UC Irvine Neuromuscular Colloquium

Case Vignette: More than Meets the Eye

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History of Present Illness

- 54 yo man developed weakness in ”left quadriceps”
- Weakness in RLE was noted in 2 weeks
- Tingling sensation and twitching in bilateral legs
- Multiple falls due to "legs giving out”
- Wheelchair depended within two months
- EMG/NCS was concerning for …

- Referred to UCI Neuromuscular Center for evaluation

- Due to worsening weakness he presented to UCIMC

ROS

Reports:

- Dizziness with change in position and nausea
- Worsening chronic tinnitus
- Tingling sensation in perianal area
- Lost 60lb in 1yr on *Weight Watchers*
- New diagnosis of gastritis 2 months prior
- Constipation

Denies:

- Urinary or bowel incontinence
- Finger and wrist weakness
- Problems with swallowing or chewing
- Double vision
- Denies new rash
- Cough or night sweats
- SOB

PMH, FM and SH

PMH

- DM2
- Psoriasis

FH

- Myasthenia gravis – distant relative
- ? Leukemia – grandfather

SH

- Occupation: HR for FICO credit score company.
- Living situation: lives in an first floor apt with his son
- Tobacco: Current Smoker, 1/2 pack PPD
- Alcohol Use: last drink 2 years ago
- Illicit Drug Use: admits daily heroin use but has not had any in 2 weeks

Pertinent Physical Examination Findings

- Fasciculations noted in bilateral lower extremity

Motor	Deltoid	Biceps	Triceps	Wrist Ext	Wrist Flx	Finger Ex	Finger Flx	APB	FDI
R	5/5	5/5	5/5	5/5	5/5	5/5	5/5	5/5	5/5
L	5/5	5/5	5/5	5/5	5/5	5/5	5/5	5/5	5/5

Motor	Hip Flx	Hip Ext	Knee Flx	Knee Ext	Dorsiflexors	Plantar Flx
R	2/5	3/5	3/5	2/5	2/5	3/5
L	2/5	3/5	3/5	3/5	3/5	3/5

- Gait: unable to ambulate
- Sensory: diminished to pinprick in upto L1
- DTRs (R/L): biceps (2+/2+), triceps (2+/2+), BR (2+/2+), patellar (2+/2+), Achilles (0/0), mute toes b/l
- Decreased rectal tone

SNC

Bilateral sural, left median, ulnar, and radial sensory potentials were normal

SNC

Nerve / Sites	Rec. Site	Onset Lat ms	Peak Lat ms	Amp µV	Distance mm	Velocity m/s	Temp. °C
L Median - Digit II (Antidromic)							
Wrist	Dig II	2.55	3.23	58.7	140	55	32.7
L Ulnar - Digit V (Antidromic)							
Wrist	Dig V	2.19	2.81	53.3	110	50	32.7
L Radial - Anatomical snuff box (Forearm)							
Forearm	Wrist	1.77	2.24	34.1	100	56	30.8
L Sural - Ankle (Calf)							
Calf	Ankle	3.49	4.48	11.6	140	40	30.3
R Sural - Ankle (Calf)							
Calf	Ankle	3.80	4.79	9.5	140	37	30.4

