

## IMAGING SPECTRUM OF CEREBRAL VASCULOPATHY

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**Target audience:** Radiologists, Emergency medicine physicians, Neurologists

### Goals and Objectives:

- To illustrate the pathophysiology, imaging appearance and differential diagnosis of cerebral vasculopathy

### Introduction:

Cerebral vasculopathy is a heterogeneous group of disorders associated with vascular wall thickening, luminal irregularity, stenosis and/or dilation. Although relatively rare compared to other cerebrovascular diseases, these disorders are important differential considerations for patients who present with acute cerebral ischemia, hemorrhage, headache or encephalopathy.

### Differential considerations:

- Primary angiitis of CNS (PACNS)
- Vasculitis related to CNS infections
- Vasculitis secondary to inflammatory, autoimmune disease or systemic vasculitis
- RCVS/PRES (Reversible Cerebral Vasoconstriction Syndrome/Posterior Reversible Encephalopathy Syndrome)
- Drug induced cerebral vasculopathy
- Radiation induced cerebral vasculopathy
- Moyamoya disease
- CADASIL (cerebral autosomal dominant arteriopathy w/subcortical infarcts & leukoencephalopathy)

### Diagnostic approaches:

- Imaging modality: CT/CTA, MRI/MRA, MR vessel wall imaging, catheter angiography
- Angiographic findings: Segmental narrowing or beading of intracranial arteries.
- CSF and blood tests
- Brain and leptomeningeal biopsy

### Case 1: Primary angiitis of CNS

History: 46 year old man with progressive weakness and mild cognitive impairment

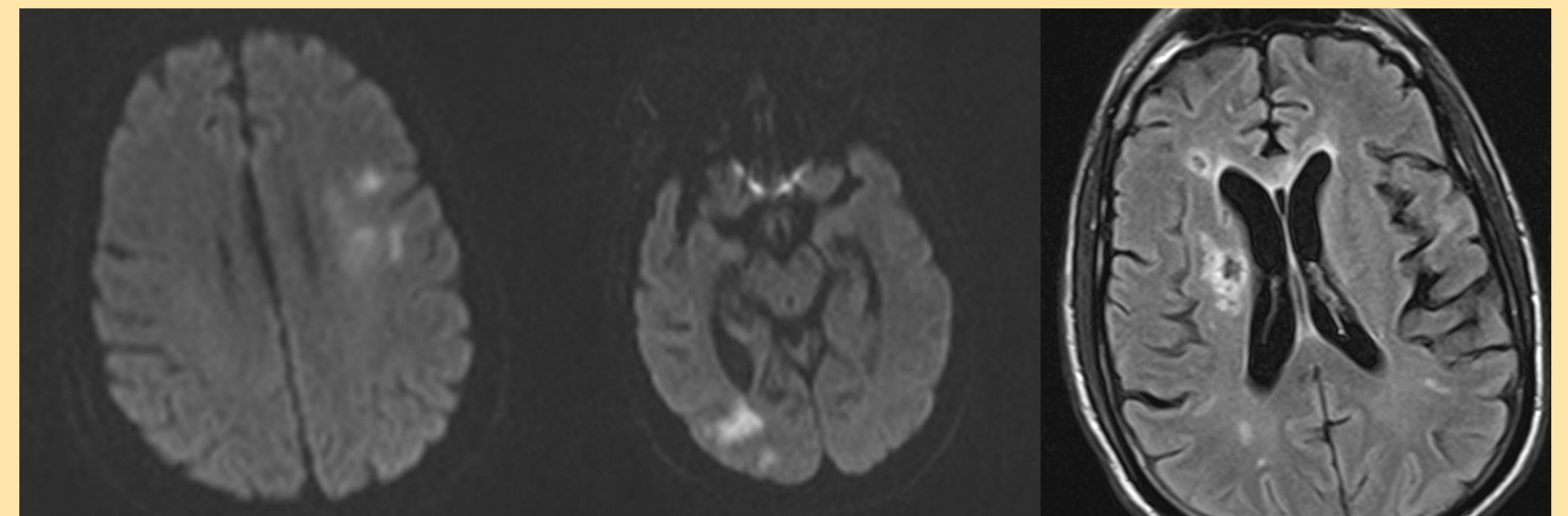


Fig 1a

Fig 1b

Fig 1c

Axial DWI (1a, 1b): restricted diffusion in the left frontal and right occipital lobes, consistent with acute or subacute infarction.

Axial FLAIR (1c): additional chronic lacunar infarctions in the right coronal radiata

3D TOF MRA (1d and 1e): multifocal arterial stenosis involving both anterior and posterior circulation, confirmed on the subsequent catheter angiogram (1g and 1f)

This patient underwent brain biopsy, which showed pathological findings consistent with primary angiitis of CNS.

## Case 2. CNS vasculitis due to infection

History: 57 year old man with recent history of streptococcal meningitis developed right side weakness.



