



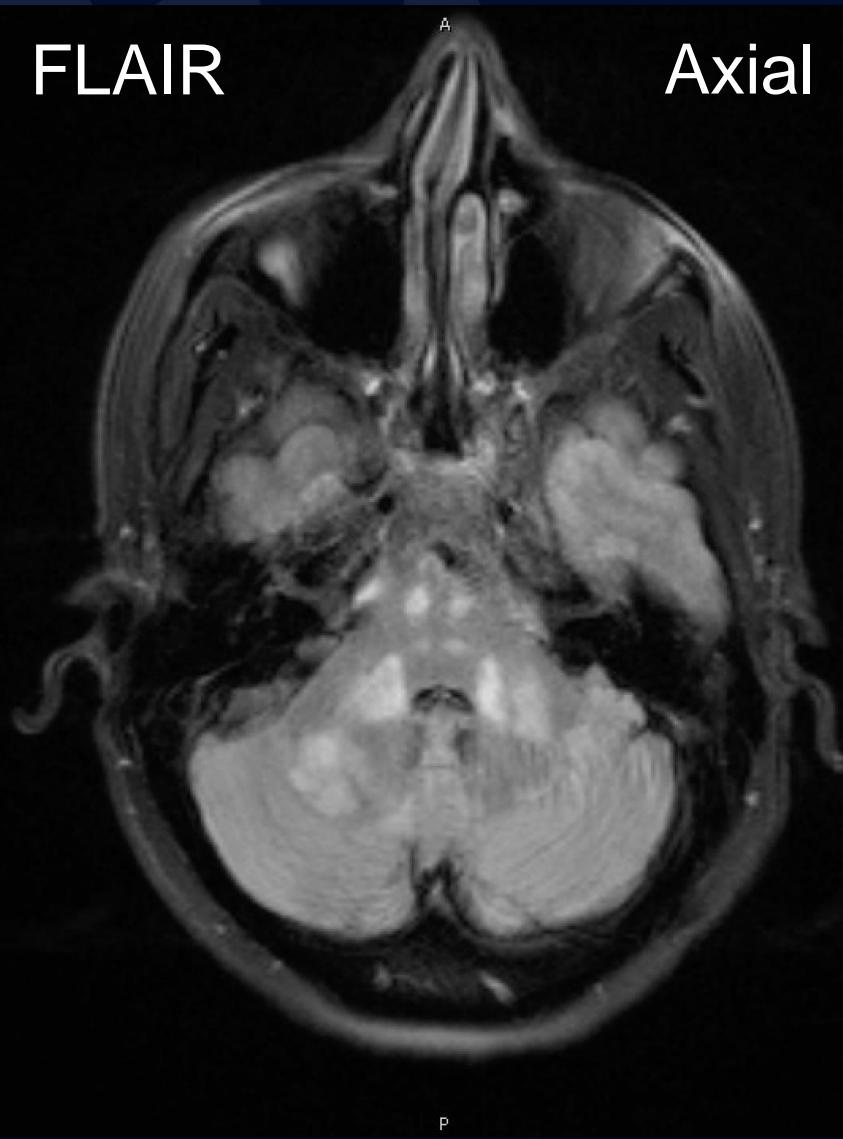
50-year-old female with  
progressive difficulty walking and  
paresthesias

