

An Unusual Case of Cognitive Decline

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A 53 years old male

- ❖ Memory disturbances
- ❖ Multiple episodes of focal seizures with impaired awareness
- ❖ Imbalance while walking
- ❖ Tremulousness of bilateral upper limbs

Subacute in onset and gradually progressive for 2 years

No H/O

- ❖ Fever / weight loss
- ❖ Headache, Nausea, Vomiting
- ❖ Blurring of Vision
- ❖ Nasal regurgitation of food
- ❖ Double vision
- ❖ Vertigo/ Tinnitus/ Hearing loss
- ❖ Limb weakness
- ❖ Loss of consciousness

Neurological examination

General Examination – Normal

**Higher mental
function**

MMSE - 21/30

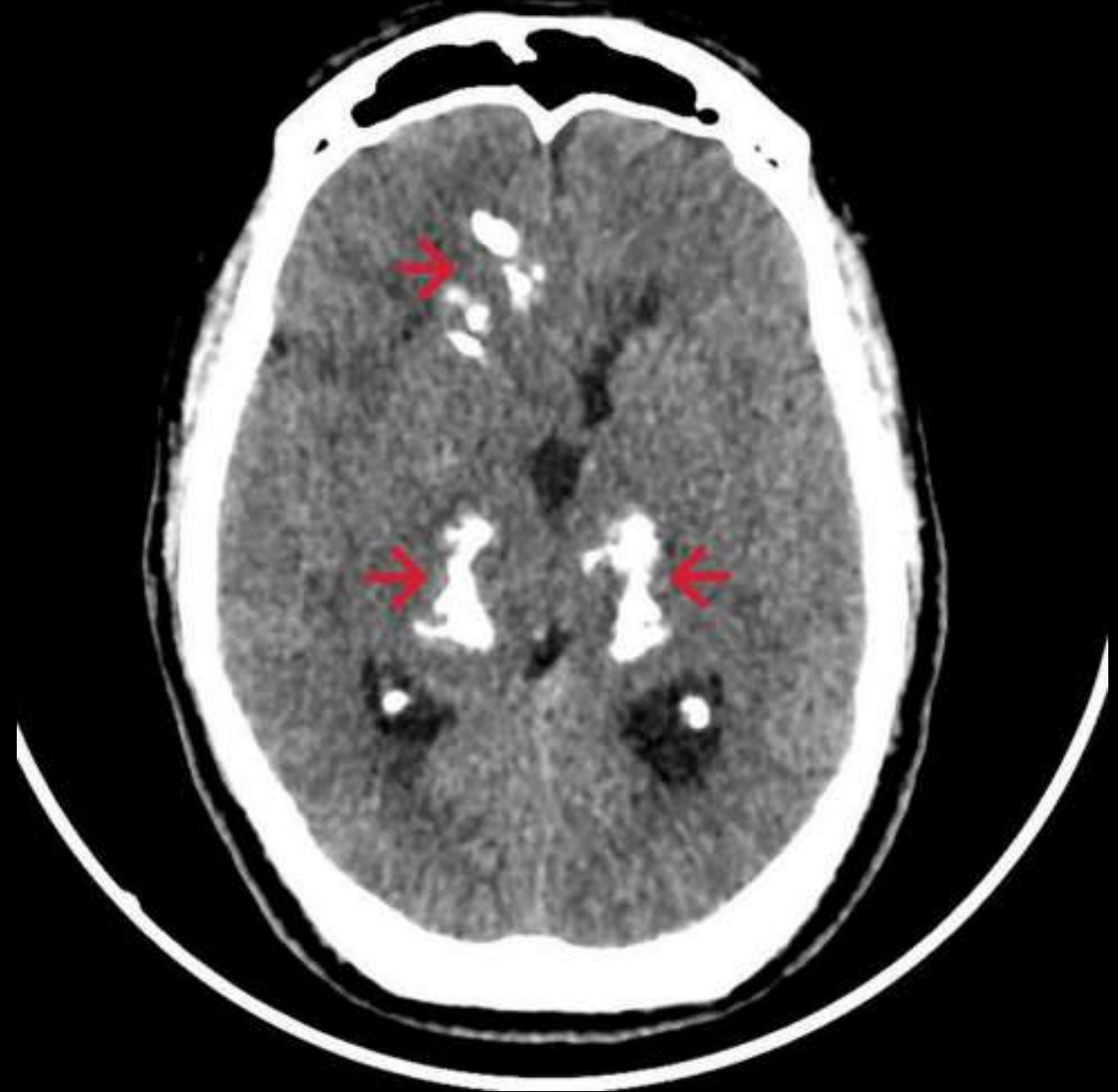
FAB – 12/18

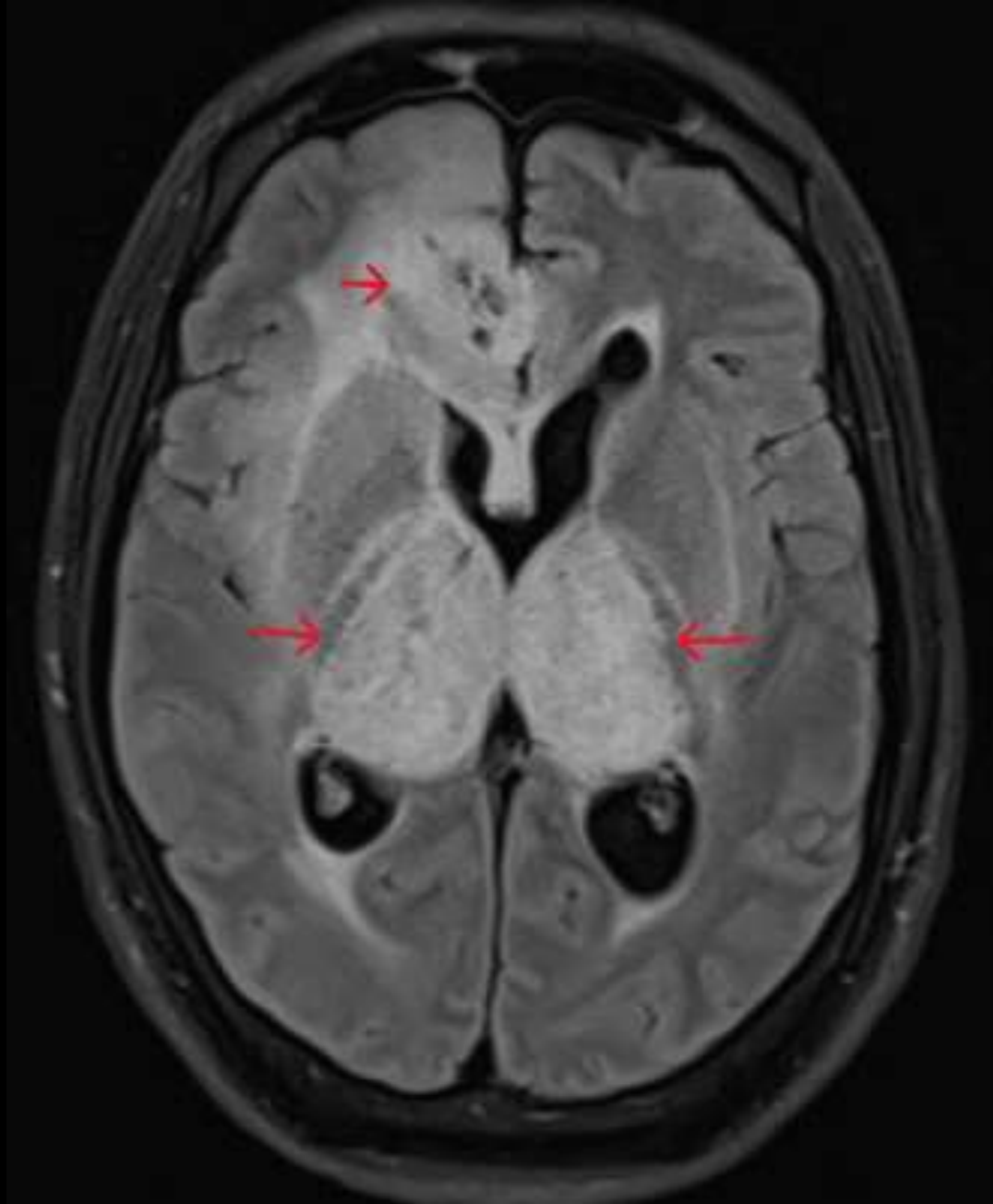
Moderate
cognitive deficit

Cranial Nerves

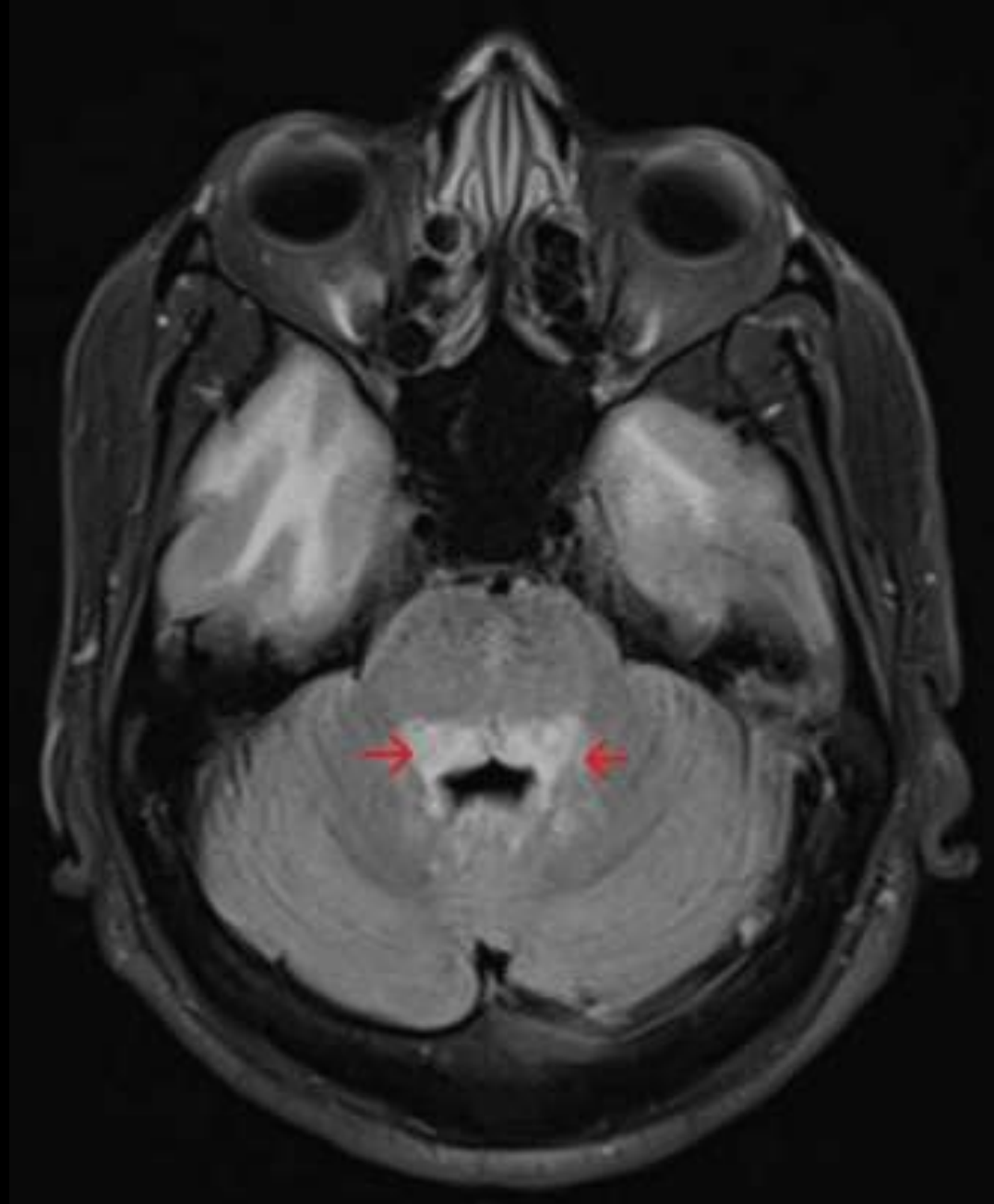
Fundus : Normal
