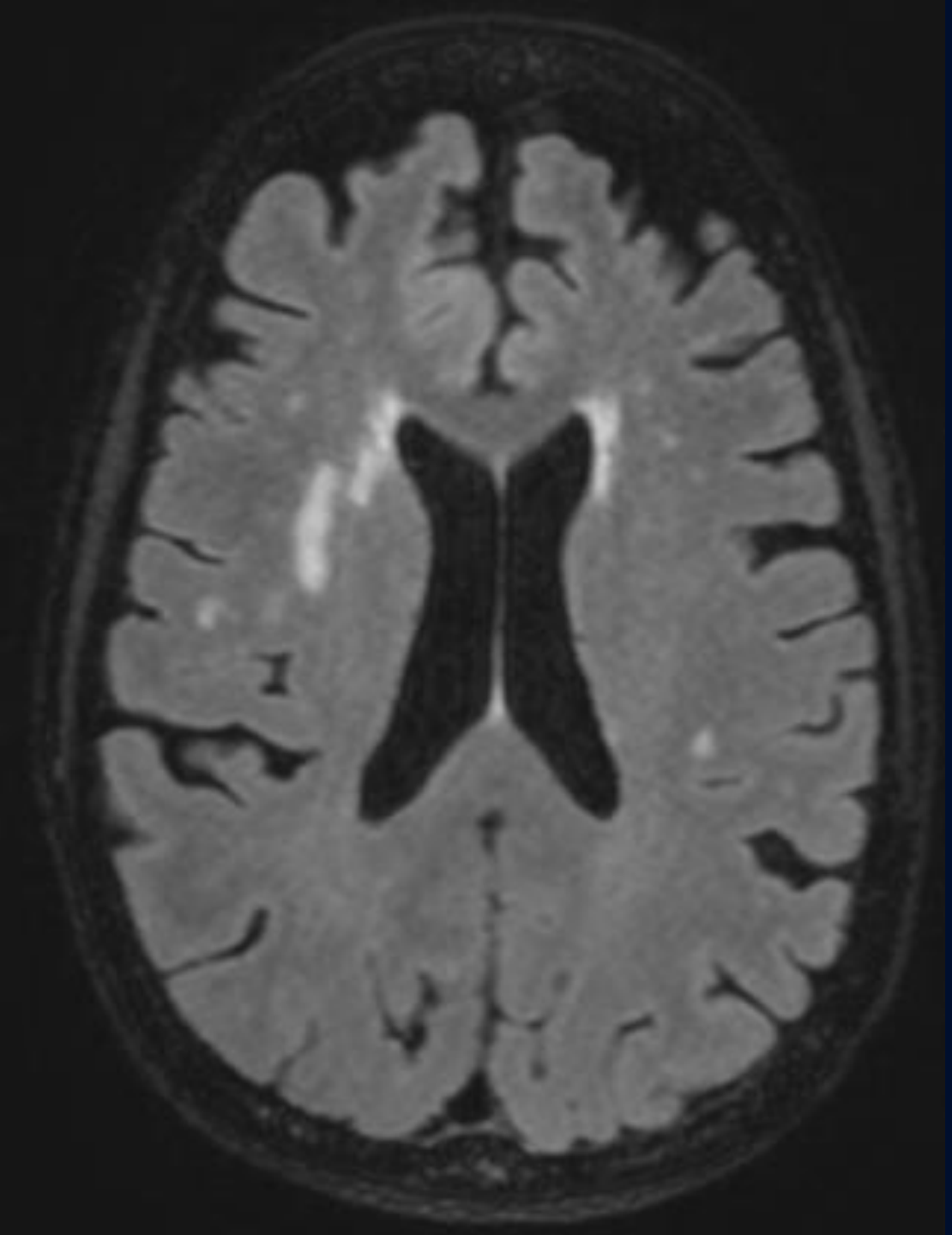

T2 Flair

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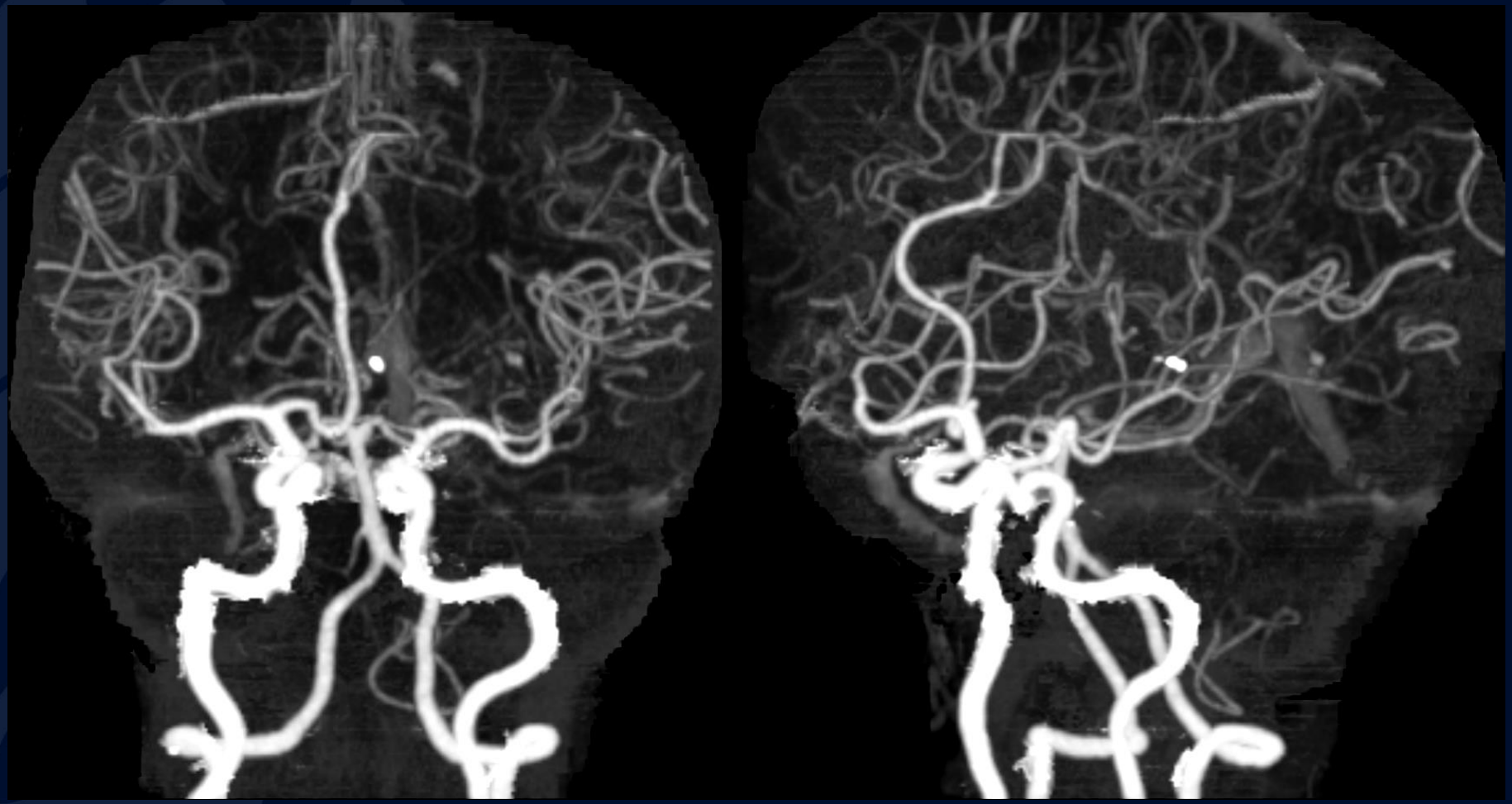
CT Angiogram



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Reversible Cerebral Vasoconstriction Syndrome (RCVS)

