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Mr. SN 43 yo man

- Presented to ED with 3 M history of left hand paresthesia, weakness and wasting
- Awoke with paresthesia L D4/5
- Progressive numbness L D1-3
- Burning pain left hand, worse at night
- Occasional shooting pain from proximal arm to hand
- Weakness left hand, difficulty grasping objects

HOPC

- Associated symptoms:
 - 15kg LOW
 - Occasional night sweats
 - Fatigue/lethargy
- Self employed, property maintenance
- Smoker 20 PY; Alcohol 4 SD/W

Referred to Neurology rapid access clinic from ED

Examination

- General examination normal
- Neuro exam normal except LUL
- Tone, reflexes normal
- Weakness median innervated muscles below and including PT
- Reduced LT/pp sensation in entire median N distribution (D₁₋₃, lateral D₄, thenar eminence)











? Differential diagnoses

? Further investigation

PMH

- Referred to neurologist in Dec 2008 for investigation of left thumb numbness and dysesthesia
- Symptoms developed after fall with traumatic pneumothorax/GA/thoracic surgery Feb 2008
- Reported numbness left dorsal thumb/index finger/distal forearm; medial aspect right forearm
- Examination normal except reduced pp in left radial and right MABC nerve territories

PMH

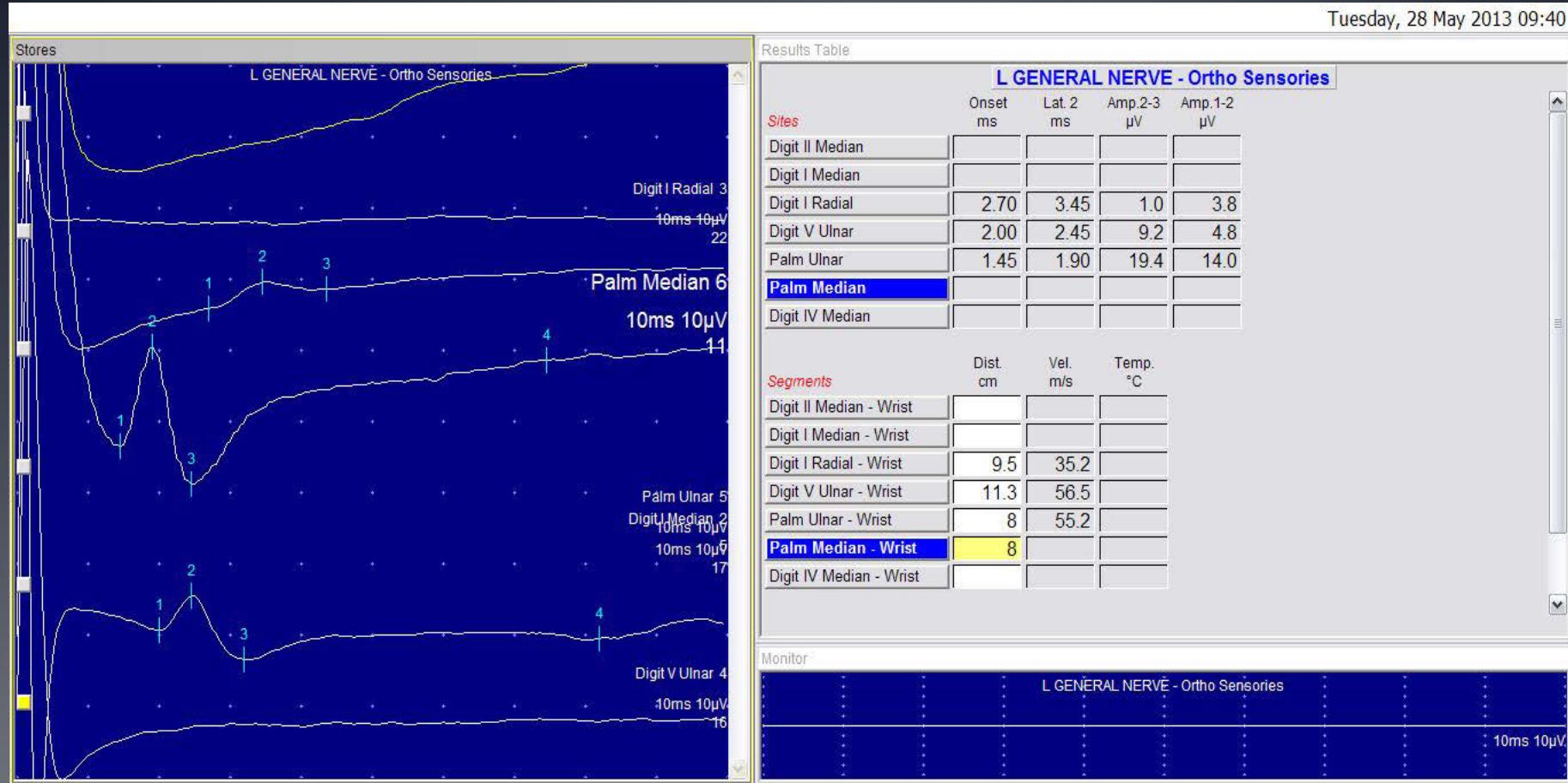
- NCS – absent left radial SNAP, MABC not tested
- MRI cervical spine/brachial plexus – normal
- Diagnosis – left radial sensory neuropathy
- Symptoms mild and stable until current presentation 5 yrs later.