MNC

Median

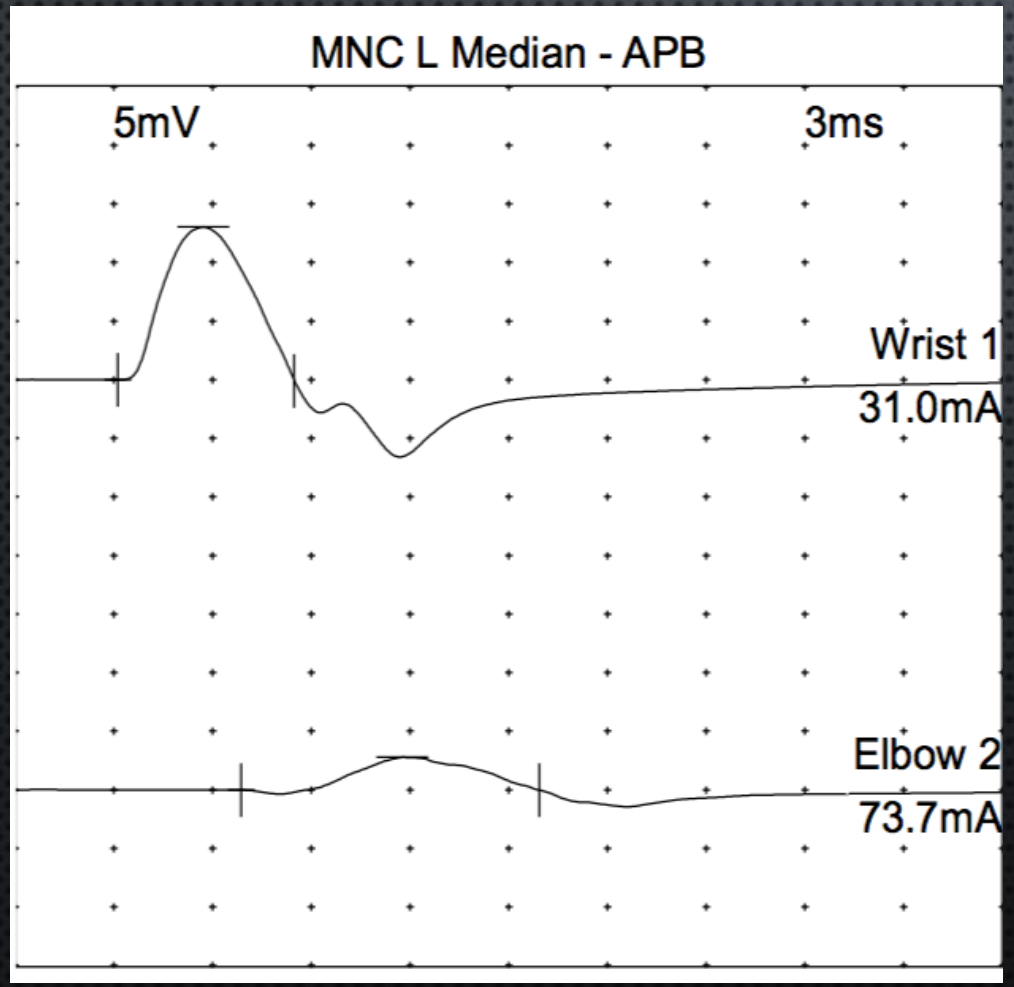
- normal distal latency and amplitude
- mildly slow conduction velocity
- F-wave latency with partial conduction block

