AANP: Diagnostic Slide Session 2014 – Case 03

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Disclosures

I have no financial relationships / conflicts of interest to disclose

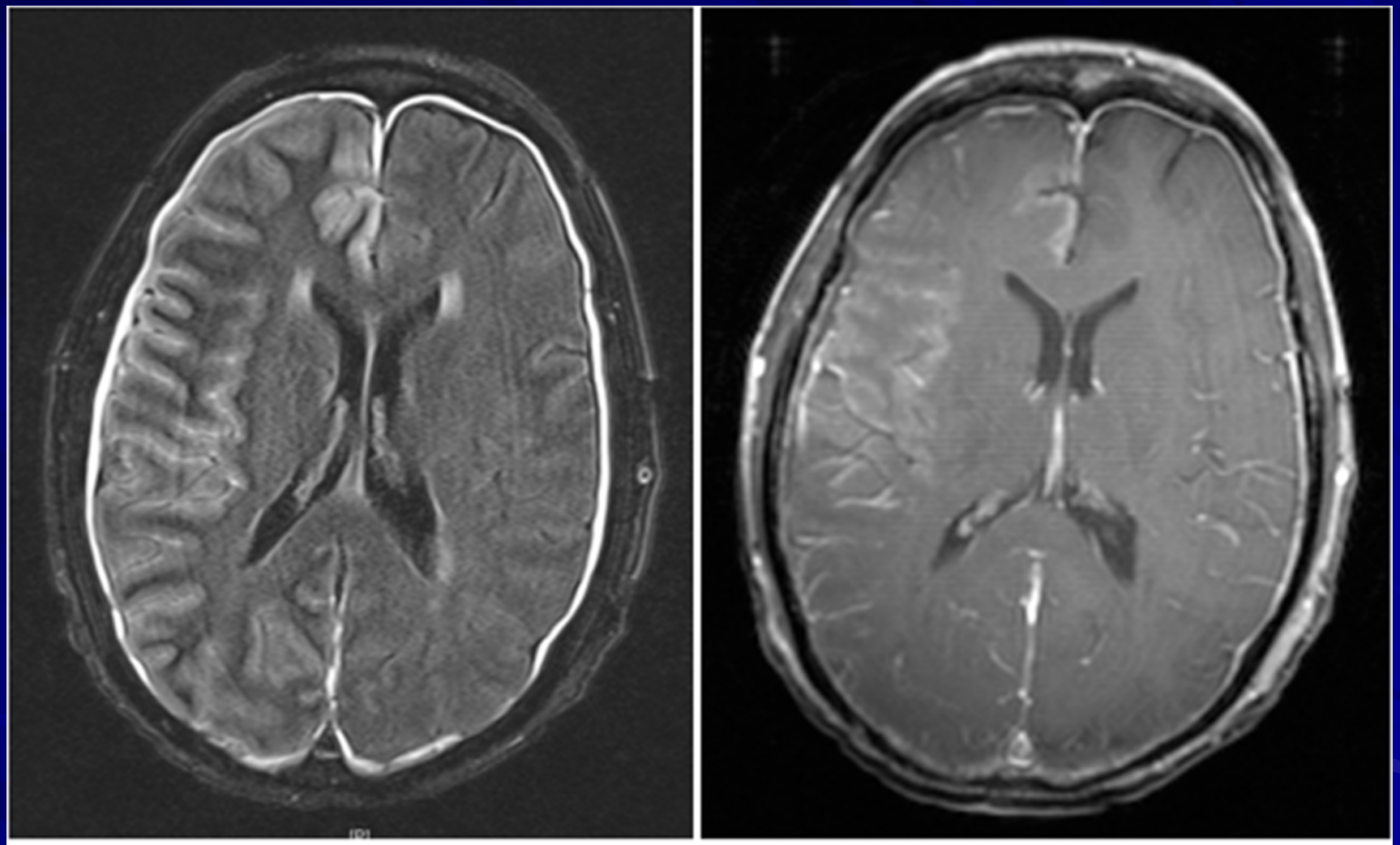
Clinical History

- 57 year-old male who presents with headache and altered mental status
  - History of NSCLC status post pneumonectomy and whole brain XRT for brain metastases, 8 yrs prior to presentation
- Neurologic exam:
  - Follow commands
  - Left facial droop, left hemiplegia, and left sided neglect with right gaze preference