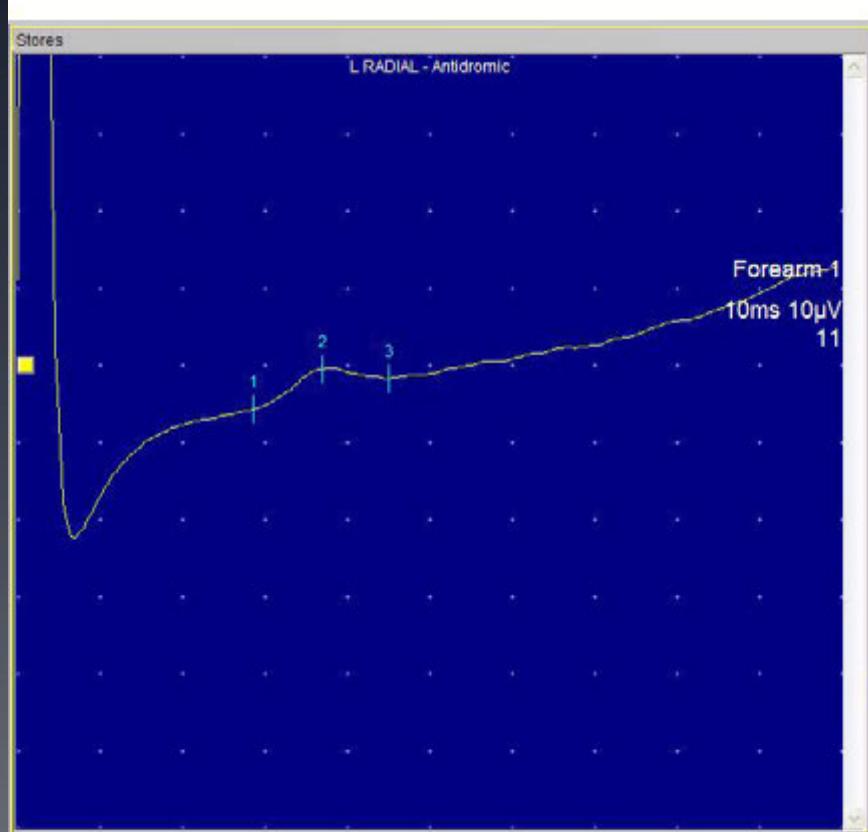
? Differential diagnoses

? Further investigation

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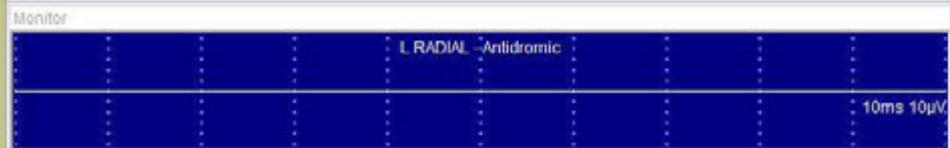
Results Table

L RADIAL - Antidromic

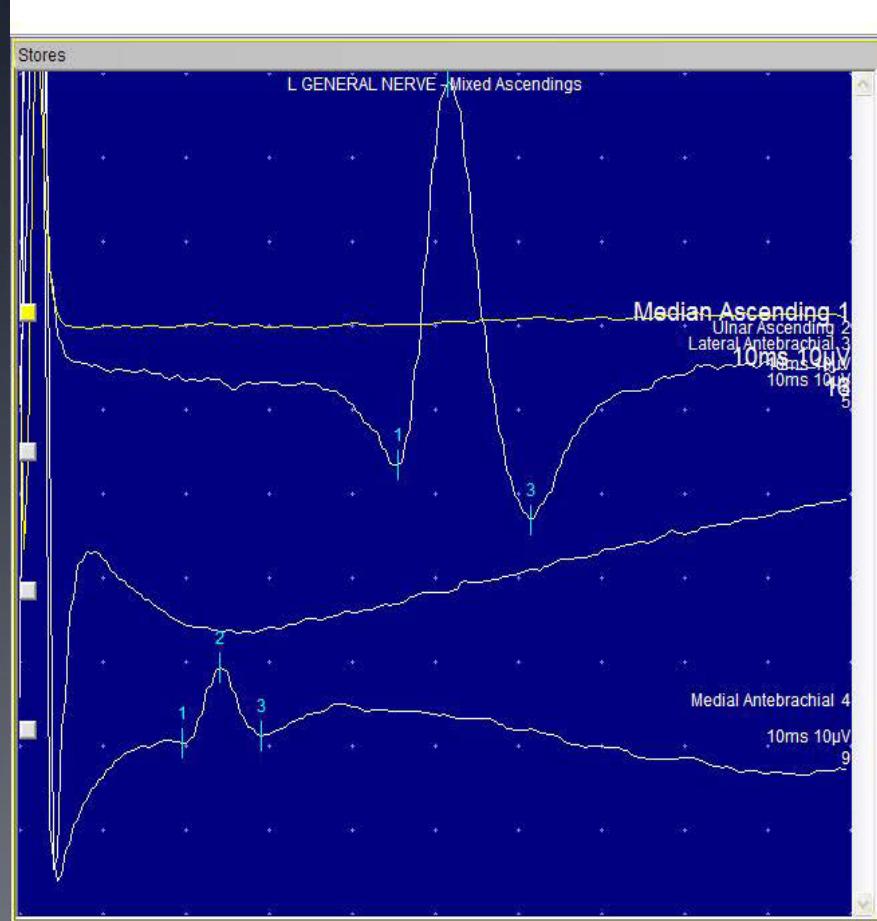
Sites	Onset ms	Lat. 2 ms	Amp 2-3 μ V	Amp 1-2 μ V
Forearm	2.85	3.70	1.2	5.2

Segments

Segments	Dist. cm	Vel. m/s	Temp. °C
Forearm - Snuff box	12	42.1	



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Results Table

L GENERAL NERVE - Mixed Ascendings

Sites	Onset ms	Lat. 2 ms	Amp.2-3 µV	Amp.1-2 µV
Median Ascending				
Ulnar Ascending	4.55	5.15	52.2	45.7
Lateral Antebrachial				
Medial Antebrachial	1.95	2.40	8.2	9.1

