The case of the 'Malingerer'

56 yr old woman

- Caring for husband with PD for last 15 yrs.
- Decreased appetite 2-3 months
- Suspiciousness 1.5 months
- Disorientation to time & place at times 3 weeks
- Sleep was reduced and she c/o pain in legs with difficulty in walking.
- There were stressors.

Examination

- She had no insight
- Marked hypophonia and did not answer most questions.
- She was disoriented to time, oriented to person and place.
- No rigidity, gait was bizarre.
- Routine blood tests were normal
- 10 days later she came with vomiting and altered sensorium of one day. She was mute, occasionally made eye contact, did not answer any questions.

Differential diagnosis of dementia

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Progression

- Rapid
- Chronic

Rapidly progressive Dementias

- Metabolic – B_{12}, CKD, CLD etc
- Infections
- Prion Disease
- Immune mediated
- Neoplastic
- NPH
- Vascular
- Degenerative

Ratnavalli E . Chapter 'Rapidly Progressive Dementias' in Tropical Neurology. Eds Chopra JS and Sawhney IM (ed), ELBS, 2015.

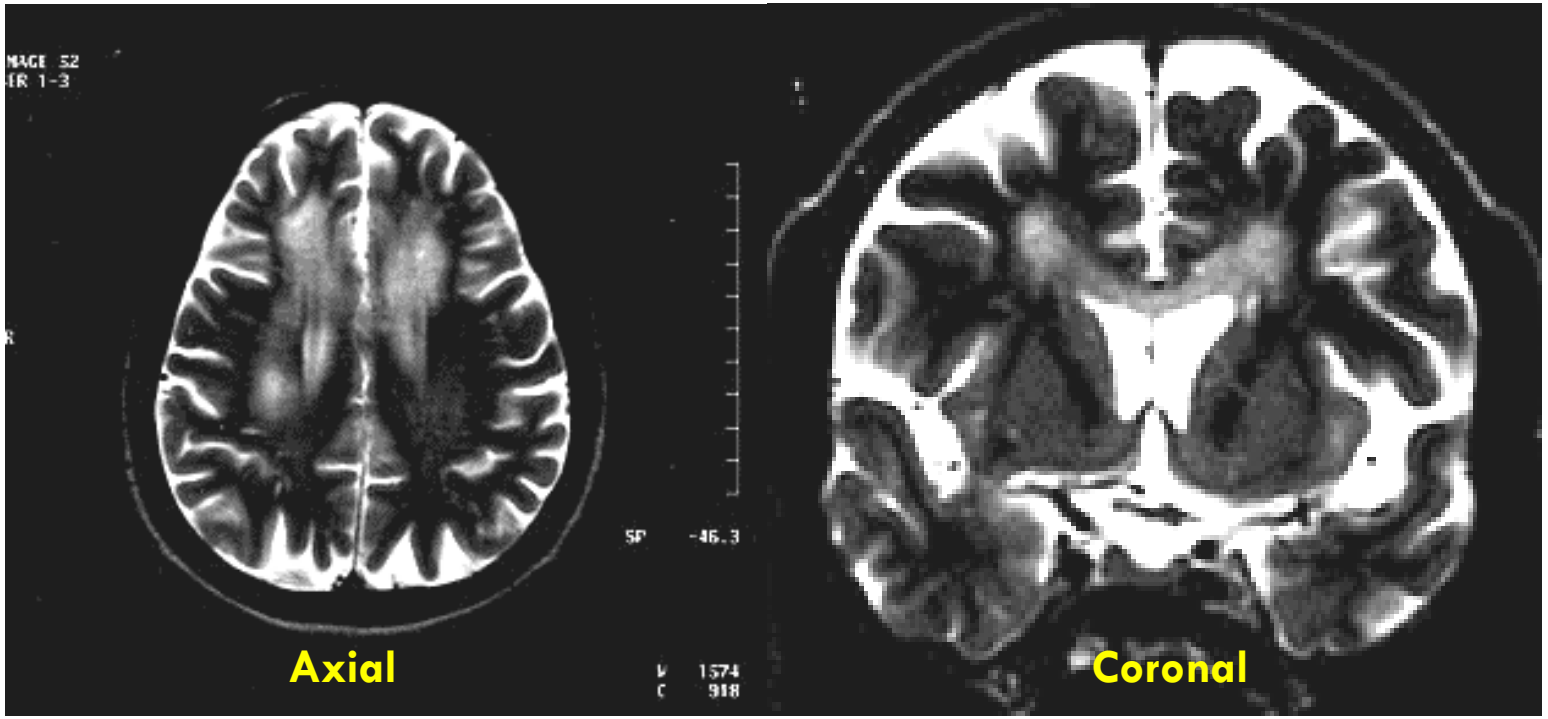
Metabolic/medical

- Onset acute/subacute
- Systemic setting
- Associated neuropathy/myelopathy/tremors
- Anemia/jaundice/knuckle pigmentation
- Associated delirium

The Case of the Uninvited Guest

- Mr.M, a 52 year old technical inspector presented with 6 months personality change
- Abnormal compulsive wandering tendency
- He would get attracted to bright lights of weddings & parties, go uninvited, have a meal and reach back home without losing way.
- He would frequently visit shops and temples and steal money.
- Became quarrelsome at work.

The case of the uninvited guest -- B_{12} Deficiency

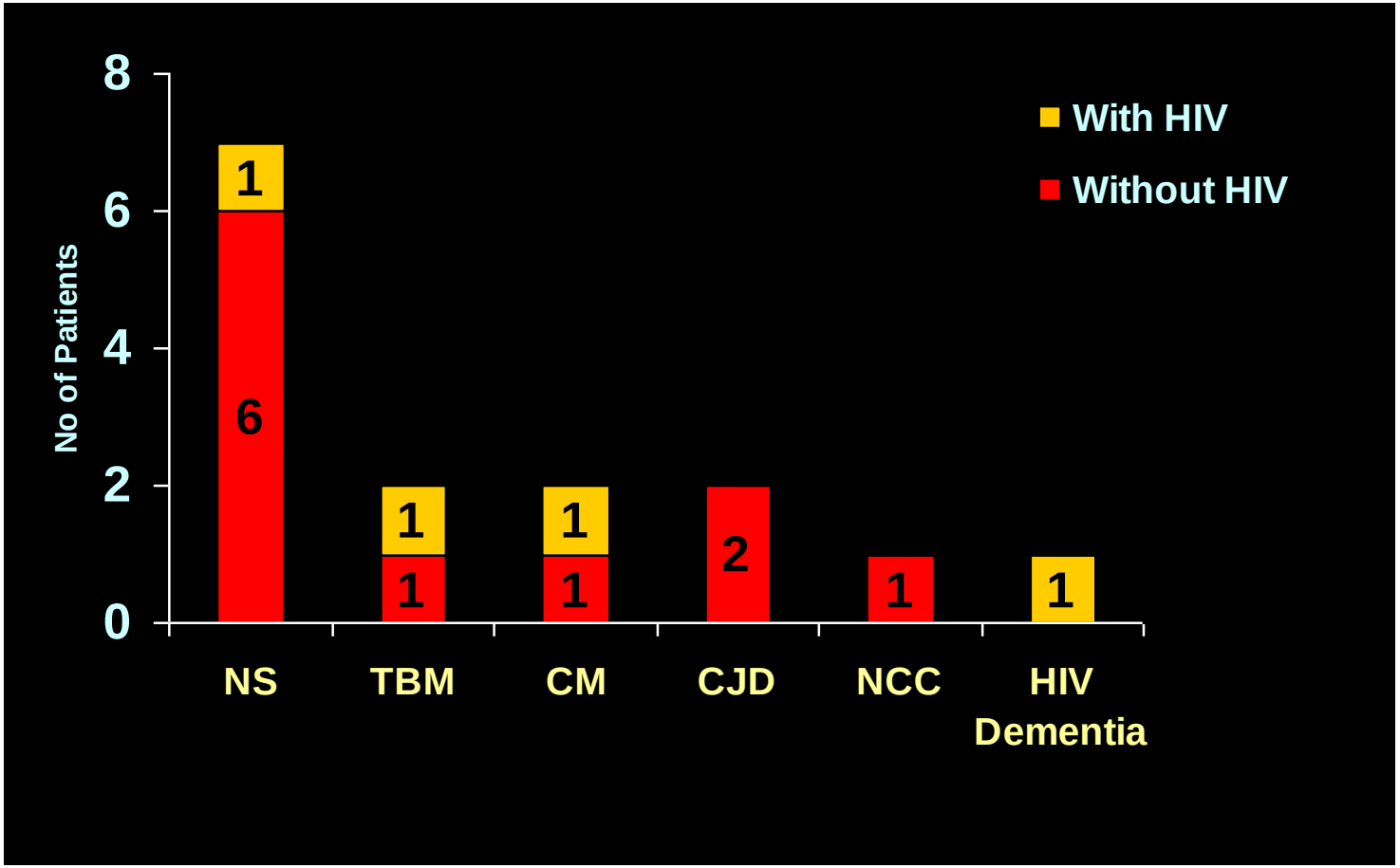


- Serum Vitamin B_{12} was low –150 pg/ml
- Serum Parietal Cell Antibody positive
- Atrophic gastritis on Upper GI Endoscopy

