60-year-old man with 2-3 year history of functional decline and behavioral changes

Anika Makol, MS3

Bharat Naraparedddy, MD

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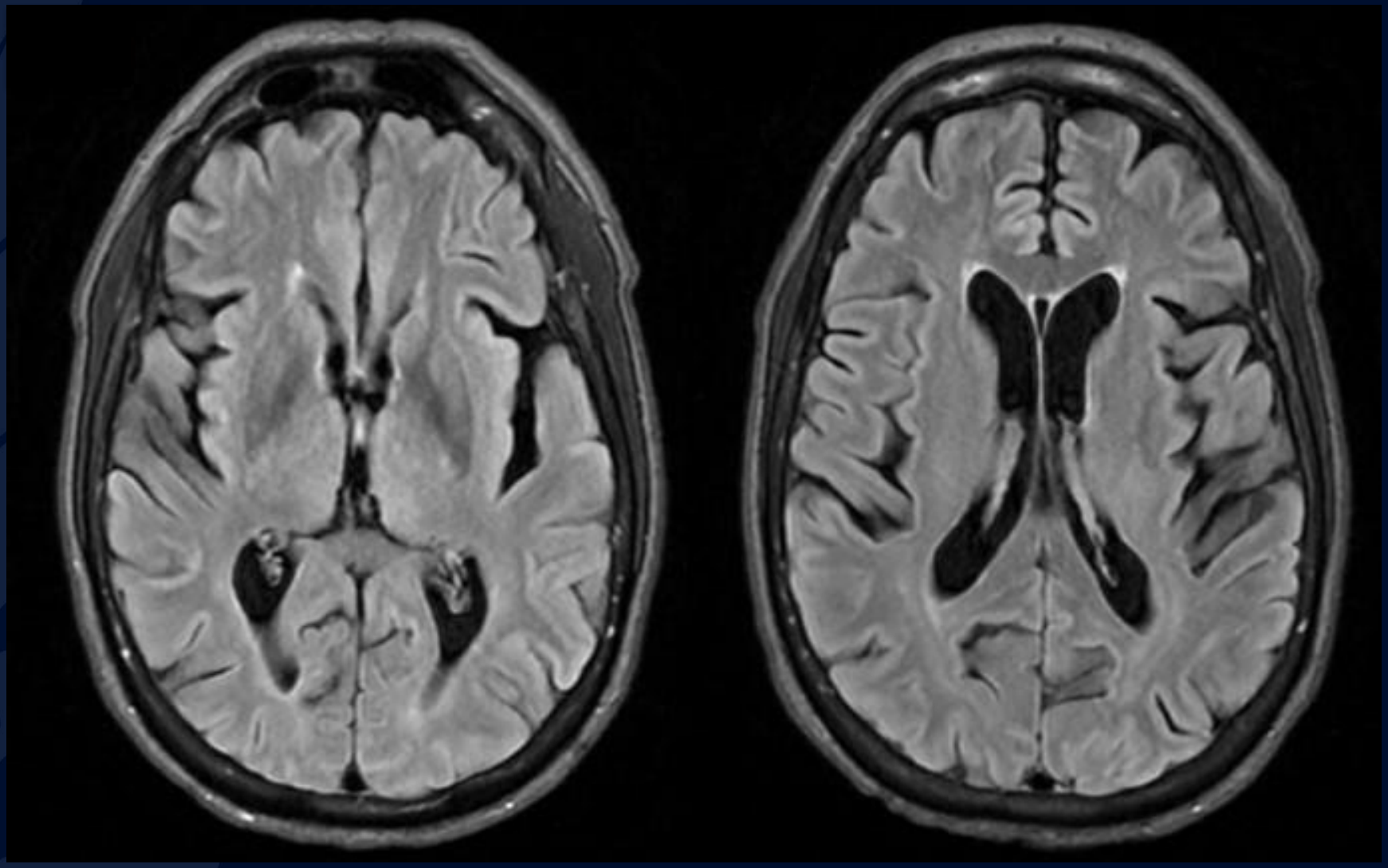
T2 FLAIR