Disc Margins

CT BRAIN –
Diffuse hyperdense
areas
s/o calcifications

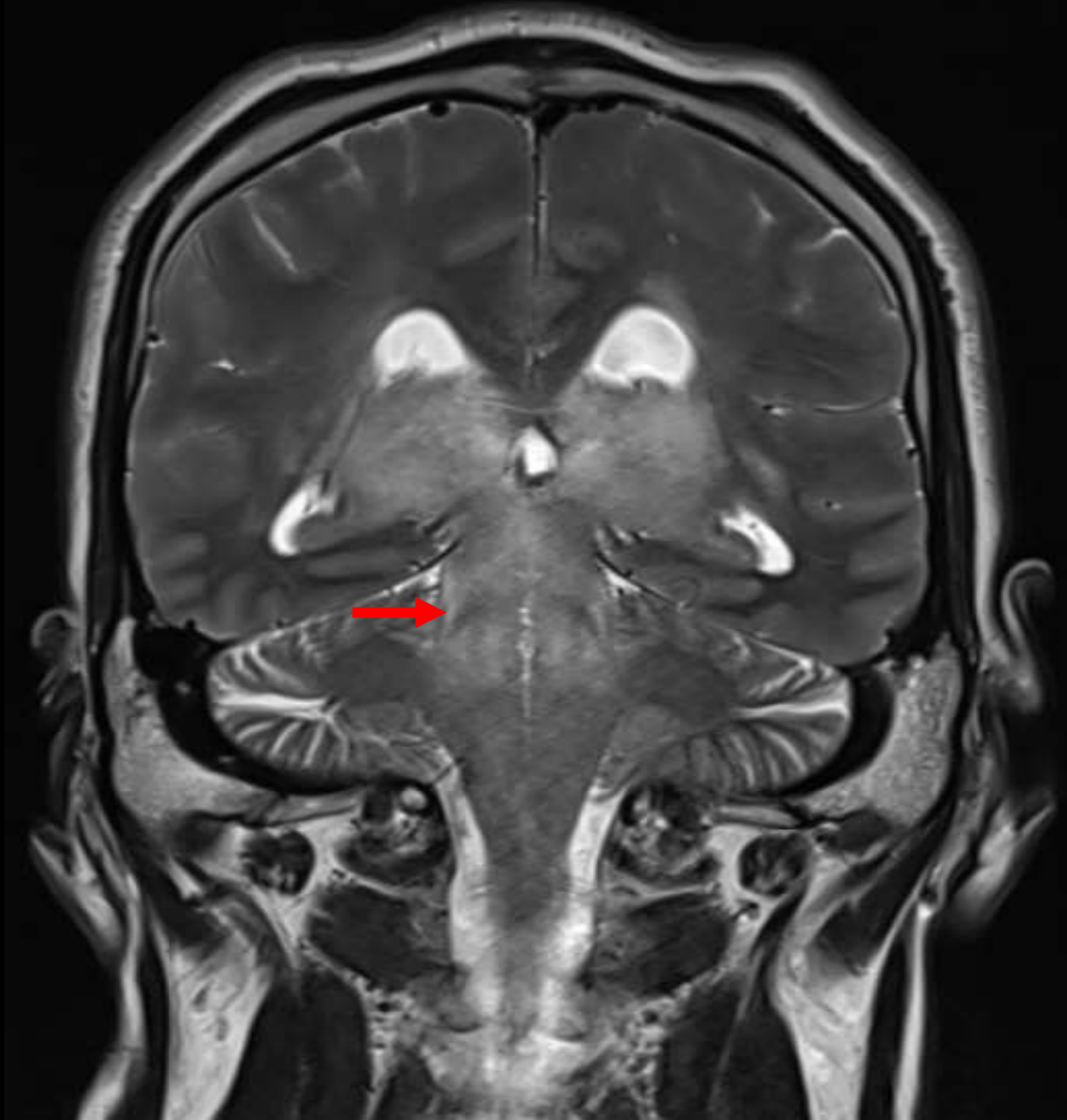




FLAIR



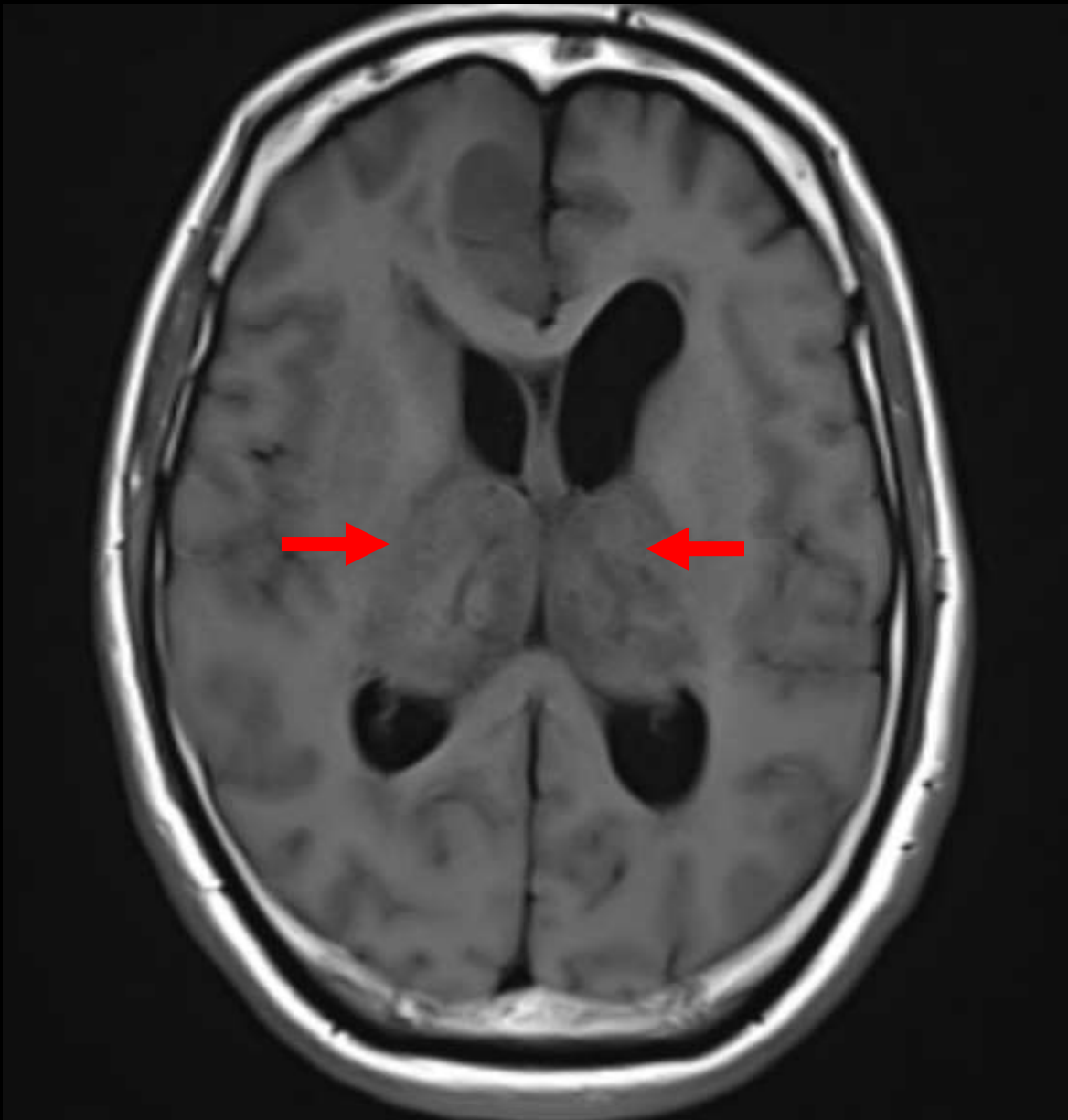
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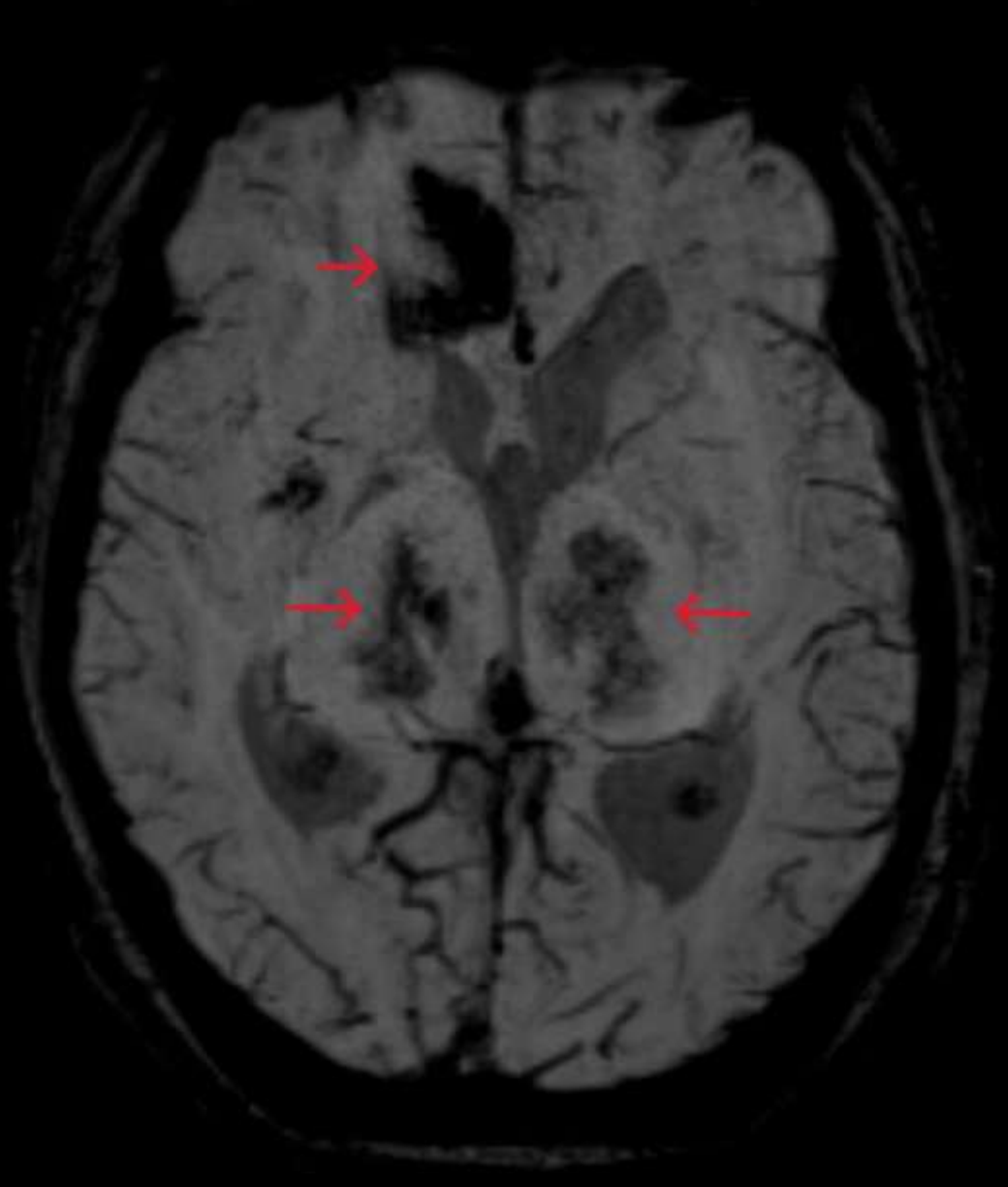
T2W - CORONAL