Fig 1d

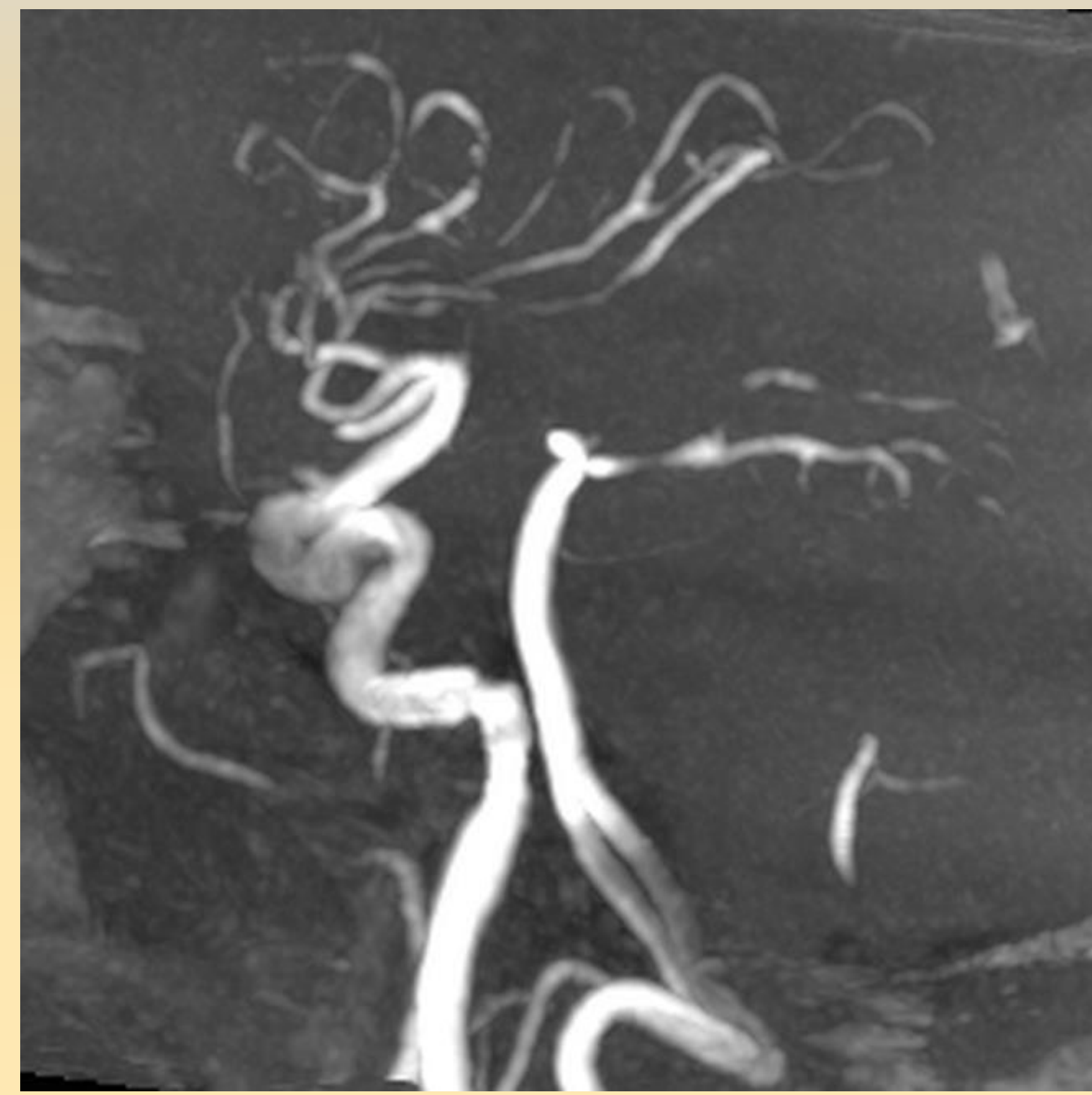


Fig 1e

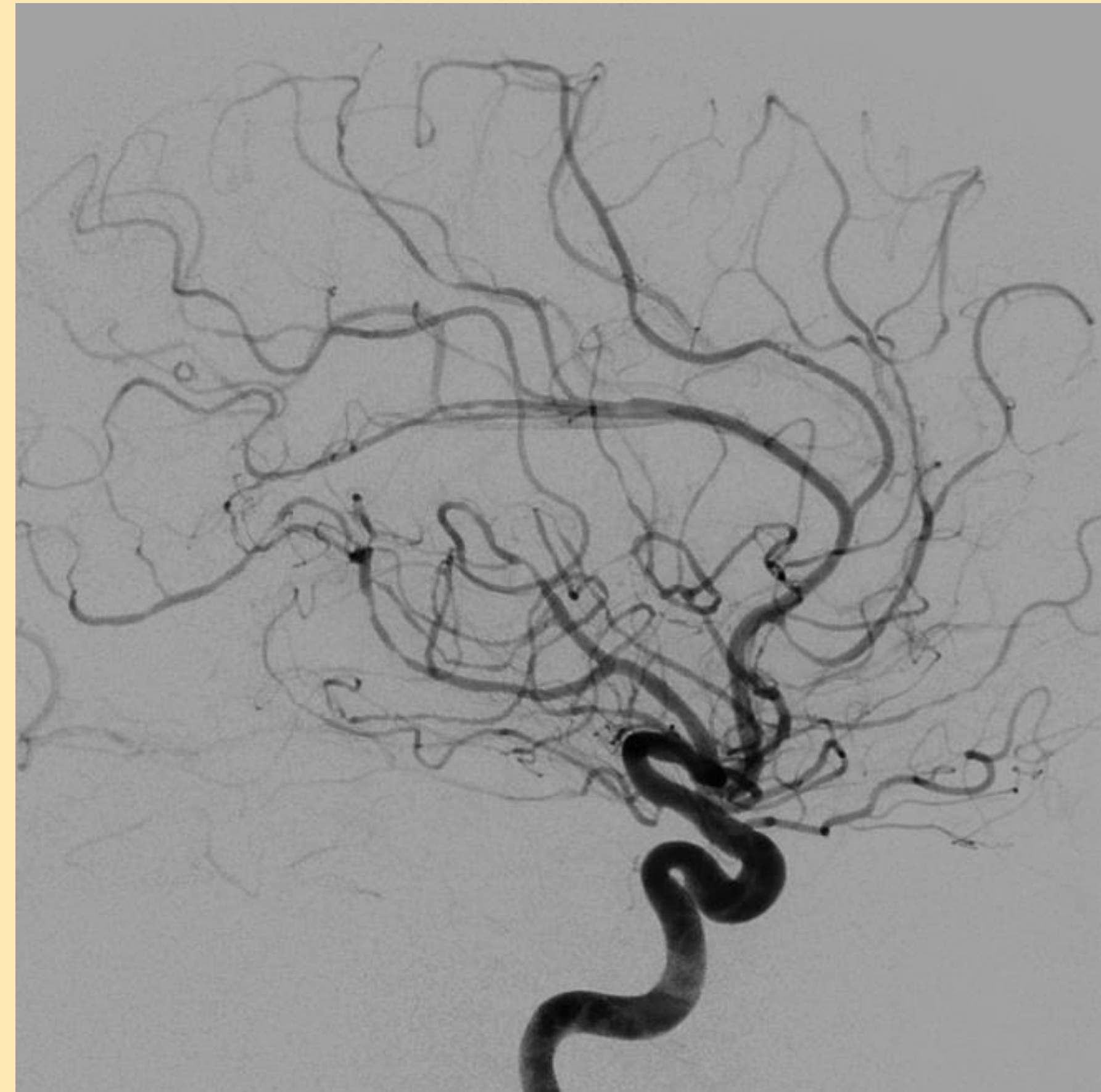


Fig 1f

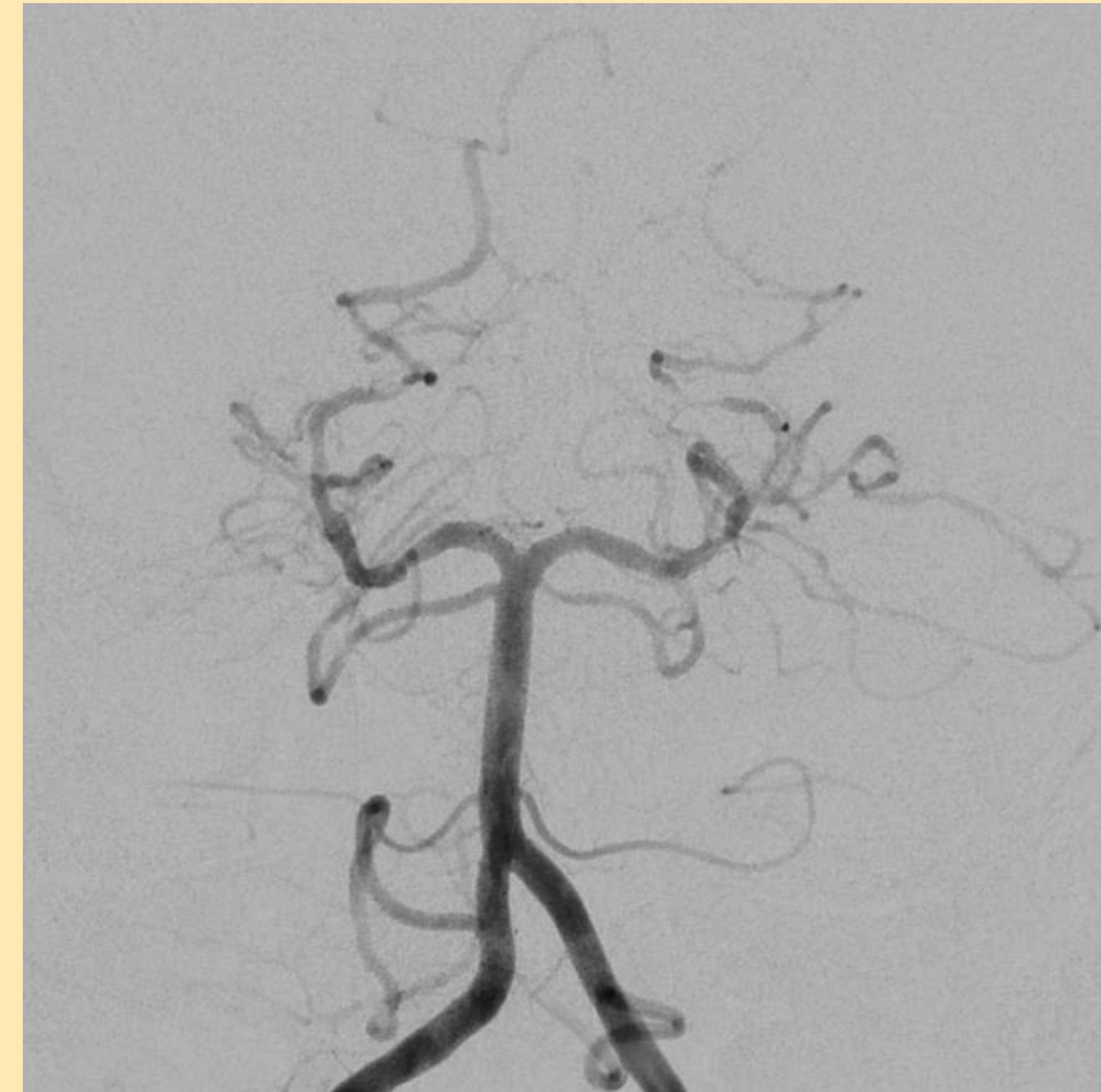


Fig 1g

- **PACNS: characterized by nonatheromatous inflammation and necrosis of the cerebral vasculature without involvement of other organs**
- **Predominantly affects small- and medium-sized arteries.**
- **MRI is sensitive although nonspecific : infarctions in multiple vascular territories and different ages. Other findings: Hemorrhage, white matter disease and parenchymal/leptomeningeal enhancement.**
- **Angiography: multifocal stenosis alternating with normal or dilated segments with “beaded” appearance.**
- **CSF: mild pleocytosis and elevated protein**
- **May require brain or leptomeningeal biopsy for definitive diagnosis**

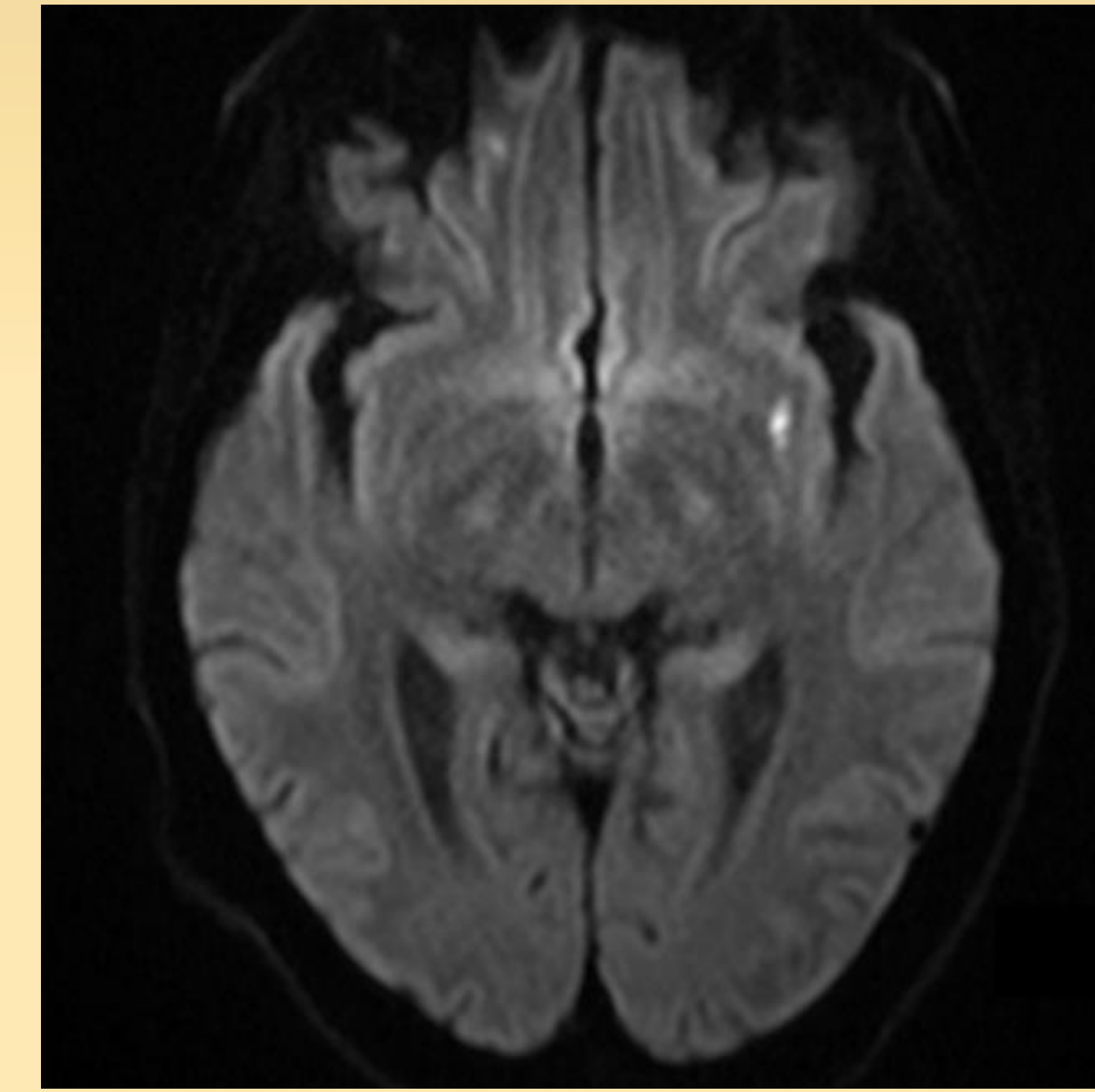


Fig 2a

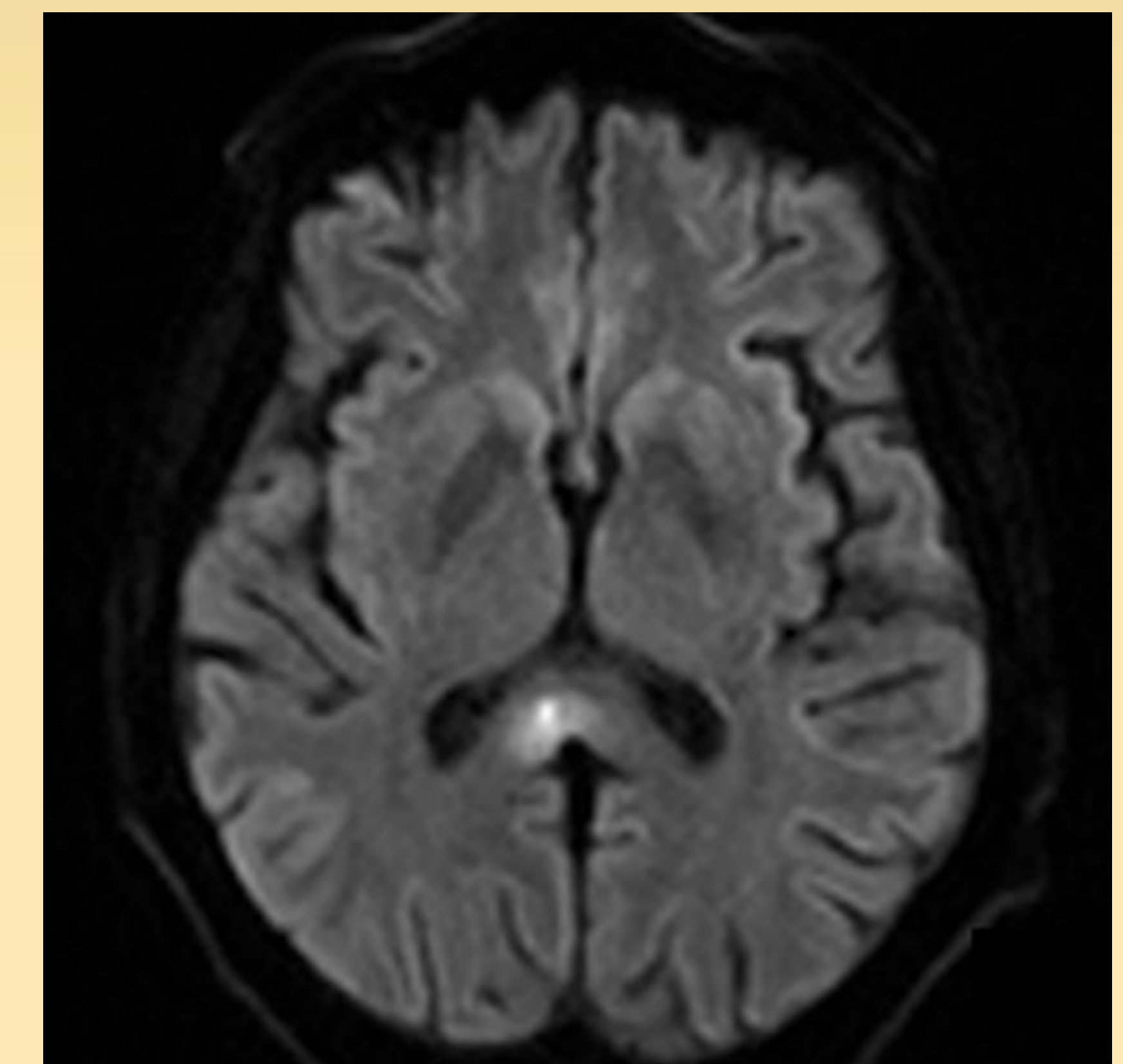


Fig 2b

DWI images (2a, 2b): acute/subacute infarction in the left subinsular white matter and splenium of corpus callosum



Fig 2c



Fig 2d



Fig 2e

TOF MRA (2c): diffuse narrowing of intracranial arteries especially supraclinoid ICAs, basilar artery, and proximal segments of MCAs, ACAs and PCAs, confirmed by catheter angiography (2d, 2e)

- **A number of CNS infections can cause vasculitis or vasospasm: HIV, syphilis, TB, septic emboli, bacterial, viral or fungal**
- **Similar appearance to PACNS on angiography**
- **CSF and blood tests are keys to differentiate**