Calvin Park, MS3

# MRI Brain without Contrast

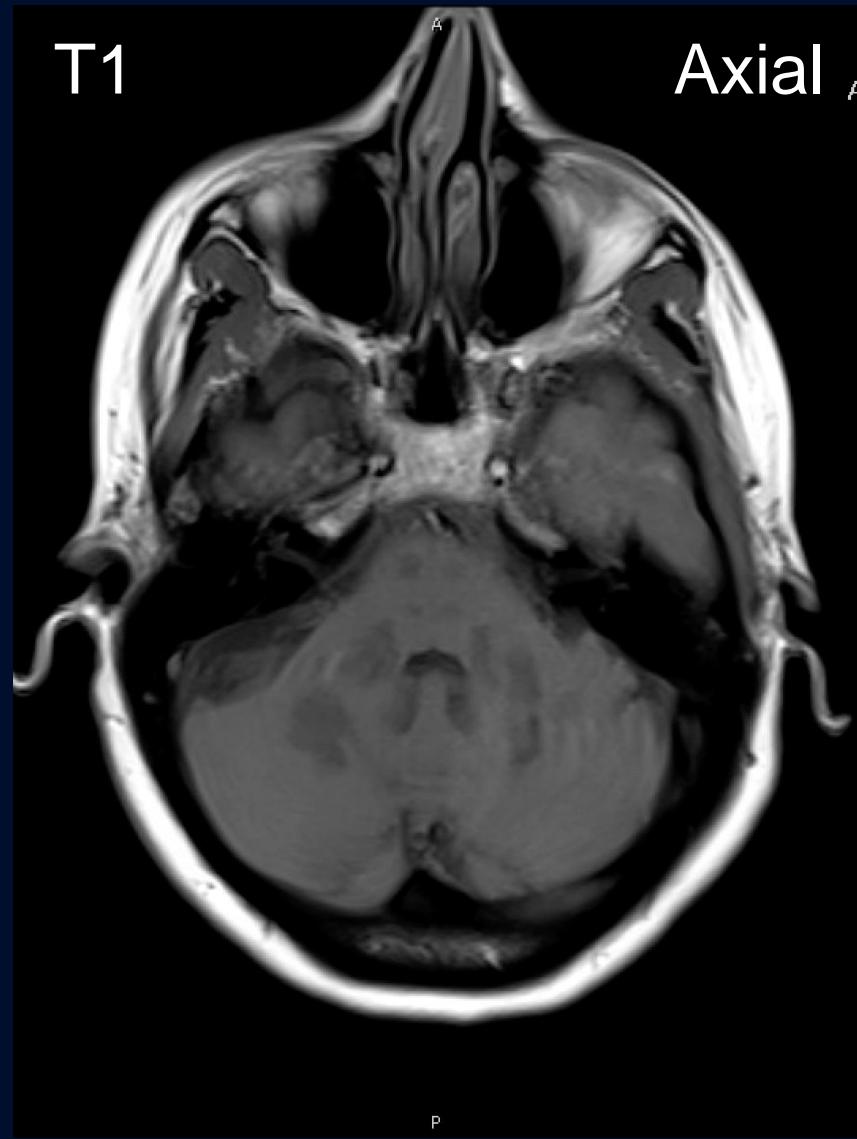
FLAIR

Axial



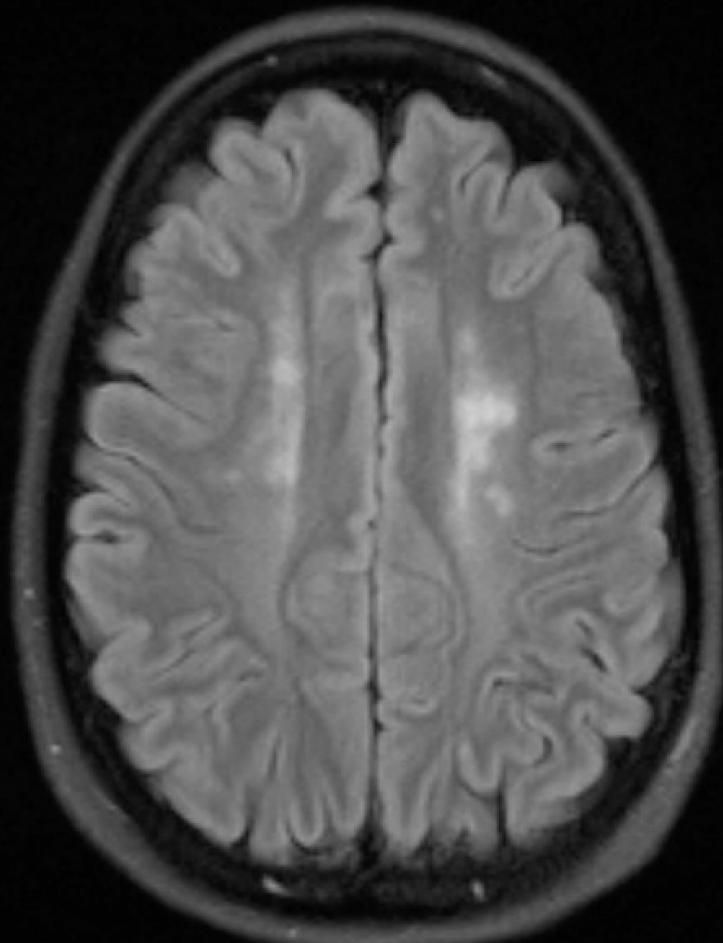
T1

Axial



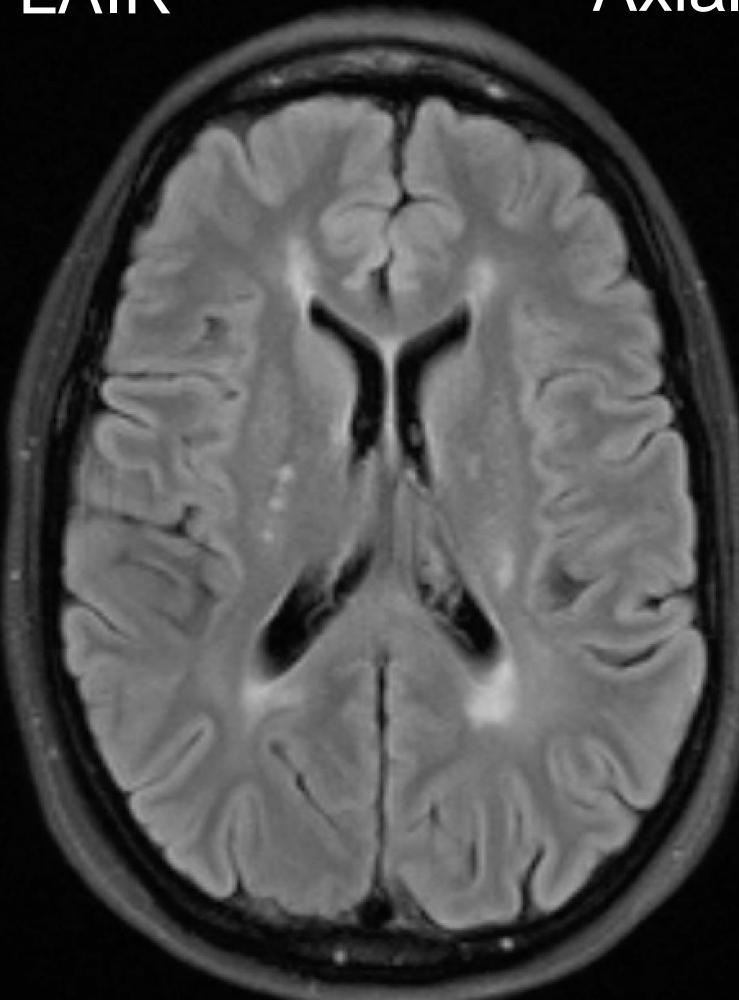