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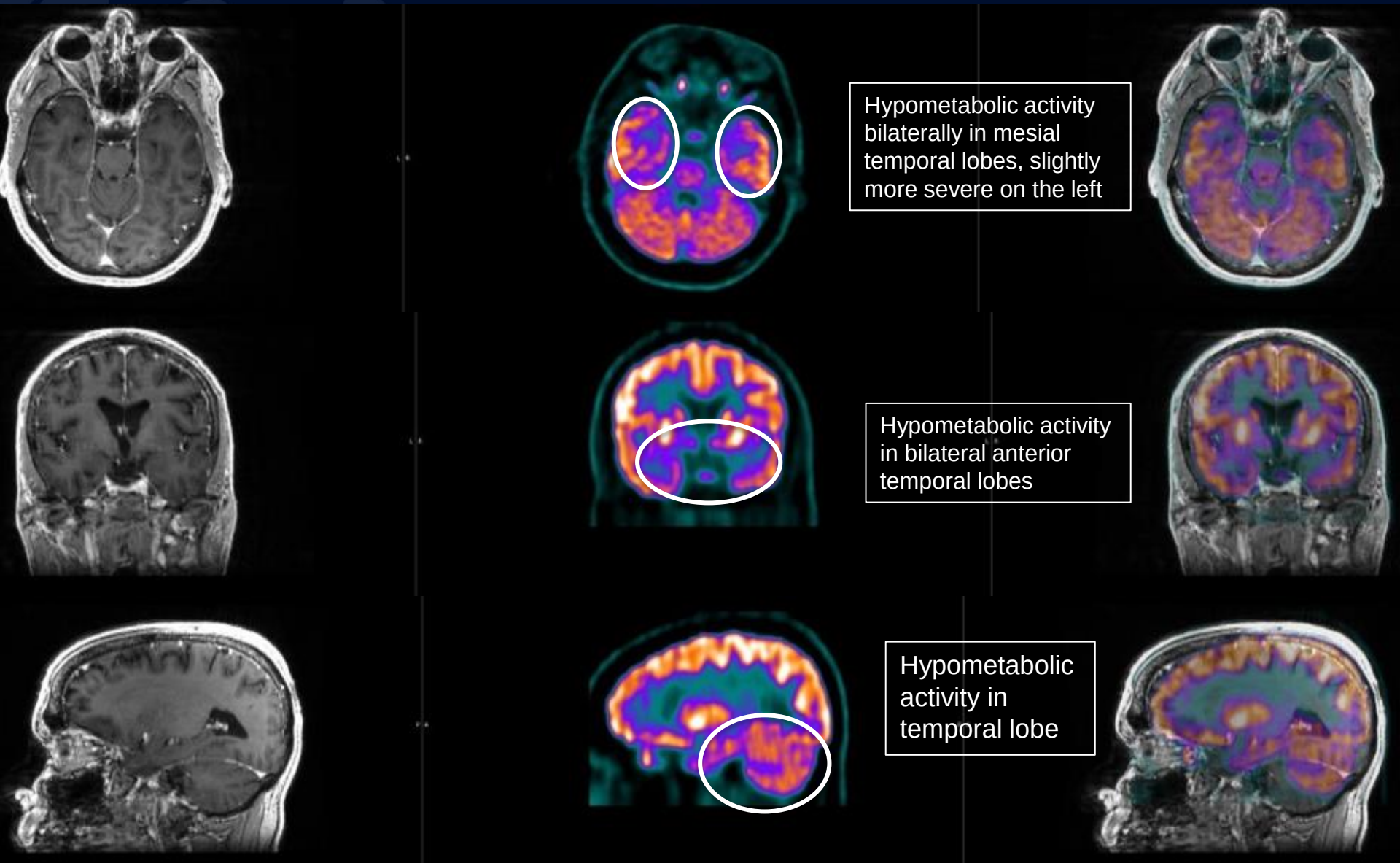
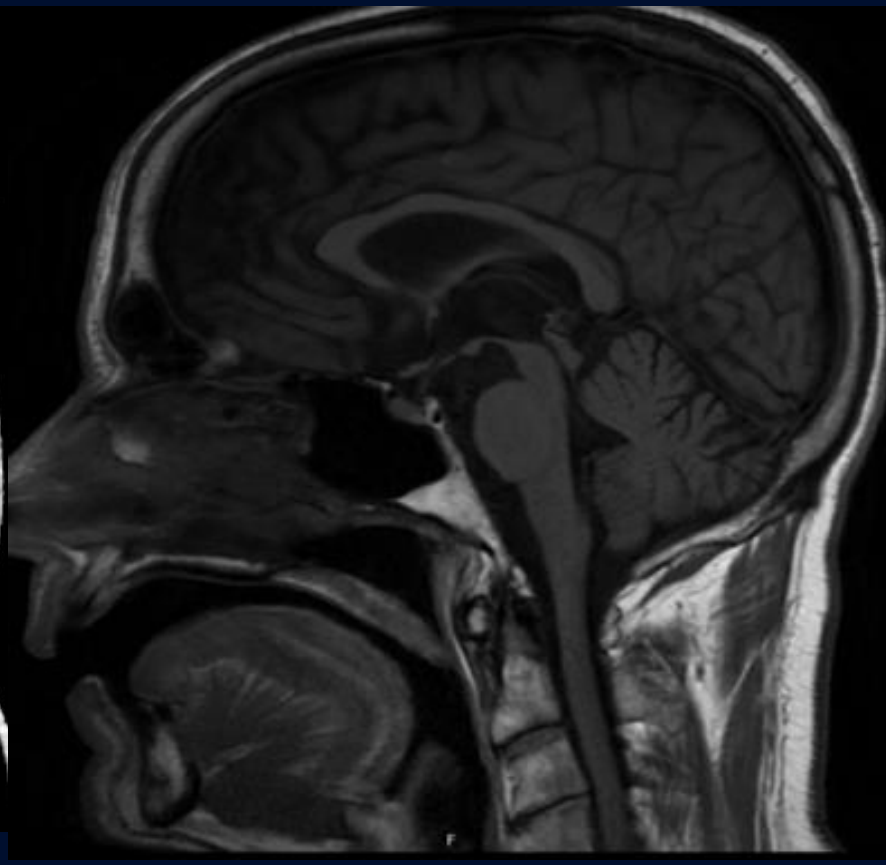
FDG-PET