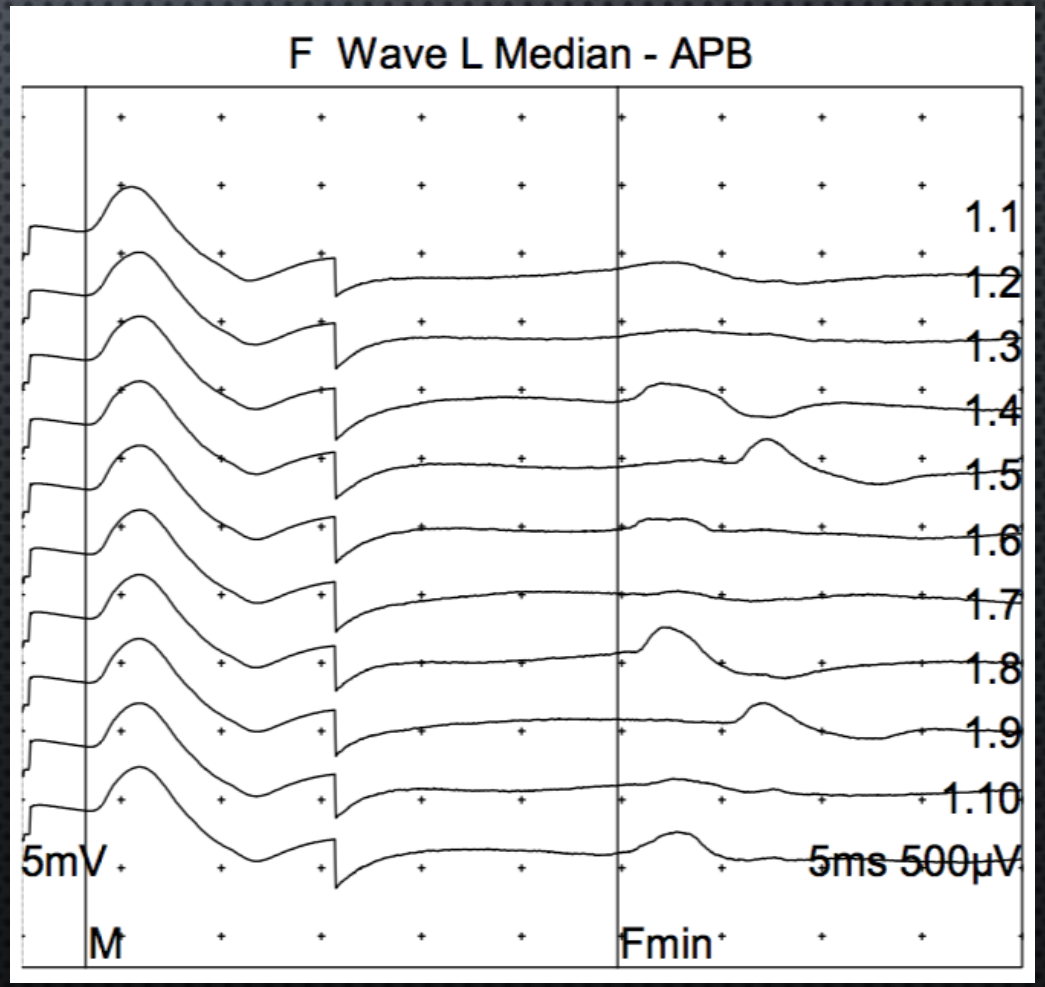
Left peroneal

- normal distal latency and amplitude
- mildly slow conduction velocity
- F-wave latency not in demyelinating range

Left ulnar and tibial

- Normal distal latency and amplitude
- Normal conduction velocity
- Normal F-wave latency





NCS

Fibrillation and fasciculation potentials were seen throughout the left leg

Needle EMG										
	Spontaneous				Volitional MUAPs					
Muscle	IA	Fibs/PSW	Fasc	Other	Dur.	Amp	Poly	Recruit	Rate	Comment
L. First dorsal interosseous	Normal	None	None	.	Normal	Normal	None	Normal	Normal	.
L. Vastus lateralis	Normal	3+	Rare	.	Long	High	Many	Mark Decr	Rapid	.
L. Tibialis anterior	Normal	3+	Few	.	Long	High	Many	Mod Decr	Rapid	.
L. Gastrocnemius (Medial head)	Normal	3+	Rare	.	Long	High	Many	Mod Decr	Rapid	.
L. Thoracic paraspinals	Normal	None	None							