# MRI Brain without Contrast

FLAIR



Axial

FLAIR

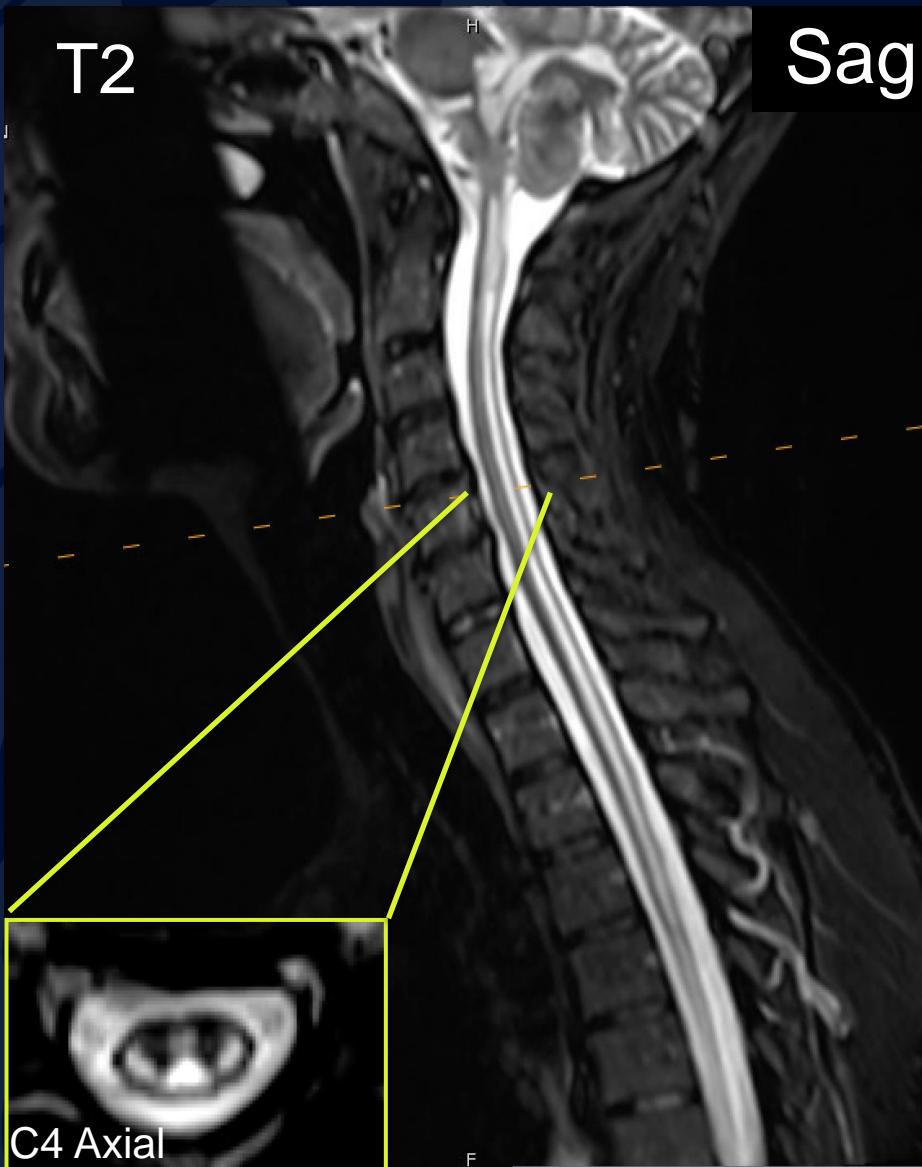


Axial

# MRI Spine without Contrast

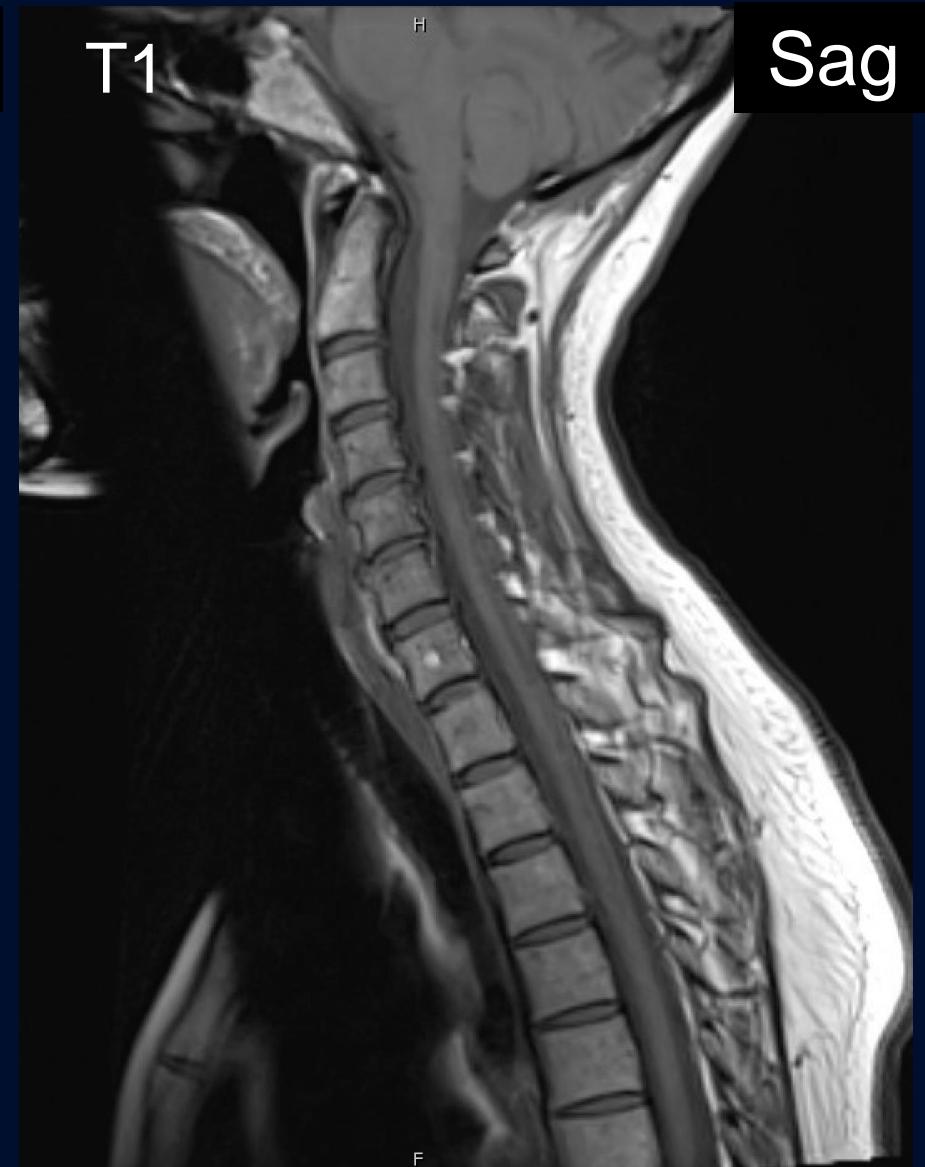