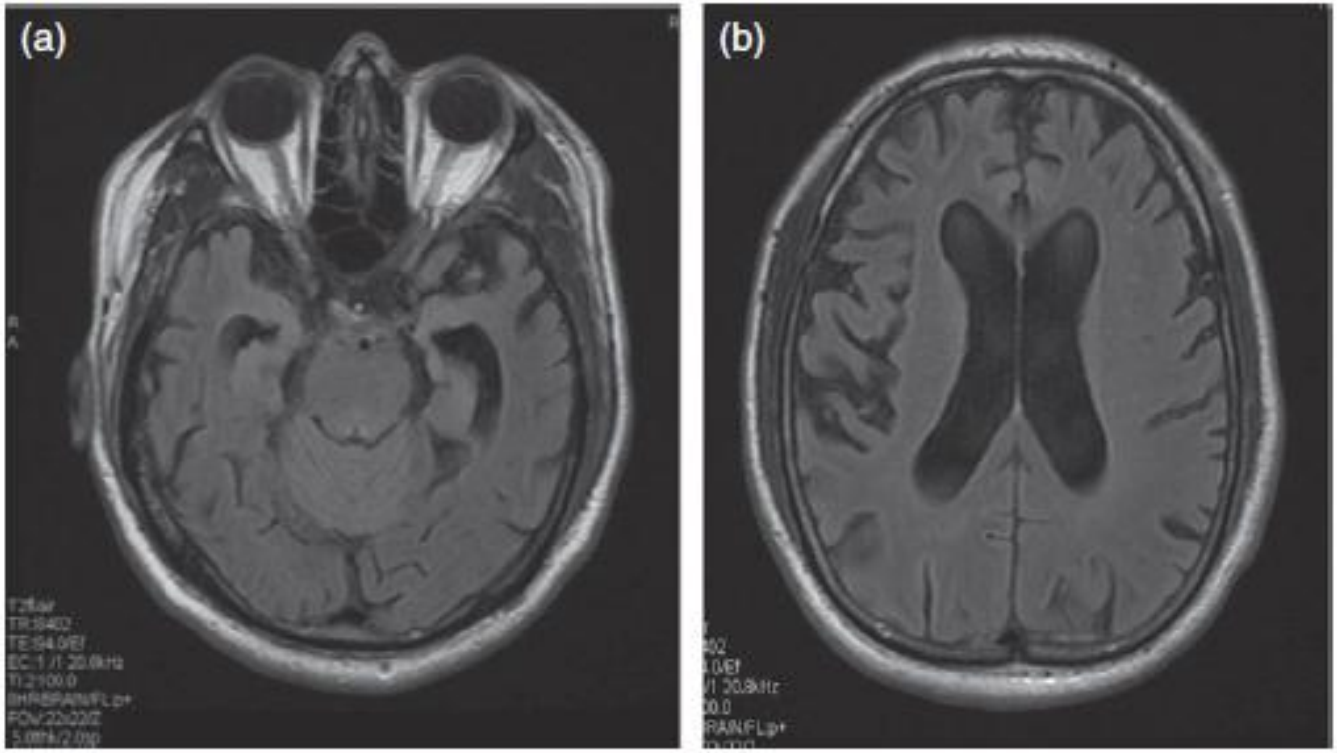
Infections

- Acute/sub acute onset
- Fever, headache, altered sensorium, seizures
- Underlying immunosuppression
- Meningeal signs
- Focal neurological deficits (FND)

Frequency of Neuroinfective dementias



Nagaraja AV, 2003



42 year old with FT syndrome of 6 months – MRI T2 FLAIR Axial shows bilateral frontotemporal atrophy.

Neurosyphilis

Ratnavalli E. Chapter 'Rapidly Progressive Dementias' in Tropical Neurology. Eds Chopra JS and Sawhney IM (ed), ELBS, 2015 in press

Immune Mediated Dementias

- Subacute and rapid progression
- Memory impairment, seizures, fluctn sensorium
- Mood and sleep disorders, hallucns
- Inflammatory CSF; CSF 14-3-3 protein and NSE
- EEG - focal epileptiform discharges
- Small cell, teratoma, testicular T, lymphoma

Ab to intracellular Ag – anti-Hu, anti-Ma2, anti CRMP5
anti- VGKC hyponatremia, dysautonomia
anti – NMDA young F, psychiatric

75-80% Patients respond dramatically to immunosuppression.

Lancet Neurol 2008; 7: 1091–98, Arch Neurol. 2008;65(10):1341-1346

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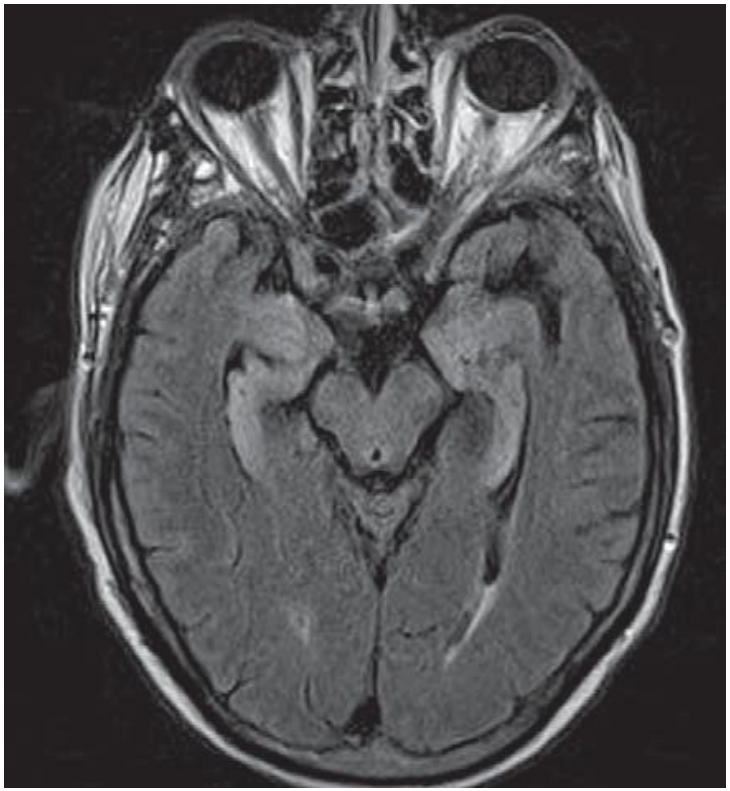
MRI FLAIR AXIAL

Increased signal in b/l hippocampi in a patient with LE



Increased signal in medial temporal lobe in a patient with HHV 6 encephalitis

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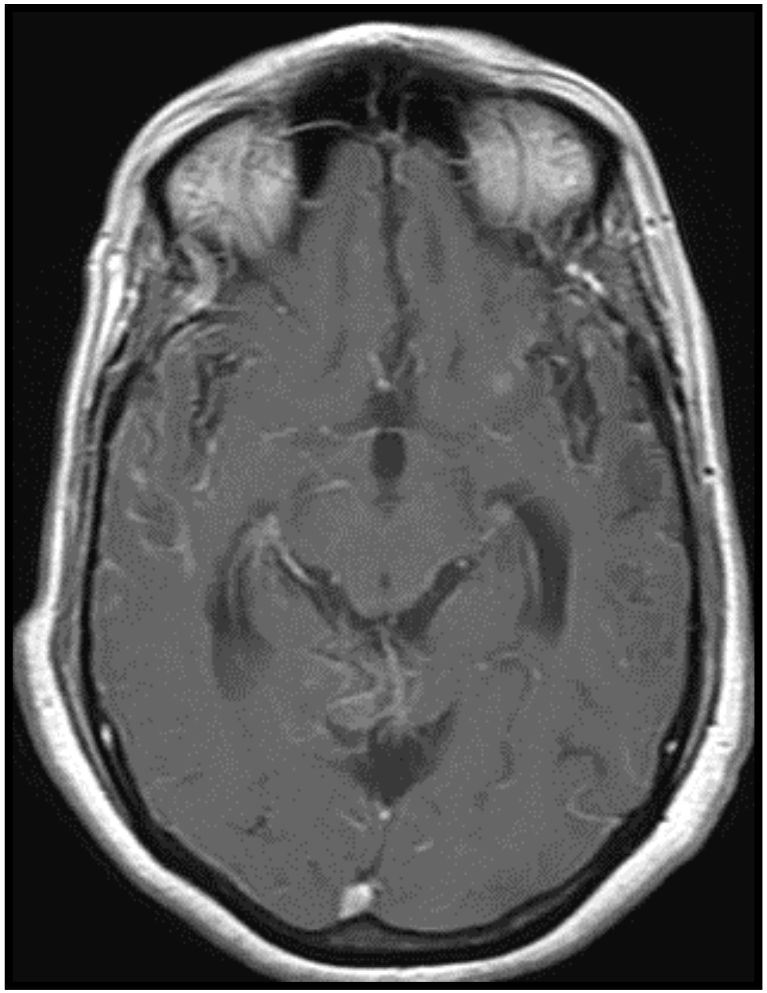
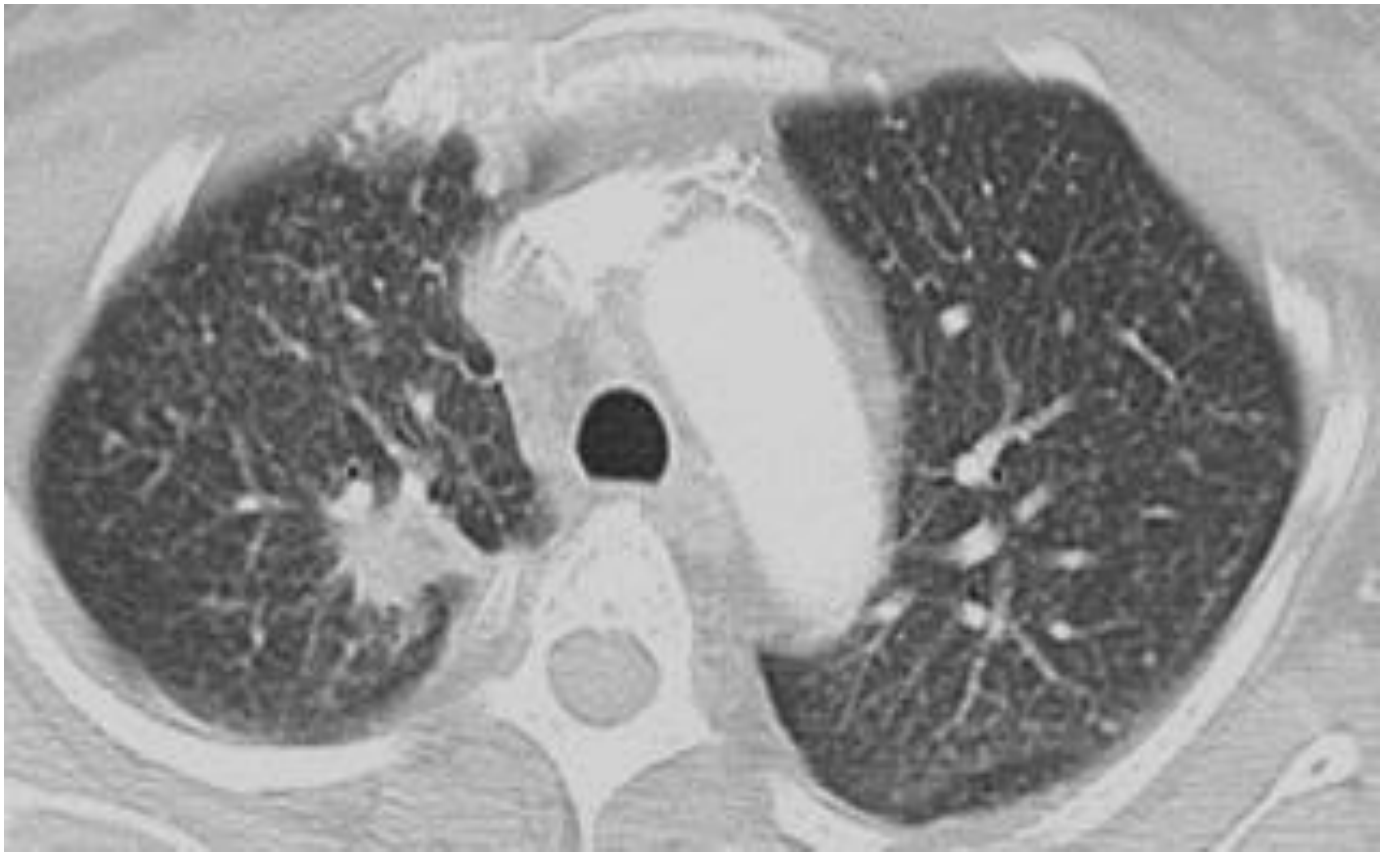
Post contrast Axial T1W image of Brain