### Case 3. Vasculitis secondary to systemic inflammatory disease

History: 35 year old woman with SLE presents with altered mental status

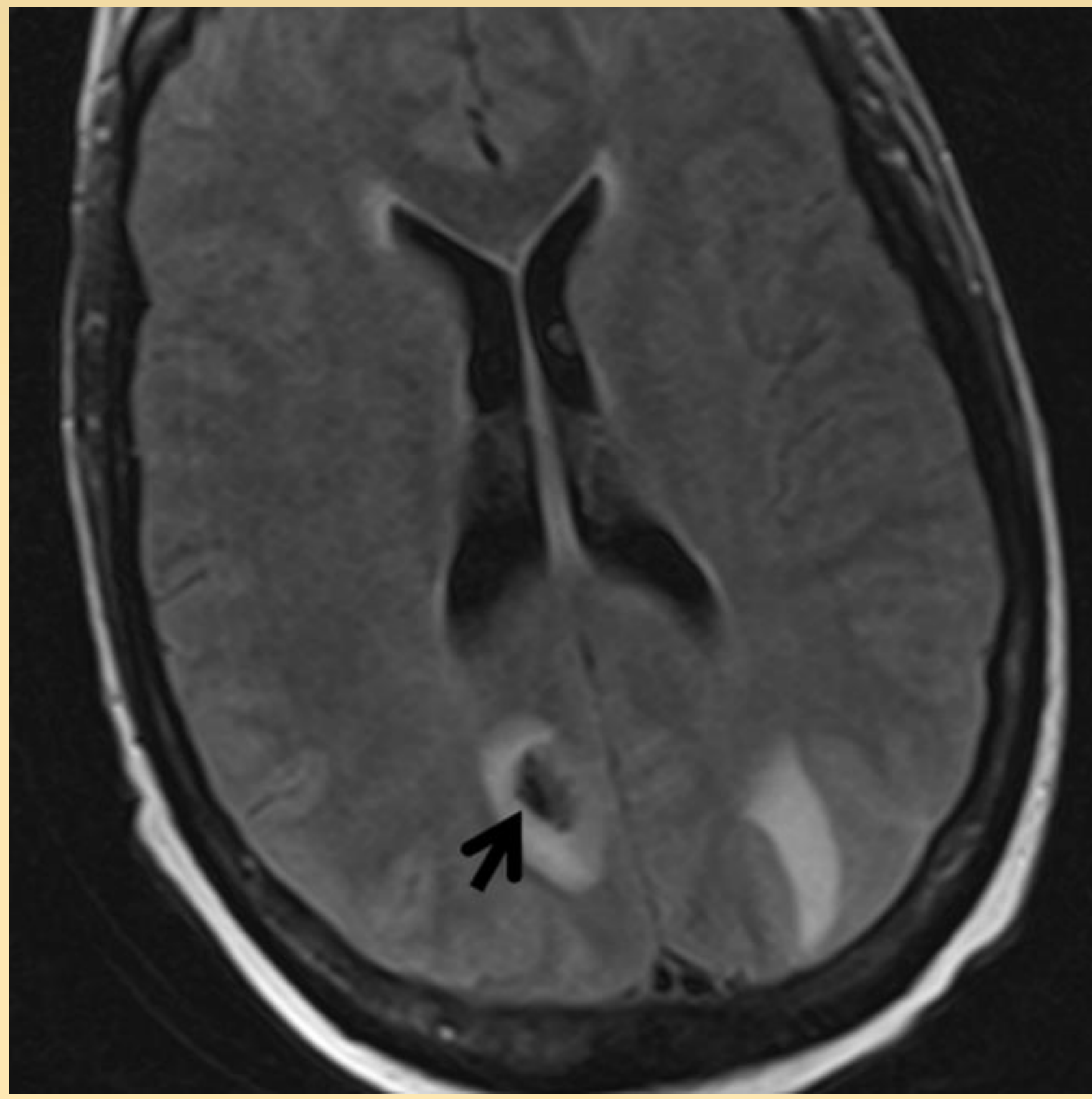


Fig 3a

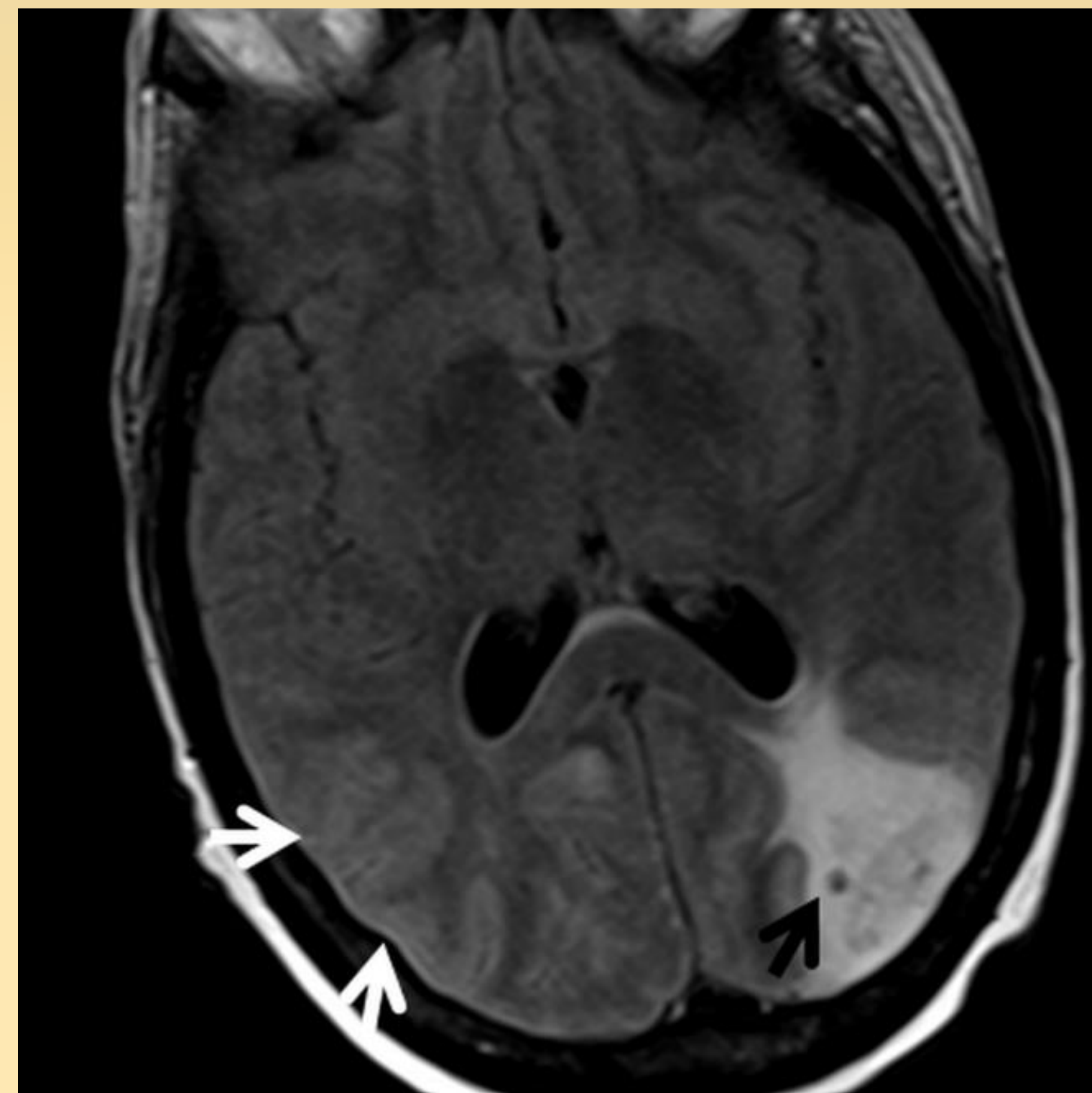


Fig 3b

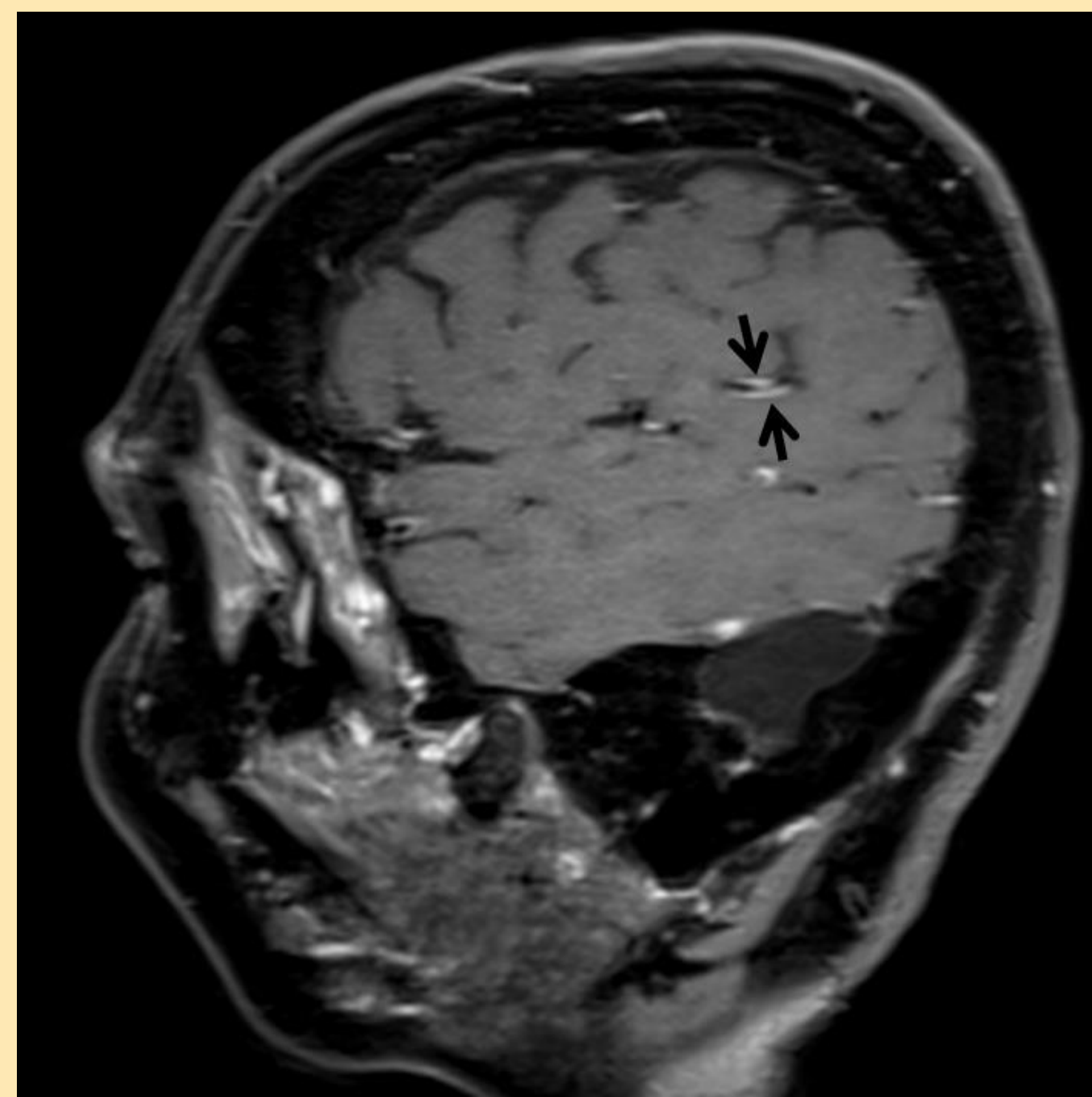


Fig 3c

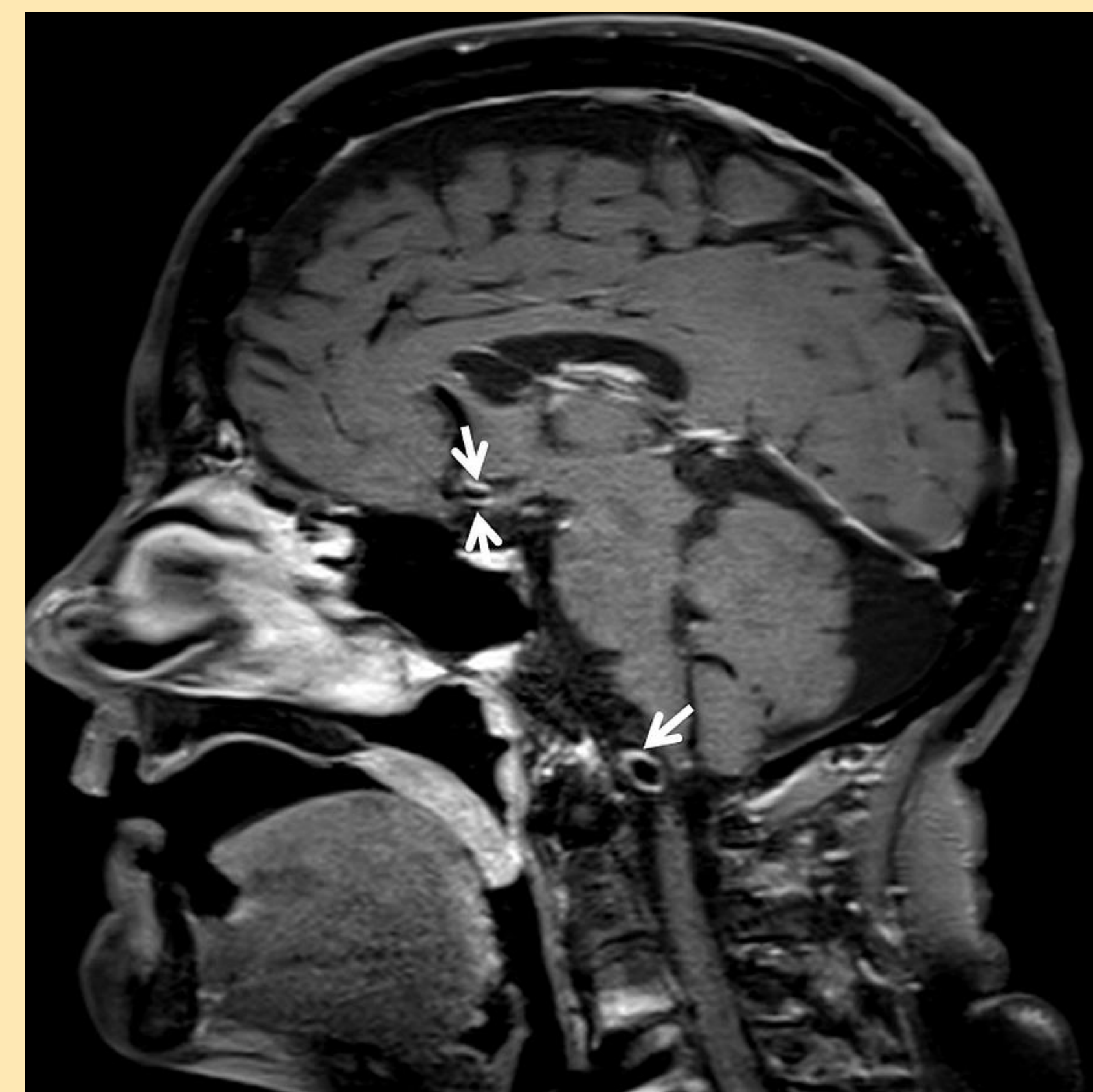


Fig 3d

Axial FLAIR (3a, 3b): edema and small foci of hemorrhage of bilateral cerebral hemispheres, consistent with lupus cerebritis. The MRA demonstrates no large vessel vasculitis (not shown), although the high resolution vessel wall imaging (3c and d) demonstrate concentric wall enhancement of MCA, ACA and basilar artery, suggestive of vasculitic involvement.