Hypometabolic activity bilaterally in mesial temporal lobes, slightly more severe on the left
Hypometabolic activity in bilateral anterior temporal lobes
Hypometabolic activity in temporal lobe

CT

T1



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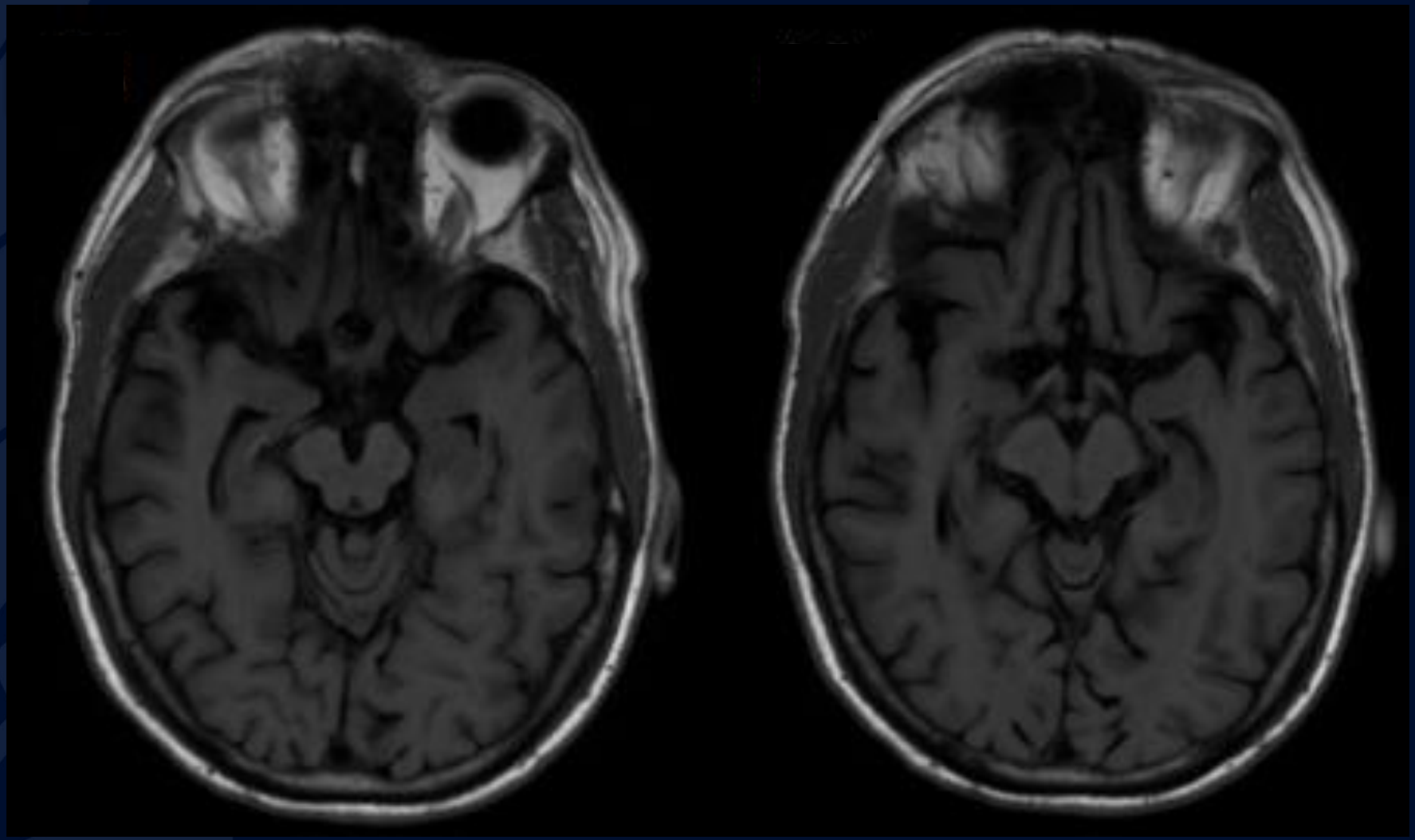
T1



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Progressive Supranuclear Palsy

