AMSER Case of the Month September 2026

67-year-old male with acute aphasia, confusion, and fever

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Patient Presentation

- 67-year-old male presenting with fever, confusion, and worsening speech disturbance over 2 days
- PMH: Atrial fibrillation on apixaban, hypertension, hyperlipidemia, prostate cancer
- Vitals: initial vitals unremarkable
- Neurologic exam: Partial global aphasia

Pertinent Labs

- Initial laboratory studies unremarkable

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant: Adult. Altered mental status. Suspected intracranial pathology or focal neurologic deficit. Initial imaging. (Scenario ID 3149724)

Procedure	Adult RRL	Peds RRL	Appropriateness Category
CT head without IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually appropriate
MRI head without and with IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate
MRI head without IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate
MRI head with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
CT head with IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually not appropriate
CT head without and with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate

CT head without IV contrast ordered first by ER physician

Initial CT head without IV contrast ordered to exclude acute hemorrhage (suspected stroke). Brain MRI with and without contrast, carotid ultrasound, transthoracic echocardiogram were subsequently ordered for further stroke work up

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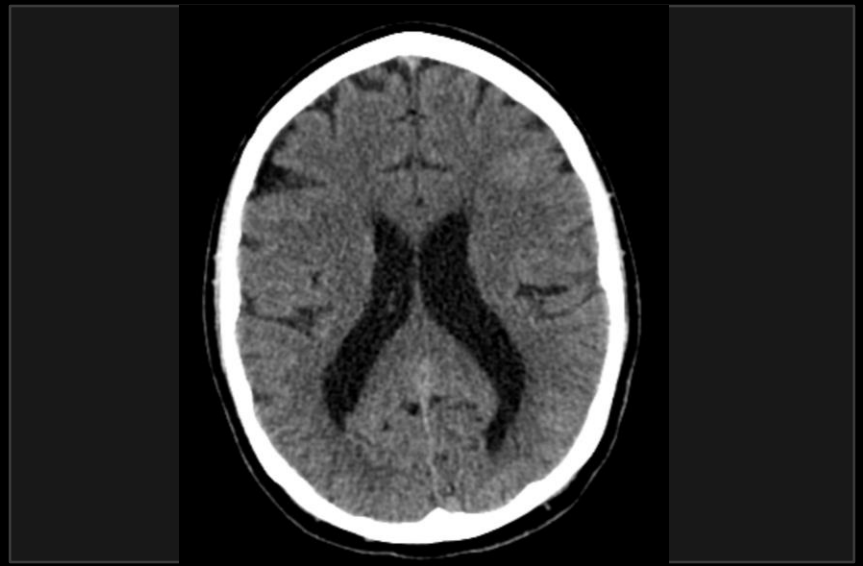
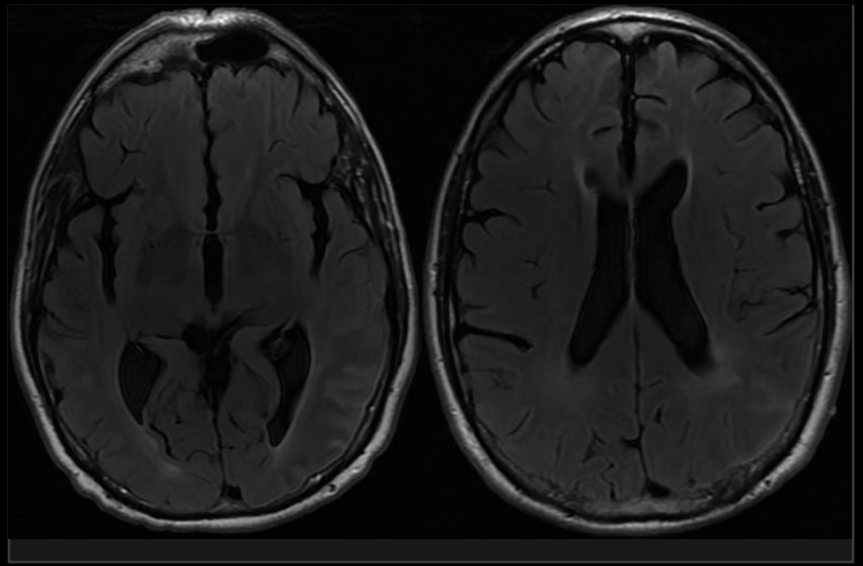
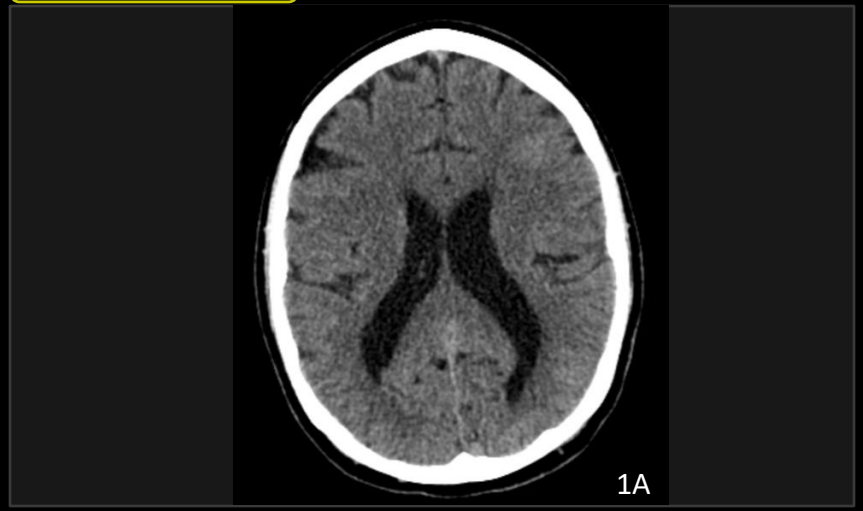
Findings (unlabeled)





