



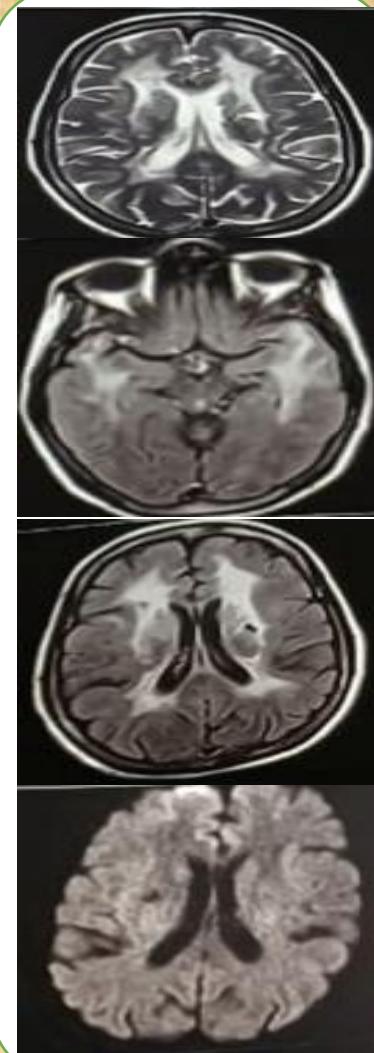
COGNITION BEYOND DEMENTIA : THE CONUNDRUM OF WHITE MATTER COGNITION DR SATYADIP SARKAR, BIN ,IPGMR,KOLKATA



THE VESSEL STORY

48 yrs /F without any comorbidity with DO of 18 months

- ❖ Progressive attention and executive dysfunction.
- ❖ Lack of motivation/apathy and impaired TOM
- ❖ Pseudobulbar affect/pathological crying for 6 months.
- ❖ No H/O memory ,language, HOVD, impulsivity or wandering, headache, seizure, motor or sensory impairment.
- ❖ **FAMILY H/O PSYCHIATRIC ILLNESS (UNDIAGNOSED)**
- ❖ ACE III : 51/100 (A 7/18; M 7/26; F 4/14; L 14/26; V 19/26)
- ❖ FAB : 6/20 (GO NO GO: 3 and prehension behavior 3)
- ❖ Routine blood parameters – WNL/SEROLOGY-NR/EEG –NAD
- ❖ ANA,ANCA profile /anti TPO ab- WNL/syphilis serology-NR
- ❖ **MRA/CTA- NAD, MR VWS-NAD.**
- ❖ AIE profile Neg/NMO &MOG Ab-Neg



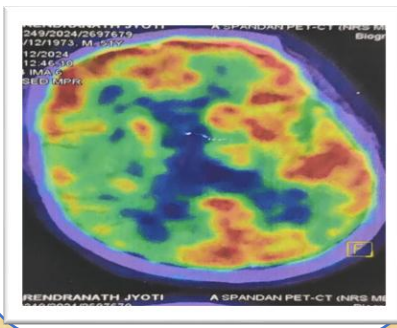
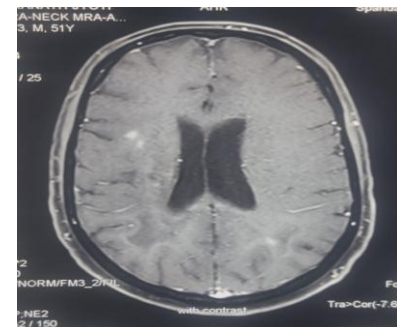
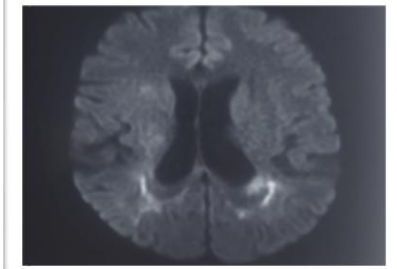
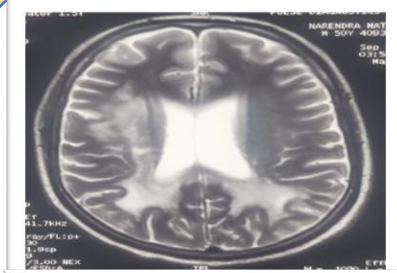
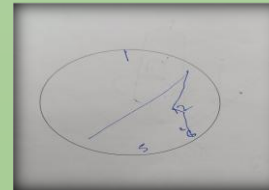
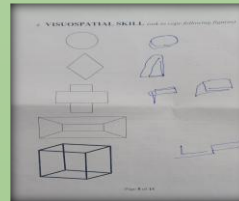
Gene# (Transcript)	Location	Variant	Zygosity	Disease (OMIM)	Inheritance	Classification ⁵
NOTCH3 (-) (ENST00000263388.7)	Exon 4	c.505C>T (p.Arg169 Cys)	Heterozygous	Cerebral arteriopathy with subcortical infarcts and leukoencephalopathy 1 (OMIM#125310)	Autosomal dominant	Pathogenic (PS3,PS4)

WHAT THE MIND DOES NOT KNOW, THE EYES CANNOT SEE

52 YRS/M without any comorbidity with DO of 6 months...

- ❖ Subacute onset left hemiparesis(resolving course)
- ❖ difficulty in driving car due to depth and motion perception
- ❖ Topographical/heading disorientation.
- ❖ Acalculia.
- ❖ No h/o object/color/face recognition.
- ❖ No h/o memory/language/attention/frontal/Dys-execution

- ❖ Left spastic hemiparesis
- ❖ MMSE 24/30,FDS 4,BDS 0
- ❖ free recall 0,DCR 12/20
- ❖ Fluency/comprehension/semantics/praxis--Normal
- ❖ DOT COUNTING/FLT--- impaired, TMT A/B ---Impaired
- ❖ DORSAL SIMULTAGNOSIA +
- ❖ LEFT HEMINEGLECT +



Gene# (Transcript)	Location	Variant	Zygosity	Disease (OMIM)	Inheritance	Classification ⁵
ABCD1 (+) (ENST00000218104.6)	Exon 1	c.838C>T (p.Arg280Cys)	Hemizygous	Adrenoleukodystrophy (OMIM#300100)	X-linked	Likely Pathogenic (PM1, PM2, PP3, PP5)

THE GREAT RADIOLOGICAL MIMICKER

61 YRS/F without any comorbidity with DO of 3 months

- ❖ Rapidly progressive multidomain cognitive deficits.
- ❖ Executive dysfunction/severe attention and working memory deficit.
- ❖ Apathy with slowed IPS WITHOUT MOTOR OR ANY SYSTEMIC SYMPTOMS.
- ❖ MMSE 16/30, ACE III 62, FDS 1, BDS 0, EOO-4, LEXICAL FLUENCY 0
- ❖ Immediate recall 04, free recall 0, cued recall 12.
- ❖ TMT A/B, GO NO GO, LURIA , Abstraction ---impaired.
- ❖ Routine blood WNL/serology-NR/vit B12/anti TPO—WNL
- ❖ EEG/NCS/VEP Study --Normal
- ❖ ANA 3+ (SPECKLED)/RO 52 +VE/SS-A TITRE inc./low C3
- ❖ NMO&MOG –Neg/AIE profile –Neg/SCHIRMER TEST +VE
- ❖ LIP BIOPSY FOCUS SCORE 2.

NO PATHOGENIC OR LIKELY PATHOGENIC VARIANTS CAUSATIVE OF THE
REPORTED PHENOTYPE WERE DETECTED