Segments

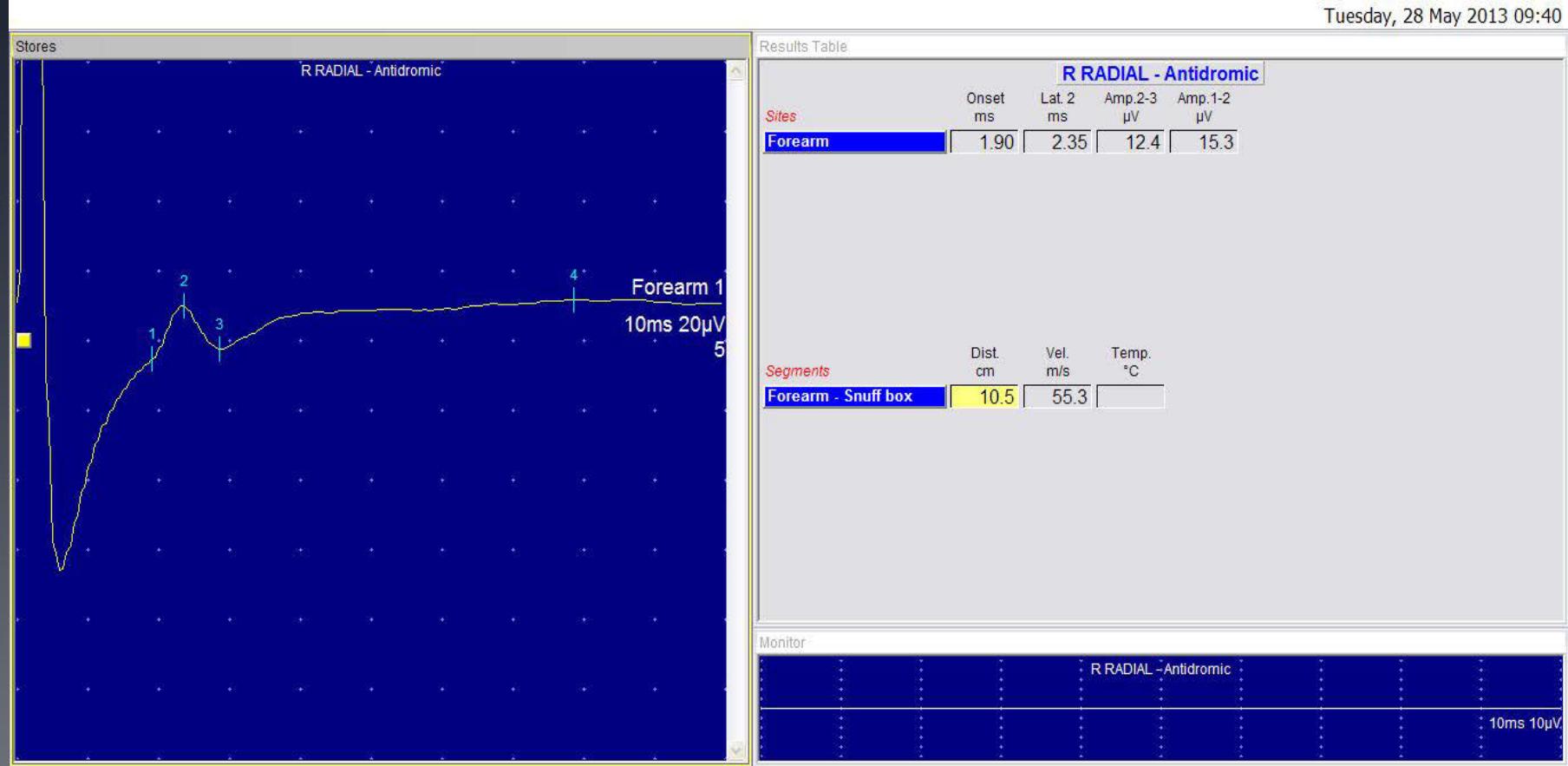
Segments	Dist. cm	Vel. m/s	Temp. °C
Median Ascendi - Elbow			
Ulnar Ascending - Elbow	25.5	56.0	
Lateral Antebrac - Forearm			
Medial Antebrac - Forearm	12	61.5	

Monitor

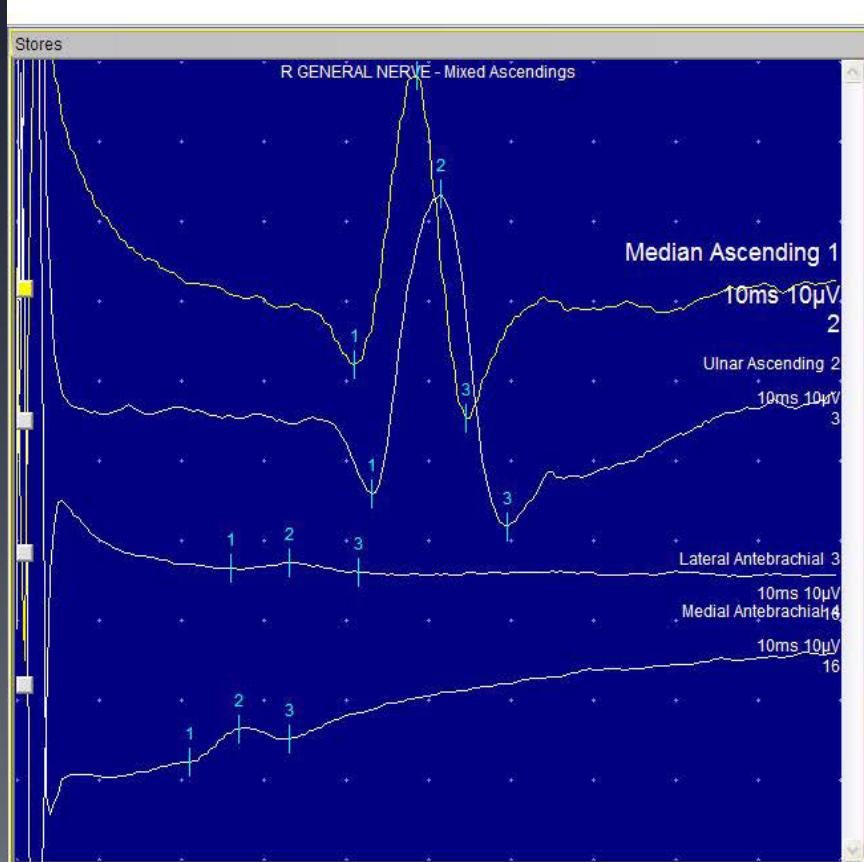
L GENERAL NERVE - Mixed Ascendings

10ms 10µV

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Tuesday, 28 May 2013 09:40



Results Table

R GENERAL NERVE - Mixed Ascendings

Sites

	Onset ms	Lat. 2 ms	Amp. 2-3 µV	Amp. 1-2 µV
Median Ascending	4.10	4.85	43.0	36.3
Ulnar Ascending	4.30	5.15	41.9	37.7
Lateral Antebrachial	2.60	3.30	1.2	0.67
Medial Antebrachial	2.10	2.70	1.2	4.1