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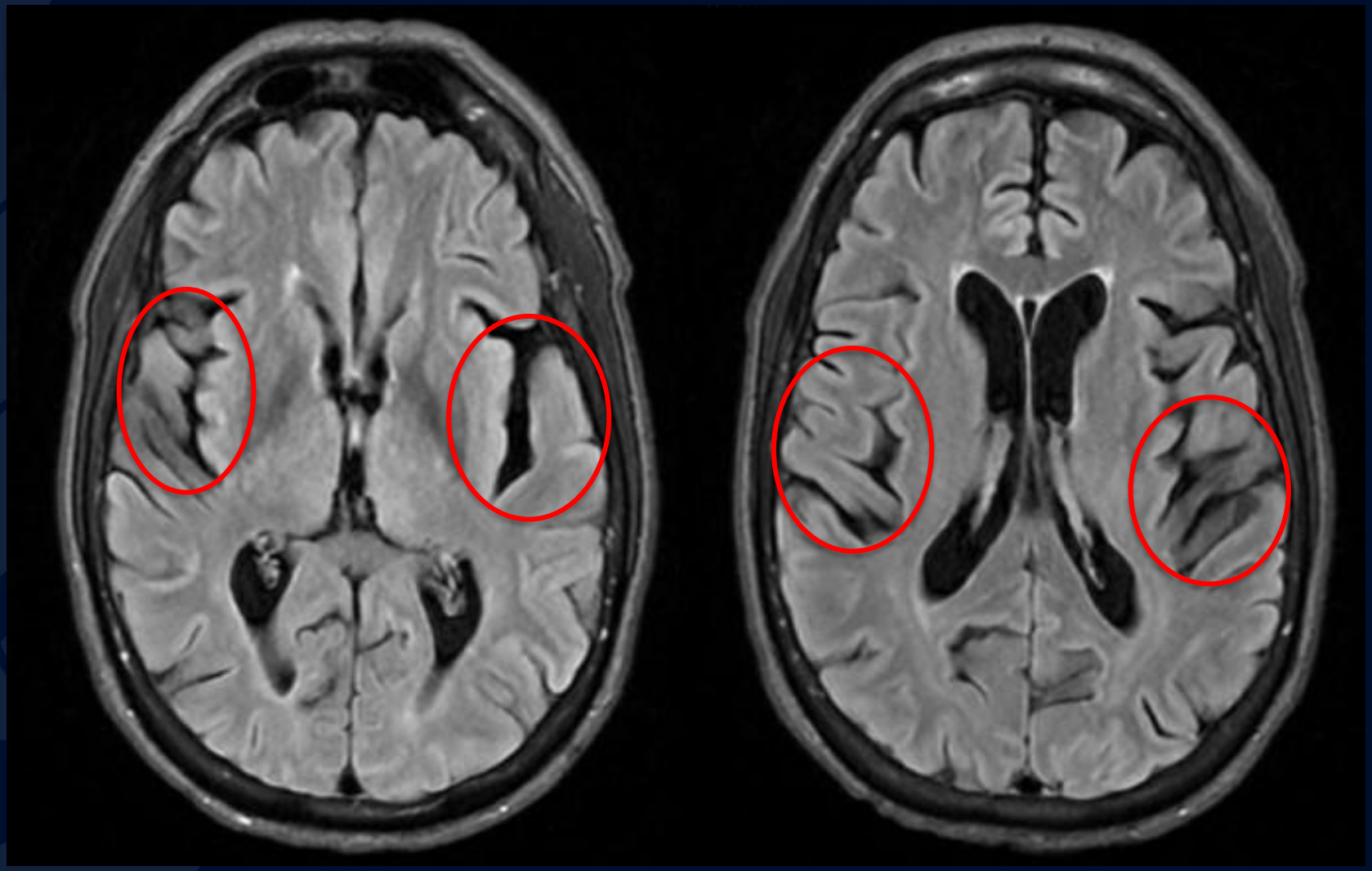
T2 FLAIR



Increased size of sulci, with left greater than right side, indicating parenchymal loss of temporal lobes