- **Many systemic vasculitides or rheumatological diseases can be associated with CNS vasculitis, commonly including SLE, Sjogren's, sarcoidosis, Wegener's, giant cell arteritis etc.**

### Case 4. Posterior reversible cerebral encephalopathy (PRES) and Reversible cerebral vasoconstriction syndrome (RCVS)

History: 28 year old female postpartum patient presents with headache and seizure

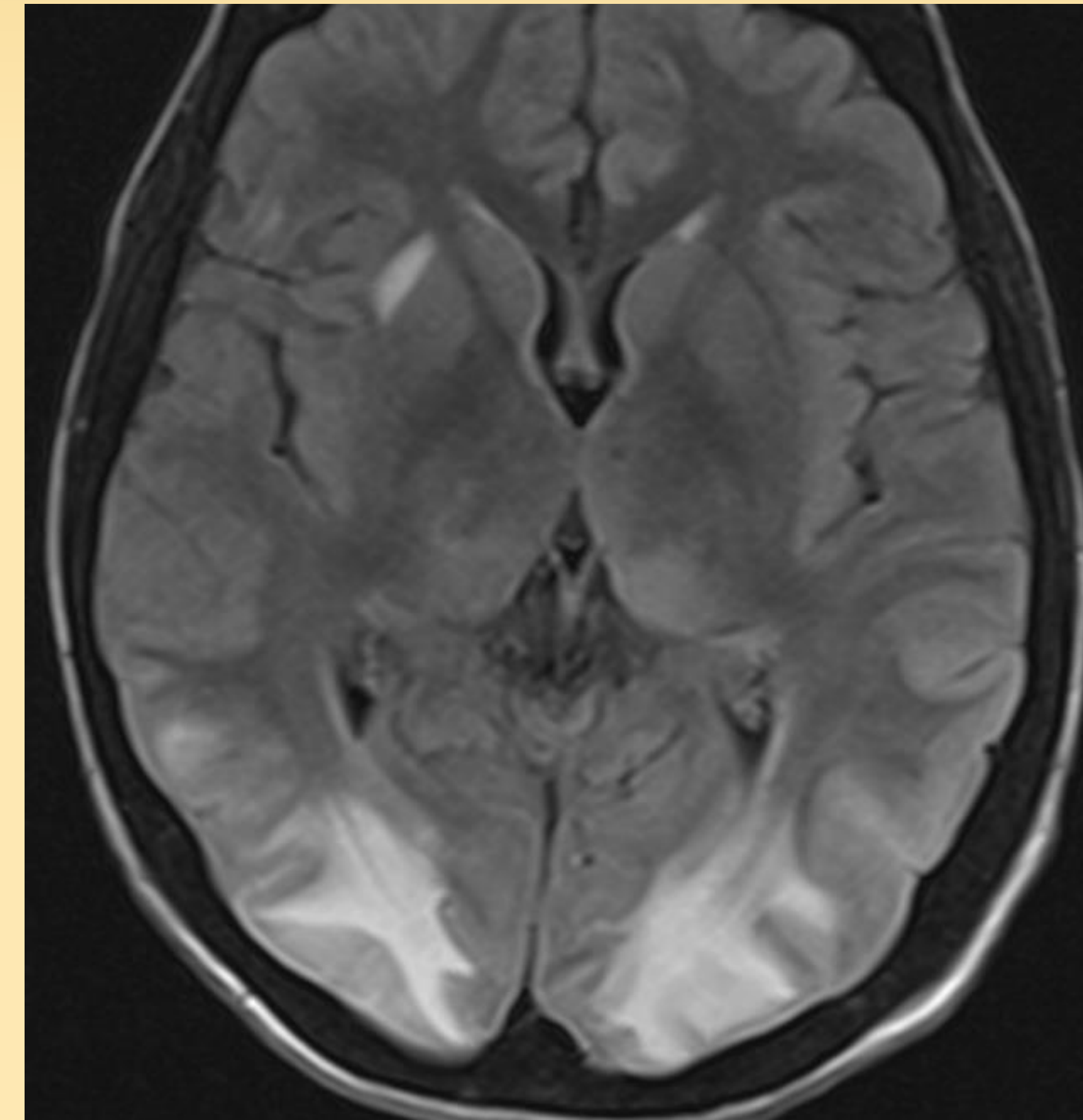


Fig 4a



Fig 4b

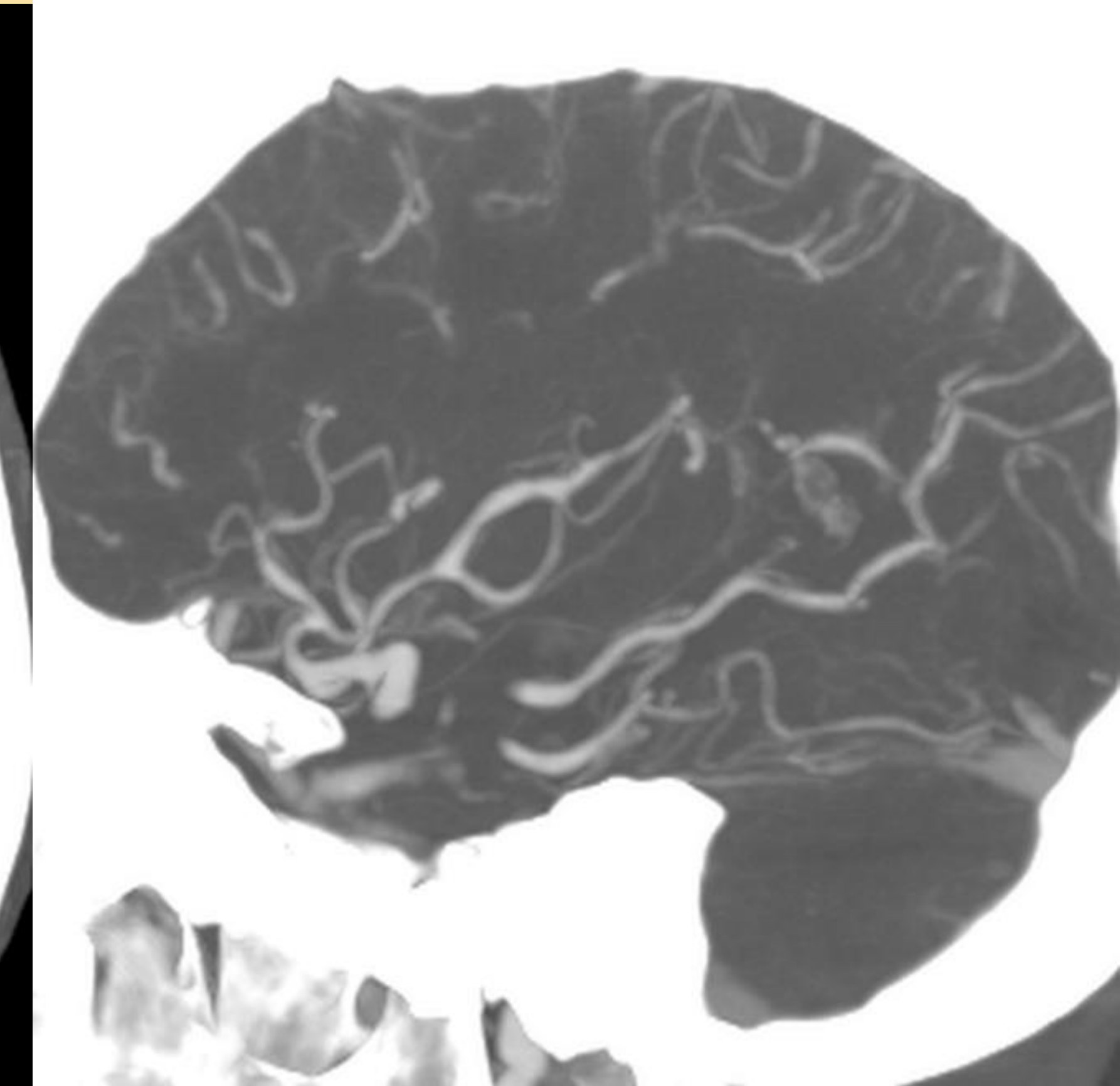


Fig 4c

Axial FLAIR (Fig 4a) demonstrates relatively symmetric vasogenic edema in the bilateral occipital lobe with the pattern consistent with PRES.

Axial and sagittal CTA (Fig 4b and 4c): multifocal arterial stenosis esp MCA branches