Segments

	Dist. cm	Vel. m/s	Temp. °C
Median Ascendi - Elbow	24.5	59.8	
Ulnar Ascending - Elbow	25	58.1	
Lateral Antebrac - Forearm	9.5	36.5	
Medial Antebrac - Forearm	10	47.6	

Monitor

R GENERAL NERVE - Mixed Ascendings

10ms 10µV

Motor NCS

Nerve / Sites	Rec. Site	Lat. ms	Amp.1-2 mV	Dist cm	Vel m/s
L MEDIAN - APB					
Wrist	APB	Absent			
R MEDIAN - APB					
Wrist	APB	3.30	6.0		
Elbow		8.10	5.5	26.5	55.2
L MEDIAN - II Lumb					
MEDIAN	II Lumb	Absent			
ULNAR	4th DI	2.45	5.5		
R ULNAR - ADM 2 point					
Wrist	ADM	2.20	10.0		
L ULNAR - ADM 3 point					
Wrist	ADM	2.25	10.4		
Below Elbow		6.35	9.5	22	53.7
Above Elbow		8.70	8.9	12	51.1
L ULNAR - FDI					
Wrist	FDI	2.95	11.2		

F Wave

Nerve	Fmin ms
L ULNAR	28.35
R MEDIAN	28.95
R ULNAR	28.90

Sensory NCS

Nerve / Sites	Onset ms	Lat. 2 ms	Amp.2-3 µV	Amp.1-2 µV	Dist. cm	Vel. m/s
L - Ortho Sensories						
Digit II Median	Absent					
Digit I Median	Absent					
Digit I Radial	2.70	3.45	1.0	3.8	9.5	35.2
Digit V Ulnar	2.00	2.45	9.2	4.8	11.3	56.5
Palm Ulnar	1.45	1.90	19.4	14.0	8	55.2
Palm Median	Absent					

Nerve / Sites	Onset ms	Lat. 2 ms	Amp.2-3 µV	Amp.1-2 µV	Dist. cm	Vel. m/s	
R - Ortho Sensories							
Digit II Median	2.30	2.80	10.2	7.5	12.7	55.2	
Digit I Radial	1.80	2.35	6.9	4.0	9.5	52.8	
Digit V Ulnar	1.95	2.45	8.3	4.6	10.8	55.4	
L - Mixed Ascendings							
Median Ascending	Absent						
Ulnar Ascending	4.55	5.15	52.2	45.7	25.5	56.0	
Lateral Antebrachial	Absent						
Medial Antebrachial	1.95	2.40	8.2	9.1	12	61.5	
R - Mixed Ascendings							
Median Ascending	4.10	4.85	43.0	36.3	24.5	59.8	
Ulnar Ascending	4.30	5.15	41.9	37.7	25	58.1	
Lateral Antebrachial	2.60	3.30	1.2	0.67	9.5	36.5	
Medial Antebrachial	2.10	2.70	1.2	4.1	10	47.6	
L RADIAL - Antidromic							
Forearm	2.85	3.70	1.2	5.2	12	42.1	
R RADIAL - Antidromic							
Forearm	1.90	2.35	12.4	15.3	10.5	55.3	

EMG Summary Table	Spontaneous					MUAP			Recruitment
	IA	Fib	PSW	Fasc	H.F.	Amp	Dur.	PPP	Pattern
L. DELTOID	N	None	None	None	None	N	N	N	N
L. BICEPS	N	None	None	None	None	N	N	N	N
L. BRACHIORADIALIS	N	None	None	None	None	N	N	N	N
L. TRICEPS	N	None	None	None	None	N	1+	N	1-
L. EXT DIG COMM	N	None	None	None	None	N	2+	2+	1-
L. FIRST D INTEROSS	N	None	None	None	None	N	1+	1+	1-
L. FLEX POLL LONG	N	4+	3+	None	None	N	N	N	No Activity
L. PRON TERES	N	4+	3+	None	None	N	N	N	No Activity
L. FLEX CARPI RAD	N	3+	3+	None	None	N	2+	2+	2-
L. ABD POLL BREVIS	N	3+	3+	None	None	N	N	N	Discrete

Motor NCS

Nerve / Sites	Rec. Site	Lat. ms	Amp.1-2 mV	Dist cm	Vel m/s
R COMM PERONEAL - EDB					
Ankle	EDB	4.55	4.9		
FibHead		11.45	5.0	31.4	45.5
Knee		13.15	5.0	7.8	45.9
L COMM PERONEAL - EDB					
Ankle	EDB	4.45	7.9		
FibHead		11.35	7.4	32.4	47.0
Knee		13.65	7.8	9.4	40.9
R TIBIAL MALLEOLUS - AH					
Ankle	AH	3.55	10.8		
Knee		12.25	8.8	39.3	45.2
L TIBIAL MALLEOLUS - AH					
Ankle	AH	3.70	10.7		
Knee		12.75	9.4	42	46.4

F Wave

Nerve	Fmin ms
R COMM PERONEAL	51.25
R TIBIAL MALLEOLUS	50.10
L TIBIAL MALLEOLUS	53.10
L COMM PERONEAL	49.40

Sensory NCS

Nerve / Sites	Onset ms	Lat. 2 ms	Amp.2-3 µV	Amp.1-2 µV	Dist. cm	Vel. m/s
R - Leg Sensories						
Sural	2.75	3.50	13.4	16.3	12.7	46.2
Superficial Peroneal	2.30	2.85	6.7	2.8	12.1	52.6
L - Leg Sensories						
Sural	3.35	4.25	12.2	9.5	16.4	49.0
Superficial Peroneal	2.00	2.70	2.2	2.3	10.5	52.5