Studies

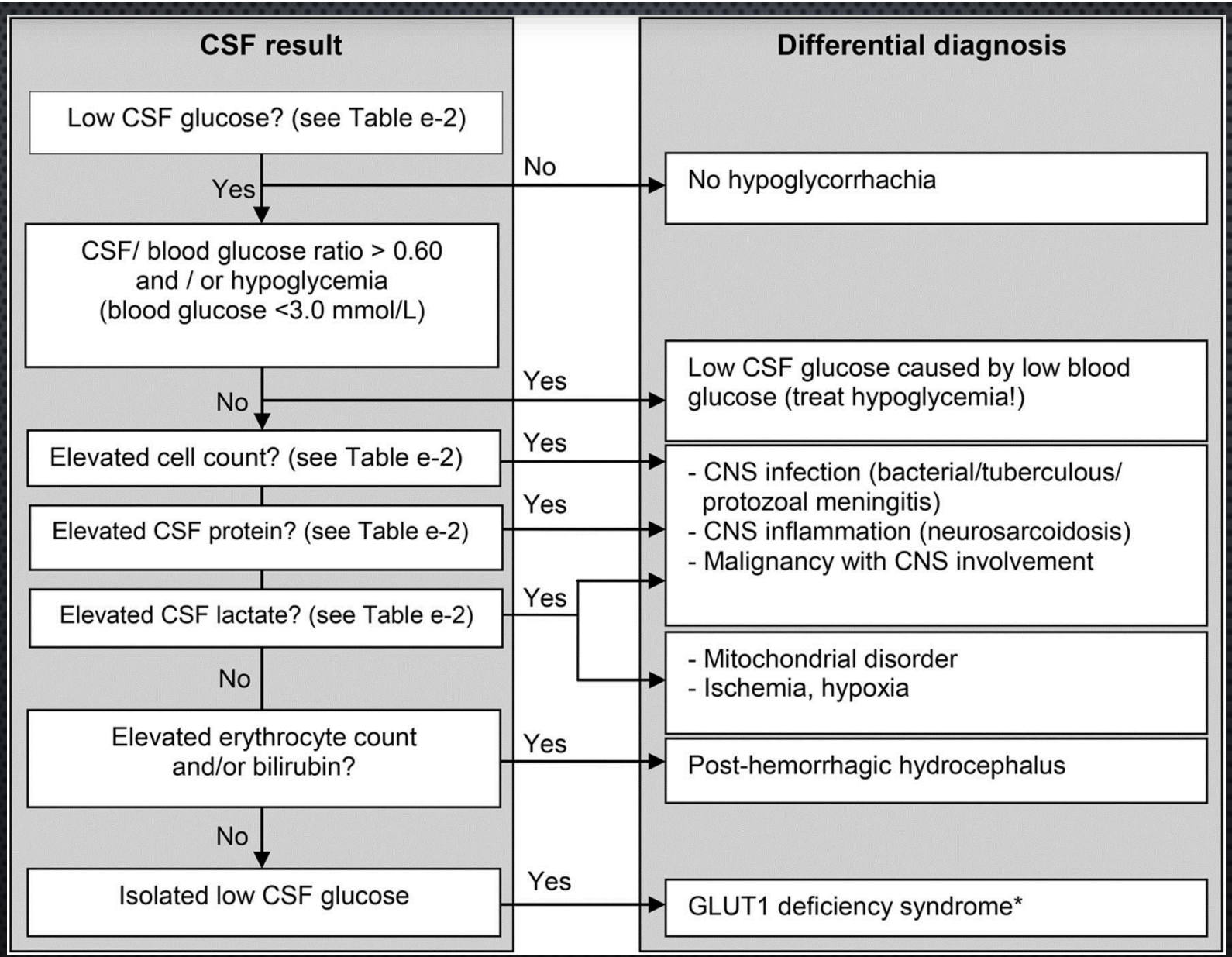
Serum	CSF
ANA – negative	Glucose – 24
A1c – 5.8%	Protein – 423
CPK - 85	Nucleated cell – 125
CRP – 0.01	Lymphocytes- 86
ESR - 18	Macrophages- 86
Folic acid 14.2	Other cells- 2
Free T4- 0.86	RBC- 0
TSH – 2.473	Paraneoplastic - negative
B12 - 211	Lyme - negative
ACE-5	VZV- negative
HIV -negative	WNV- negative
Hep panel - negative	TB - negative
T pali – negative	Crypto - negative
Paraneoplastic – negative	VDRL - negative

Differential Diagnosis

Differential Diagnosis?

What would you do next?