The case of the ‘Malingerer’

The case of the ‘Malingerer’

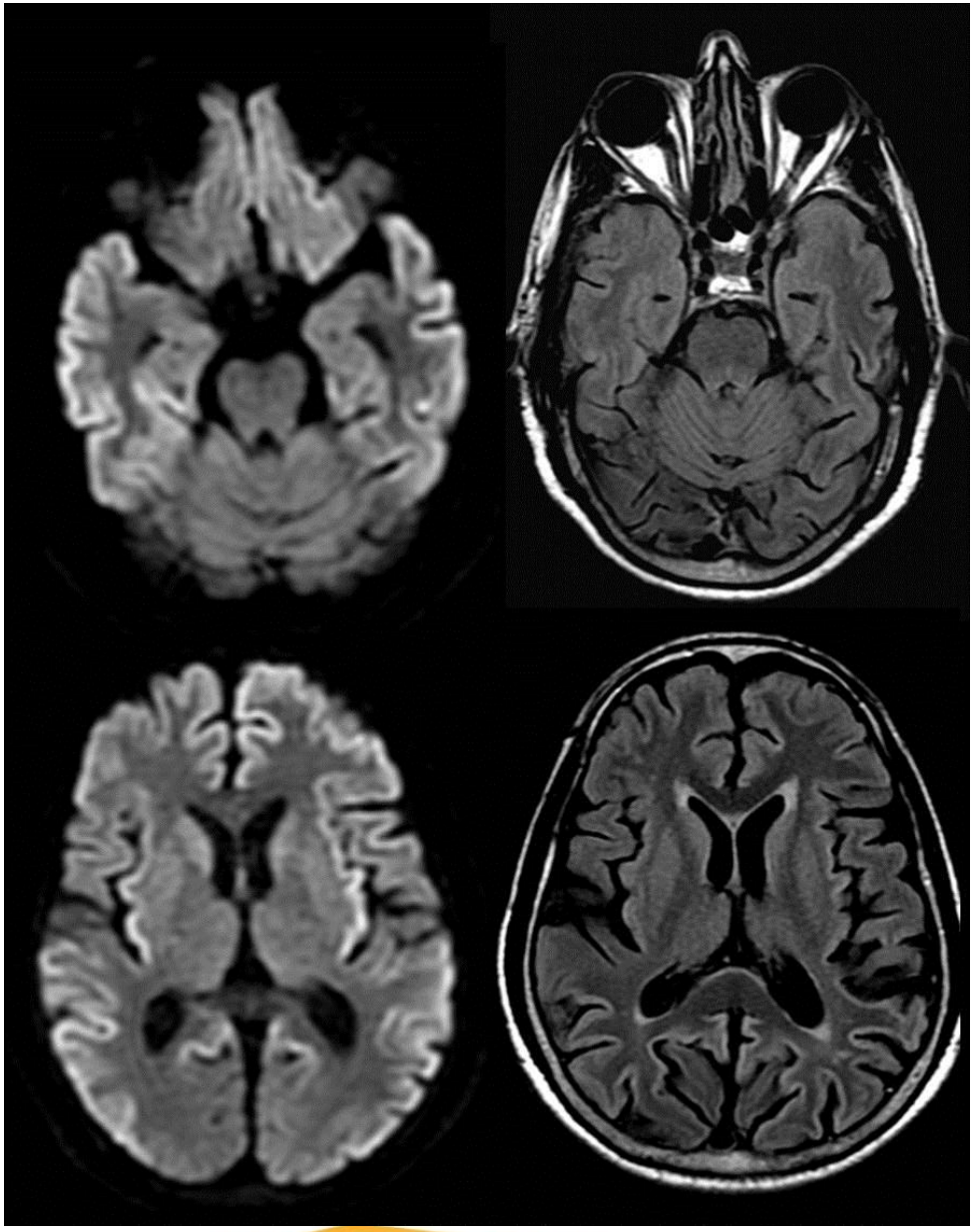


Axial Contrast CT of chest

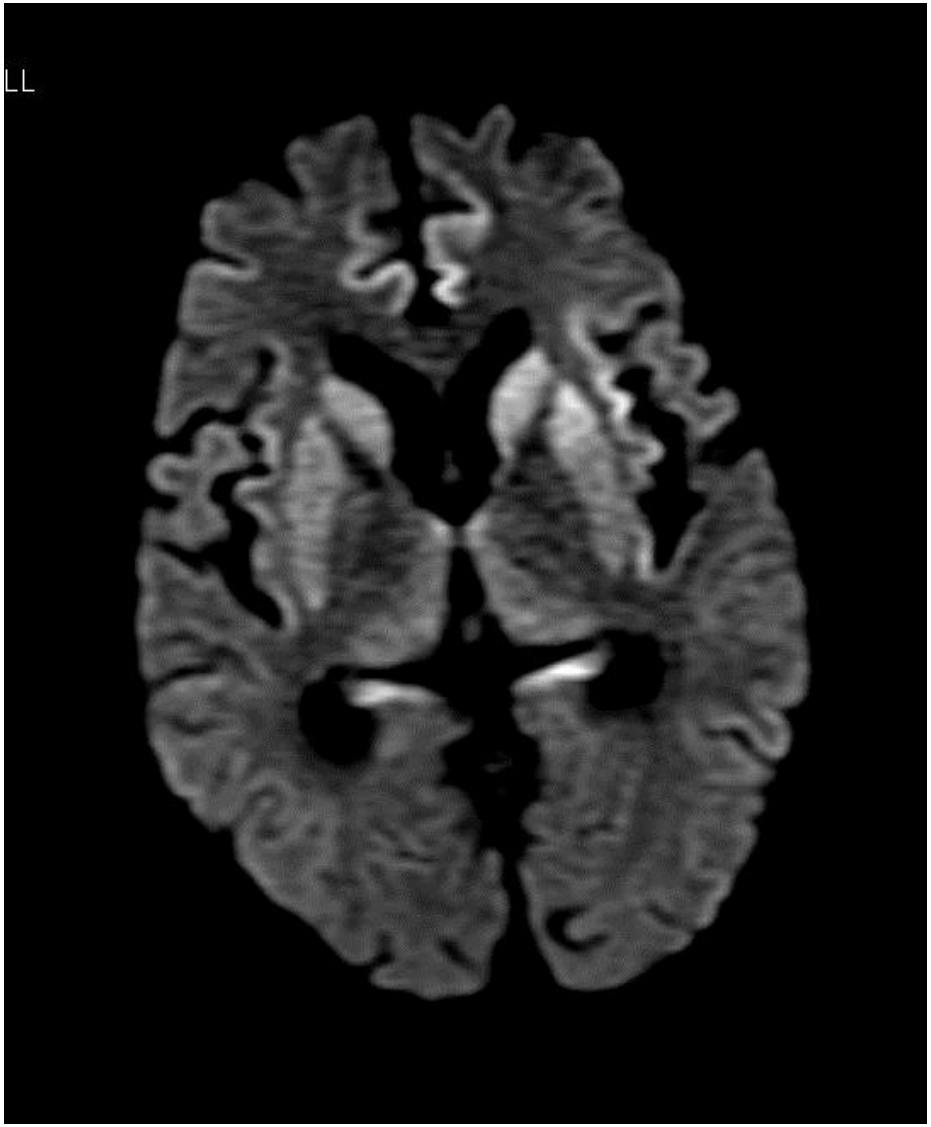
Prion Disease - sCJD

- 50-70 yrs
- Cognitive, cerebellar, behavioural, constitutional, visual, sensory
- Myoclonus, E/P, cortical blindness
- EEG
- CSF
- MRI- DWI and FLAIR- >90% Sen and Sp
- Hypointensity on ADC