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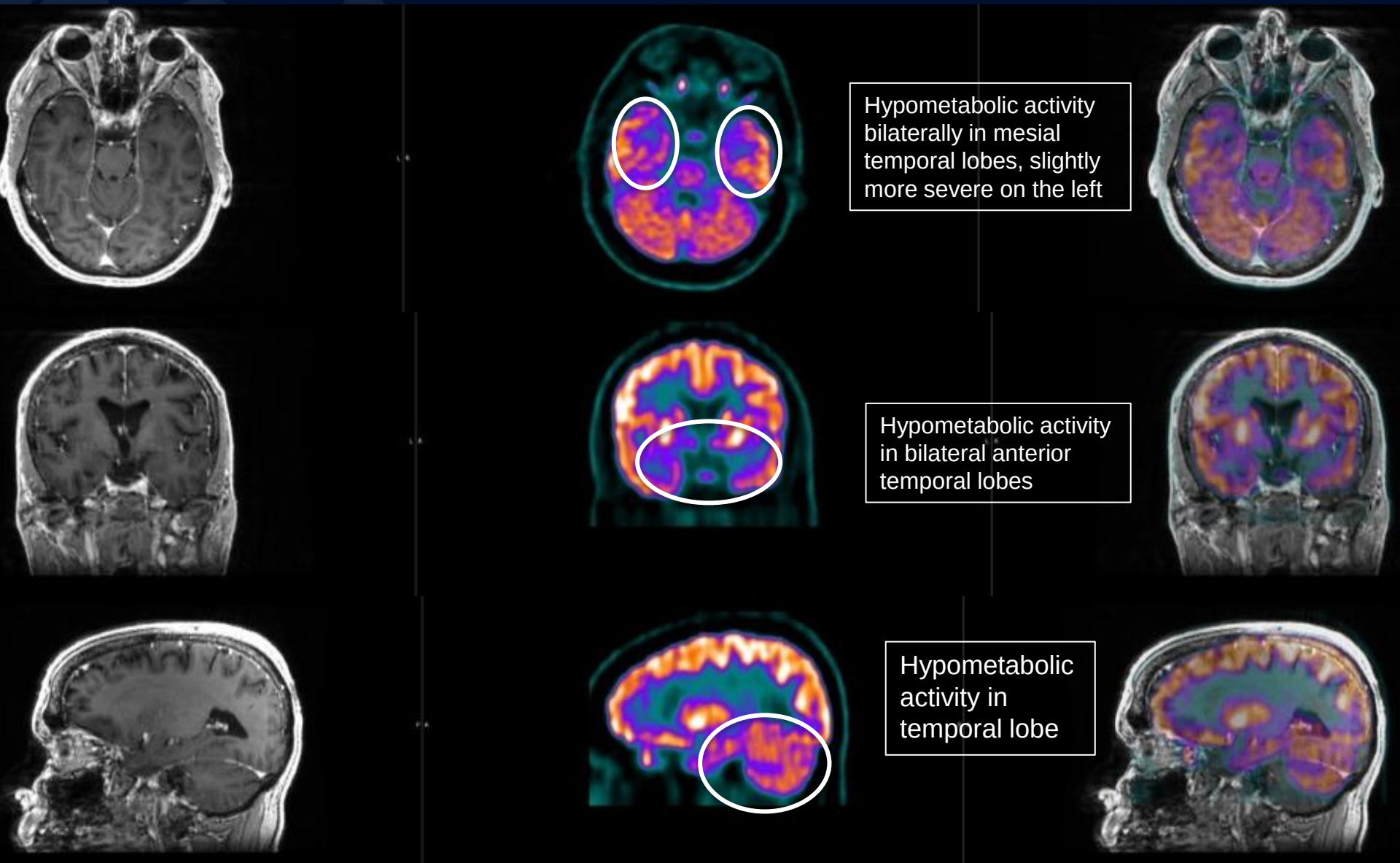