T2

Sag



T1

Sag





?

**UCONN**  
**HEALTH**  
RADIOLOGY

# Leukoencephalopathy with Brainstem and Spinal Cord Involvement and Lactate Elevation (LBSL)

# MRI Brain without Contrast

FLAIR

Axial

T1

Axial

FLAIR hyperintensities with corresponding T1 hypointensities in the:

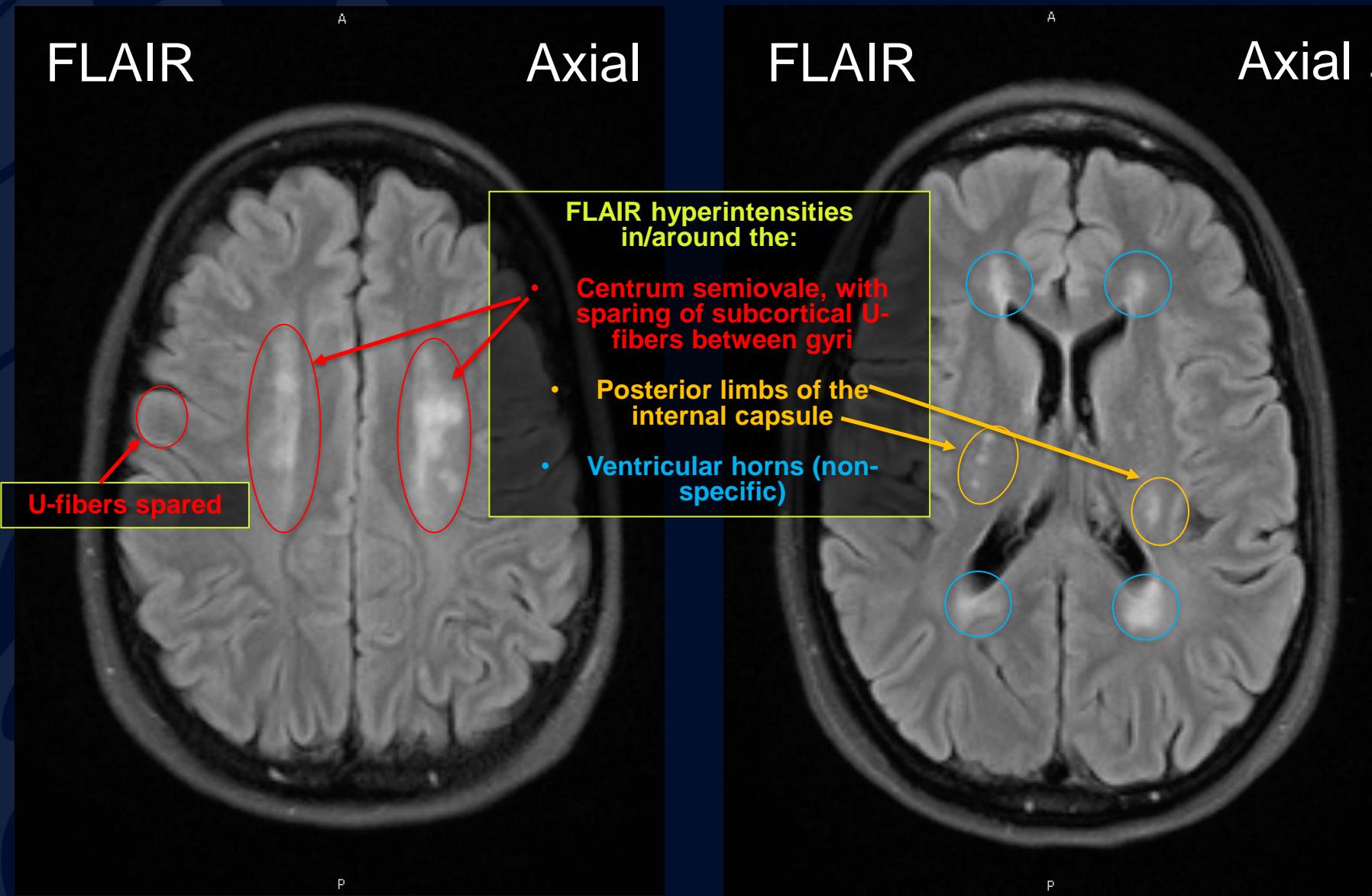
• Medullary pyramids  
• Medial lemniscus