Findings (labeled)

CT — Day 0



MRI — Day 0

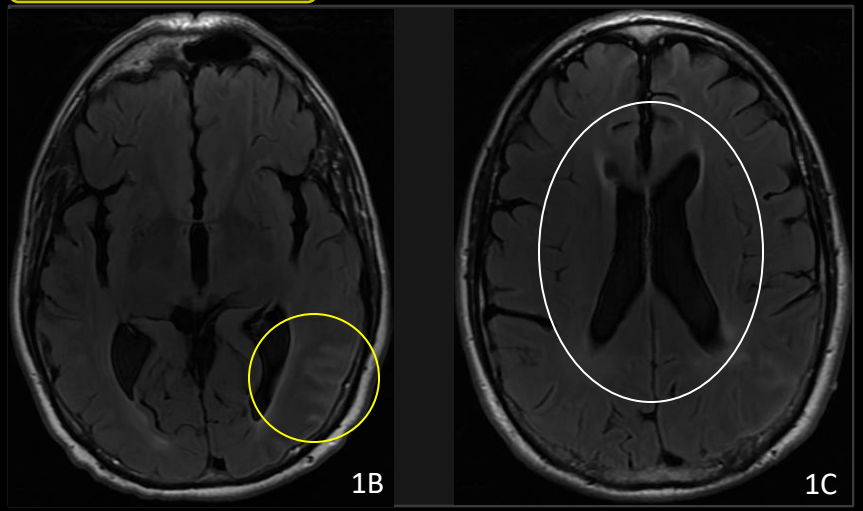


Figure 1.

- (A) CT head without contrast (Day 0): No acute intracranial abnormality
- (B, C) MRI (Day 0): Increased FLAIR signal in the left temporal cortex and subcortical white matter (yellow circle), and periventricular white matter T2 hyperintensities (white oval)

Leading Dx on discharge:
subacute infarct
(apixaban failure)