Radiologic Studies

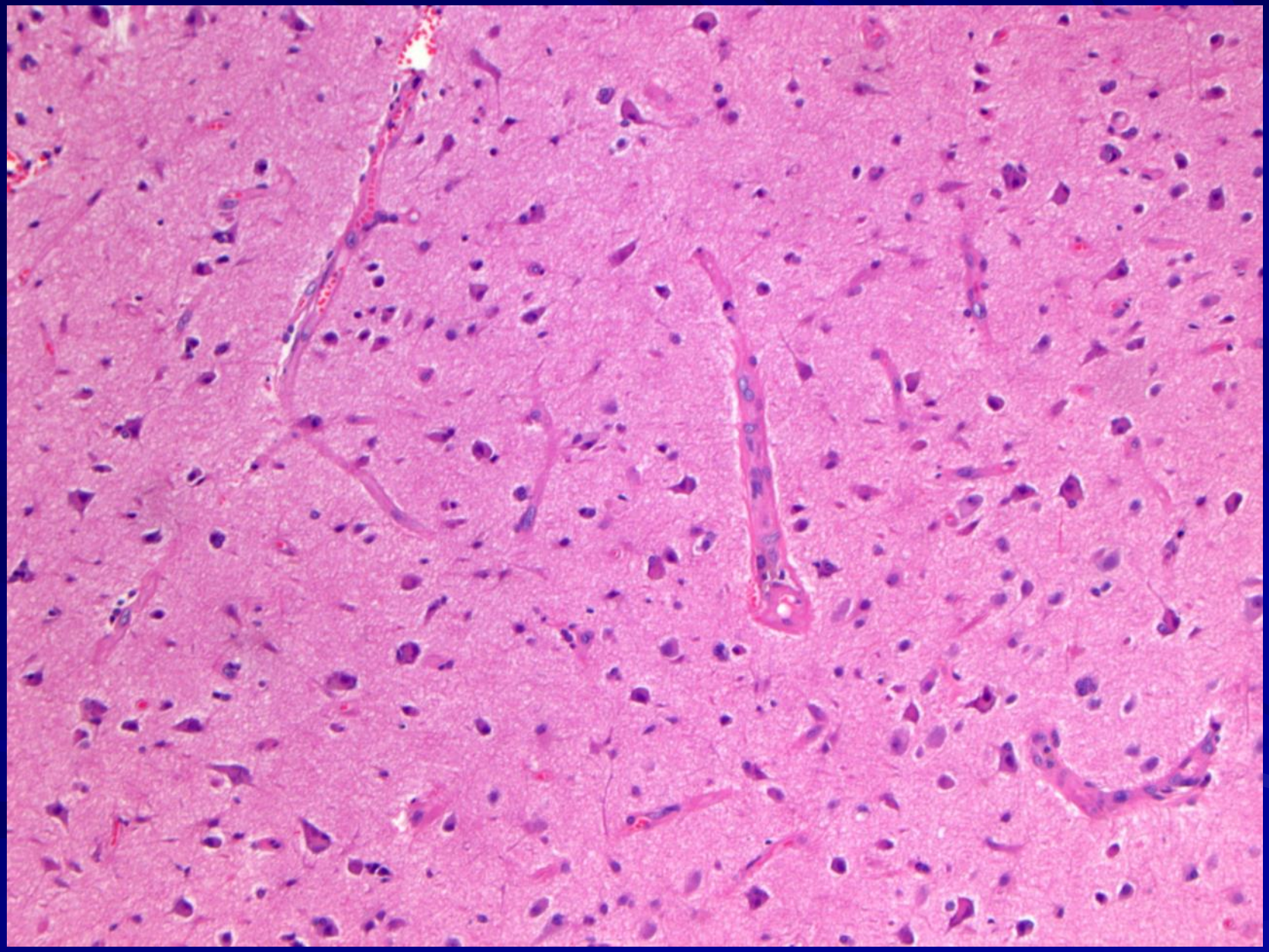
- Head CT: Old lacunar infarct but no acute hemorrhage