Repeat CTA (4d and 4e) in 3 months shows interval resolution of stenosis



Fig 4d

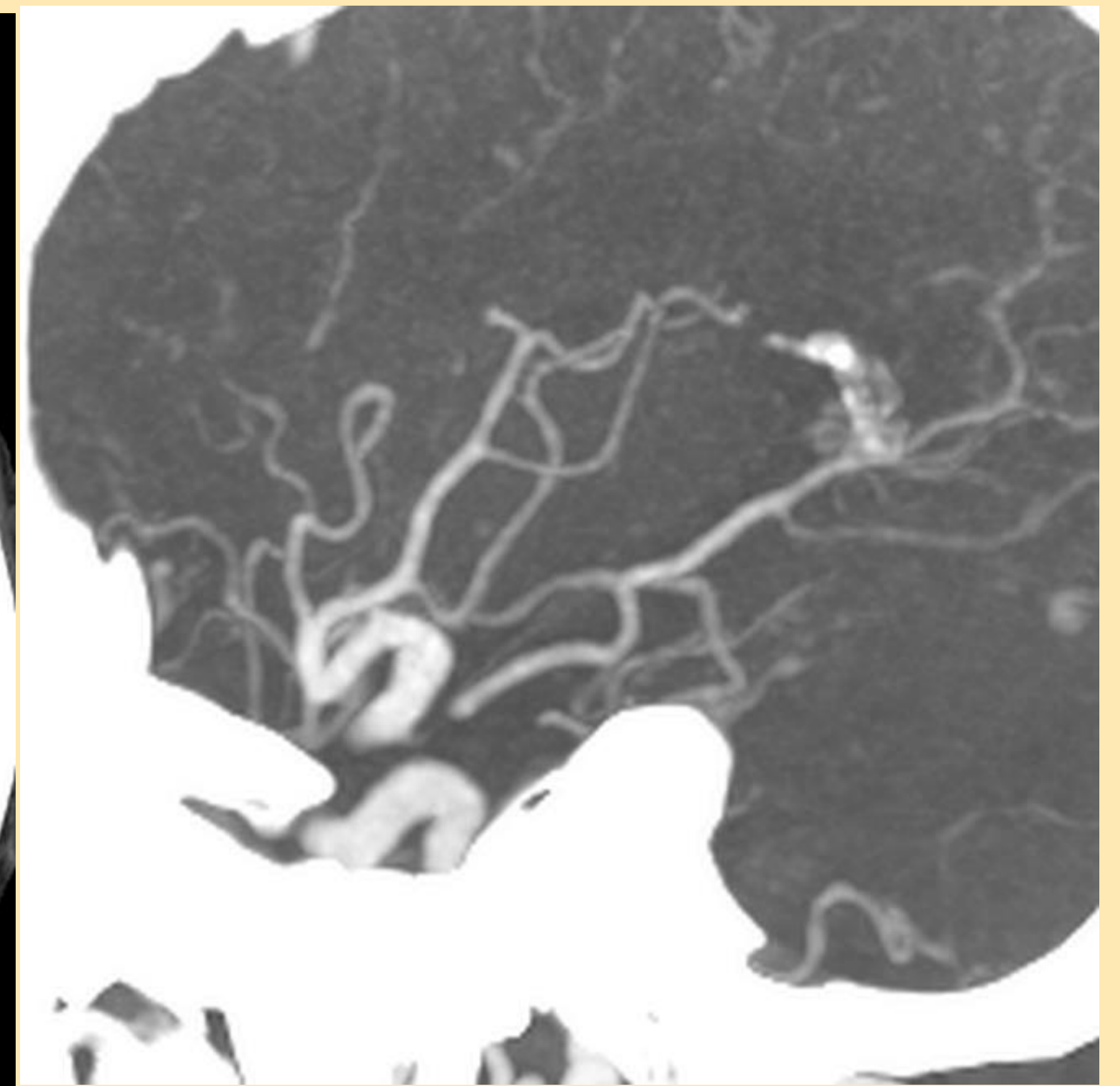


Fig 4e

- **Most frequent mimicker of true CNS vasculitis**
- **Triggering factors include sympathomimetic or vasoactive agents, peripartum, strenuous exercise, sexual activity and excessive alcohol drinking etc**
- **Severe "thunderclap" headache**
- **Similar angiographic finding to vasculitis**
- **Resolution within 3 months**
- **Treated with removal of triggers and calcium channel, not steroid**

## Case 5 Drug induced vasculitis

History: 35 year old man with history of cocaine use presents with seizure and altered mental status