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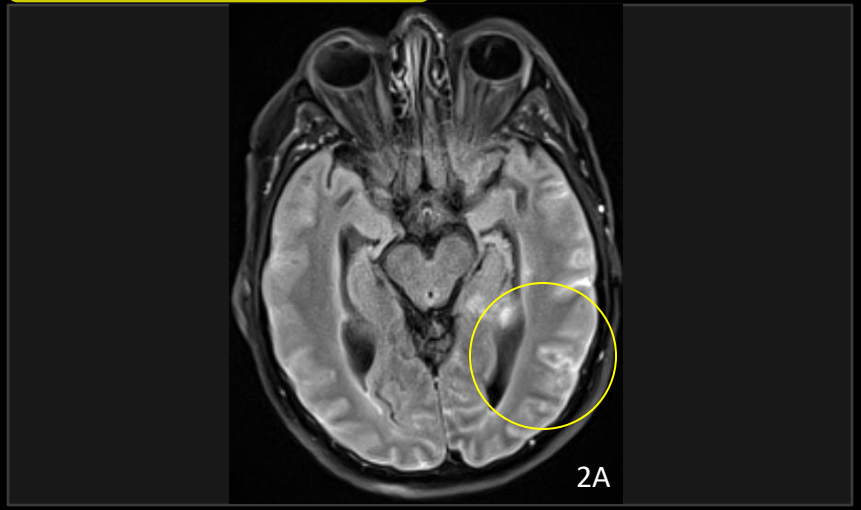
Patient Presentation

- Re-presentation 10 days later with worsening expressive aphasia and persistent left-sided headache
- Serial MRI examinations over the next ~2 months demonstrated asymmetrical cortical and subcortical FLAIR hyperintensities involvement and an interval increase in punctate low-signal foci on susceptibility-weighted imaging
- Serial post-contrast FLAIR images demonstrated interval progression of leptomeningeal enhancement

Patient Presentation

MRI — Day 17



MRI SWI — Day 17

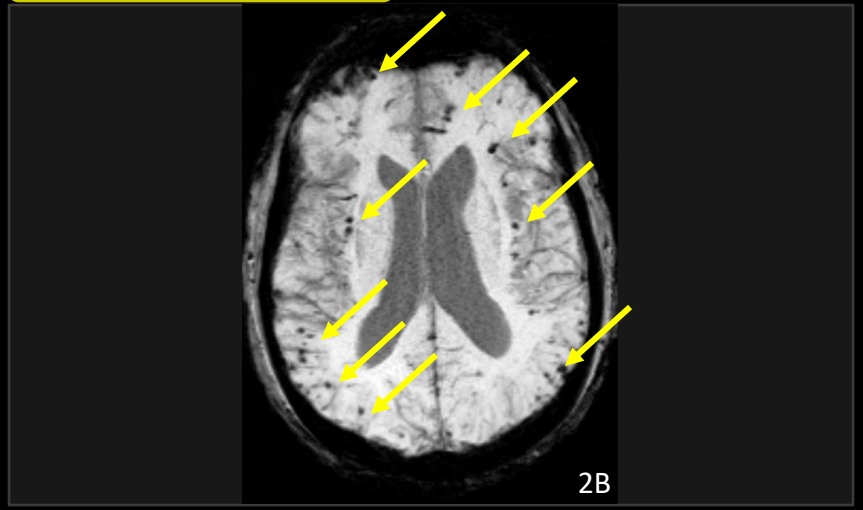


Figure 2.

- (A) MRI Axial FLAIR (Day 17): Additional mild degree of scattered punctate white matter FLAIR hyperintensities in the cortical and subcortical (yellow circle)
- (B) MRI (Day 17): New development of numerous punctate low-signal foci (microbleeds, yellow arrows) throughout bilateral supratentorial hemispheres

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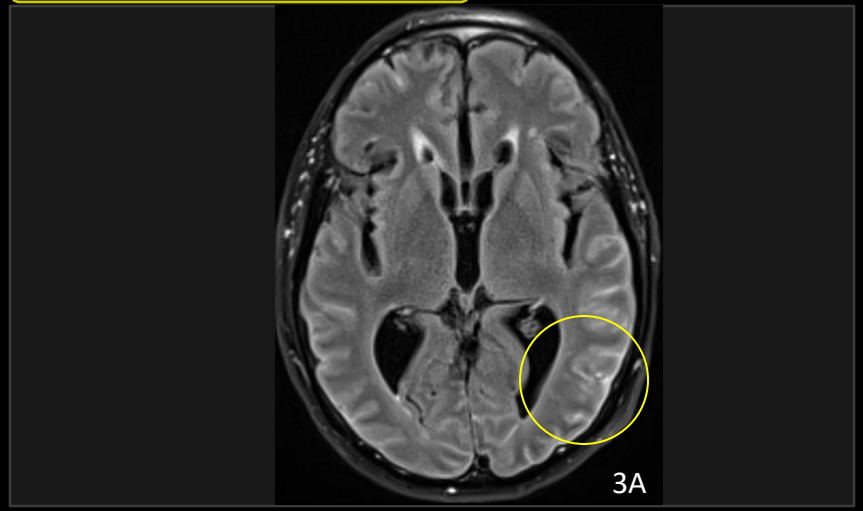
Patient Presentation