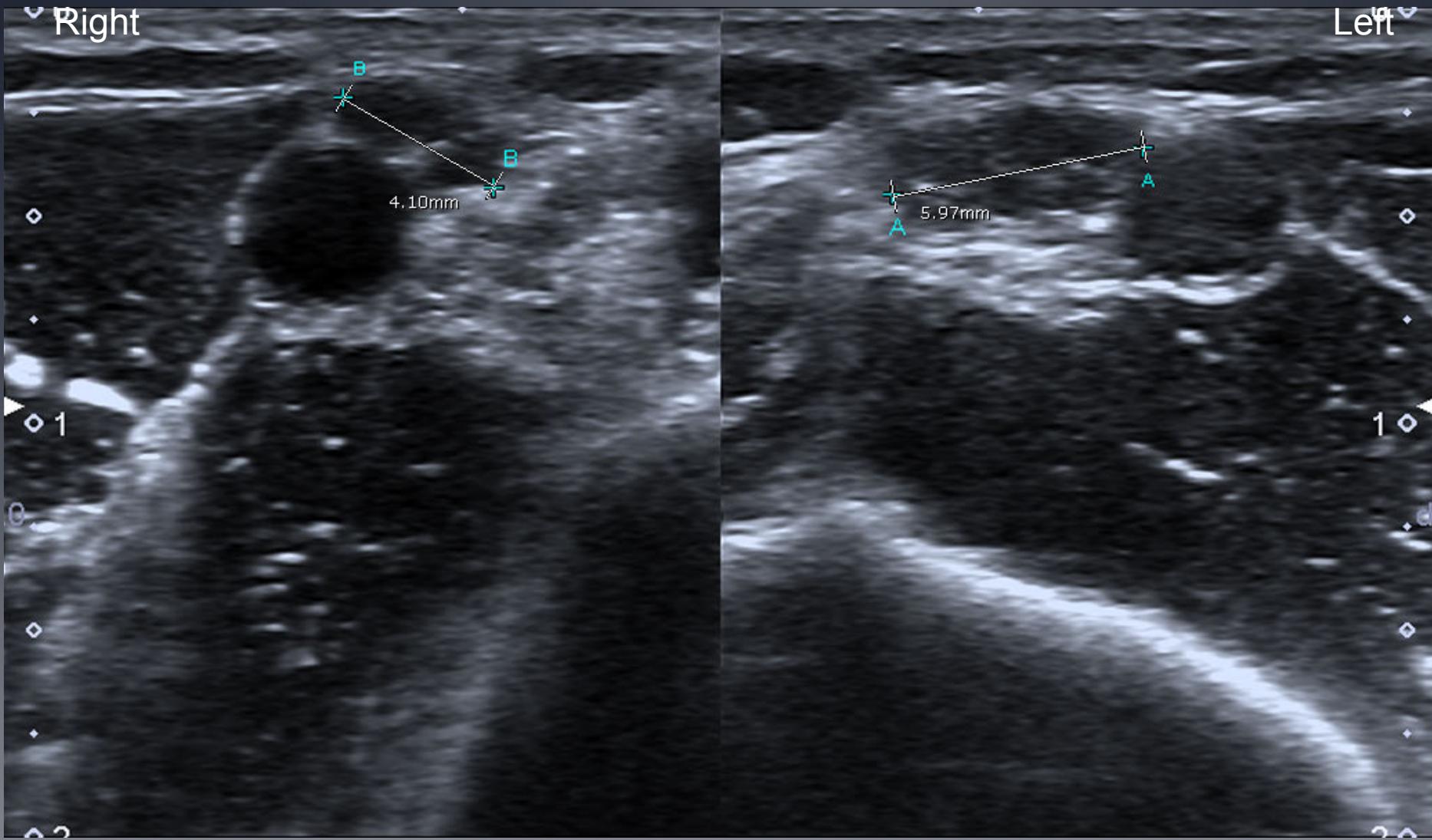
? Differential diagnoses

? Further investigation

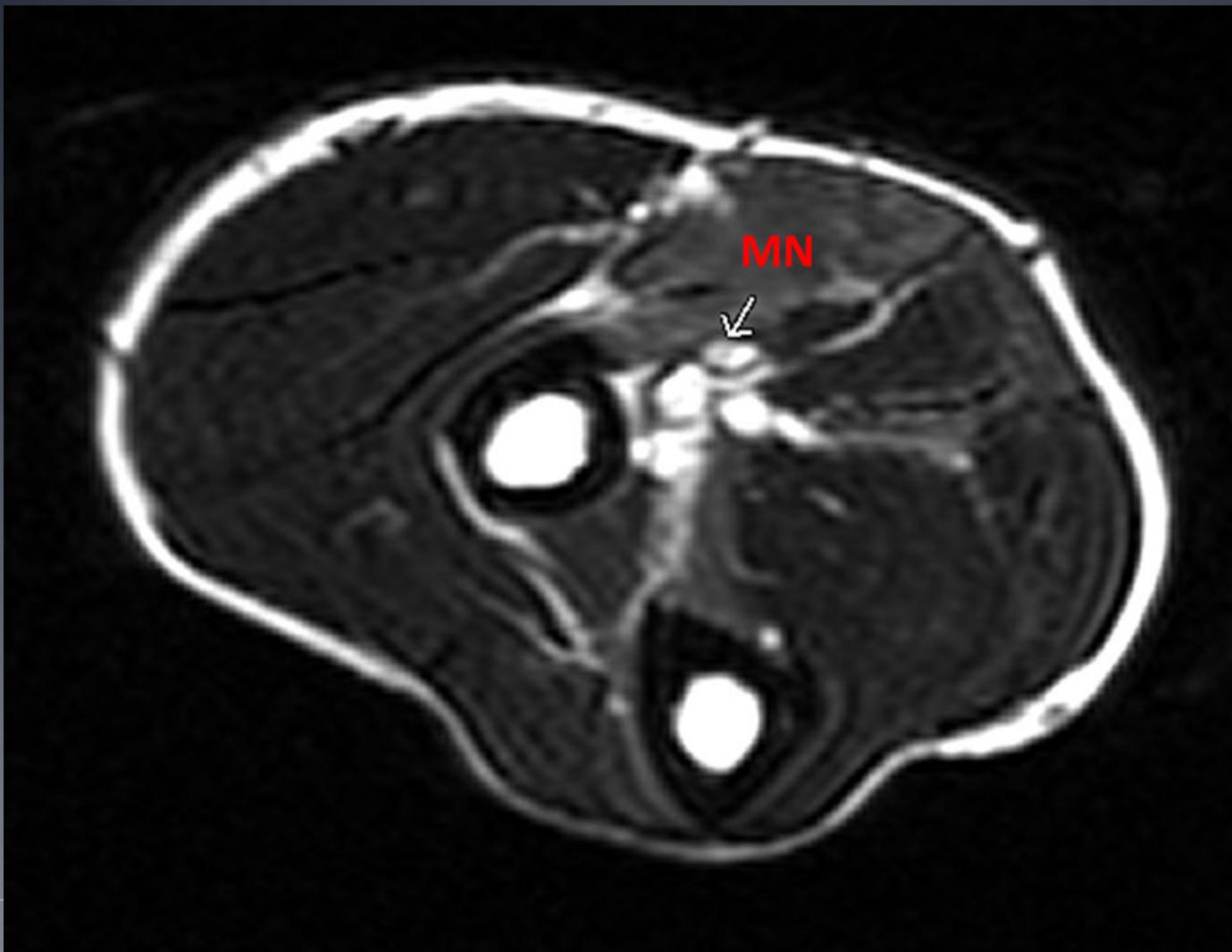
Investigations

- CRP 16 (<5), ESR 2
- C₃ 0.71 (0.9-1.8), C₄ 0.10 (0.16-0.50)
- Vasculitic screen (RF, ANA, ANCA, ENA anti-dsDNA, urine microscopy/cold agglutinins) negative
- Anti-neuronal Ab/tumor markers negative
- HIV/Hep B,C/syphilis/CMV/EBV negative
- LDH 520 (210-420), β₂ microglobulin normal
- SPEP: IgMκ paraprotein 3g/L; BJ protein positive in urine (monoclonal free kappa light chains)
- Anti-MAG negative