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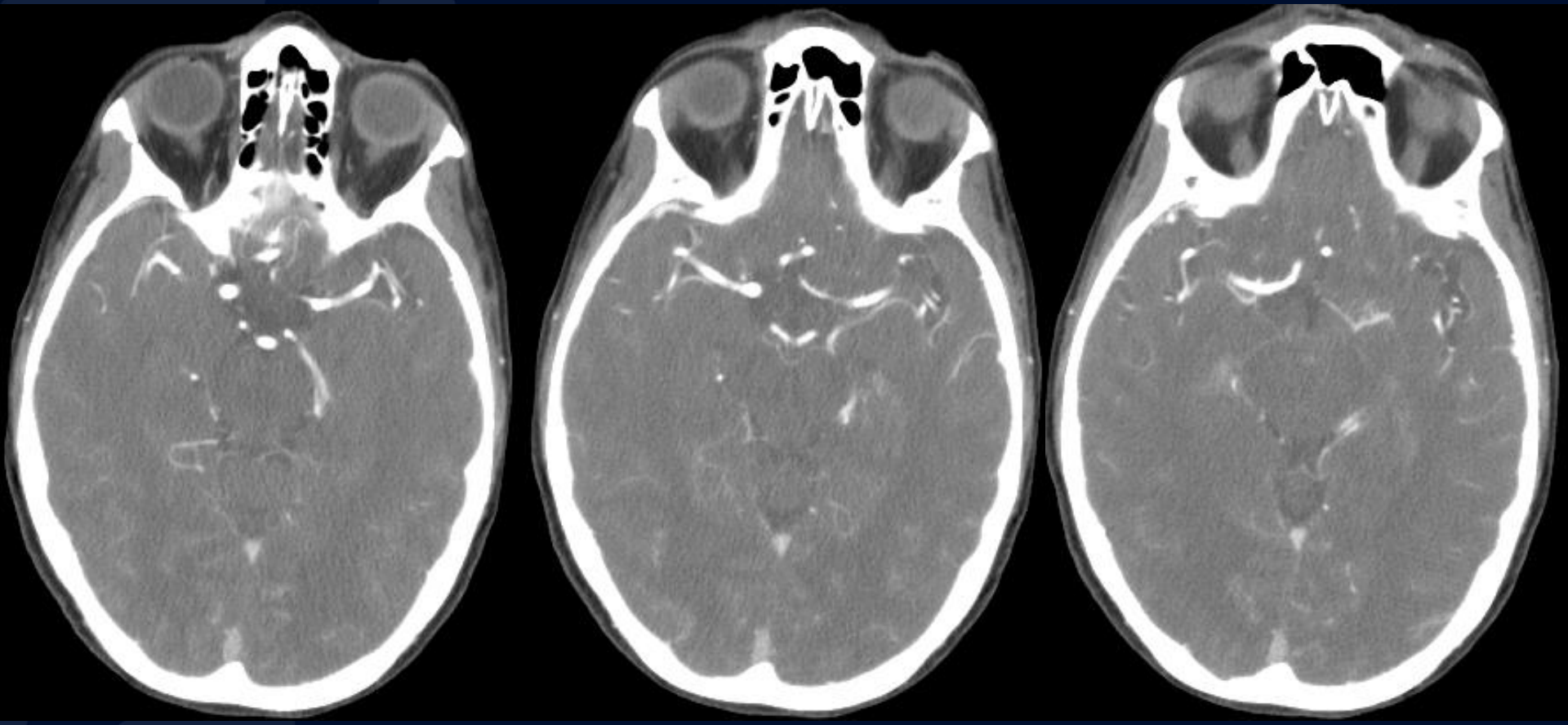
CT Angiogram



Patent circle of Willis; no high-grade focal stenosis, dissection, or aneurysm

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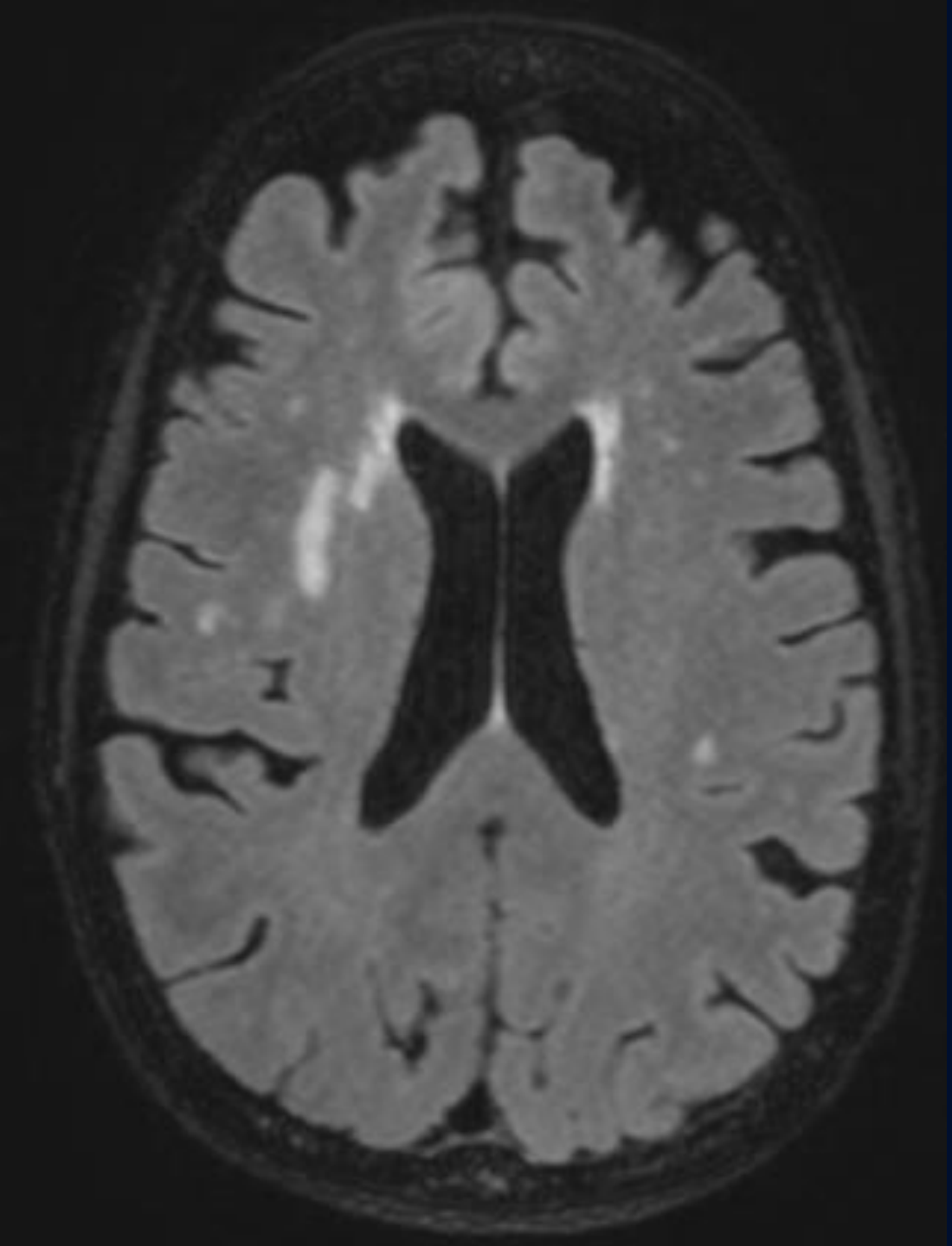

