Interesting Neuro Case

- Mohull Mehta, Wayne State

Case Presentation

HPI:

- 73-year-old woman presented after mechanical fall from bed
- Also noted to have 2-3 months of progressive confusion; history obtained from partner
- **PMH**: subarachnoid hemorrhage in 2010 with residual right-sided weakness, HTN
- **Social hx**: Lives with partner, was independently feeding, bathing, ambulating until last month.

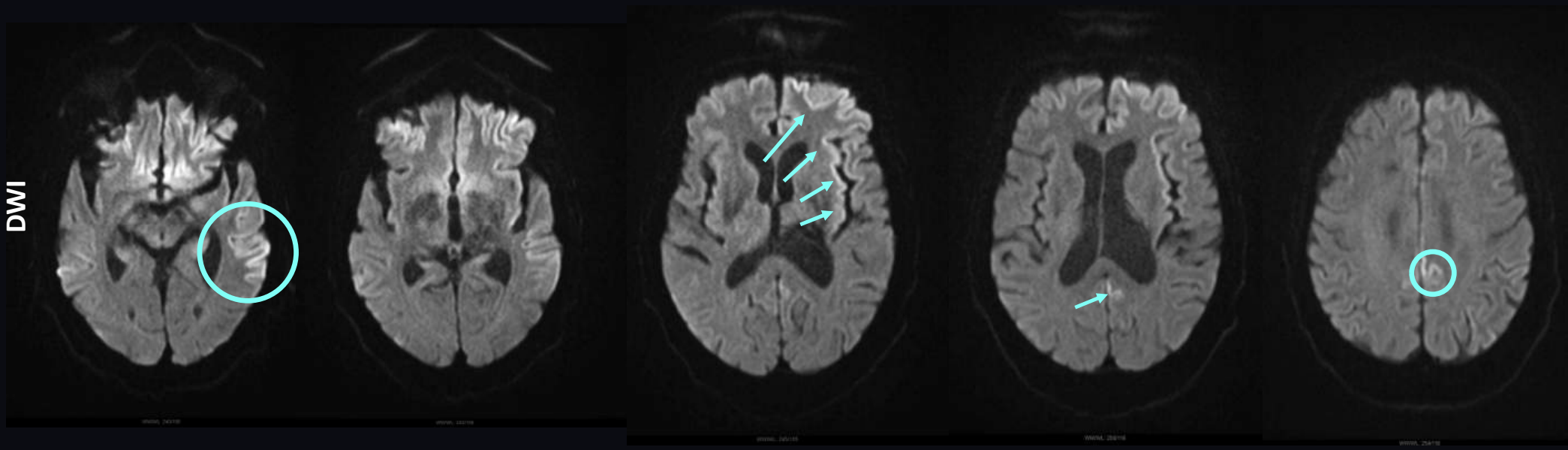
Case Presentation

- **Physical exam**: Limited due to altered mentation.
  - Patient **arousable to self only**, perseverative speech, mild dysarthria, attention and orientation impaired.
  - RUE rigid and contracted, no purposeful movement.
- **ED course**: hemodynamically stable, CT head negative, echo showed moderate PFO with R → L shunt