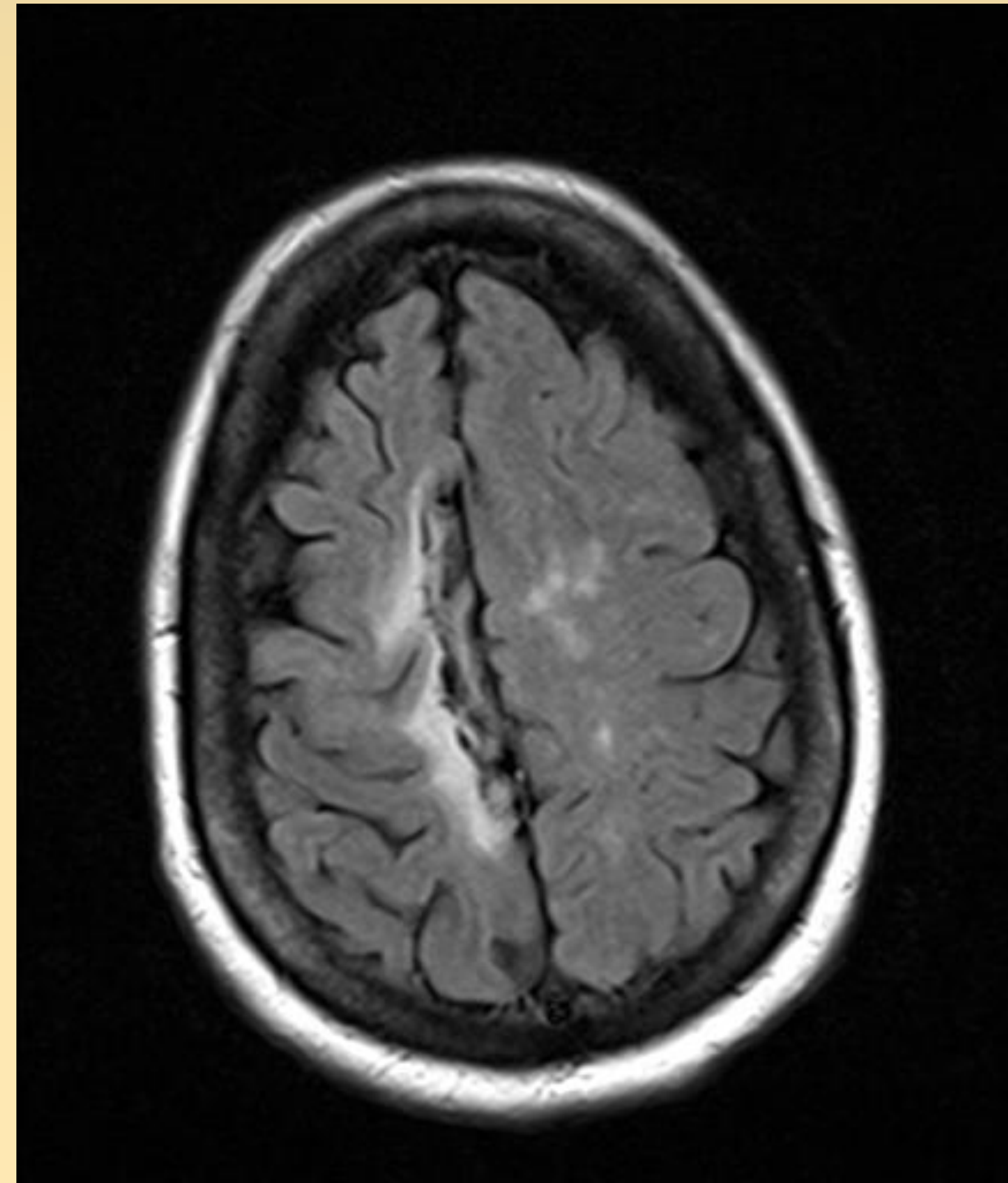


Fig 5a

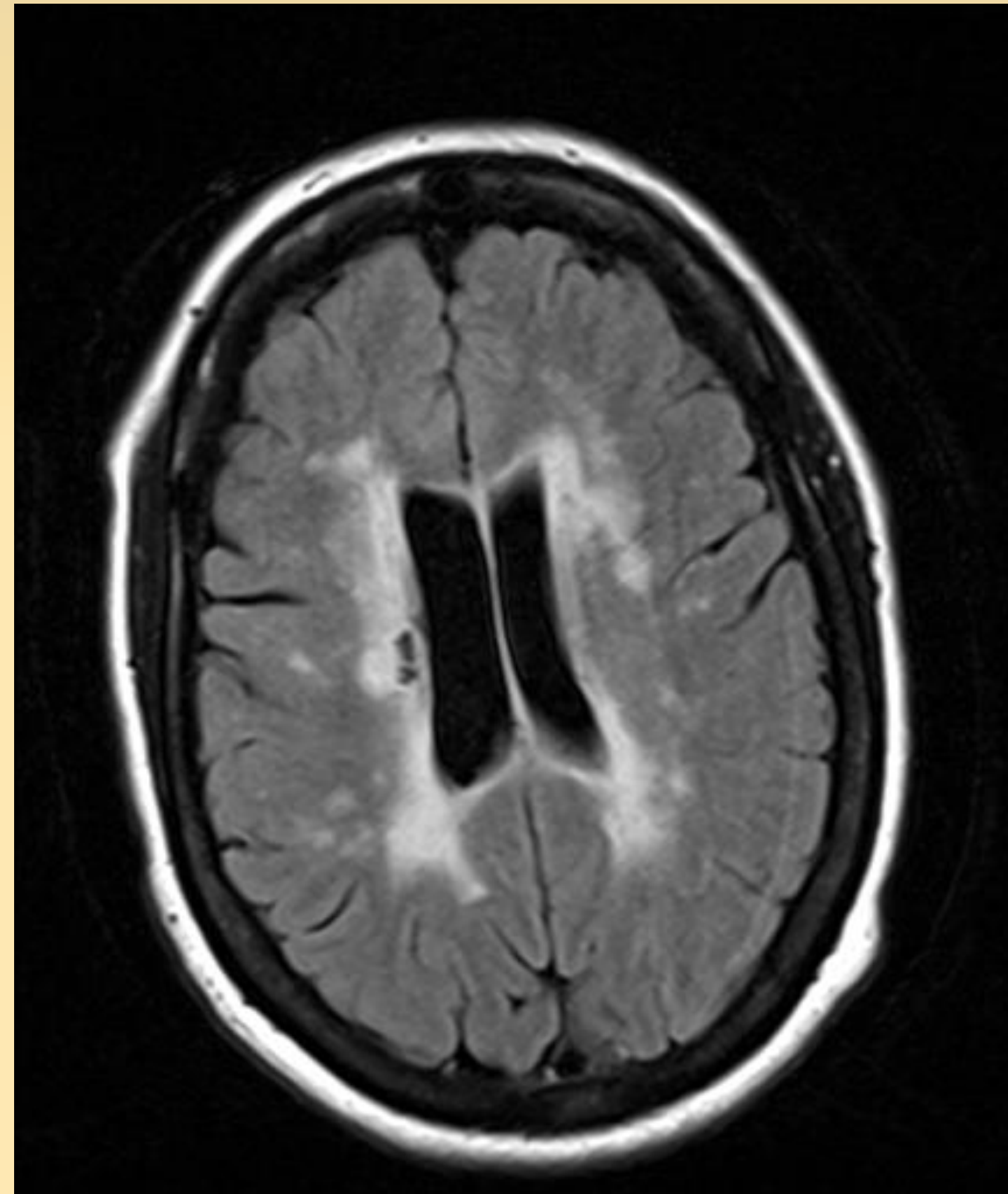


Fig 5b

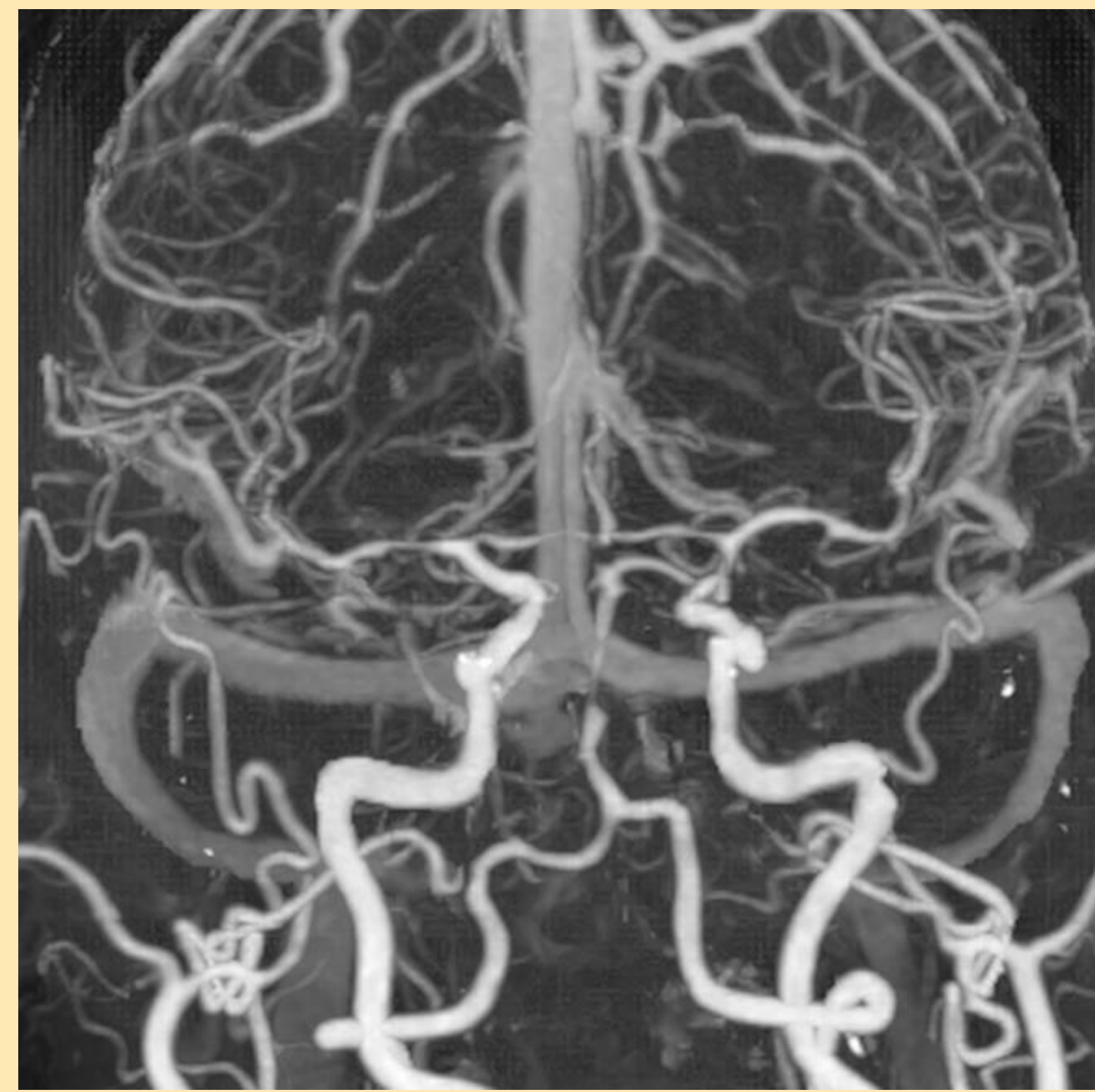


Fig 5c



Fig 5d

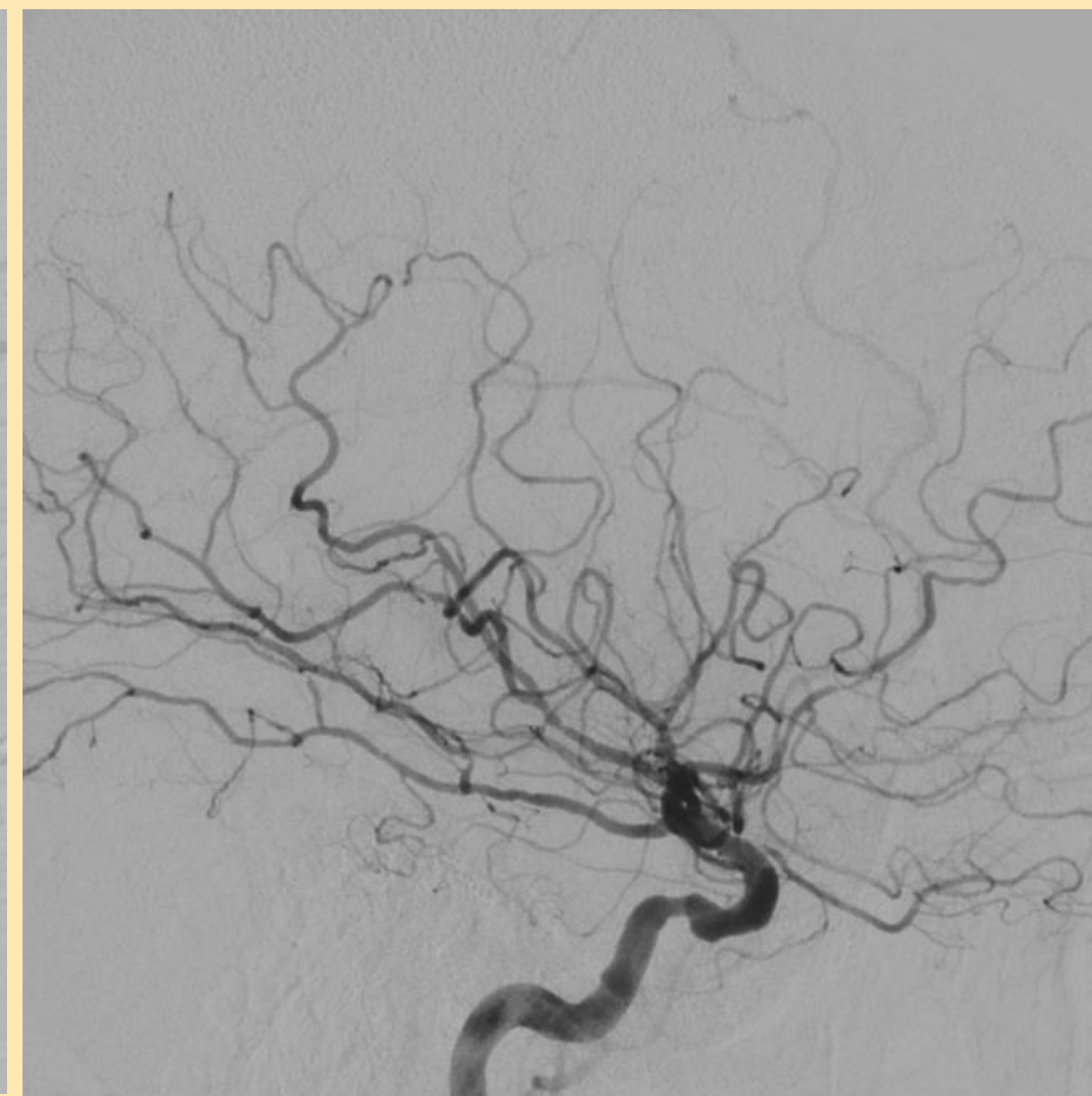


Fig 5e

Axial FLAIR (5a and 5b): chronic right frontal ACA infarction and extensive chronic white matter ischemic changes  
CTA (5c): diffuse stenosis in the basilar artery, MCA and ACA, confirmed by catheter angiography (5d, e)

- **Addictive drugs (cocaine, amphetamine, heroin), and many medications such as antibiotics and chemotherapy agents can cause vasculitis or vasospasm.**

## Case 6 Radiation induced vasculitis

History: 26 year old man with history of medulloblastoma during childhood treated with surgery and radiation, presents with recurrent stroke