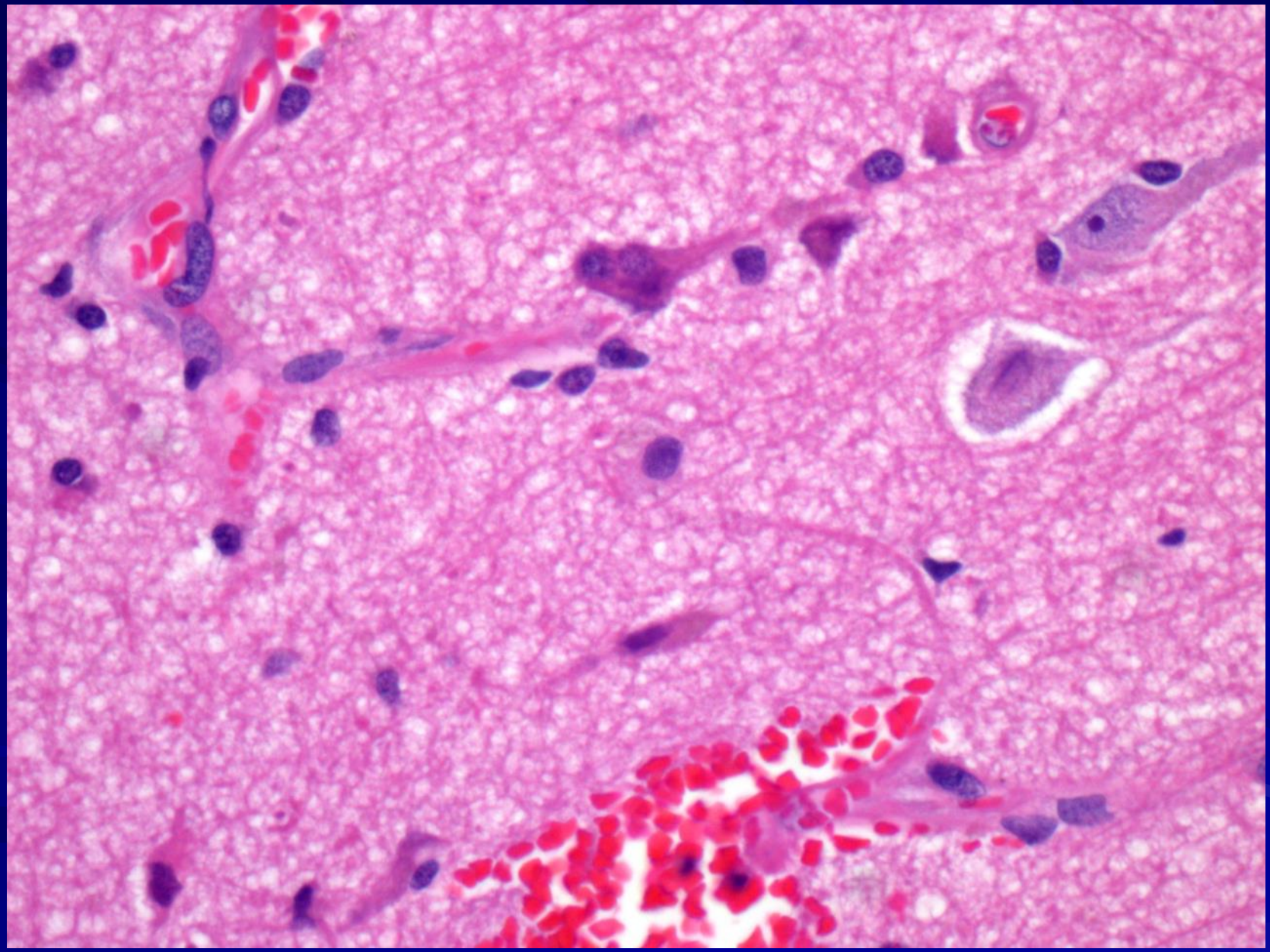
- Brain MRI:
  - Diffuse right hemisphere cortical expansion with abnormal T2/FLAIR signal and associated restricted diffusion.
  - Diffuse leptomeningeal enhancement in the right cerebral hemisphere