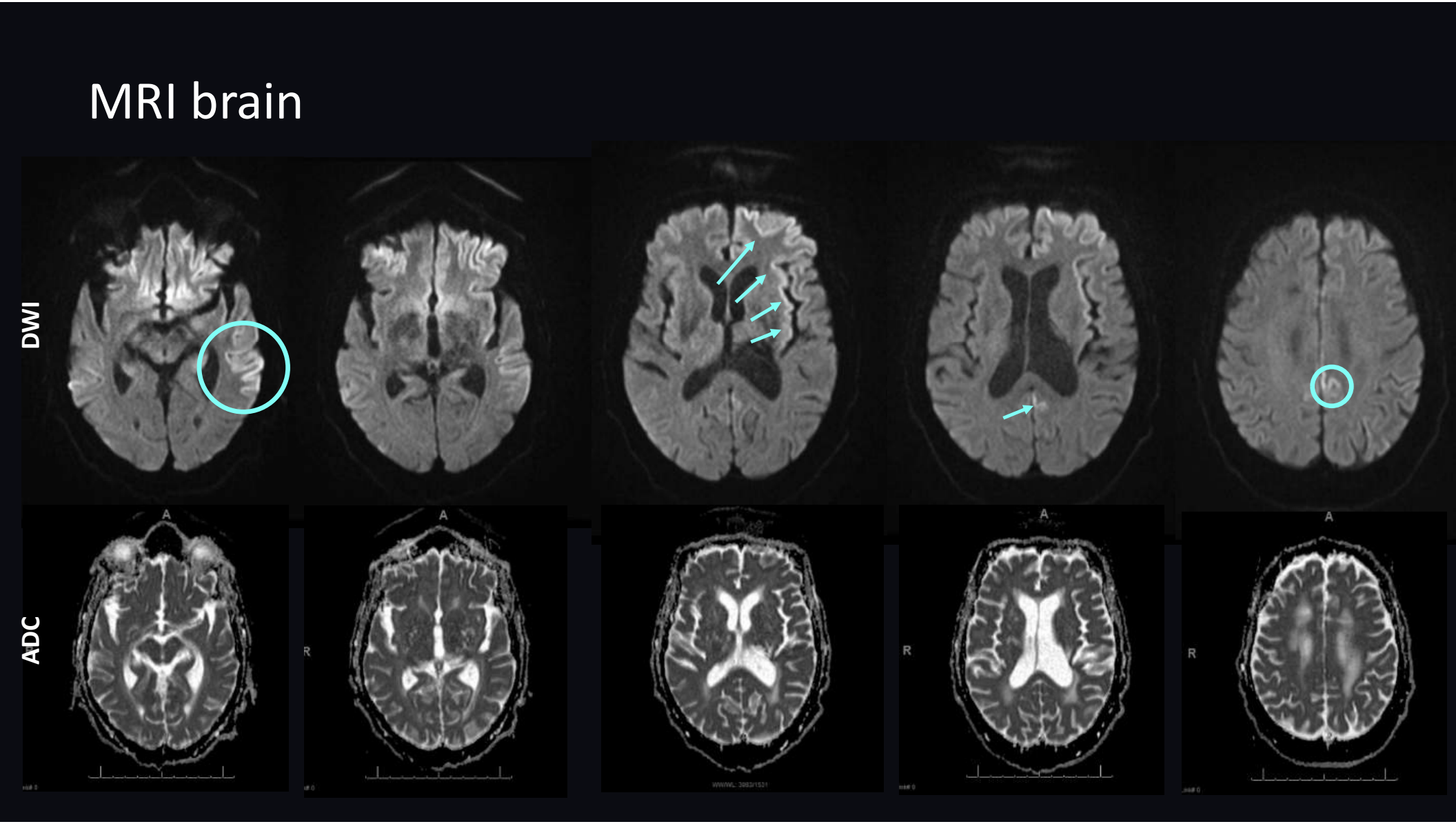
Case Presentation

- **Hospital Day 2**: acute decline
  - unresponsive, RUE contracture, left facial twitching
  - Code Stroke activated
  - CT head / CTA / CTP: no acute ischemic changes, no large vessel occlusion, no perfusion mismatch
  - MRI brain ordered for further workup