Differential Diagnosis of Low Glucose in CSF



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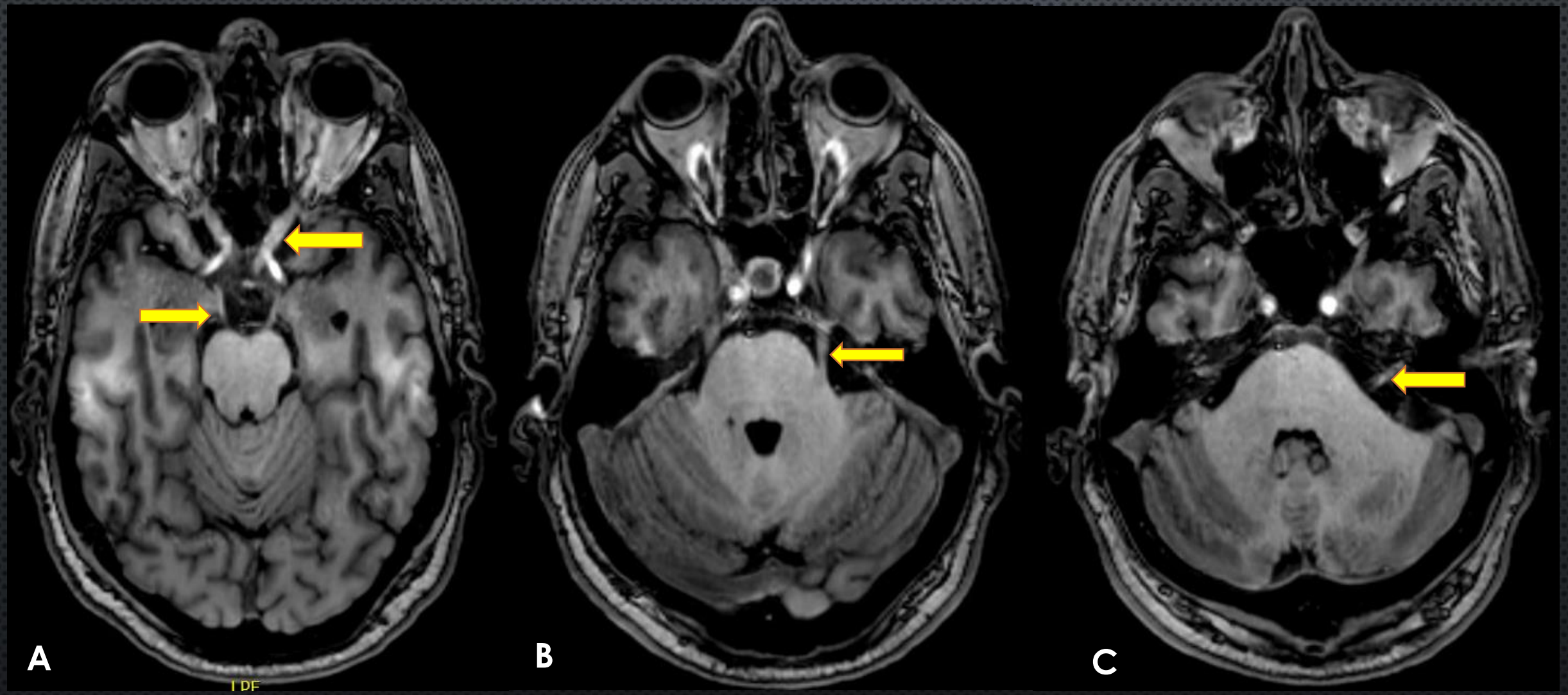