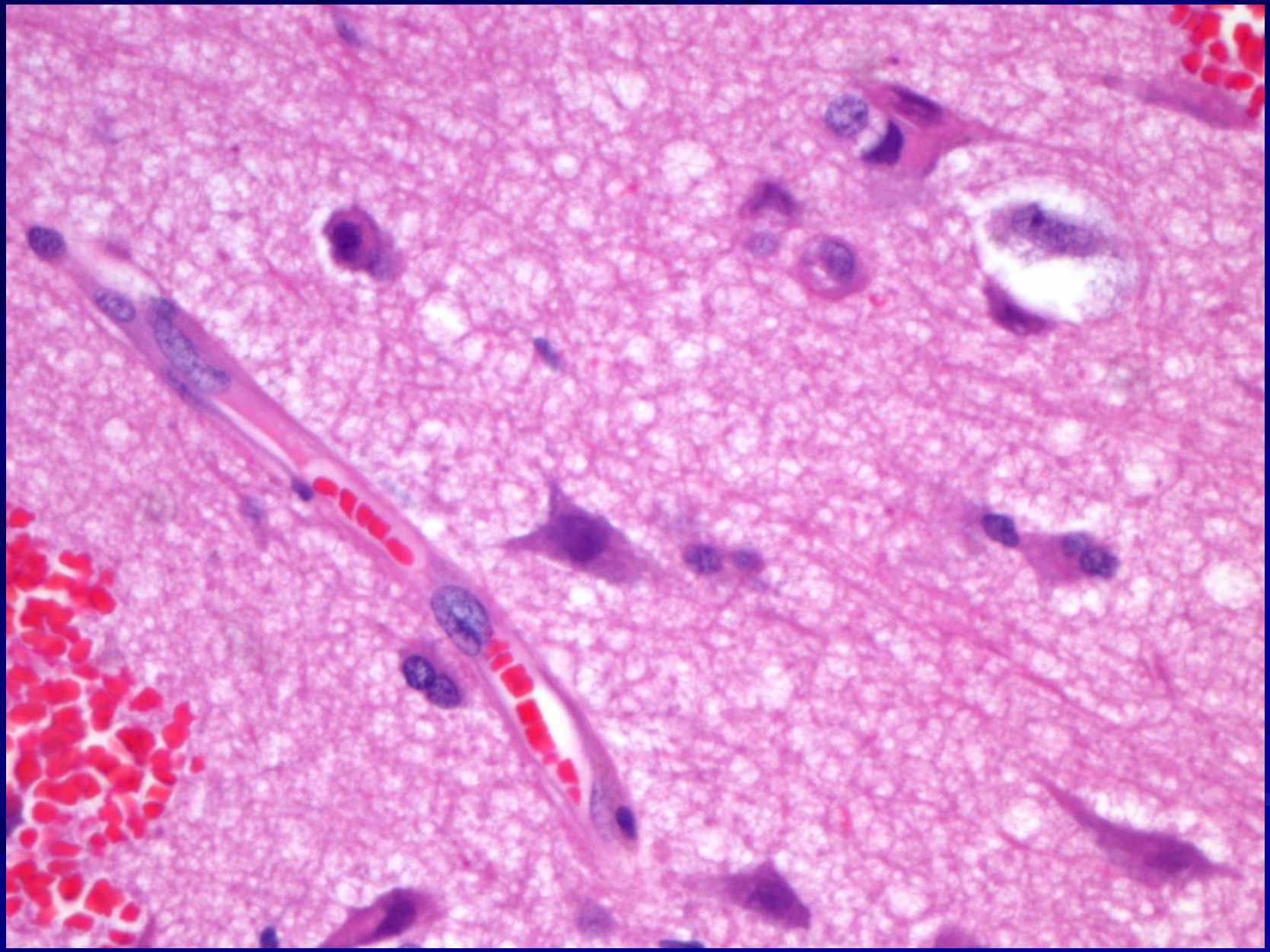


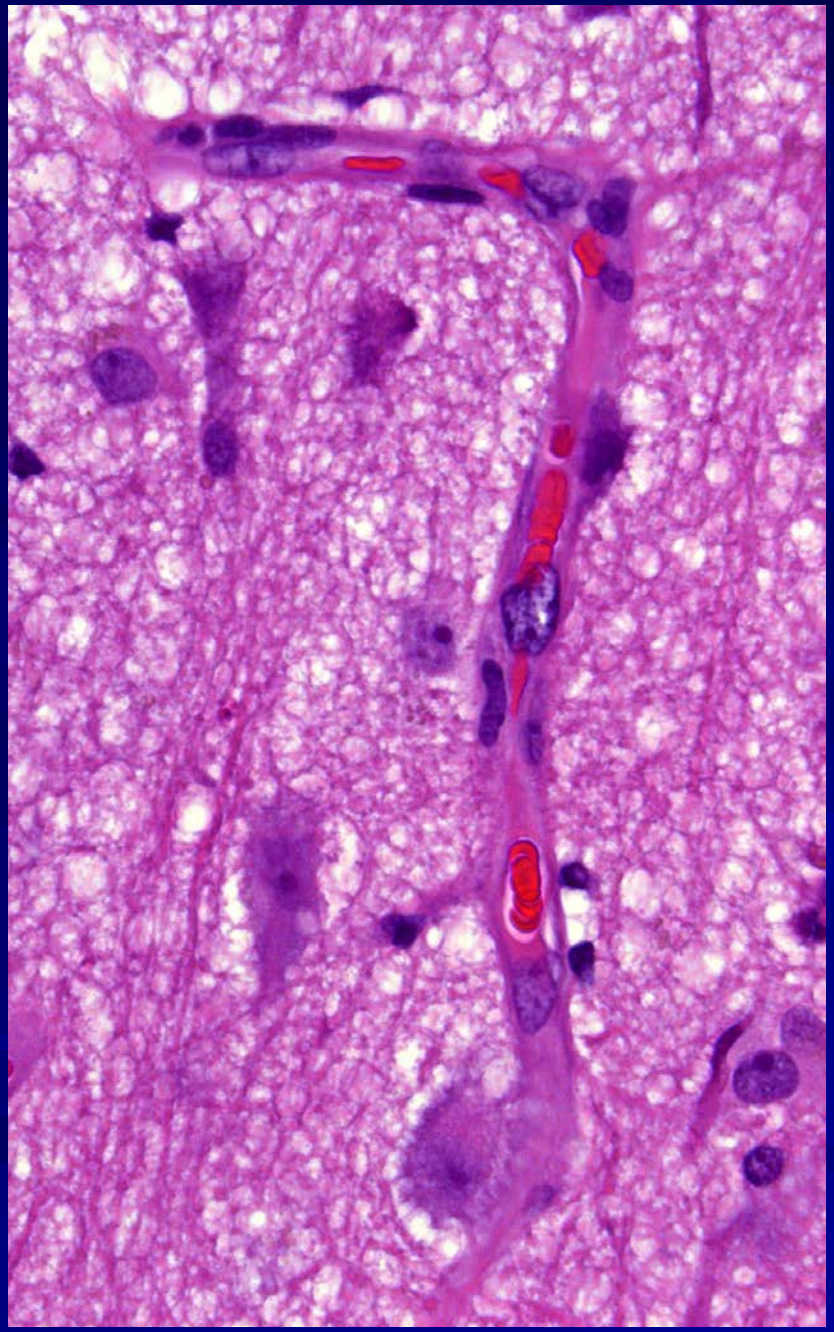
Brain MRI. Left: Axial T1 Post contrast, Right: Axial Flair T2

A biopsy was performed of the “Right frontal lesion”