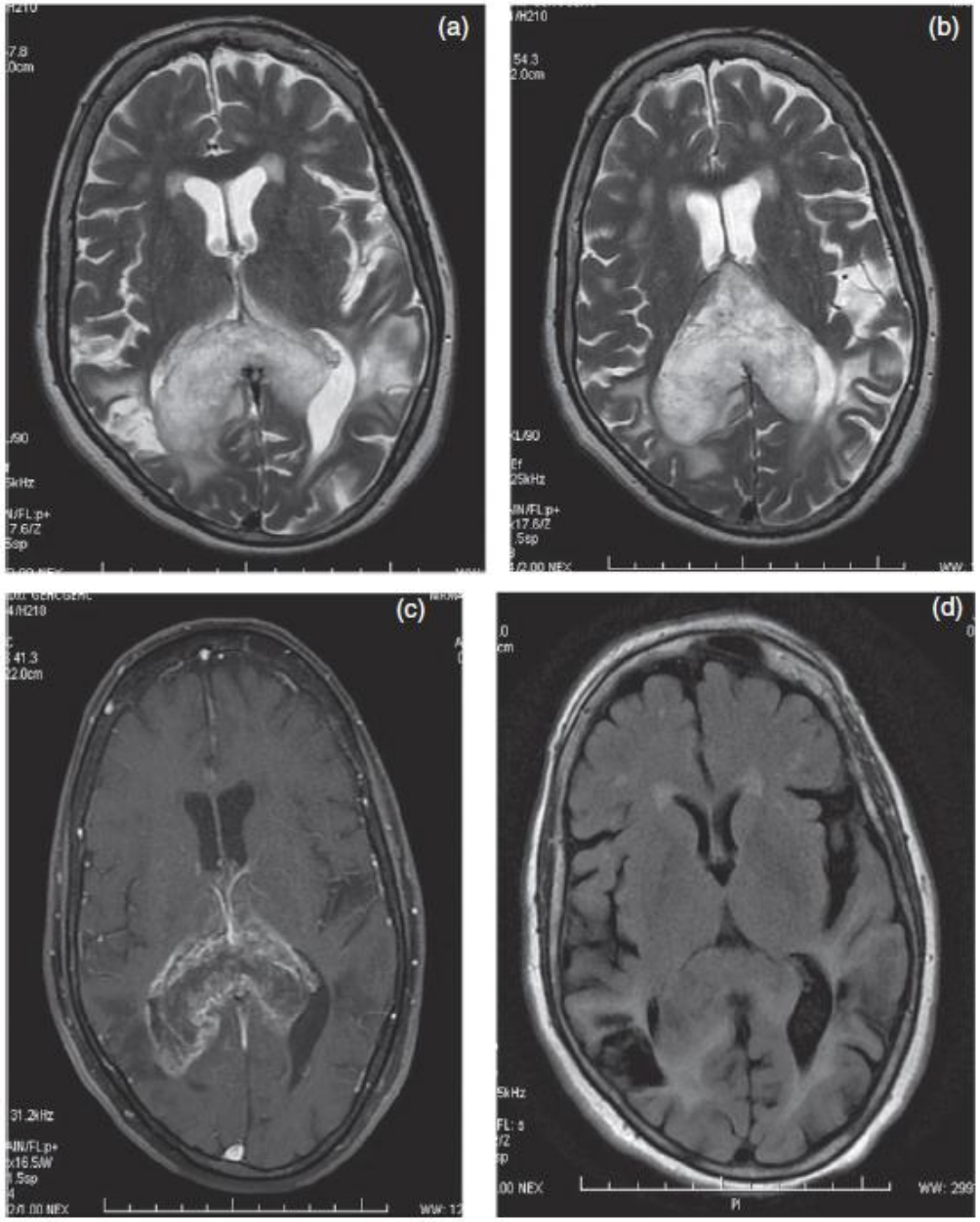
Periodic complexes on EEG in a sporadic CJD patient



DWI Brain – cortical hyper intensities with mild diffuse cerebral atrophy



Axial DWI- hyperintensities b/l insula, basal ganglia and pulvinar (hockey stick appearance) in a sporadic CJD patient

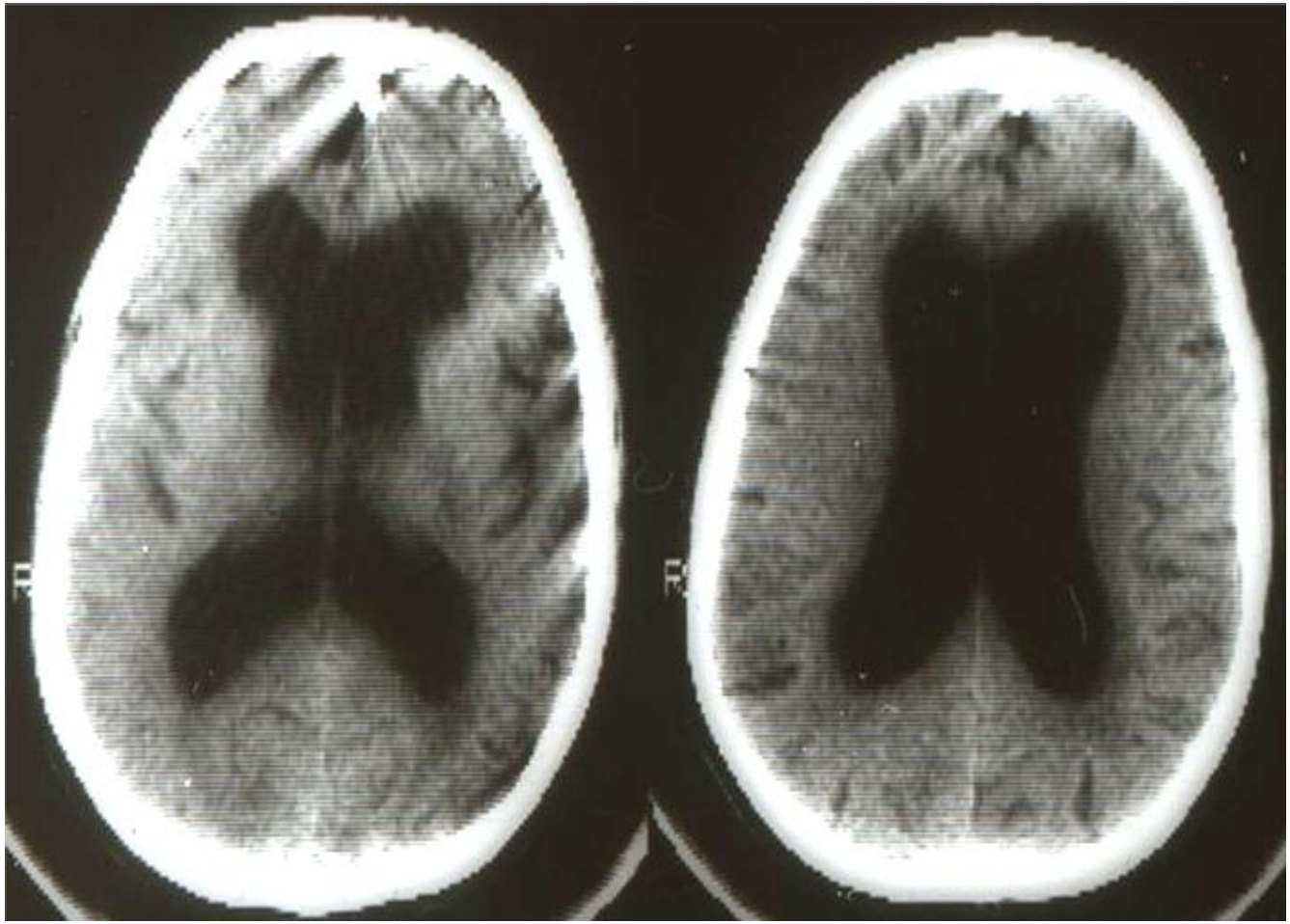


T2 Axial

T2 Axial

68 year old lady with VaD and IHD.
Worsening of cognitive impairment since one month.

T1 FLAIR Axial



CT Scan in Normal Pressure Hydrocephalus

Do Not Miss

- SDH
- Wernicke's encephalopathy (dementia, ataxia and opthalmoplegia)
- Multiple sclerosis

A comparison of cortical and subcortical dementia according to neuropsychological profile

Characteristic	Cortical	Subcortical
Speed of cognitive processing	Normal	Slowed
Planning, problem solving, initiative (frontal "executive" abilities)	Preserved in early stages	Impaired from onset
Personality	Intact until late, unless frontal type	Apathetic, withdrawn
Memory	Severely amnesic	Forgetful
Language	Aphasia	Normal except for dysarthria and reduced output
Visuospatial and perceptual difficulties	Impaired	Impaired
Mood	Depression not uncommon in early Alzheimer's disease	Depression common
Agnosia/prosopagnosia	Often present	Not usually seen

Cortical Vs Subcortical

- Both immediate and delayed free and cued recall deficits in AD and HD. (Delis et al 1991).
- Recognition was better in HD.
- Accelerated forgetting over a 20 minute delay in AD (20%) than in HD (70%).
- Retrieval deficits more common in HD than PD.

  (Zizak et al 2005).
- 25% non demented PD retrieval deficits, 25% like cortical and 50% had no memory deficits

  (Filoteo et al 1997).

Qualitative Analysis

- Drawing to command Vs copying
- AD were significantly worse in command condition but HD impaired in both
- AD pts made conceptual errors while HD made graphic, visuospatial and planning errors. Rouleau et al 1992
- AD were impaired on extrapersonal spatial (RCFT) orientation abilities and HD in personal spatial orientation (money road map test). Brouwers et al 1984

Cortical Vs Subcortical

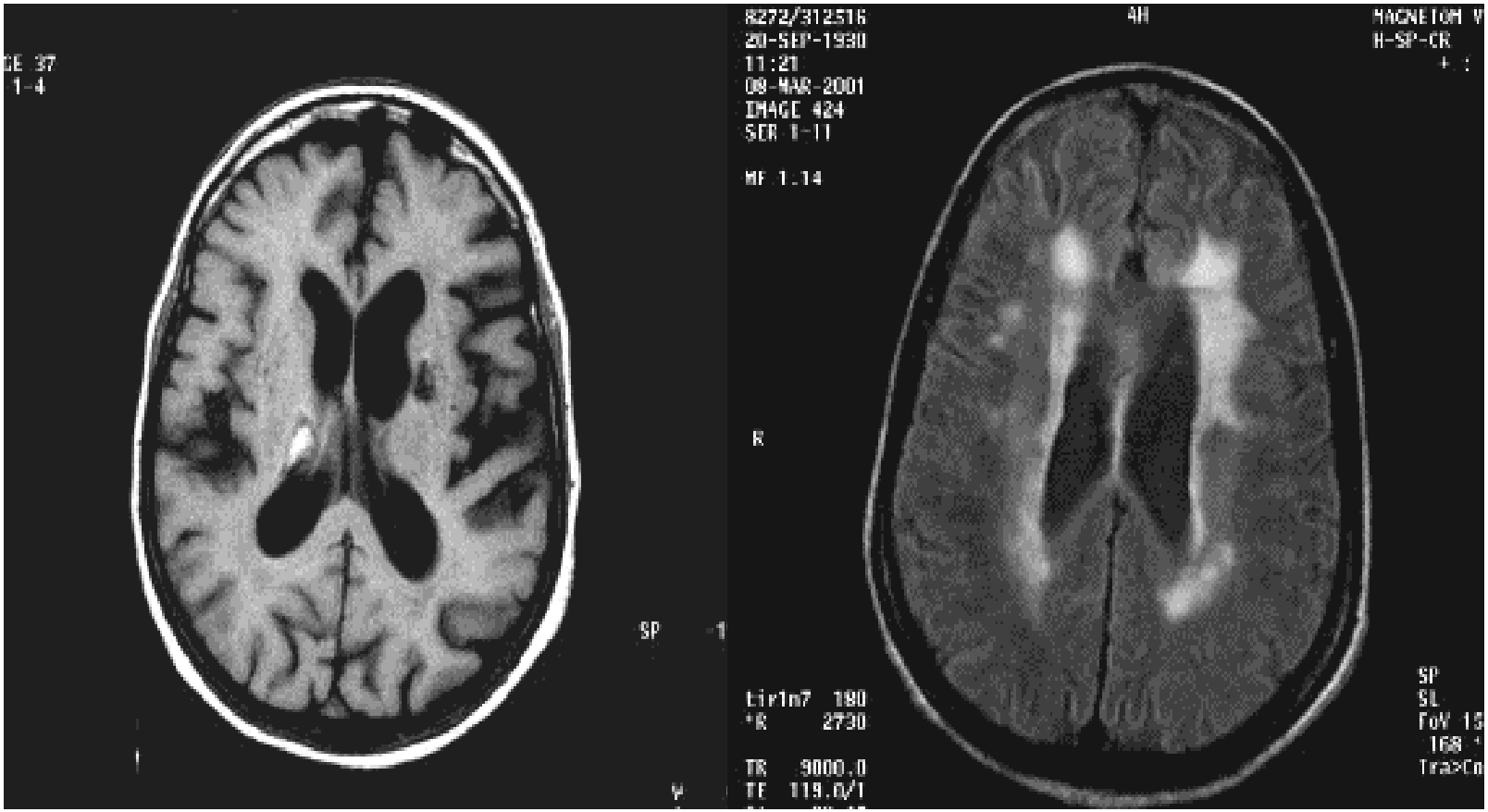
- Animal and letter fluency were both significantly reduced in HD as compared to AD (familial).
- Did not differ on any other measures.