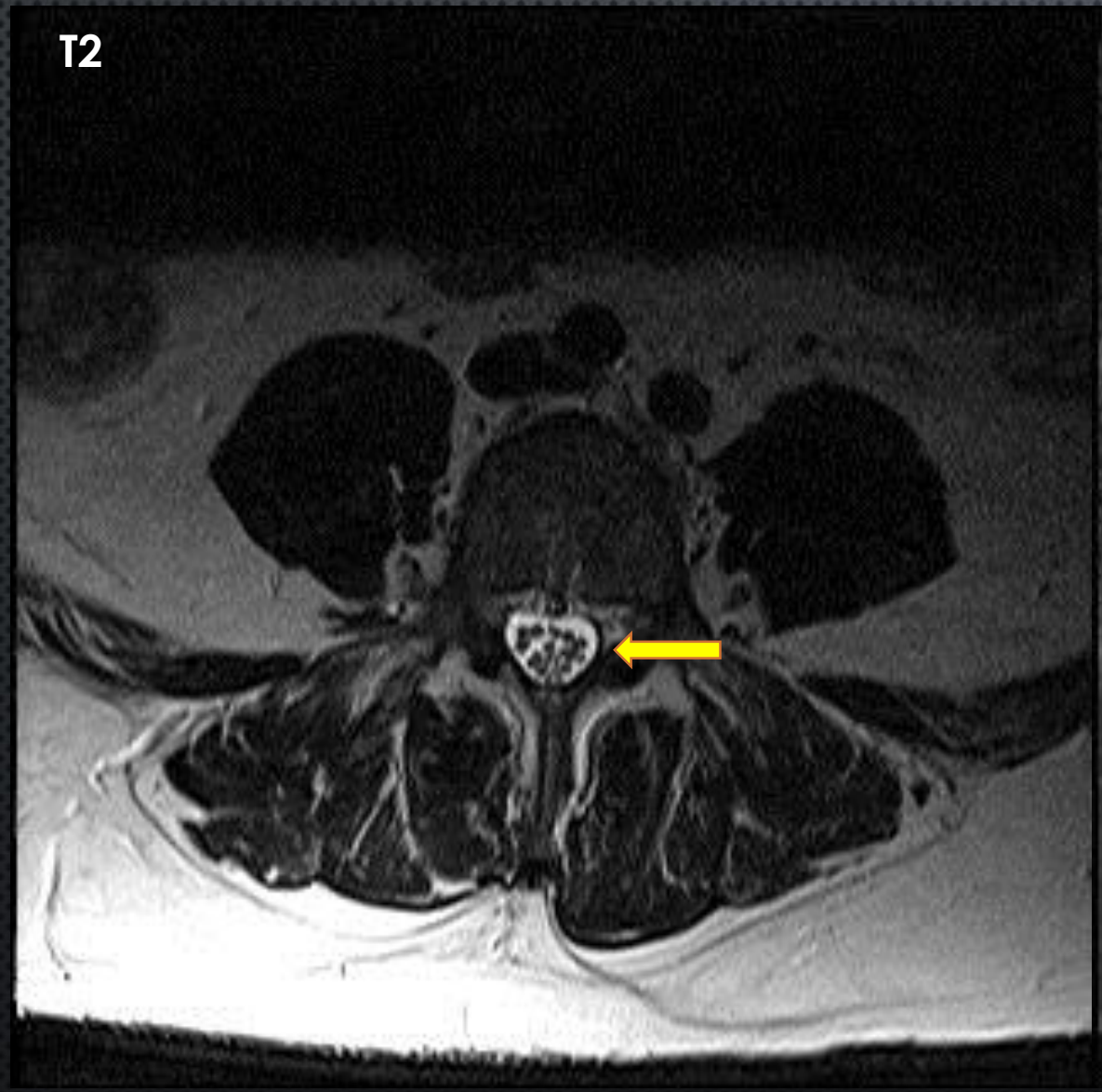
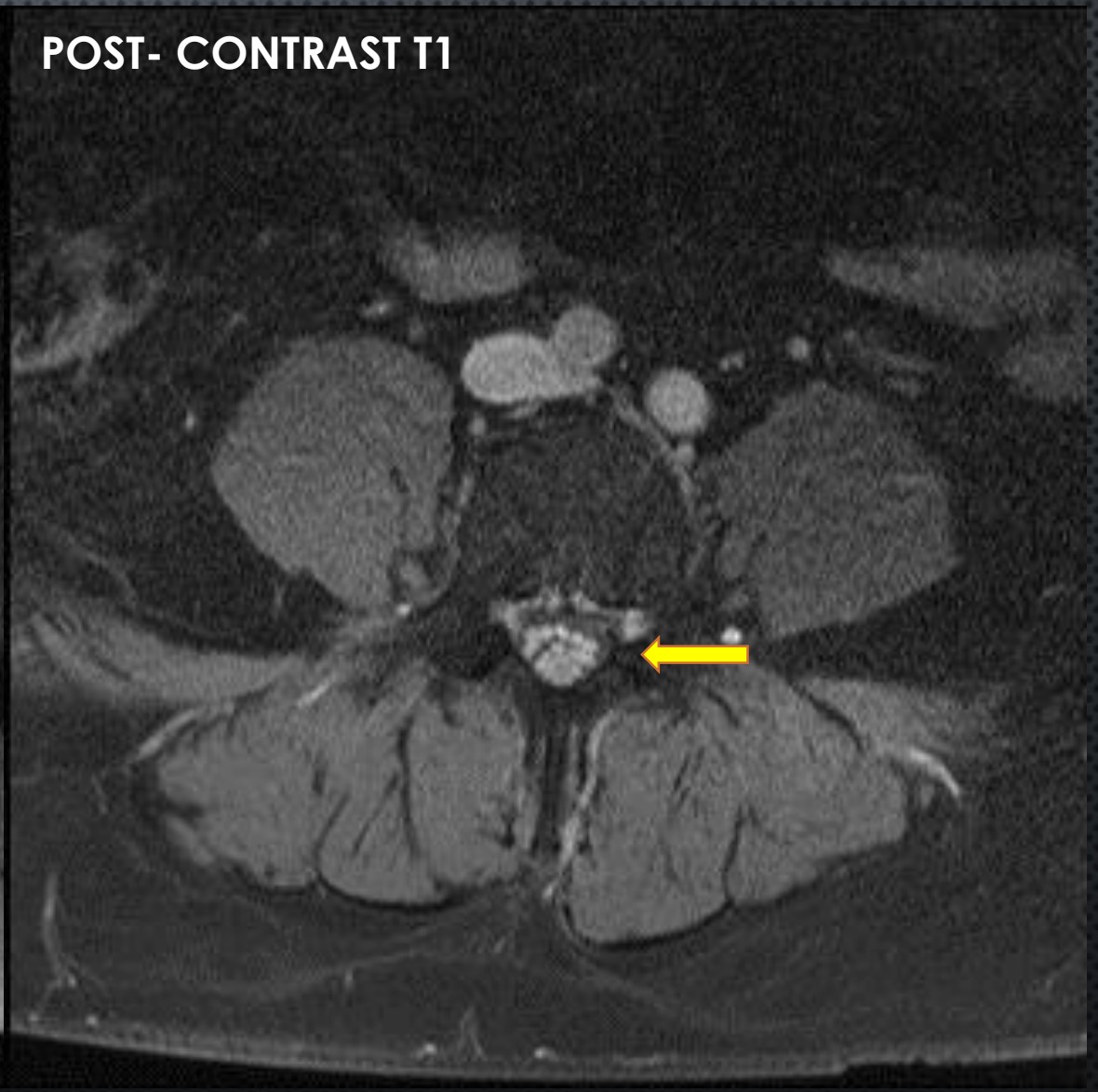
MRI BRAIN W/WO CONTRAST