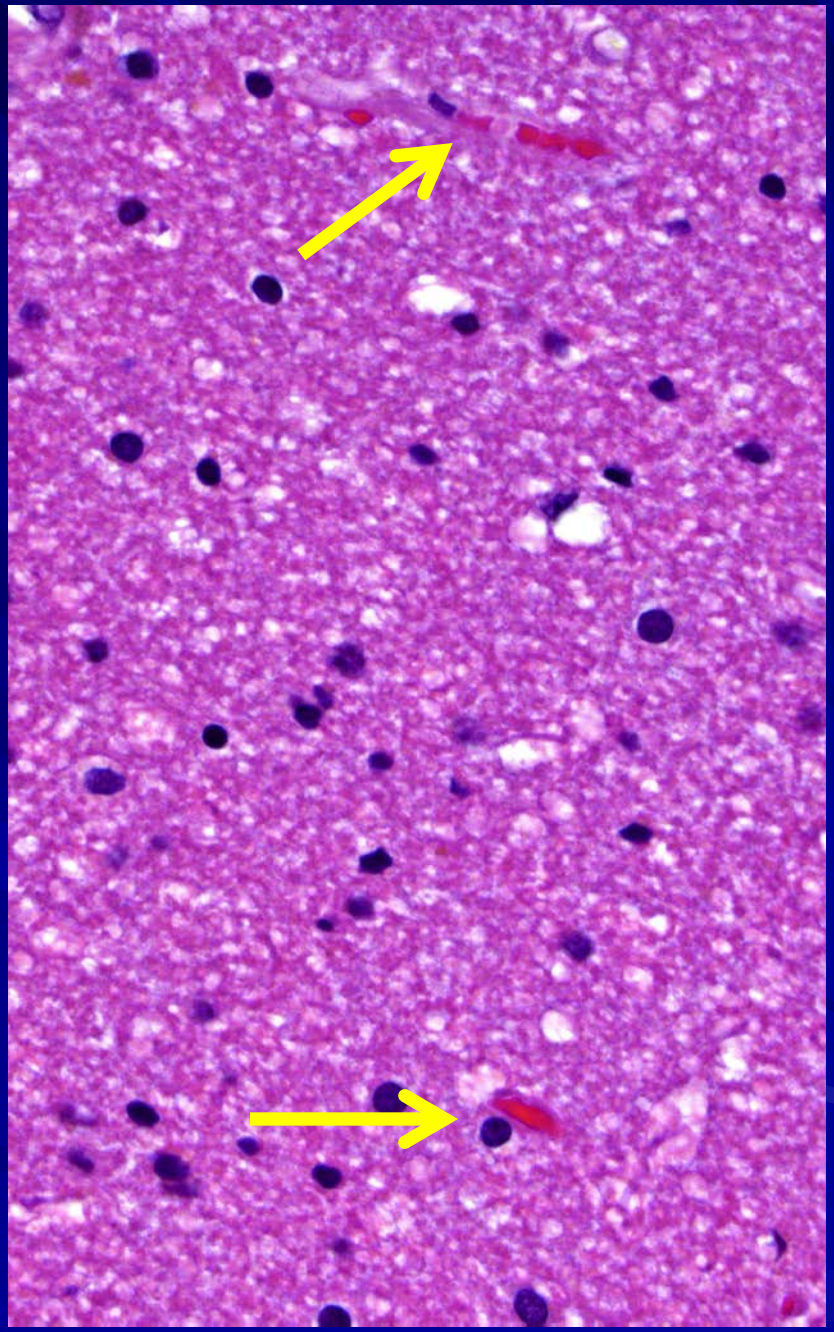




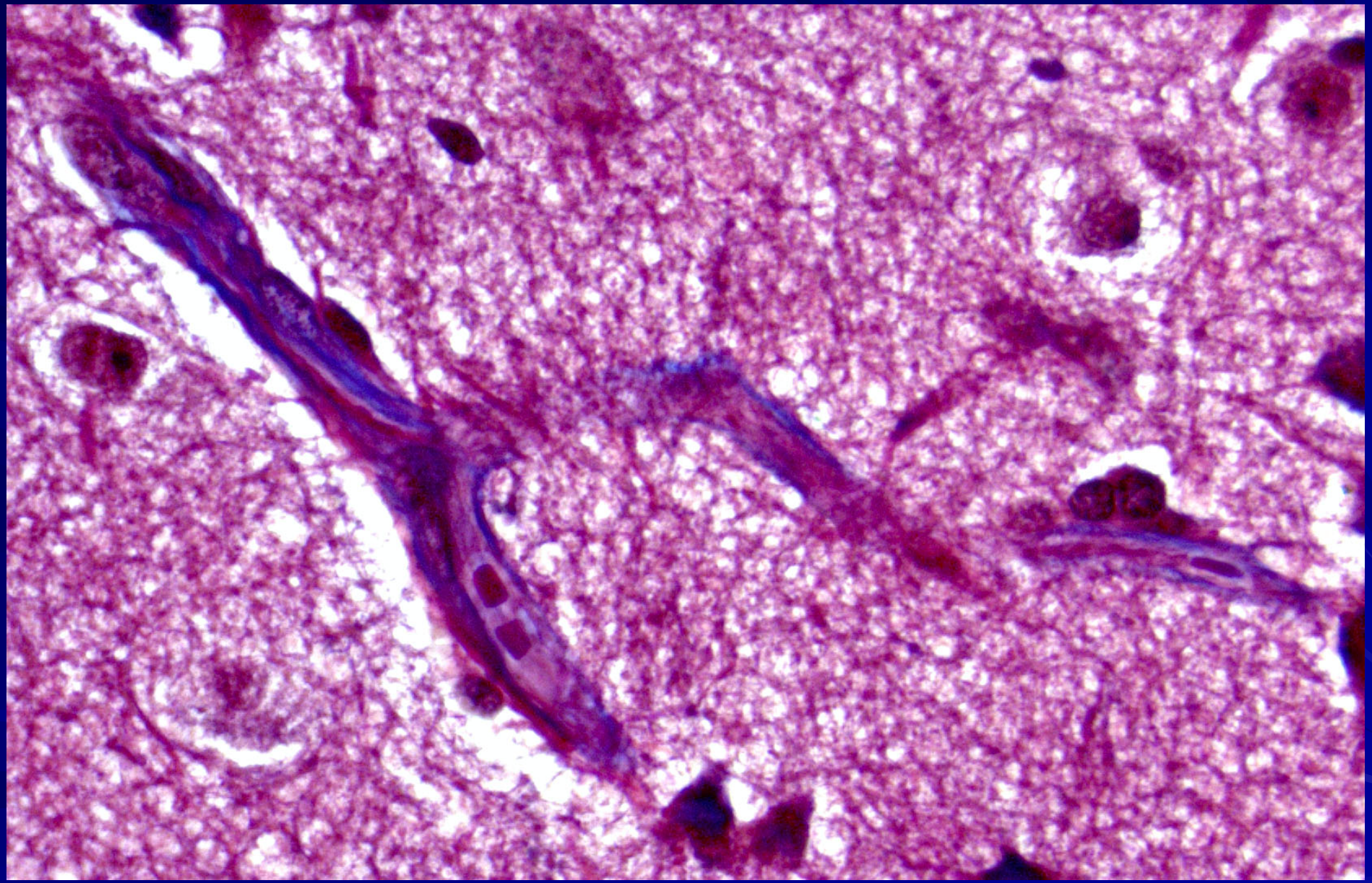




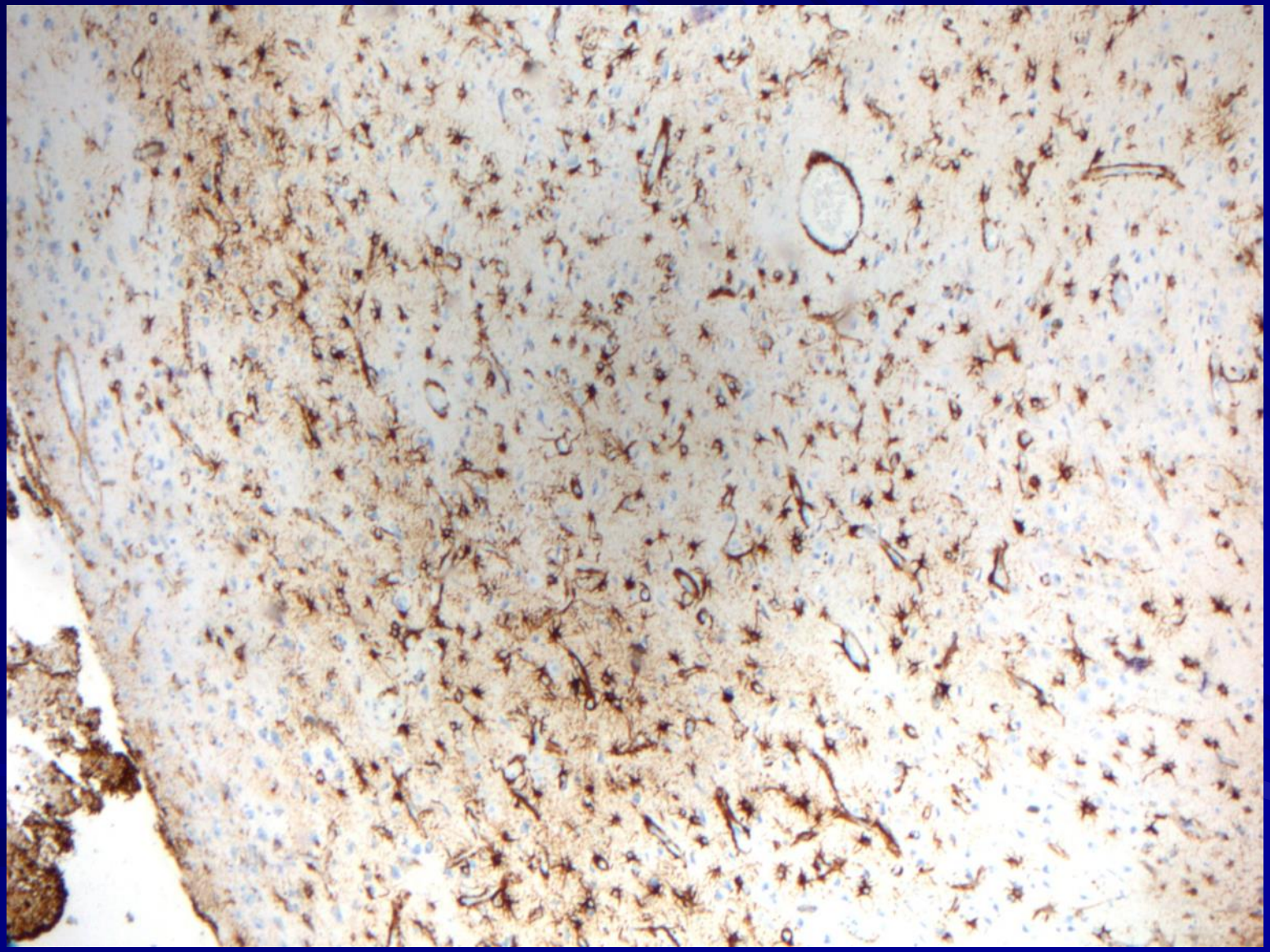
Cortex



Underlying white matter



trichrome



GFAP

Diagnosis

- Brain tissue with vasculopathy and severe gliosis, see note.
  - Changes of capillary fibrosis and endothelial reactive atypia are likely secondary to prior radiation therapy.
  - Given the clinical history and neuroimaging characteristics, the entity known as SMART syndrome (stroke-like migraines after radiation therapy) should be considered