(Arango-Lasprilla et al 2006)

Vascular Dementia

- Sudden/Subacute/chronic onset
- Step wise/Fluctuations/
- FND, Pseudobulbar palsy, early bladder
- Gait disorders, Parkinsonism
- Risk factors for stroke
- Neuroimaging - Infarcts, WM changes

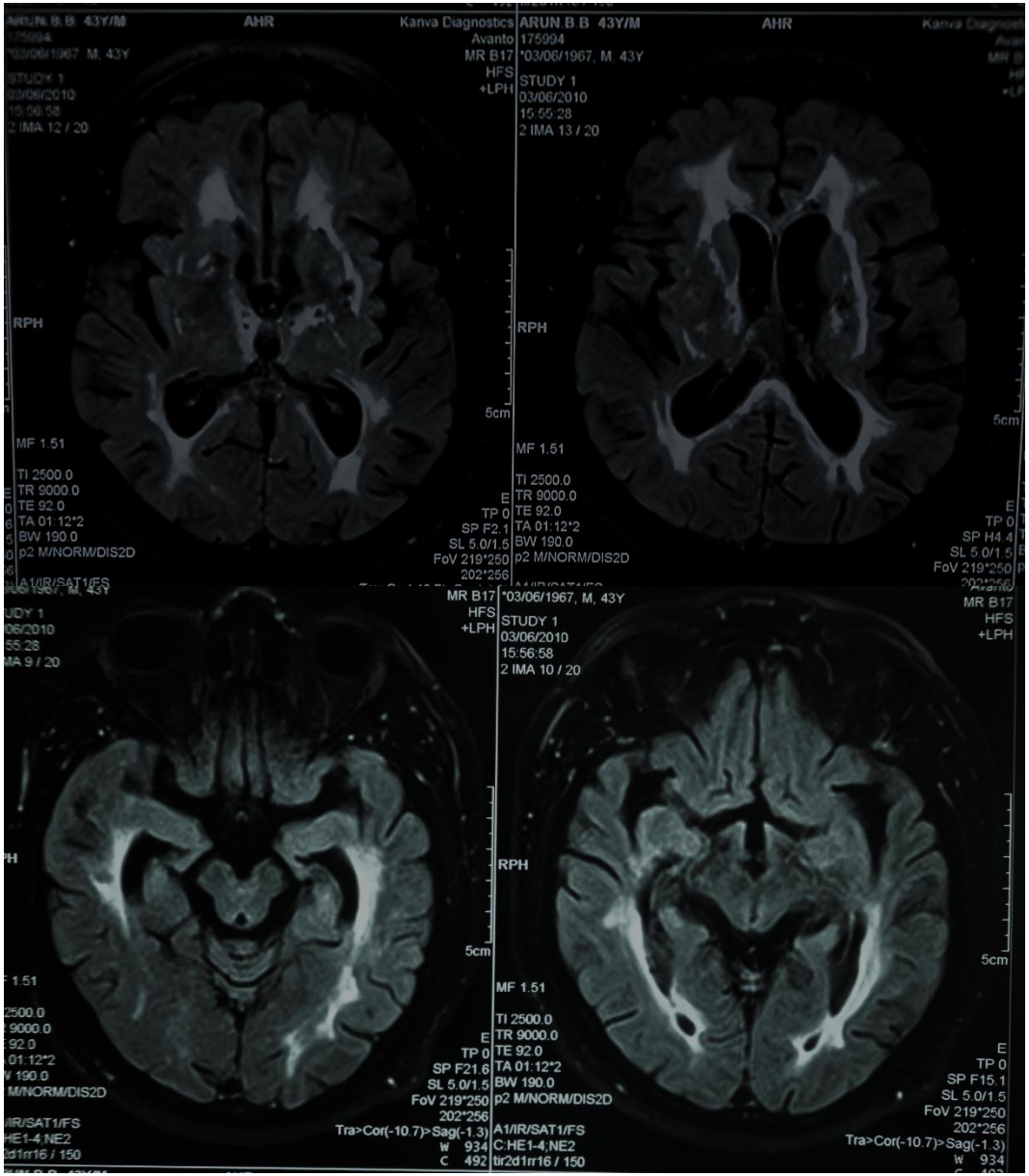
MRI - VaD



T1

Flair

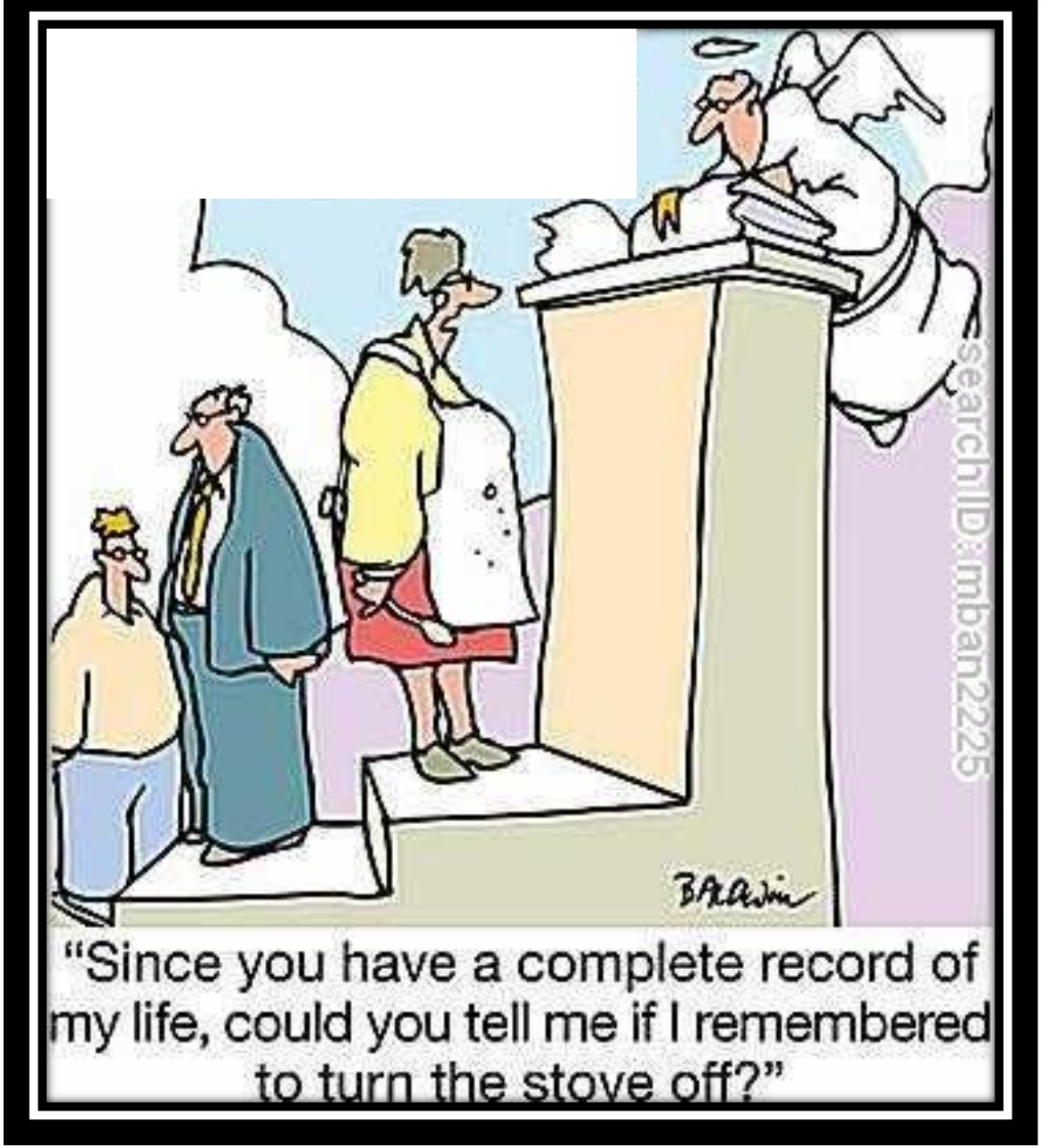
CADASIL



MRI T2 FLAIR
AXIAL

Slowly Progressive

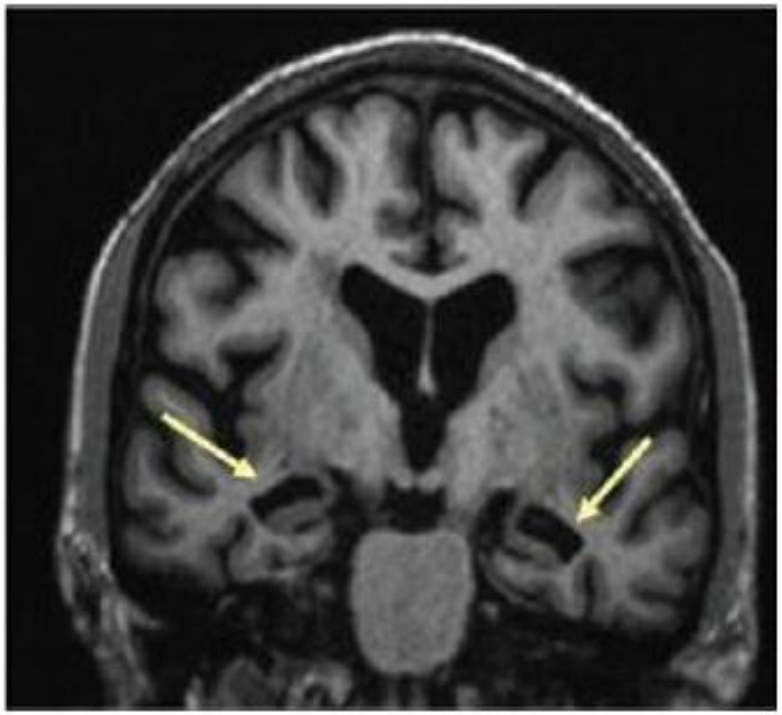
- Amnestic – AD
- Behaviour – bvFTD
- Aphasia – AD (logopenic) FTD (PNFA, SD)
- Extrapyramidal – PSP, CBD, PD, DLB
- Fluctuations – VaD, DLB
- Psychosis – DLB, bvFTD


searchID:mban2225
"Since you have a complete record of my life, could you tell me if I remembered to turn the stove off?"