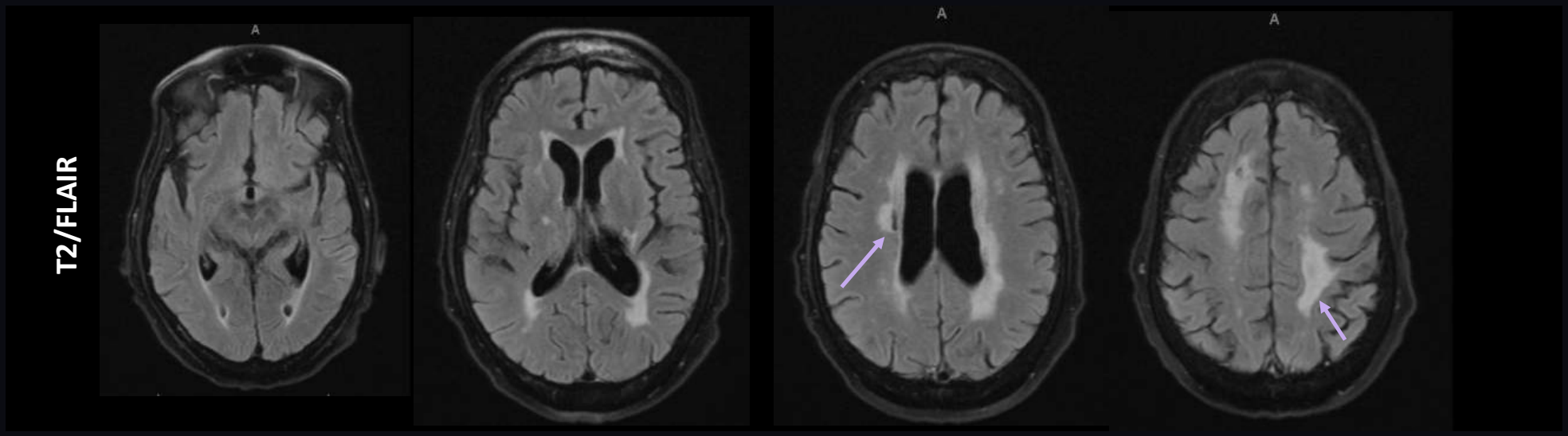
MRI brain



MRI brain

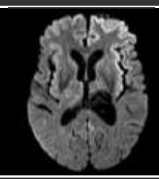
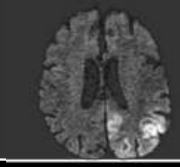
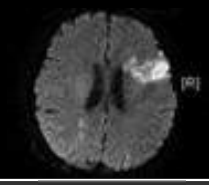
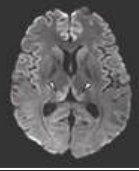
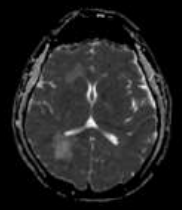


MRI brain



Imaging findings

- CT head / CTA / CTP: no acute ischemic changes, no large vessel occlusion, no perfusion mismatch
- MRI brain:
  - Multifocal **cortical restricted diffusion in the left hemisphere**
  - T2 FLAIR hyperintensity in the left temporal lobe
  - No gyral swelling or pathologic enhancement
  - Chronic findings: left thalamic encephalomalacia, white matter ischemic changes, old and new lacunar infarcts

Differential diagnosis of cortical diffusion restriction			
Diagnosis	DWI findings		Distinguishing Feature
Creutzfeldt-Jakob Disease	**Cortical ribboning**		• No swelling, no enhancement • No white matter involvement
Post-ictal changes	Cortical restriction **(can mimic CJD)**		• Resolves in 24–48 hrs • Can have gyral swelling and leptomeningeal enhancement
Multifocal embolic infarcts	DWI bright		• Follows vascular territories • Involves white matter
Metabolic/toxic encephalopathy	Variable (seen in up to 73%)		• Gyral swelling + hemorrhage • Specific anatomic patterns + clinical context
Autoimmune encephalitis	No diffusion restriction		• Mesial temporal FLAIR signal • Contrast enhancement • CSF shows pleocytosis

Additional diagnostic workup

- EEG/LTM: diffuse background slowing, no seizures or epileptiform activity
- LP: no organisms, no pleocytosis, normal glucose/protein
- **CSF RT-QuIC: positive → confirmed probable CJD**
- Patient transitioned to hospice care