MRI — Day 27



MRI SWI — Day 27

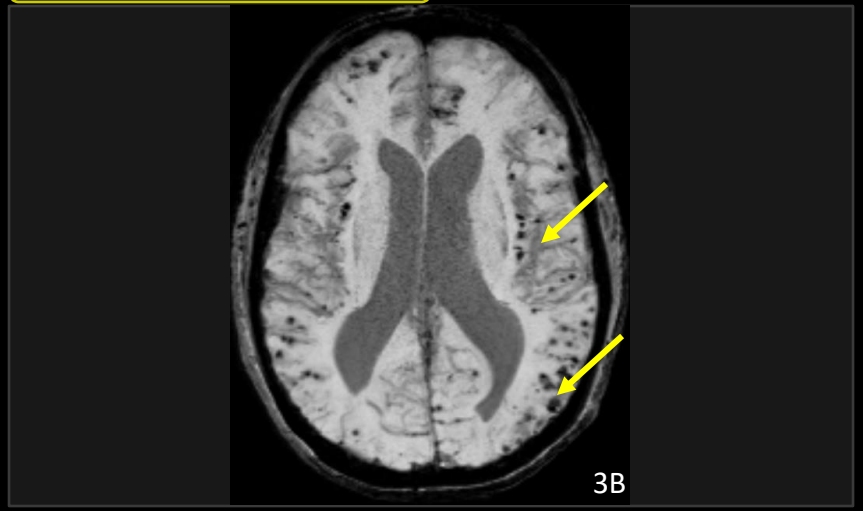


Figure 3.

- (A) MRI Axial Flair (Day 27): Persistent asymmetrical cortical and subcortical white matter FLAIR hyperintensities involving the left temporal lobe (yellow circle)
- (B) MRI (Day 27): Continued development of numerous punctate low-signal foci (microbleeds, yellow arrows) throughout bilateral supratentorial hemispheres

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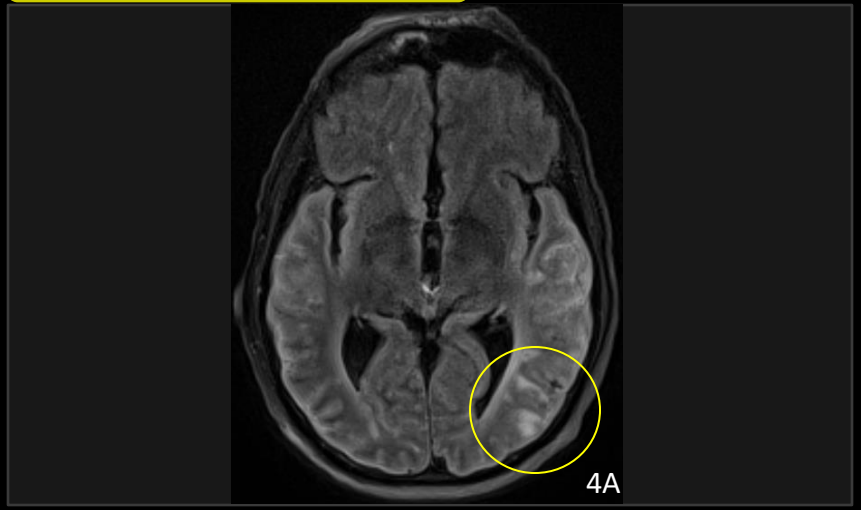
Patient Presentation