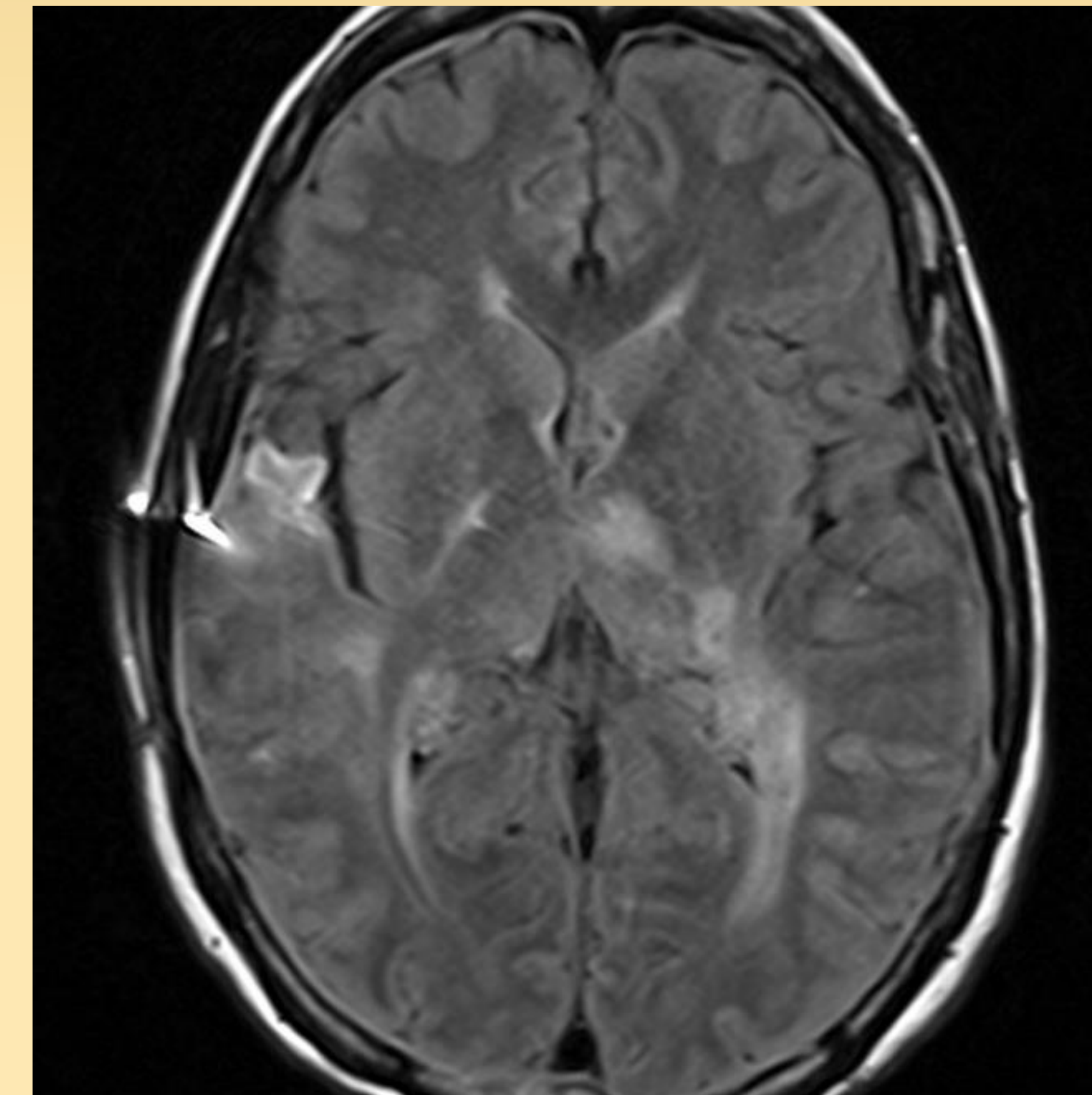


Fig 6a

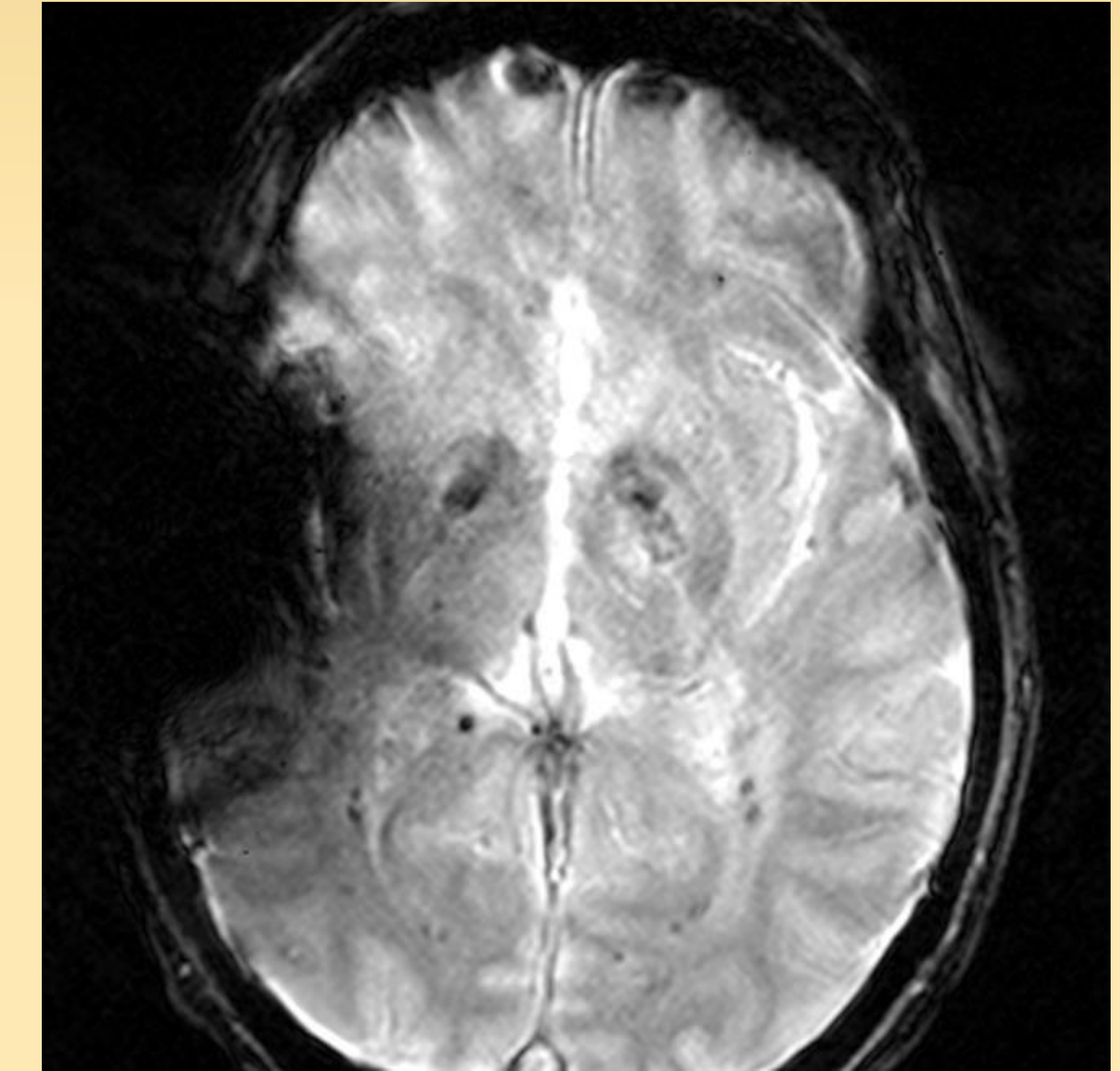


Fig 6b



Fig 6a

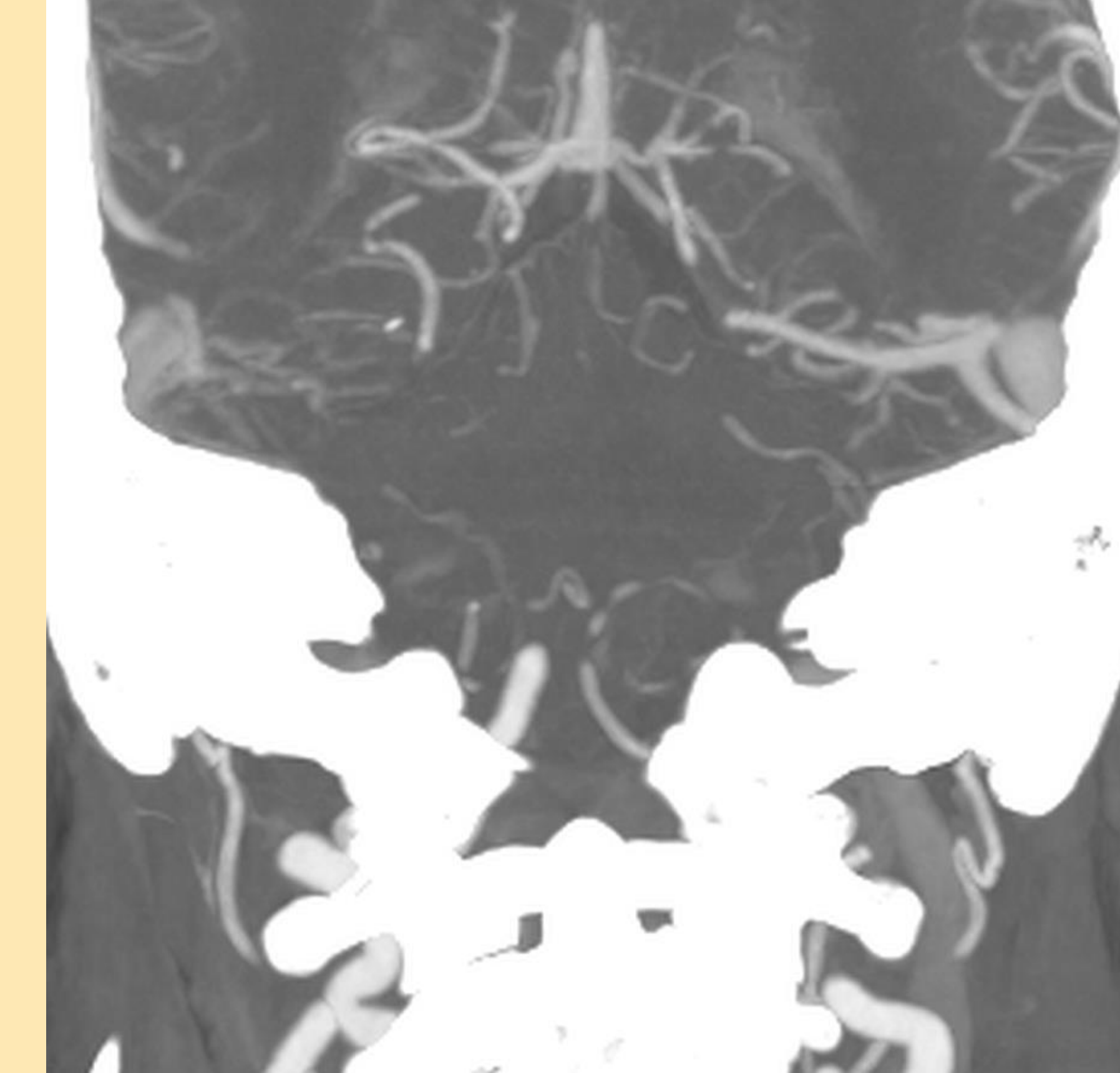


Fig 6b

Axial FLAIR (6a) shows infarction of left thalamus and chronic white matter ischemic changes  
Axial GRE (6b) shows multiple foci of microhemorrhages from previous radiation  
CTA (6c) demonstrates segmental stenosis of bilateral MCA branches, right PCA and left PICA

- **Late effect of cranial radiation**
- **May lead to ischemia, hemorrhage, moyamoya vasculopathy, vascular malformation (cavernoma, capillary telangiectasia)**