Creutzfeldt-Jakob Disease

- Fatal prion disease: normal prion protein (PrP^{c}) misfolds into pathologic form (PrP^{sc}), triggering a chain reaction of further misfolding
- Causes spongiform encephalopathy and neuronal loss
- Sporadic CJD = 85-90% of cases; incidence 1-2 per million/year
- **Rapidly progressive dementia** (weeks to months)
  - myoclonus, visual/cerebellar signs
  - akinetic mutism
- Diagnosis: clinical criteria + MRI + CSF RT-QuIC
  - definitive diagnosis can only be made via biopsy/autopsy

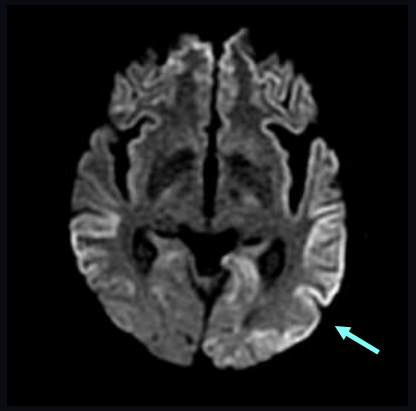
Why CJD was favored in this case

- Cortical restricted diffusion in multiple regions, non-vascular distribution
- **No gyral swelling or enhancement!**
- Persistent DWI abnormalities
- Subacute, rapidly progressive dementia preceding acute presentation
- Normal CSF cell count/protein, no evidence of seizures on EEG/LTM
- Positive RT-QuIC confirmed diagnosis

CJD Imaging Patterns by Subtype

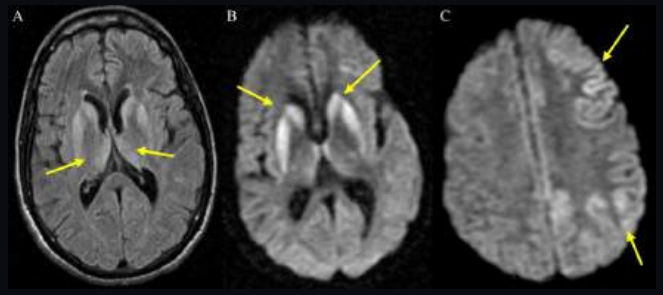
MM1 subtype

- most common (70%)
- **asymmetric cortical ribboning** ± caudate involvement



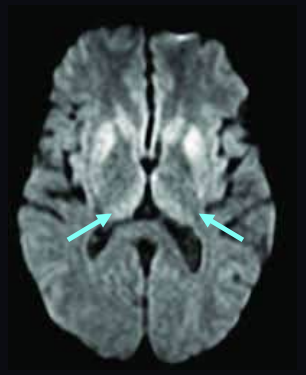
VV2 subtype

- predominantly cerebellar ataxia
- **less cortical involvement** (symmetric caudate, putamen, thalamus)



variant CJD

- "mad cow disease"
- characteristic pulvinar **"hockey stick" sign**



ACR Appropriateness Criteria

Variant 10. Adult. Rapidly progressive dementia. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRI functional (fMRI) head without IV contrast	Usually Not Appropriate	O
Amyloid PET/CT brain	Usually Not Appropriate	☢☢☢
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT brain	Usually Not Appropriate	☢☢☢
SPECT or SPECT/CT brain perfusion	Usually Not Appropriate	☢☢☢
SPECT or SPECT/CT brain striatal	Usually Not Appropriate	☢☢☢
Tau PET/CT brain	Usually Not Appropriate	☢☢☢

Management & outcome

- No disease-modifying treatment exists for CJD; most patients die within 12 months of diagnosis
- Supportive and palliative care is the standard of care
- This patient's course:
  - **RT-QuIC positive** on hospital day 20 → confirmed probable sCJD
  - Family elected comfort-focused care: code status changed to DNAR Select, discharged to hospice
  - Passed away 13 days after discharge
- Total illness duration from first cognitive symptoms to death: ~3 months

Key takeaways

- CT is normal in CJD, **MRI with DWI is essential for diagnosis**
- Always check ADC maps to confirm true restriction vs. T2 shine-through
- The presence of cortical ribboning should always be an indication for CSF RT-QuIC analysis
- Early radiologic recognition expedites diagnosis and guides family conversations

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