MRI FLAIR — ~2 months



MRI SWI — ~2 months

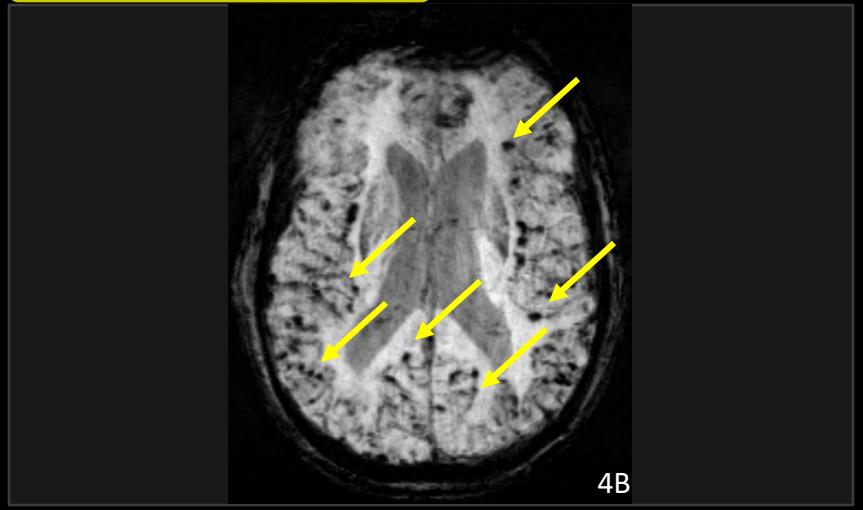


Figure 4.

- (A) MRI Axial FLAIR (~2 months): Increased cortical/subcortical FLAIR hyperintensities, particularly in the left temporo-occipital region (yellow circle)
- (B) MRI (~2 months): Continued interval increase in new punctate low-signal foci throughout bilateral supratentorial hemispheres (yellow arrows)

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Patient Presentation

Subsequent workup in the meantime:

- CSF (lumbar puncture): Pleocytosis and elevated protein; no evidence of infection
- Positive antinuclear antibody with elevated titers
- Negative GAD65 and NMDA receptor antibodies; extensive infectious workup negative
- Echocardiography and carotid ultrasound: Unrevealing for cardioembolic source
- Carotid cerebral angiogram: Low suspicion for vasculitis (small-vessel disease not fully excluded)