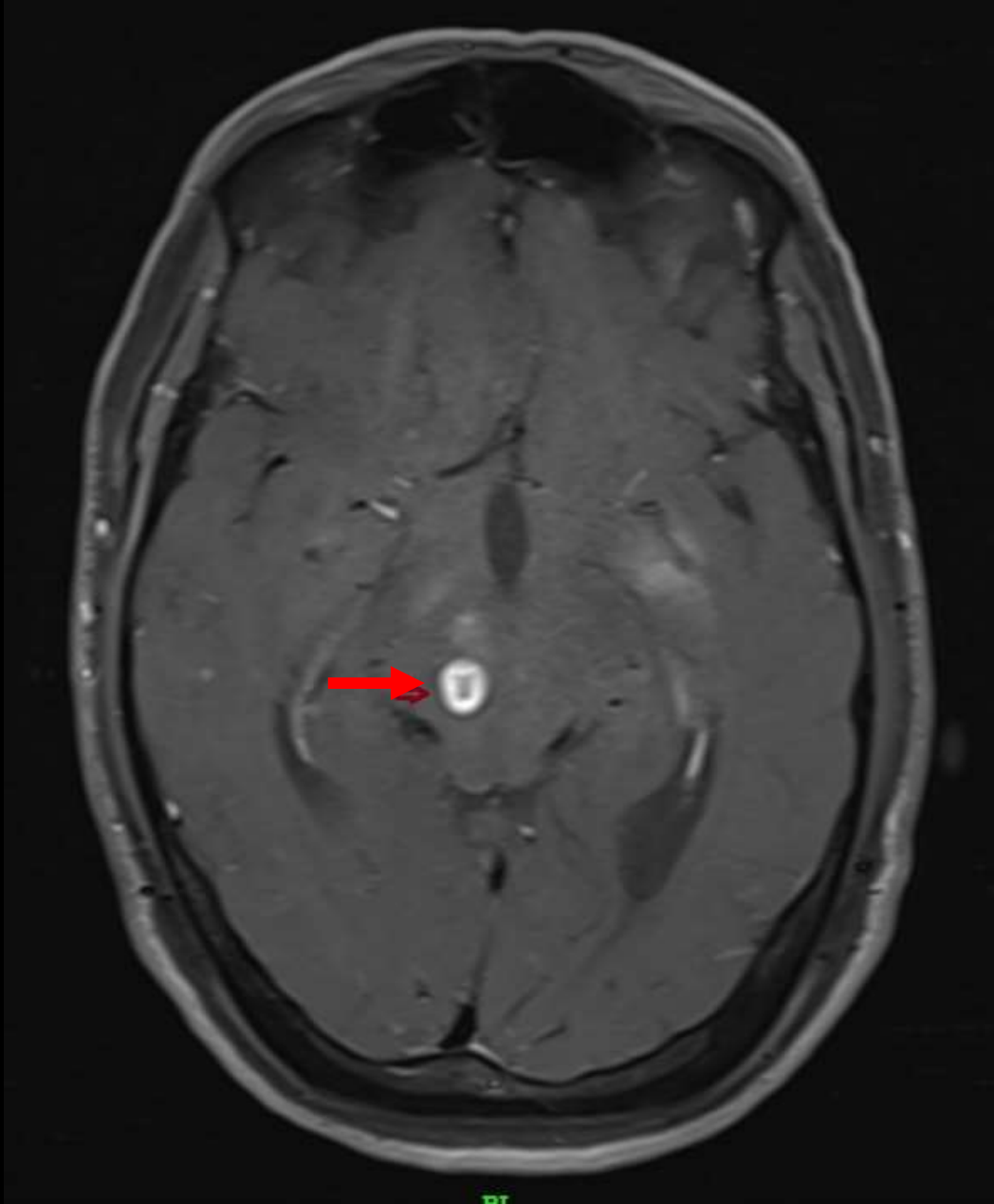
T2W - AXIAL



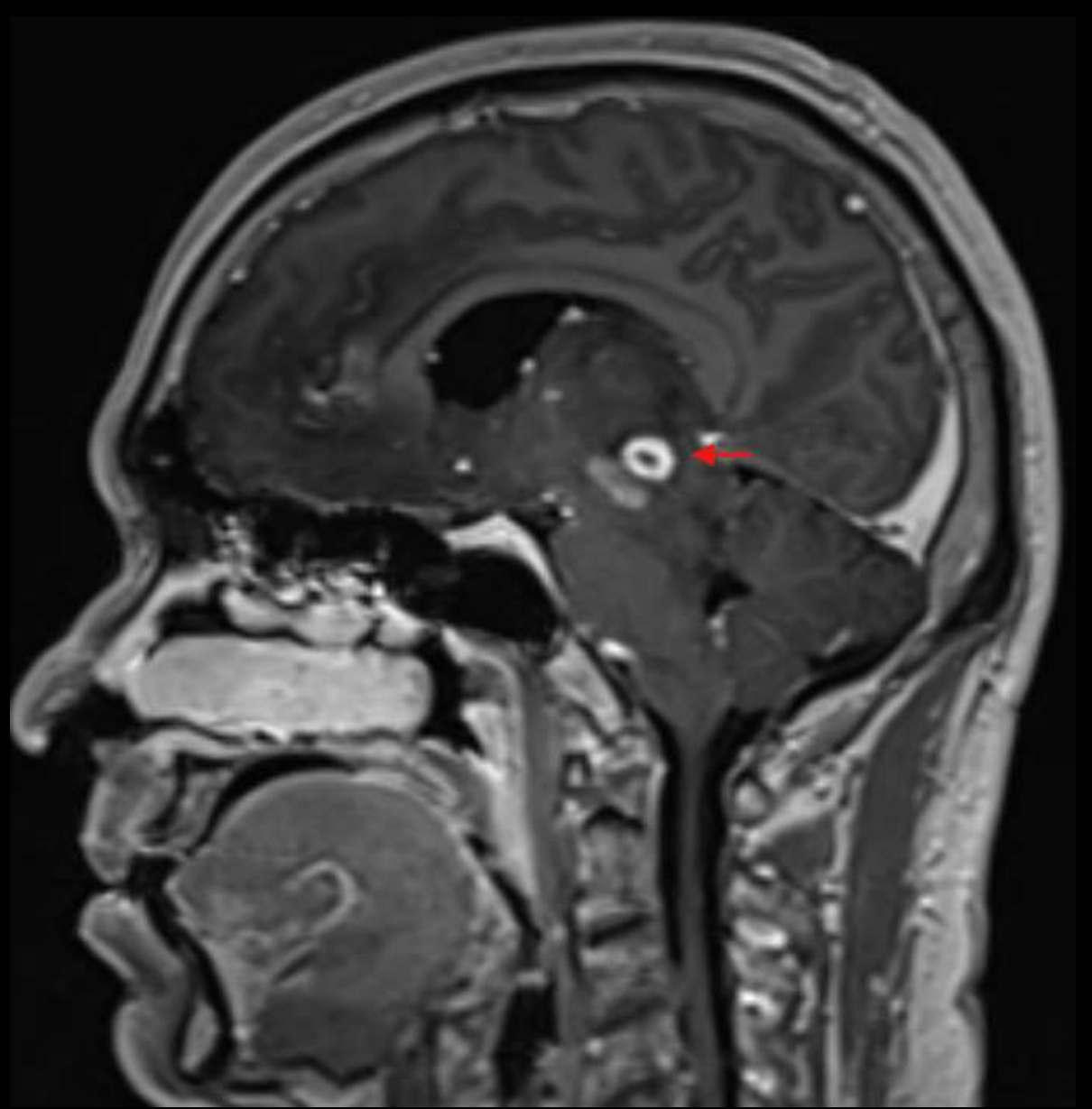
T1 -AXIAL



SWI



T1 – Post contrast – Axial



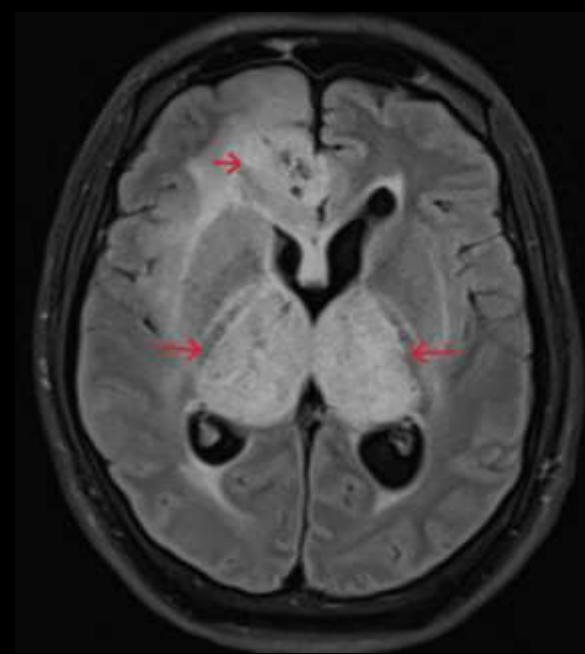
T1 – Post contrast – Sagittal

White matter changes

Labrune
syndrome

Cystic lesions

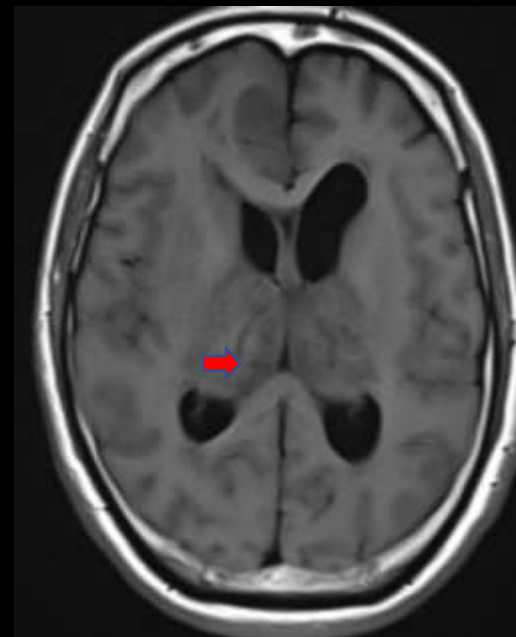
Cerebral Calcification



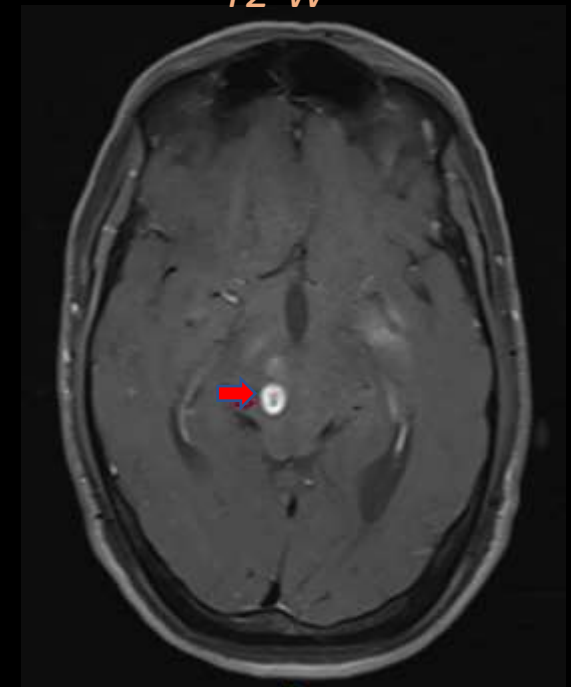
FLAIR



T2-W



T1-W



T1W - POSTCONTRAST

Typical Radiological features:

- T2 hyperintensity in bilateral cerebral white matter is the most observed neuroimaging features.
- The corpus callosum is typically unaffected.
- Brain calcifications are asymmetrically distributed and usually appear as small punctate foci or larger confluent regions within the cerebral white matter or deep grey nuclei.
- They are rarely located in the cerebellum.
- They are typically found in the supratentorial region.