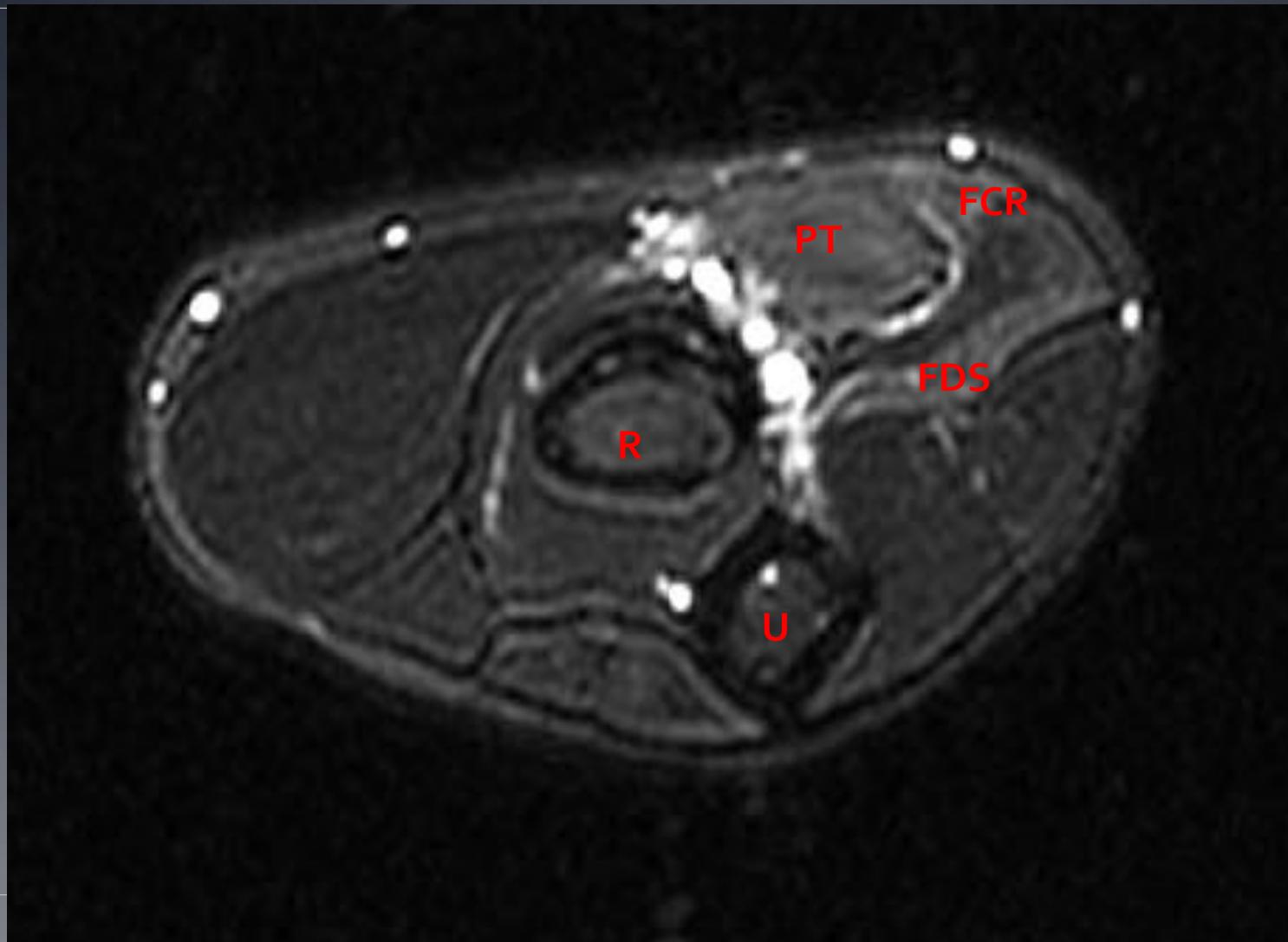
US left arm – mid humerus level



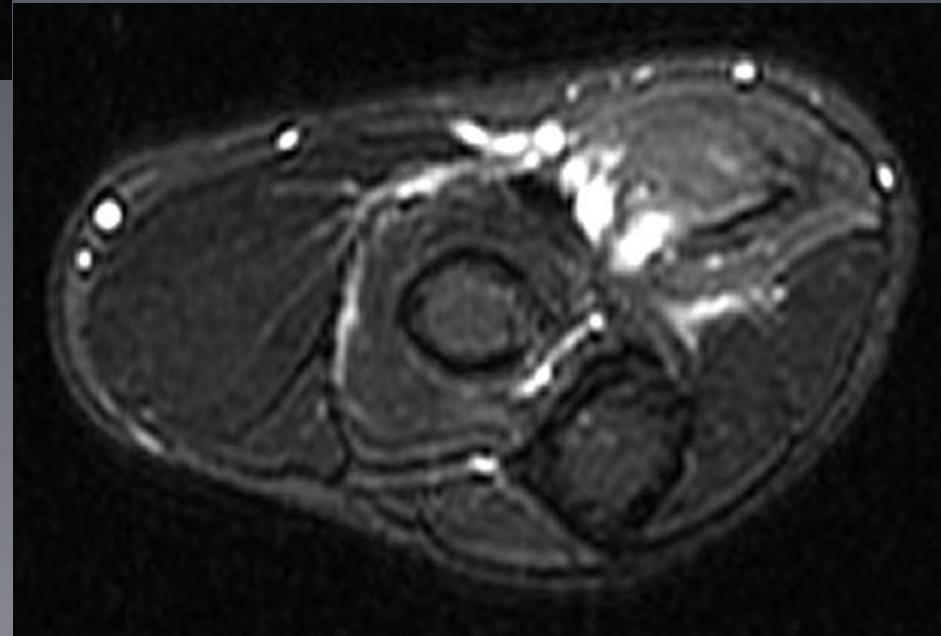
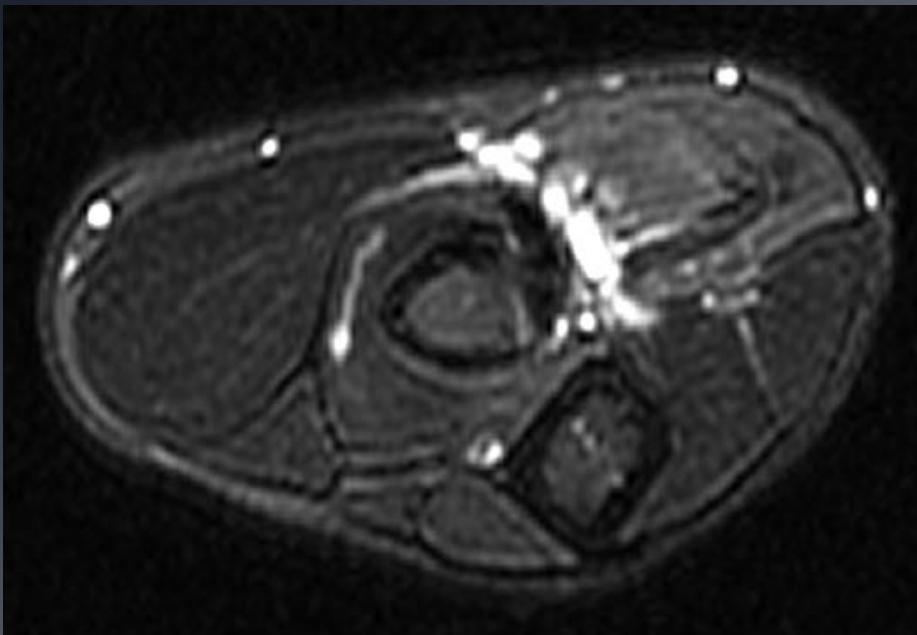
MRI left forearm