Serial MRI: Hyperintensities Progression

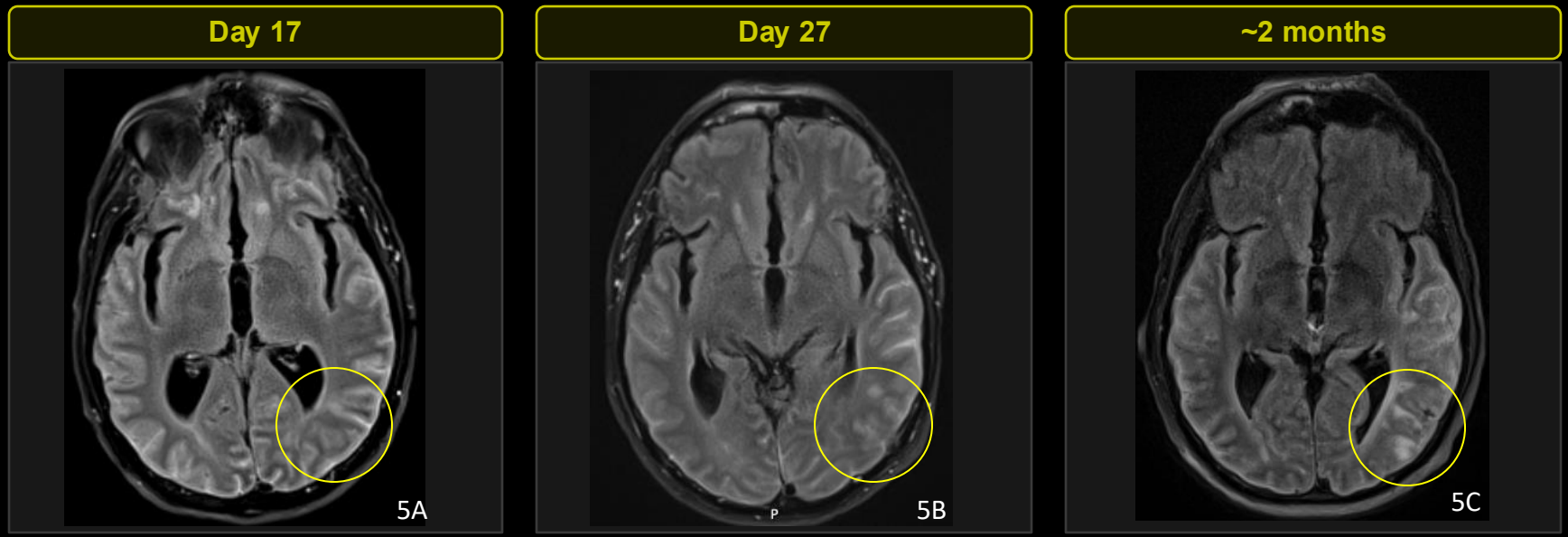

Figure 5. Axial FLAIR at comparable anatomic levels

- (A, B, C) Serial axial FLAIR MRI demonstrating cortical and subcortical white matter hyperintensities progression (yellow circles)

Serial MRI: Microbleed Progression (SWI)

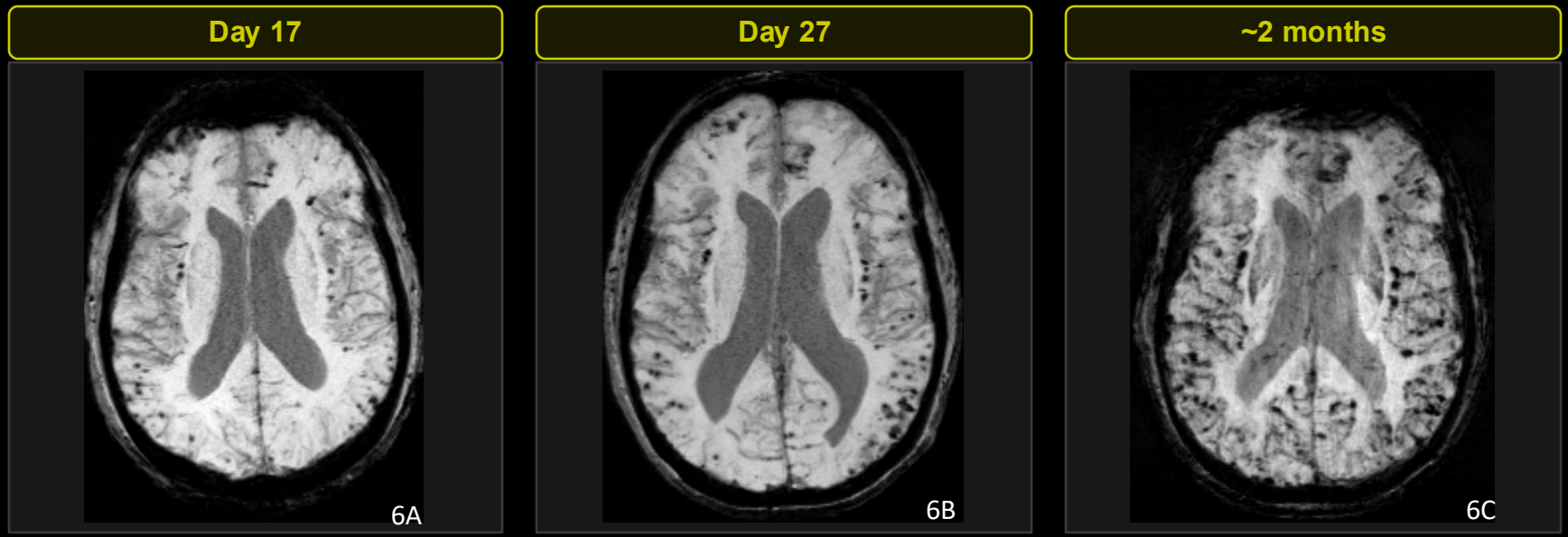

Figure 6.