Diagnosis

- Whole-genome sequencing - **SNORD 118 - Negative**

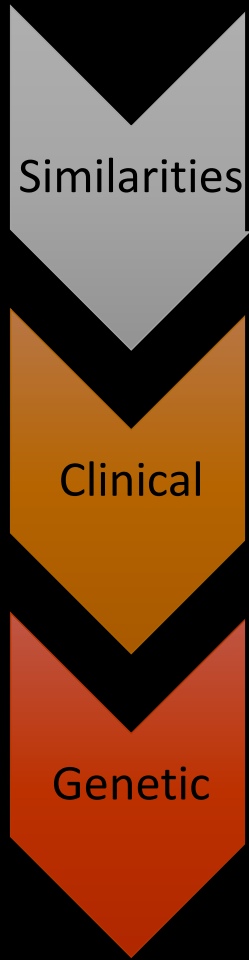


Management

- Surgical - removal of cysts.
- Antiepileptic medications.
- Antipsychotic drugs.

- The patient was treated with **antiepileptics** and **supportive care** for seizures along with **physiotherapy** and **gait balancing exercises**.

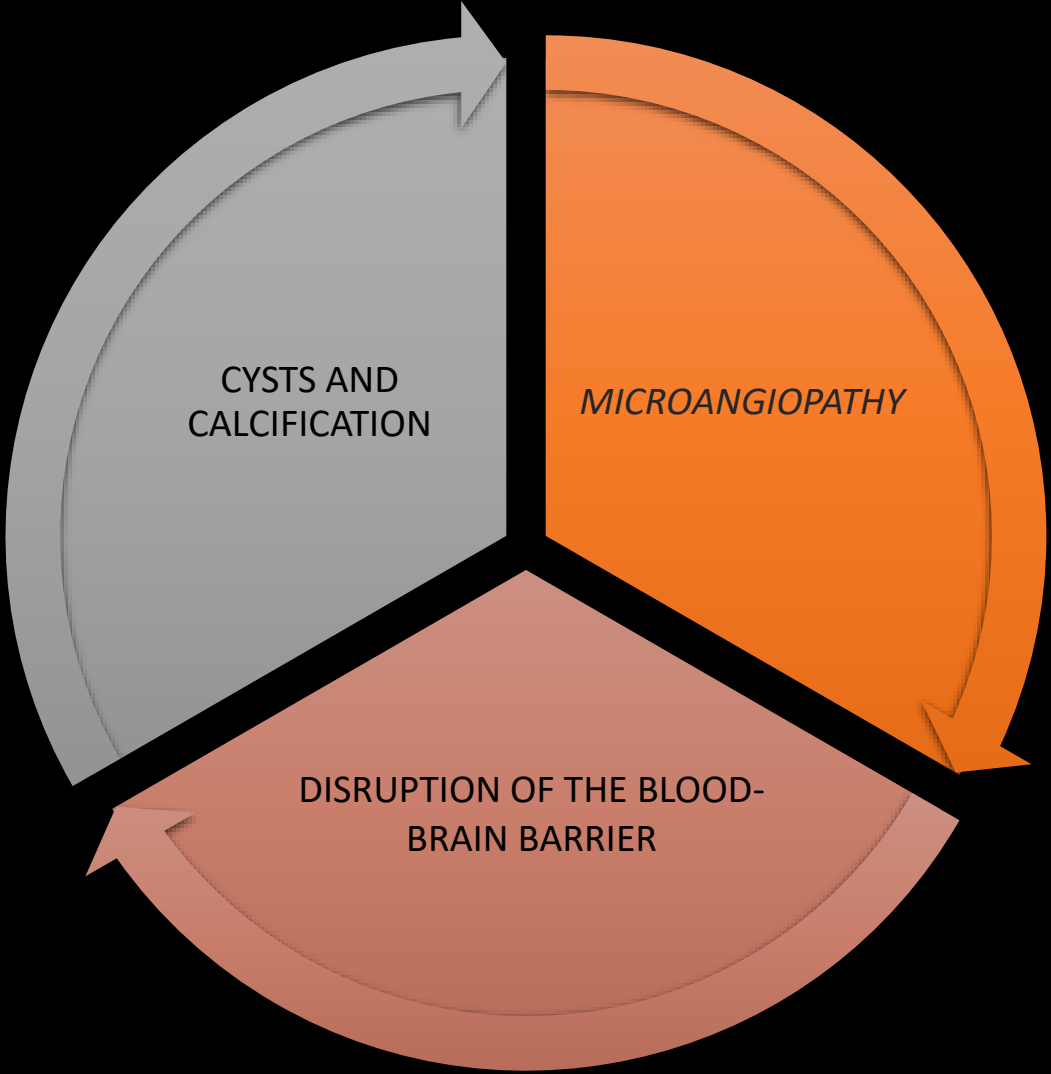
Radiological differential – Coats Plus Syndrome



Other D/ds

- Adult-onset leukoencephalopathy with axonal spheroids and pigmented glia (ALSP)
- Neurocysticercosis
- Aicardi-Goutieres syndrome
- Astrocytoma