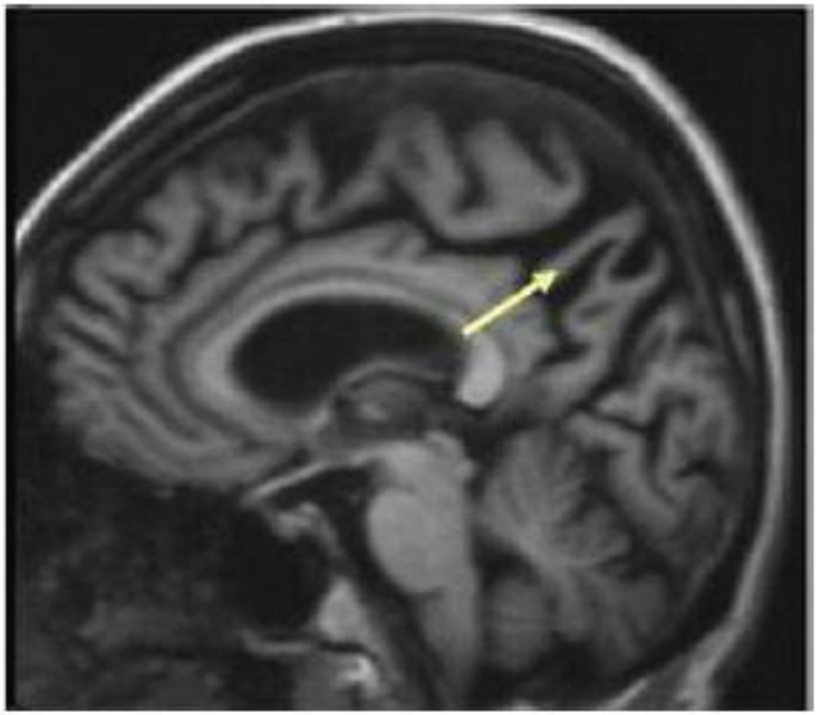
Diagnosis

- Temporal profile of cognitive deficits.
- Early Memory impairment
- Variants (visuospatial, aphasia, apraxia and frontal)
- Onset after 60 yrs.
- Normal Neurological Examination.
- Exclusion of VaD & secondary D.
- CT/MRI SPECT/PET