T2 Flair

Supratentorial white matter lesions, consistent with patient's history of MS and unchanged from previous MRI 6 years prior

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CT Angiogram

Azygous anterior cerebral artery

M2 branches with beaded contour irregularity R > L

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RCVS

Epidemiology

- Incidence of 3 per 1 million adults
  - Age 20-50 years, females > men (F:M = 2.4:1)

Pathophysiology

- Overactivated systemic sympathetic system – sudden release of vasoconstrictors such as catecholamines, neuropeptide Y or endothelin-1 resulting in abrupt dysregulation of cerebral vascular tone

Clinical Presentation

- Thunderclap headache which may be associated with photophobia, nausea and/or vomiting
- Less often focal neuro deficits secondary to comorbid pathology (e.g., ischemia, hemorrhage, demyelination)
- Associated with various vasoactive substances (e.g., cannabis, SSRIs, etc.)
- Normal CSF studies

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RCVS

Diagnosis

- Recurrent thunderclap headaches or single thunderclap headache
- Normal neuroimaging study

Treatment / Prognosis

- Spontaneous resolution
- Complete long-term resolution without neuro-deficits in up to 90% of patients

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Imaging Findings

CT

- Often normal

MRI

- Hyperintensities may represent:
  - Vasogenic edema* (38%)
  - Watershed infarct (29%)
  - Convexity subarachnoid hemorrhage (22-34%)
  - Lobar hemorrhage (6-20%)

Angiography

- Narrowing and dilation (“string and beads”) of second- or third-order branches is most characteristic findings.

*Early findings may include isolated cortical vasogenic edema and hyperintense vessel sign, when observed within hours of headache onset.

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References

- Gaillard F. Reversible cerebral vasoconstriction syndrome | Radiology Reference Article | Radiopaedia.org. Radiopaedia. Accessed June 24, 2023. https://radiopaedia.org/articles/reversible-cerebral-vasoconstriction-syndrome-2?lang=us
- Magid-Bernstein J, Omran SS, Parikh NS, Merkler AE, Navi B, Kamel H. RCVS: Symptoms, Incidence, and Resource Utilization in a Population-Based US Cohort [published online ahead of print, 2021 May 28]. Neurology. 2021;97(3):e248-e253. doi:10.1212/WNL.0000000000012223
- Ducros A, Boukobza M, Porcher R, Sarov M, Valade D, Bousser MG. The clinical and radiological spectrum of reversible cerebral vasoconstriction syndrome. A prospective series of 67 patients. Brain. 2007;130(Pt 12):3091-3101. doi:10.1093/brain/awm256
- Singhal AB, Hajj-Ali RA, Topcuoglu MA, et al. Reversible cerebral vasoconstriction syndromes: analysis of 139 cases. Arch Neurol. 2011;68(8):1005-1012. doi:10.1001/archneurol.2011.68
- Murase S, Gon Y, Watanabe A, et al. Isolated cortical vasogenic edema and hyperintense vessel signs may be early features of reversible cerebral vasoconstriction syndrome: Case reports. Cephalalgia. 2018;38(6):1207-1210. doi:10.1177/0333102417731779
- Case courtesy of David Cuete, Radiopaedia.org. From the case rID: 23768

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