MRI L-SPINE W/WO CONTRAST

Diffuse thickening/enhancement of the cauda equina nerve roots





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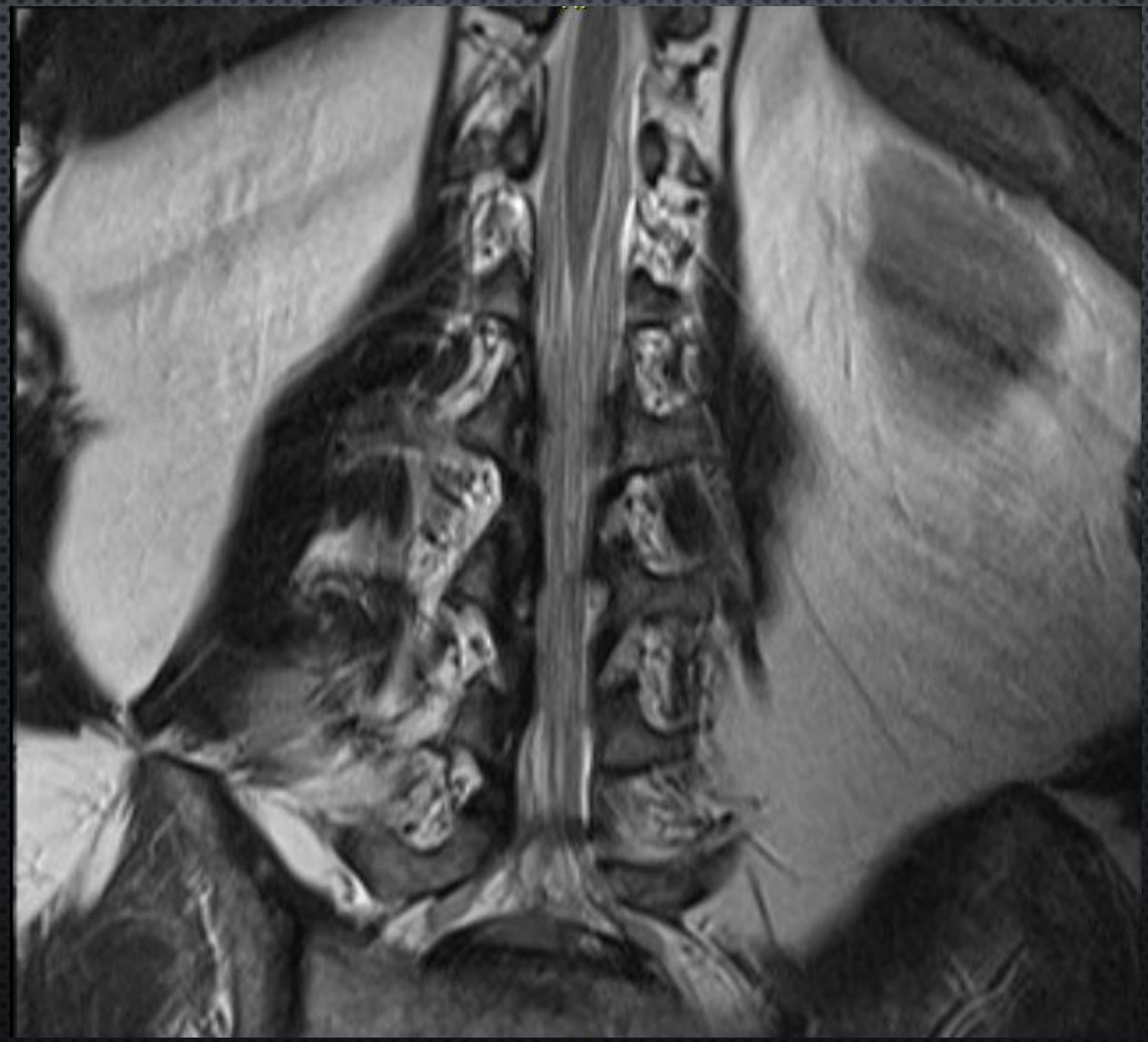