AD

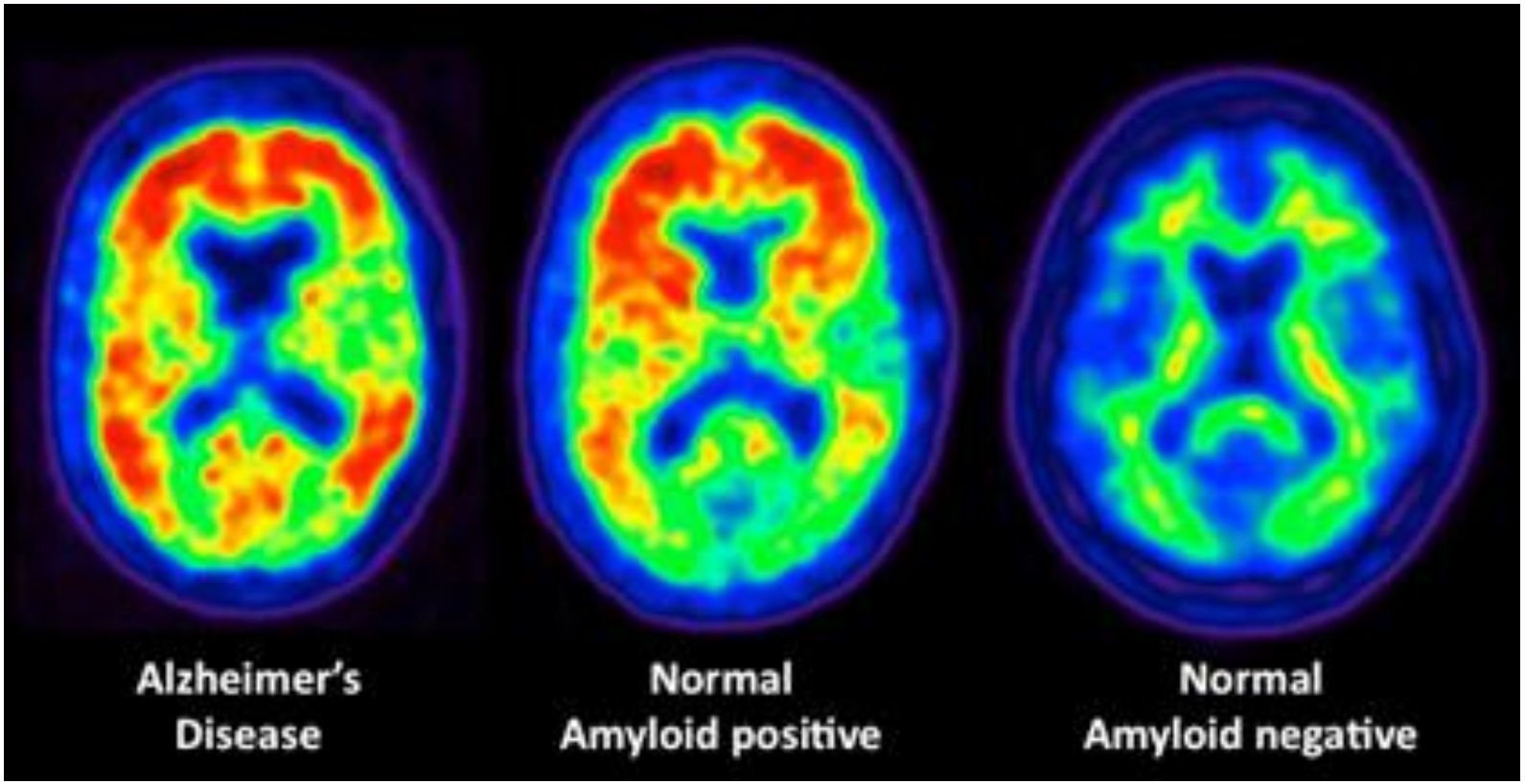


MRI T1 Coronal
B/L medial temporal atrophy



T1 Sagittal
Posterior cortical atrophy

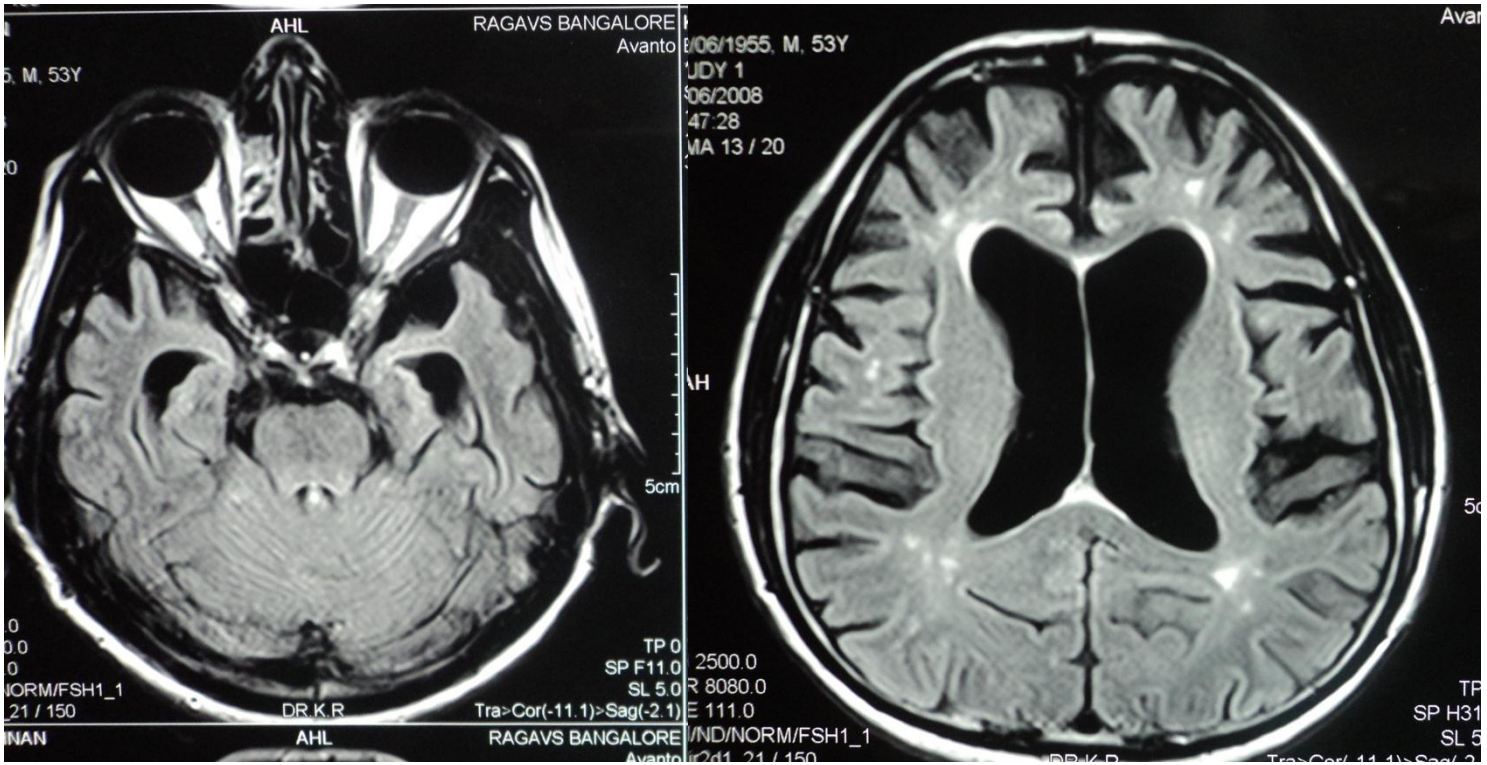
PET- Amyloid Imaging



FTD

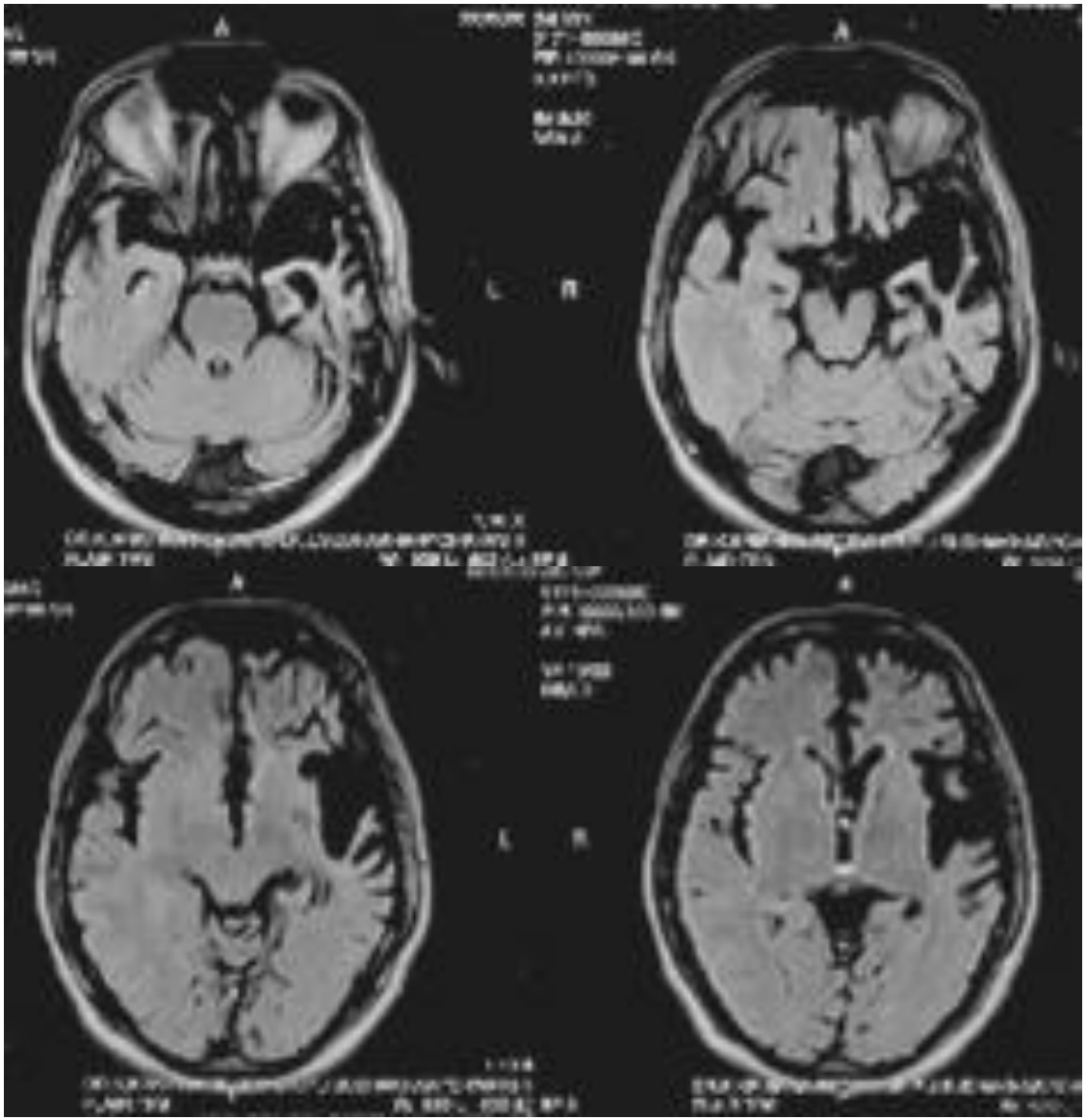
- <65 yrs
- bvFTD- Personality change (disinhibition, apathy, lack of empathy, OCD)
- Aphasia- PNFA or SD
- Associated MND or E/Pyramidal
- Early urinary incontinence
- Loss of insight
- Release reflexes
- Normal EEG
- Asymmetric frontotemporal atrophy

FTD



MRI T2 FLAIR Axial – B/L Frontal and temporal atrophy

MRI T1
Axial



SD

Patient VS - Predominant left anterior temporal atrophy with Dilatation of Sylvian Fissure

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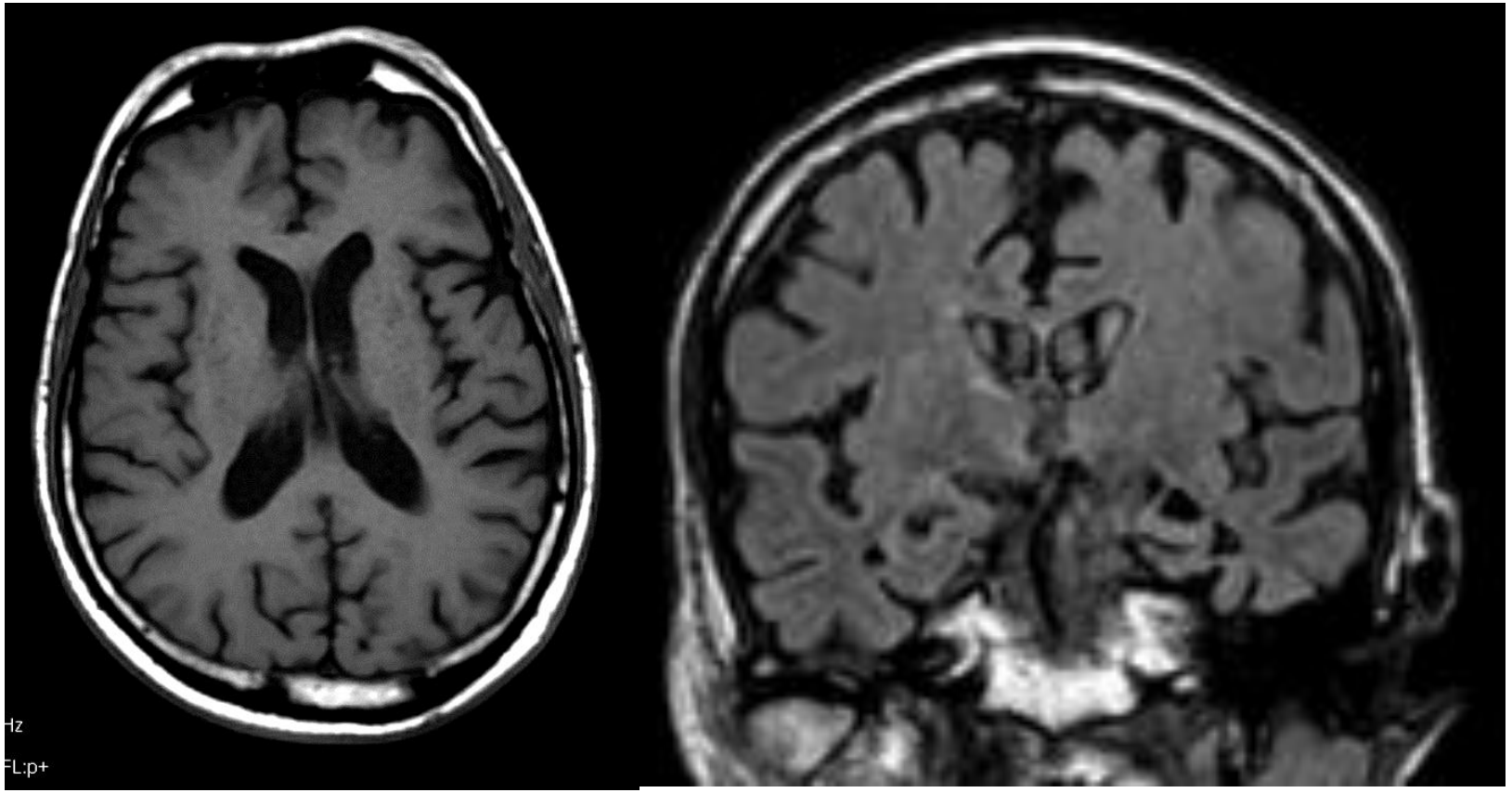
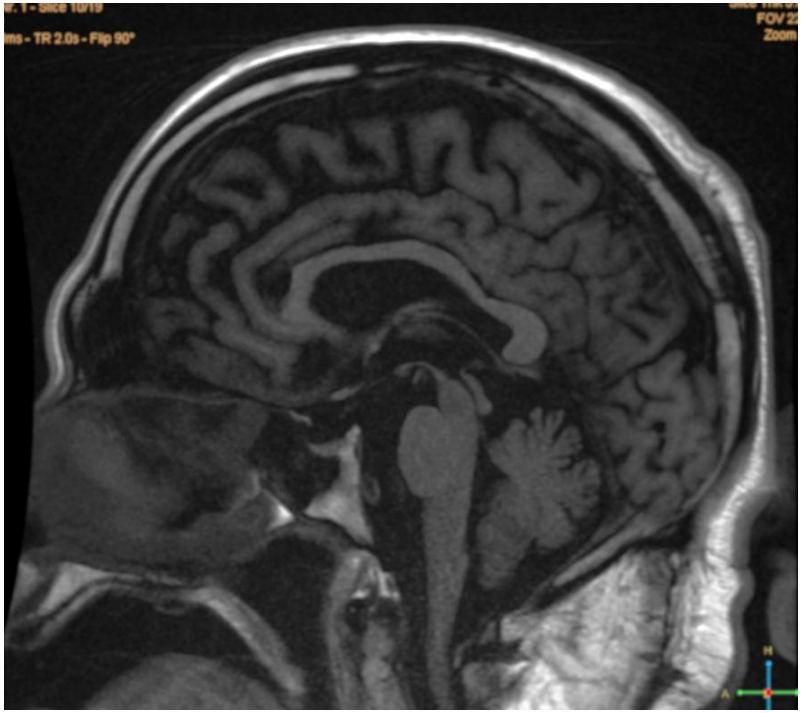