Patient follow-up

- 6 months post biopsy, the patient shows significant improvement
  - Cognitive
  - Motor function
- No longer using a wheelchair; walks well with a walker, transitioning to a cane
- Follow-up MRI demonstrated radiation related changes

Final Diagnosis:

SMART SYNDROME

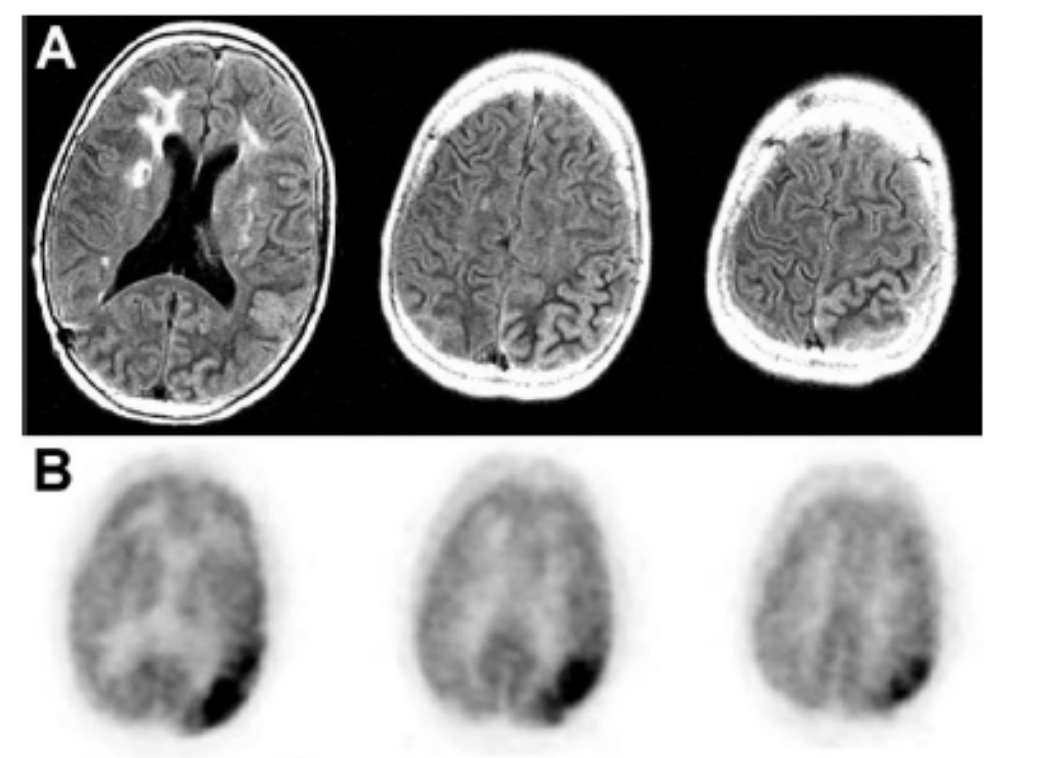
SMART Syndrome

- Episodes of reversible neurological dysfunction
  - Migraine headache ± aura
  - Stroke-like deficits (aphasia, hemiparesis, hemisensory deficits, homonymous hemianopsia)
- Remote history of external beam cranial XRT (dose ≥ 50 Gy)
- Transient, diffuse unilateral cortical gadolinium enhancement on MRI within previous radiation field

Pathophysiology – SMART-ER?

- Pathologic substrate for SMART has not been well-described
  - Gliosis
- Pathophysiology poorly understood

Pathophysiology – SMART-ER?



Pruitt A et al *Neurology* 2006

Hypothesis: Cerebral hyperexcitability with impaired autoregulation, perhaps due to endothelial damage

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