FDG-PET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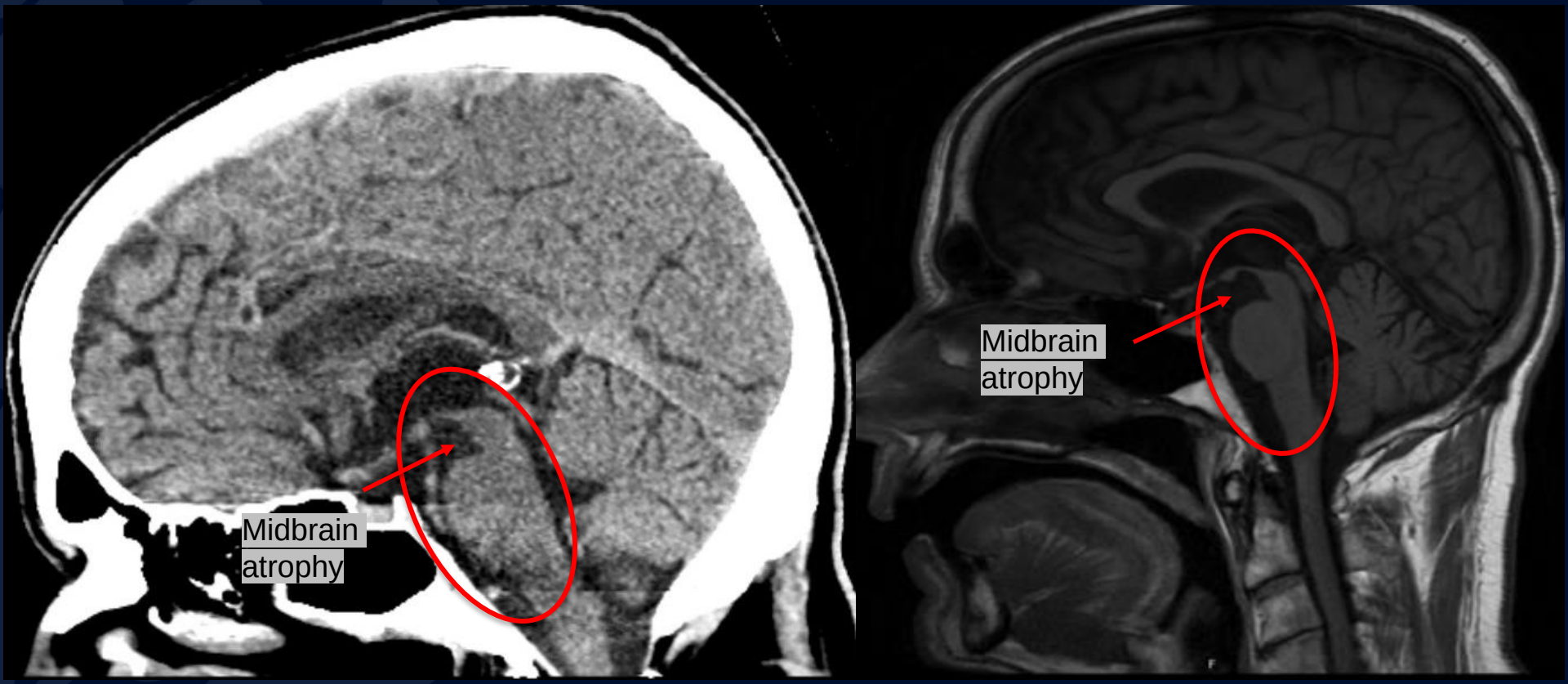