68 yr old man with PNFA – left perisylvian and left temporal lobe atrophy on MRI T1 Axial and Coronal

Ratnavalli E – Ann Indian Acad NeuroSci 2010

FTD overlap syndrome

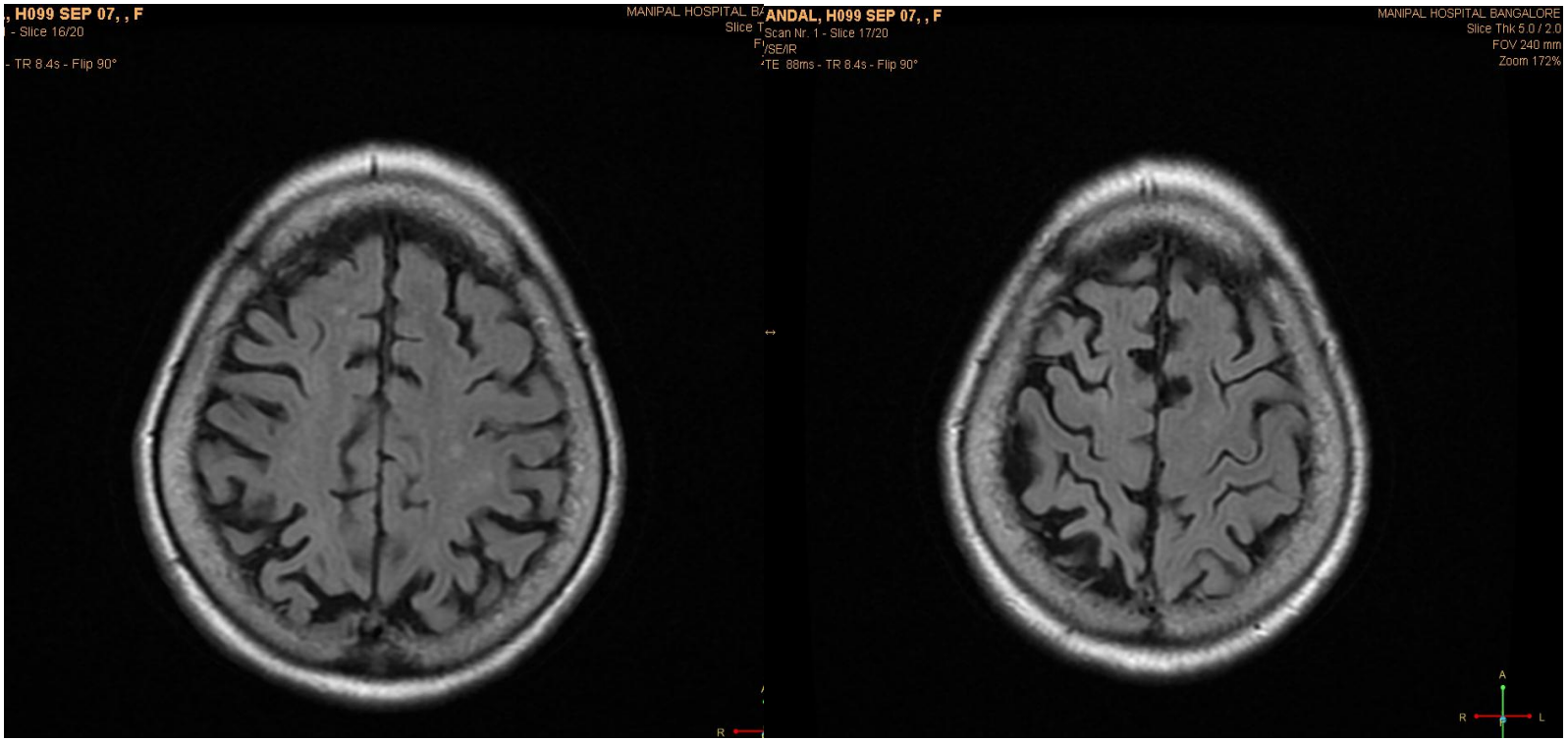
Progressive Supranuclear Palsy (PSP)



MRI T1 Sagittal – Midbrain atrophy ‘Hummingbird sign’ in a 67 yr old man presenting with progressive nonfluent aphasia (PNFA)

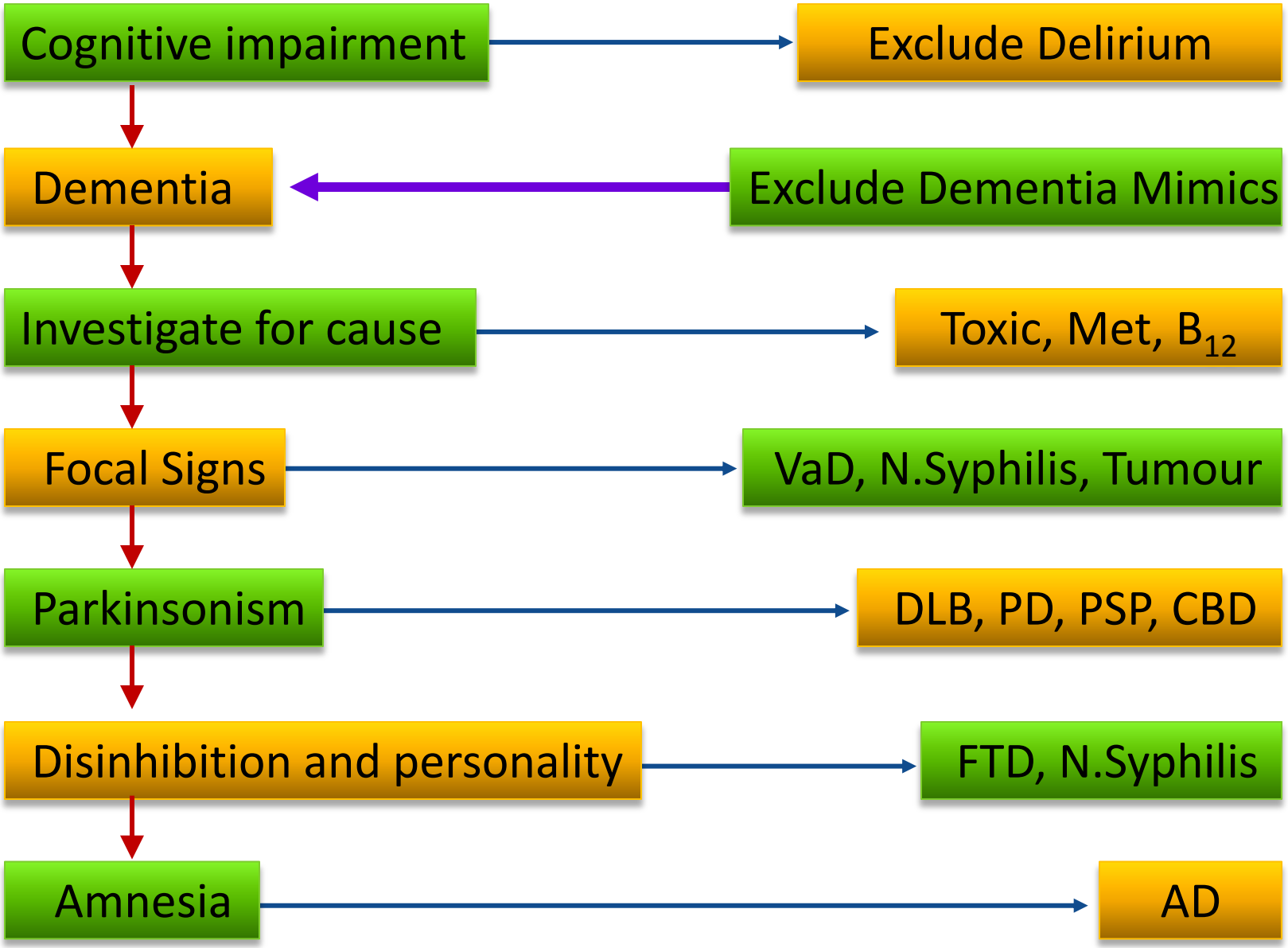
Ratnavalli E – Ann Indian Acad NeuroSci 2010

FTD overlap syndrome
Corticobasal Syndrome (CBS)



MRI T2 FLAIR Axial – Asymmetrical Parietal Atrophy (Rt>Lt)

	AD	FTD	DLB	VaD
Attention	N	Impaired	N/impaired	May be Impaired
Fluency	Letter F better	Category F better	Decreased	Decreased
Visuospatial	Yes	Preserved	Striking	Maybe
Executive	Later	Impaired	Impaired	Impaired
Hallucinations	Yes		Prominent	Yes
Disinhibition		Yes		
Eating disorder		Yes		
Associated Deficits		E/P, MND	E/P, REM	FND, Gait, early incontinence


Cognitive impairment
Exclude Delirium
Dementia
Exclude Dementia Mimics
Investigate for cause
Toxic, Met, B_{12}
Focal Signs
VaD, N.Syphilis, Tumour
Parkinsonism
DLB, PD, PSP, CBD
Disinhibition and personality
FTD, N.Syphilis
Amnesia
AD

Conclusions

- A step-by –step approach is required.
- Neuropsychological evaluation is useful in degenerative dementias.
- Longitudinal evaluation is important for accurate diagnosis.
- All RPDs need fast and thorough investigations.
- All efforts should be made to identify and subtype the dementia …..this is important for prognosis and treatment.

Acknowledgments

- Patients and their families
- Teachers - Prof.Ravi Nehru, Dr.Frank Benson
- Colleagues for referrals – Uday Muthane, Uday Murgod, Murali S, TG Suresh, Vikram Kamath, Raghavendra, Pramod Pal, Soumya Hegde, Roop Gursahni, Sudhir Kothari, Shobha, Anuradha and many others
- Prof.Shankar, NIMHANS for pathology slides.

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