## Case 7 Moyamoya disease

History: 13 year old with recurrent TIA symptoms

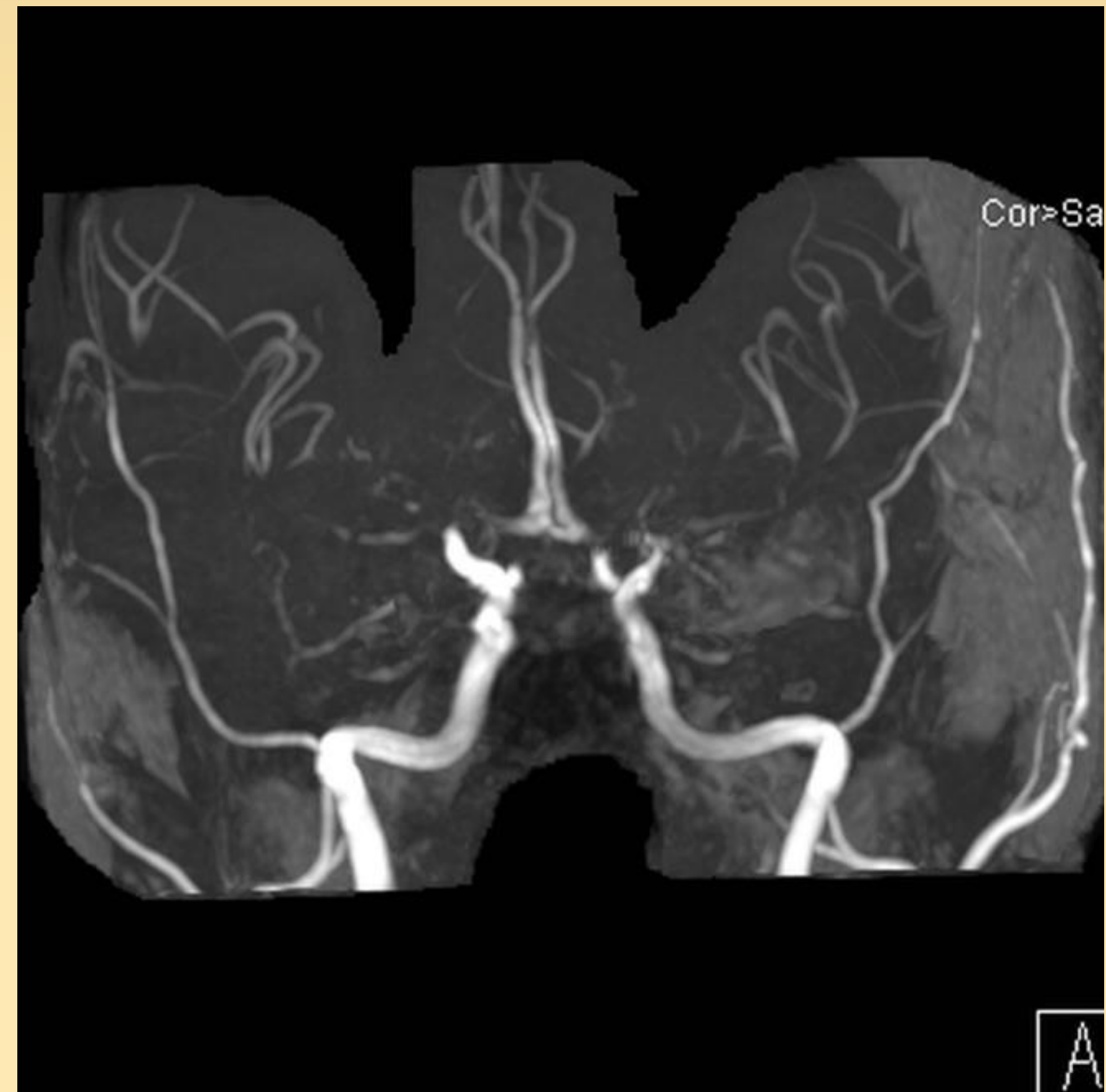


Fig 7a

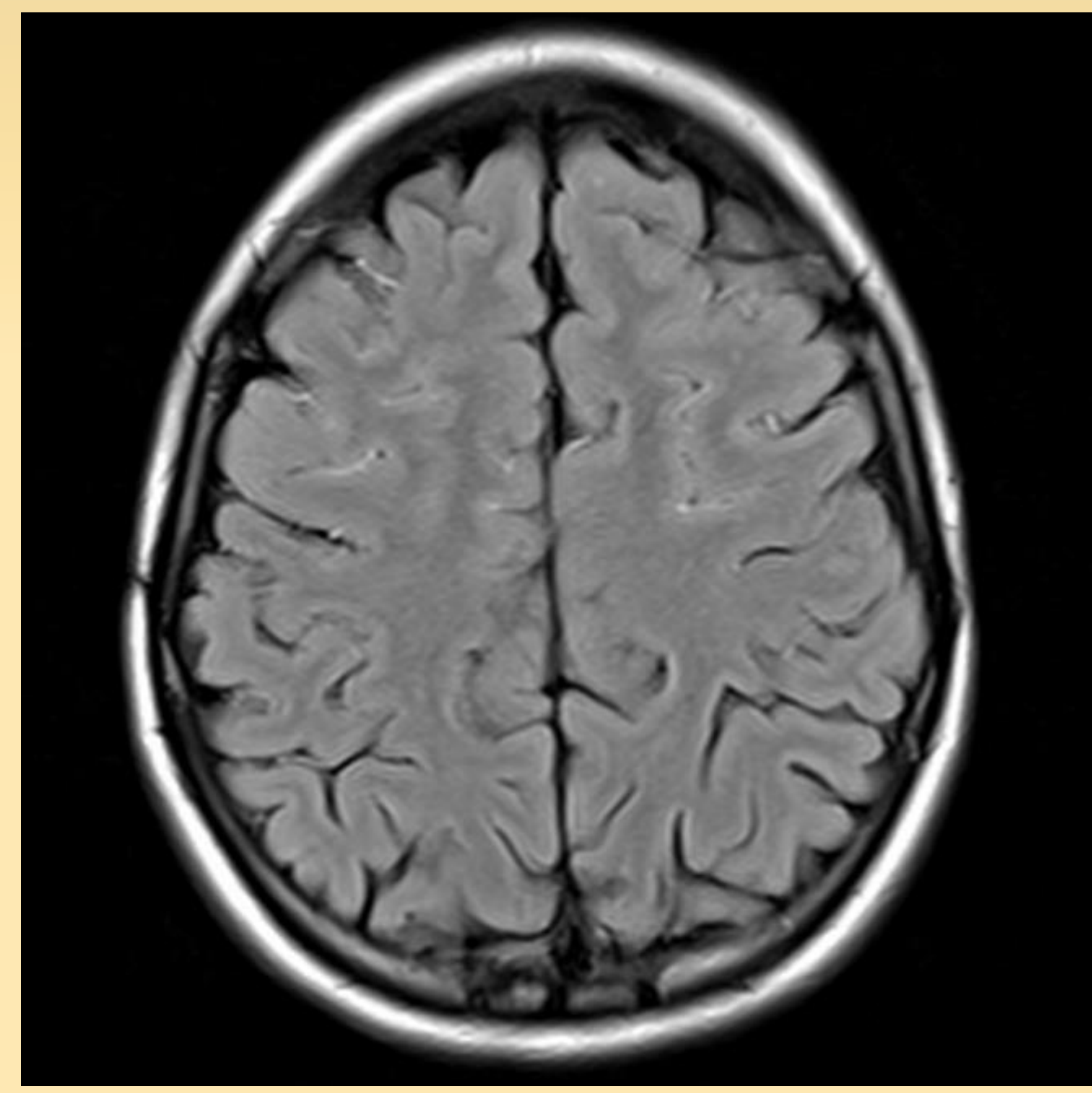


Fig 7b

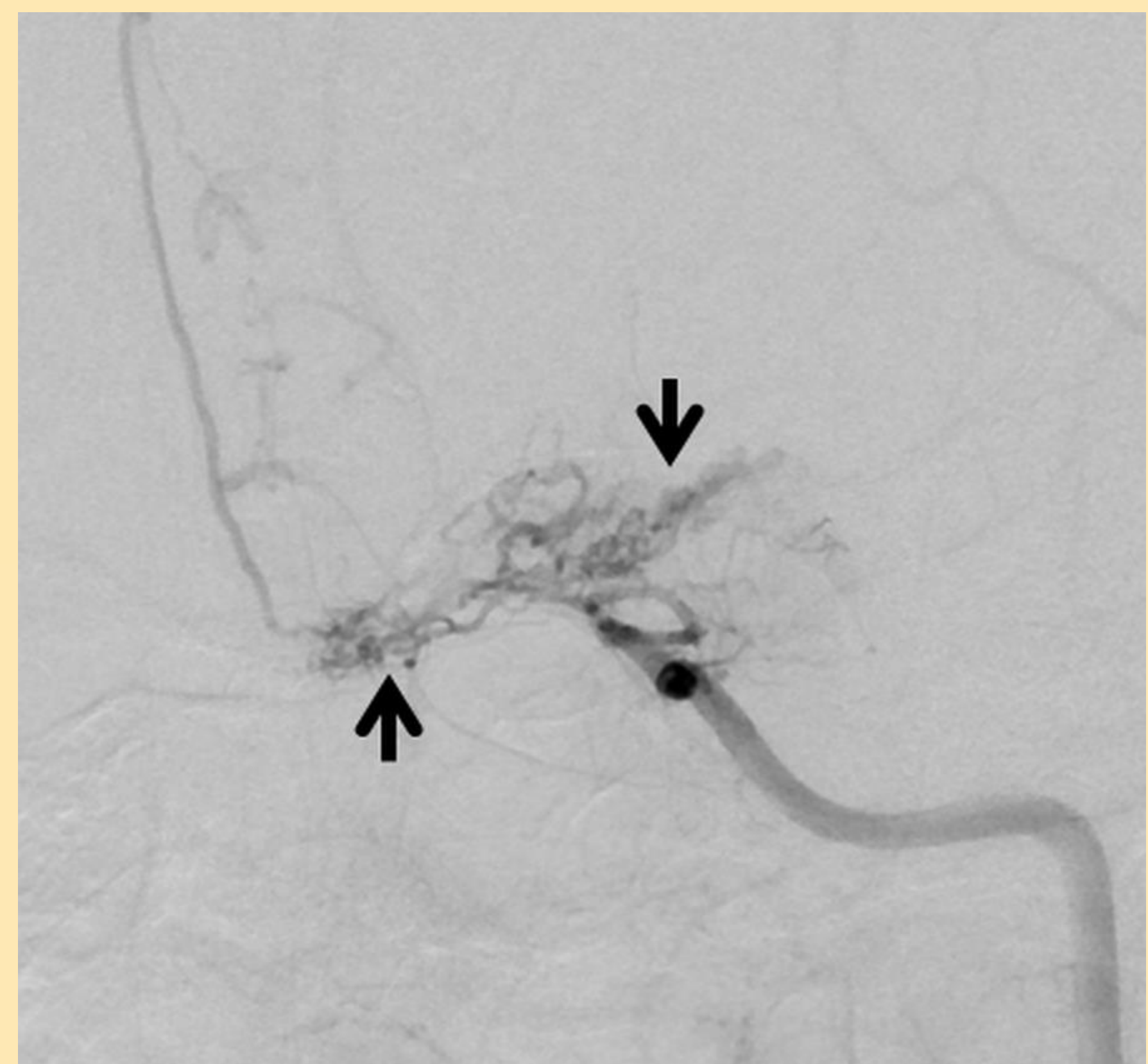


Fig 7c

3D TOF (7a): Occlusion of bilateral ICA termini, M1 and A1 segments  
Axial FLAIR (7b): leptomeningeal ivy sign from collaterals  
Catheter angiogram (7c): right ICA injection shows occlusion of ICA, MCA and ACA with characteristic lenticulostriate collaterals

## Case 8 CADASIL

53 year old woman with mild cognitive impairment and recurrent TIAs

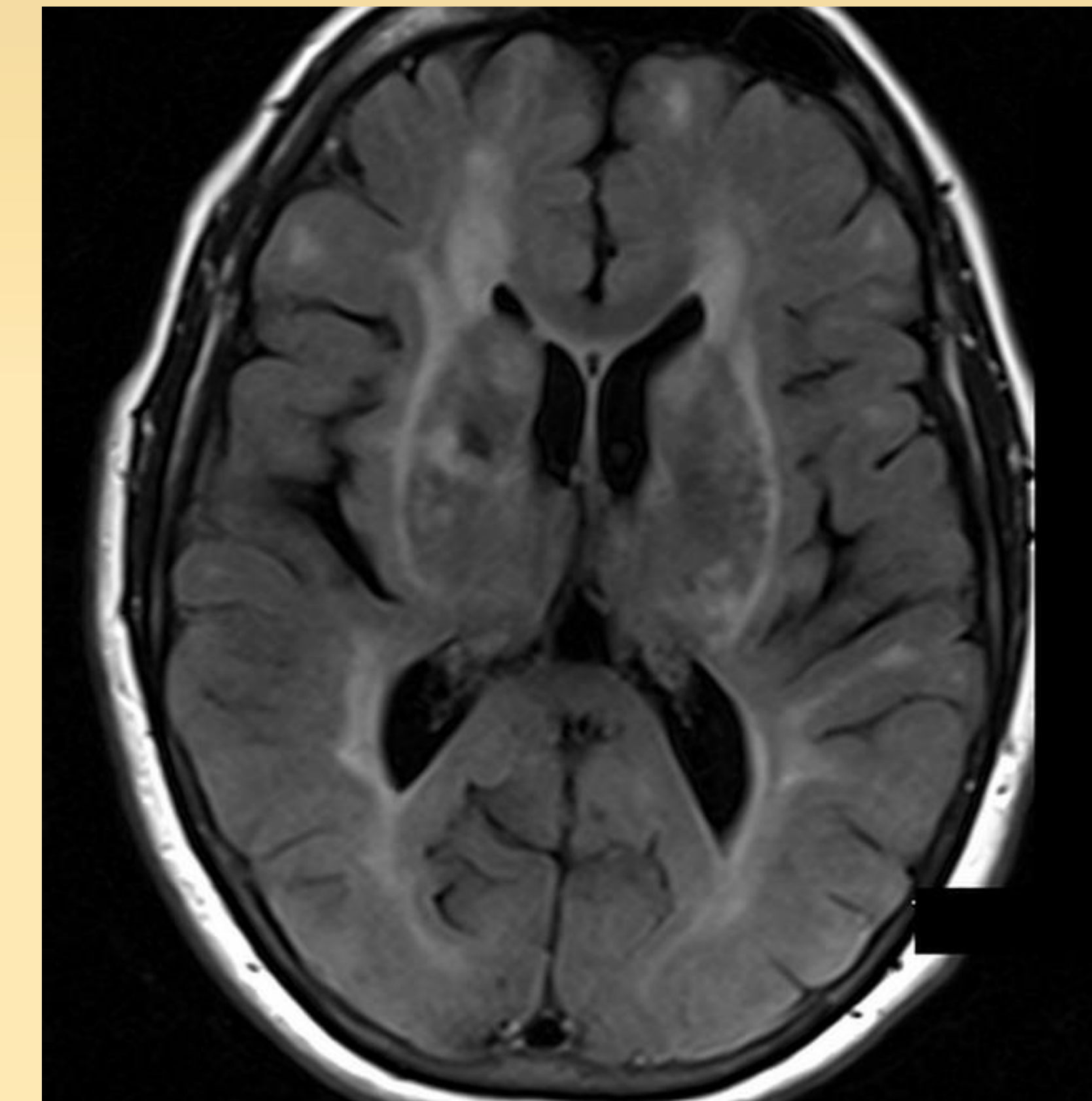


Fig 8a

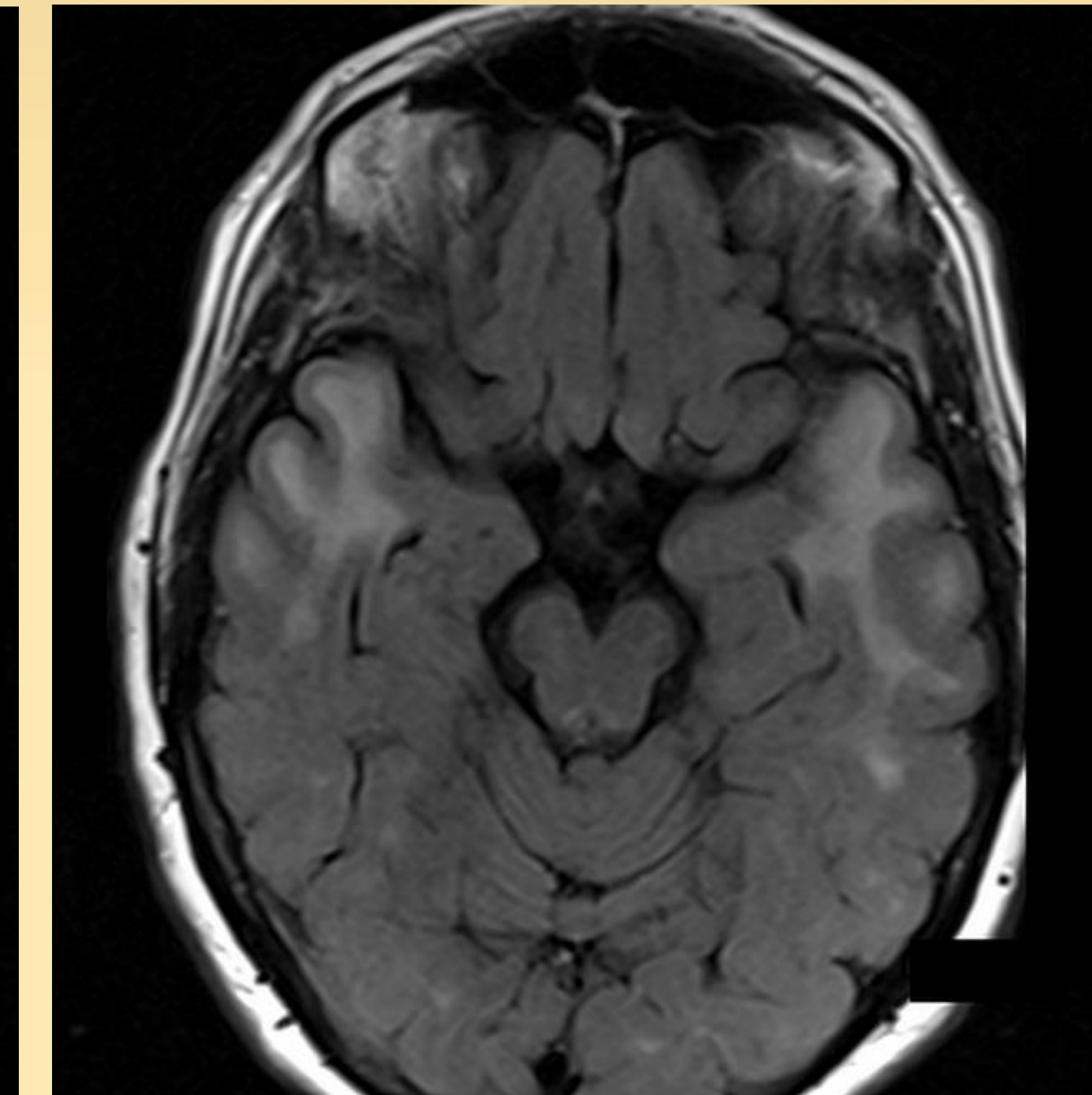


Fig 8b



Fig 8c

Axial FLAIR (8a,b) shows extensive white matter disease with characteristic involvement of external capsules and anterior temporal lobes, suggestive of CADASIL.

CTA (8c) shows multifocal arterial stenosis

- **Hereditary microvasculopathy caused by mutation of Notch 3 gene on chromosome 19**
- **Recurrent lacunar and subcortical white matter ischemic strokes and vascular dementia in young and middle age patients without known vascular risk factors**
- **Characteristic white matter involvement of external capsules and anterior temporal lobes on MRI**
- **Angiography may be normal or shows arterial stenosis**

### Conclusion:

- Cerebral angiopathy encompasses a large number of diseases, with similar angiographic appearance.
- Be suspicious in patients with infarctions or hemorrhages in different vascular territories and ages.
- Important to recognize the findings on CTA and MRA, esp distal branches
- Clinical history and lab tests are essential for differential diagnosis

### References:

1. Abdel Razek, A. A. K., Alvarez, H., Bagg, S., Refaat, S. & Castillo, M. Imaging Spectrum of CNS Vasculitis. *RadioGraphics* **34**, 873–894 (2014).
2. Birnbaum, J. & Hellmann, D. B. Primary Angiitis of the Central Nervous System. *Arch. Neurol.* **66**, 704–709 (2009).
3. Cappelen-Smith, C., Fracp, M., Calic, Z. & Cordato, D. Reversible Cerebral Vasoconstriction Syndrome: Recognition and Treatment. *Curr Treat Option*

- **Chronic cerebrovascular disease characterized by progressive stenosis or occlusion of the distal ICA and proximal Circle of Willis vessels with collateral formation.**
- **"Moyamoya" means puff of smoke in Japanese, describe the characteristic angiographic appearance of the collateral network.**
- **A number of disease can also cause moyamoya type of pattern: sickle cell disease, atherosclerosis, meningitis, Down syndrome, Neurofibromatosis type I, and cranial radiation**
- **Treatment: surgical revascularization (EDAS)**