Hummingbird sign:

- Pons forms the body
- Atrophic midbrain forms the head
- Beak is formed by the optic chiasm

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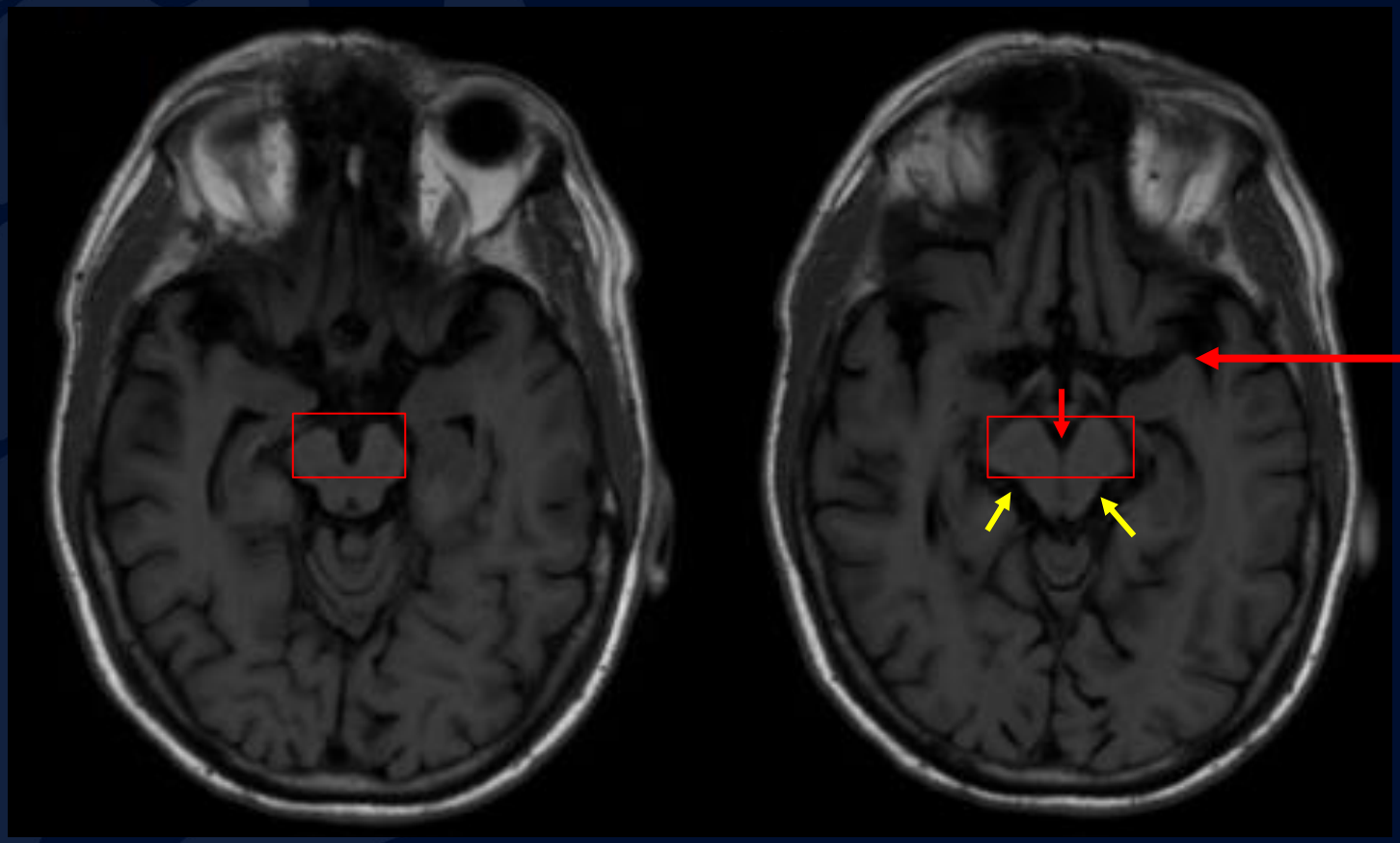
T1



Morning Glory/Mickey Mouse Sign:

- Loss of lateral convex margin of tegmentum of midbrain (golden) with deep interpeduncular cistern (red)

Mickey mouse sign

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Progressive Supranuclear Palsy with Frontal Lobe Presentation

- **Epidemiology:**
  - ~6 per 100,000 persons
- **Etiology:**
  - Build-up of tau protein aggregates that may be due to genetic mutations, toxins, or infectious agent
- **Presentation:**
  - Early: gait difficulty and falls, non-specific dizziness, generalized motor slowing, personality change, executive dysfunction, resting tremor, insomnia
  - Later: worsening parkinsonism, dysarthria, dysphagia, frontal cognitive difficulties, eye movement abnormalities
- **Diagnosis:**
  - Challenging as the variant shares a similar presentation to behavioral variant FTD
  - MRI:
    - Atrophy of midbrain: Hummingbird sign/morning glory sign, Midbrain to pons ratio < 0.52
    - Atrophy of frontal and temporal lobe: hypometabolism on FDG-PET
- **Treatment:**
  - no curative treatment
  - Physical and occupational therapy – management of cognitive, motor, and gait aspects

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