• Middle and inferior cerebellar peduncles

Cerebellar dentate nuclei/WM

No corresponding enhancement on Gd imaging

# MRI Brain without Contrast



# MRI Spine without Contrast

T2

Sag

T1

Sag

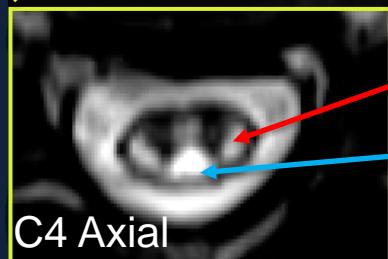
Longitudinally extensive segment of T2 hyperintensity and corresponding T1 hypointensity spanning throughout the cord down to the conus medullaris

Axial cut through cervical spine at C4 level shows bilateral T2 hyperintensity in the:

Lateral corticospinal tracts (continuation of medullary pyramids)

Dorsal columns (continue to medial lemniscus)

No corresponding lesion enhancement with Gd



# Leukoencephalopathy with Brainstem and Spinal Cord Involvement and Lactate Elevation (LBSL)

- LBSL is a rare, recently (2003) identified inherited leukodystrophy
  - Mutation in DARS2 gene encoding mitochondrial aspartyl-tRNA synthetase (mt-AspRS)
- Most commonly manifests in children as ataxia, spasticity and loss of proprioception
- Progresses with age, with later onset associated with better prognosis
- Definitive diagnosis by brain and spine MRI plus genetic testing
- Patients are treated symptomatically

# Imaging Findings

MRI Changes	LBSL ( <i>n</i> = 83) <sup>a</sup> , proportion of patients (%)
<b>Supratentorial</b>	
Cerebral WM with subcortical sparing*	92.8
Posterior part of the corpus callosum	77.1
Posterior limb of the internal capsule	81.9
<b>Infratentorial</b>	
Lateral corticospinal tracts or dorsal columns of the spinal cord	89.2
Pyramidal tract and/or medial lemniscus of the brainstem	94.0
Superior cerebellar peduncles	73.5
Inferior cerebellar peduncles	69.9
Intraparenchymal trajectories of the trigeminal nerve	63.9
Mesencephalic trigeminal tracts	47.0
Anterior spinocerebellar tracts of the medulla oblongata	32.5
Cerebellar white matter	88.0

Highlighted features were  
pertinent to this case

Muthiah et al. 2021

# References

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