Oncological Studies

- **CSF FLOW CYTOMETRY:**
  - Lymphocytosis with a small Kappa-monotypic B-cell population CSF involvement by a mature CD5-/CD10 - B-cell neoplasm
- **CYTOLOGY-** Atypical cells
- **BONE MARROW BIOPSY:**
  - Normocellular marrow with active trilineage hematopoiesis
  - Rare small reactive-appearing lymphoid aggregate
  - No evidence of lymphoma
- **GI BIOPSY:**
  - No evidence of lymphoma within the above biopsies

Neuropathy Associated with Lymphoma

- Occurs in 5% of patients with lymphoma
- Common types:
  - Neurolymphomatosis
  - Paraneoplastic neuropathy
  - Sensory ganglionopathy
  - Vasculitic neuropathy
  - Multiple mononeuropathy
  - Polyneuropath

Neurolymphomatosis

- NL characterized by infiltration of malignant lymphocytes into the peripheral nervous system
- Subacute to chronic in onset

- PRESENTATION PATTERNS:
  - painful involvement of nerves or roots
  - cranial neuropathy with or without pain
  - painless involvement of peripheral nerves
  - painful or painless involvement of a single peripheral nerve

- Nerve biopsy – gold standard

- MRI FINDINGS:
  - nerve enlargement
  - isointensity to muscle on T1
  - hyperintensity on T2 or STIR sequences
  - significant focal or diffuse gadolinium enhancement

Table 2 Neuropathic features of neuropathy associated with lymphoma

Patients	Progression*	Type of neuropathy	Cranial nerve involvement	Muscle weakness	Sensory disturbance		Spontaneous pain	Autonomic failure	EFNS/PNS CIDP electropiagnostic criteria**
					Superficial sensation	Deep sensation			
Neurolymphomatosis***									
1	Subacute	MM	V	3+	2+	2+	3+	–	Possible
2	Subacute	MM	VI	2+	1+	3+	3+	–	
3	Chronic	MM	III VI VII	3+	2+	0	1+	Adie pupil	Possible
4	Subacute	MM	V VII	3+	2+	2+	–	–	Definite
5	Chronic	MM	–	2+	2+	3+	–	–	Possible
6	Chronic	MM	–	3+	1+	0	3+	–	
7	Chronic	MM	XII	3+	3+	2+	1+	–	Possible
8	Subacute	PN	–	1+	2+	3+	–	–	Definite
9	Chronic	PN	–	3+	3+	3+	1+	–	Definite
10	Chronic	MM	–	3+	3+	3+	2+	–	Definite
11	Chronic	MM	–	3+	1+	1+	1+	–	Probable
12	Chronic	MM	V	3+	2+	0	1+	–	
13	Subacute	MM	VII	3+	2+	3+	3+	–	
14	Chronic	MM	–	3+	3+	3+	2+	–	
15	Chronic	MM	VII	3+	2+	1+	3+	–	Definite

EMG Features in Neurolymphomatosis

- Reduced compound muscle and sensory nerve action potentials
- Focal demyelination proximal to the nerve trunk
- Subsequent axonal degeneration in the portion distal to the demyelination site

- Electrodiagnostic studies have the potential to detect aberrations in peripheral nerves owing to macroscopic derangements as well as anomalies occurring at the molecular level that imaging may miss. Therefore, electrodiagnostic studies should be considered for patients suspected to have NL.

BRAIN
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