- (A, B, C) Serial axial SWI demonstrating increasing bilateral microbleeds

Final Dx:

Biopsy-confirmed Cerebral Amyloid Angiopathy-Related Inflammation (CAA-ri)

- Brain biopsy: Perivascular and intramural lymphocytic inflammation, rare multinucleated giant cells, and beta-amyloid deposition in cortical vessel walls on immunohistochemistry — confirming definite CAA-ri

Case Discussion

- CAA-ri is a rare inflammatory subtype of CAA characterized by vascular inflammation with giant-cell reaction among amyloid-laden cerebral vessels.[1]
- Typical presentation: subacute cognitive decline, seizures, or behavioral changes in older patients; may fluctuate over weeks to months.[2]
- Probable CAA-ri defined by Auriel et al. (2016) clinicoradiologic criteria (Table 1); brain biopsy is the gold standard for definite diagnosis.[3]
- Differential: PRES (typically bilateral but asymmetric parieto-occipital vasogenic edema, associated with systemic trigger) and primary CNS vasculitis (infarction, hemorrhage, increased signal intensity on T2/FLAIR, and leptomeningeal enhancement; normal angiogram cannot exclude primary CNS vasculitis).[4,5]

- Treatment: Immunosuppression is associated with meaningful clinical and radiographic improvement; early recognition is essential.[6]

Case Discussion

Diagnostic Component	Possible CAA-RI	Probable CAA-RI
Age	≥ 40 years	≥ 40 years
Clinical Features	≥ 1 symptom: headache, decrease in consciousness, behavioral change, or focal neurological deficits and seizures	≥ 1 symptom: headache, decrease in consciousness, behavioral change, or focal neurological deficits and seizures
MRI Findings	White matter hyperintensities extending to the immediately subcortical white matter	Unifocal or multifocal white matter hyperintensities extending to the immediate subcortical white matter with asymmetric distribution not attributable to prior intracerebral hemorrhage.
Hemorrhagic Lesions	≥ 1 corticosubcortical hemorrhagic lesions: cerebral macrobleed, cerebral microbleed, or cortical superficial siderosis	≥ 1 corticosubcortical hemorrhagic lesions: cerebral macrobleed, cerebral microbleed, or cortical superficial siderosis
Alternative Diagnosis	Excluded	Excluded
Pathology	Not required	Not required

Table 1. Criteria for the Diagnosis of CAA-ri.

Adapted from Auriel et al. [3].

Case Discussion

- Role of biopsy: Pursued given atypical presentation, rapidly evolving imaging, and inconclusive non-invasive workup. Hemorrhagic risk must be weighed against diagnostic benefit.
- Treatment and outcome: Steroid therapy initiated; despite treatment, hospital course complicated by altered mental status, urinary retention, dysphagia, and electrolyte derangements. Family elected comfort-focused care based on guarded prognosis; patient transitioned to home hospice and died ~5 months after initial presentation.
- Take-home: Cerebral amyloid angiopathy–related inflammation is a rare and potentially reversible neurologic condition, often resembling common pathologies such as ischemic stroke upon initial presentation. This case emphasizes the diagnostic importance of serial MRI in identifying the characteristic radiological findings of CAA-ri. Early recognition of CAA-ri is critical, and timely initiation of immunosuppressive therapy can result in meaningful clinical improvement.

References:

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3. Auriel E, Charidimou A, Gurol ME, et al. Validation of clinicoradiological criteria for the diagnosis of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol.* 2016;73(2):197-202. doi:10.1001/jamaneurol.2015.4078
4. Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions. *Lancet Neurol.* 2015;14(9):914-925. doi:10.1016/S1474-4422(15)00111-8
5. Salvarani C, Brown RD Jr, Hunder GG. Adult primary central nervous system vasculitis. *Lancet.* 2012;380(9843):767-777. doi:10.1016/S0140-6736(12)60069-5
6. Regenhardt RW, Thon JM, Das AS, et al. Association between immunosuppressive treatment and outcomes of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol.* 2020;77(10):1261-1269. doi:10.1001/jamaneurol.2020.1782

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