Pathophysiology

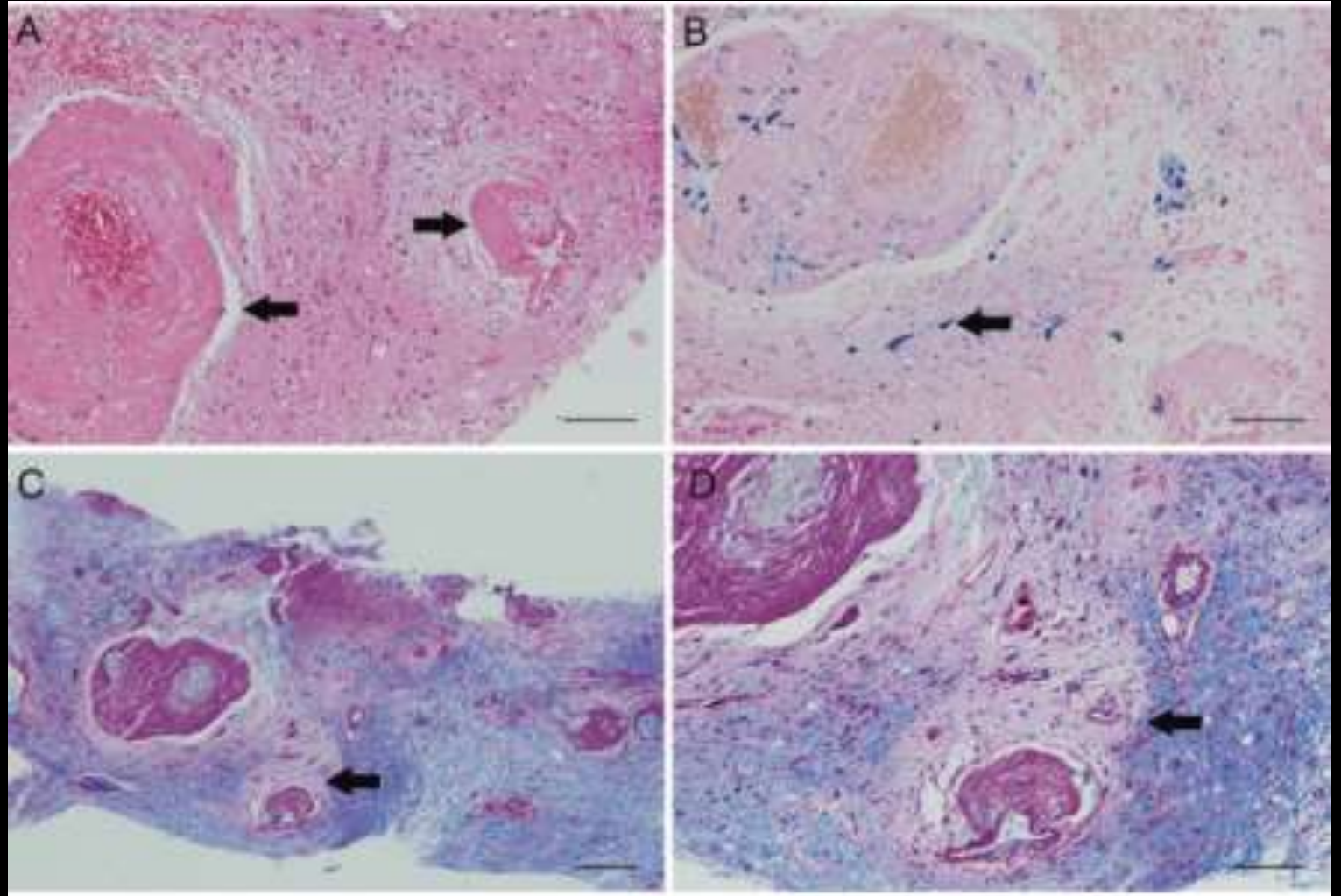


Histopathology

A} Blood vessels with hyaline degeneration.

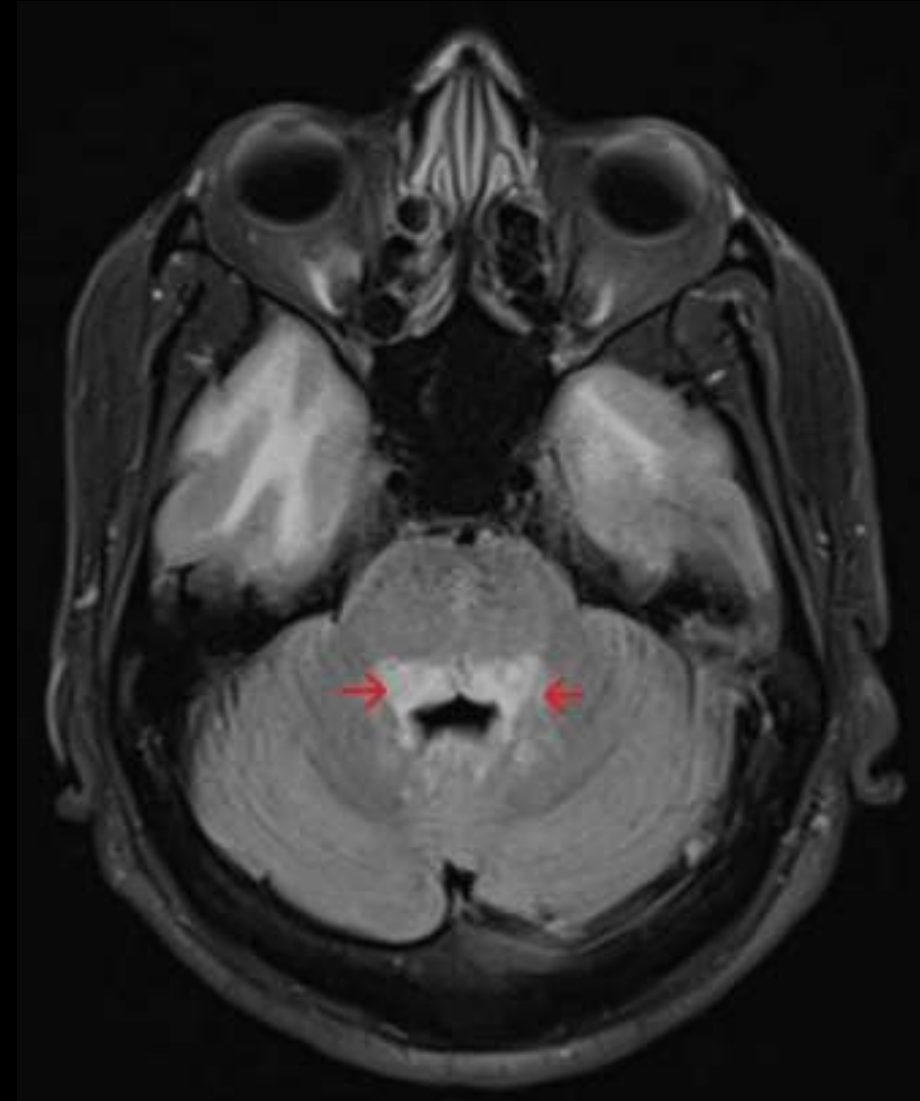
B} Signs of micro-bleedings.

C} and D} Perivascular loss of myelin



What is unique In our case ?

SNORD 118 negative.
Infratentorial brain
parenchyma



Thank you