MRI left forearm



MRI left forearm



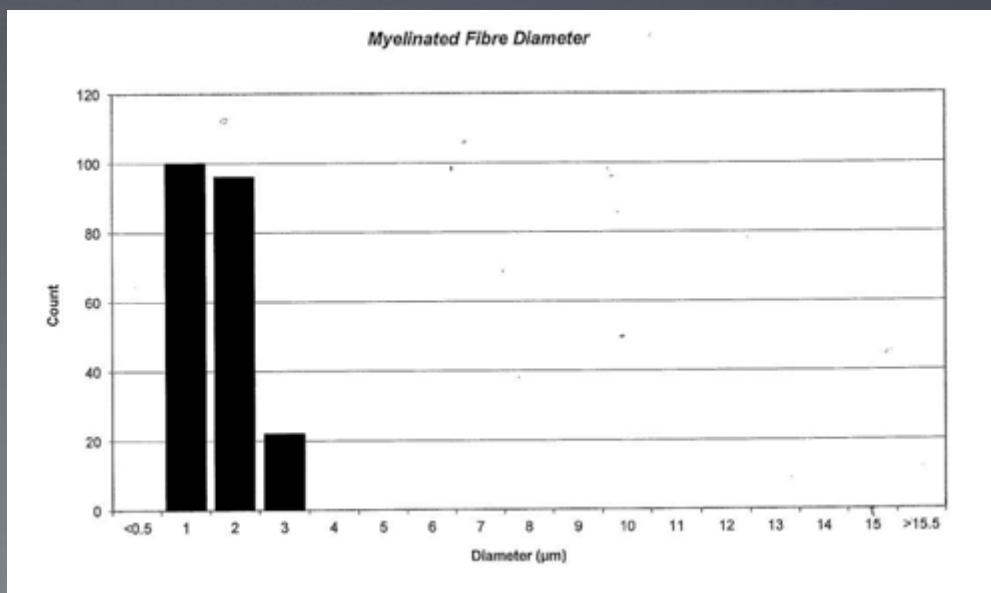
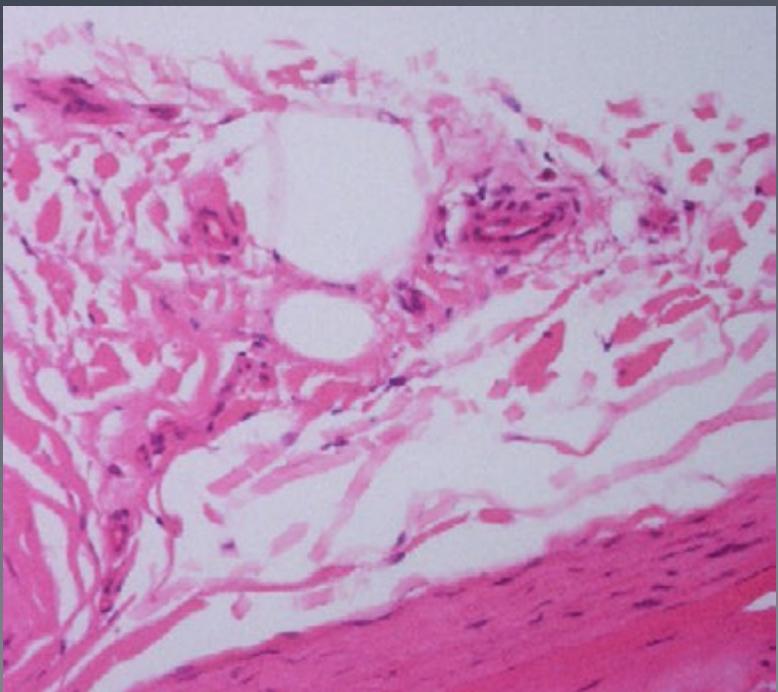
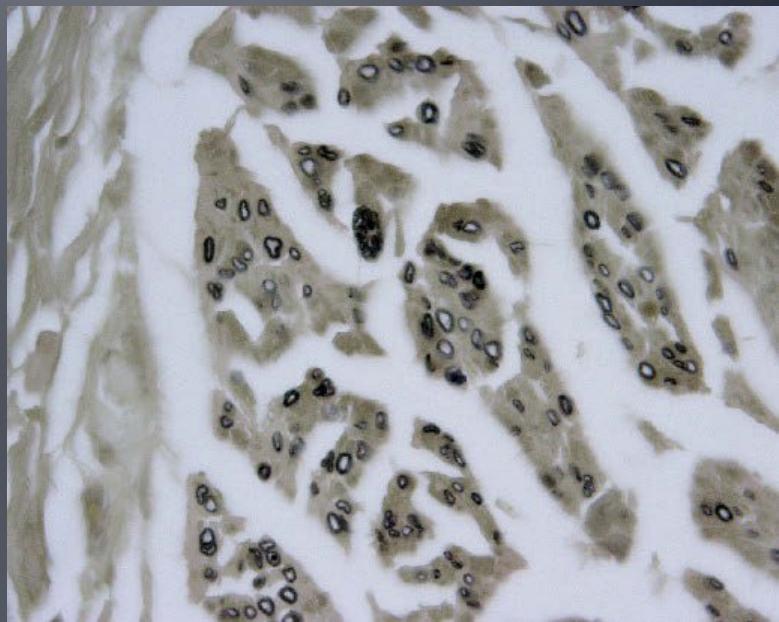
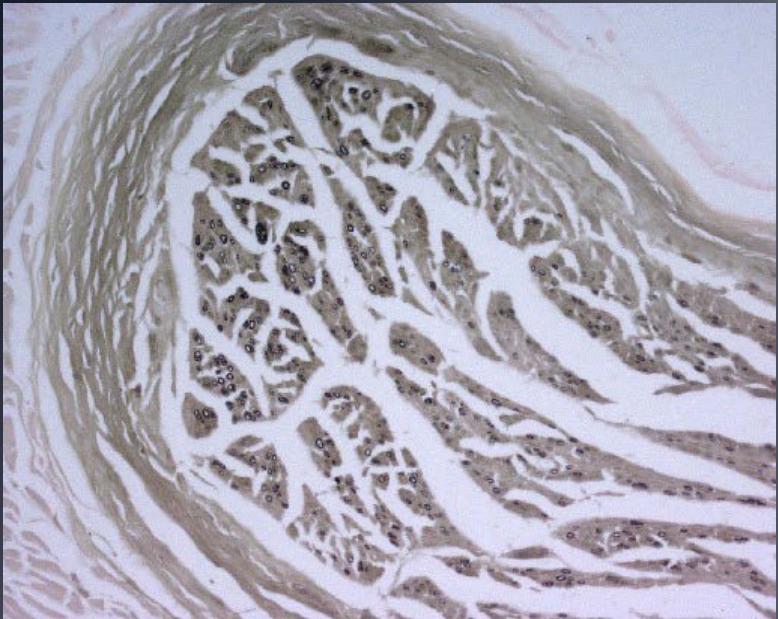
Investigations

- CT chest/abdomen/pelvis – normal
- Skeletal survey normal
- BMAT – aberrant population plasma cells (~5 %), no abnormal lymphocyte population



? Further investigation

? To treat or not to treat



Progress

- No treatment
- Repeat NCS/EMG unchanged
- Clinically stable – severe, left median neuropathy/allodynia; multiple cutaneous mononeuropathies

J Peripher Nerv Syst. 2010 Sep;15(3):176-84. doi: 10.1111/j.1529-8027.2010.00281.x.

Peripheral Nerve Society Guideline on the classification, diagnosis, investigation, and immunosuppressive therapy of non-systemic vasculitic neuropathy: executive summary.

Collins MP¹, Dyck PJ, Gronseth GS, Guillemin L, Hadden RD, Heuss D, Léger JM, Notermans NC, Pollard JD, Said G, Sobue G, Vrancken AF, Kissel JT; Peripheral Nerve Society.

Nonsystemic vasculitic neuropathy: insights from a clinical cohort.

Collins MP¹, Periquet MI, Mendell JR, Sahenk Z, Nagaraja HN, Kissel JT.

Clin Exp Rheumatol. 2008 May-Jun;26(3 Suppl 49):S118-30.

Isolated vasculitis of the peripheral nervous system.

Collins MP¹, Periquet MI.

Front Neurol Neurosci. 2009;26:26-66. doi: 10.1159/000212368. Epub 2009 Apr 6.

Nonsystemic vasculitic neuropathy: update on diagnosis, classification, pathogenesis, and treatment.

Collins MP¹, Periquet-Collins I.

Primary Amyloidosis Presenting as Upper Limb Multiple Mononeuropathies

Jennifer A. Tracy, M.D., Peter J. Dyck, M.D., and P. James B. Dyck, M.D.

Peripheral Neuropathy Research Laboratory, Department of Neurology, Mayo Clinic, 200 First Street SW, Rochester, Minnesota, USA 55905

- Patient with step-wise progressive, multiple upper limb mononeuropathies (left median, bilateral radials) over a 2 year period
- Monoclonal IgM λ (1g/dL)
- Left superficial radial N Bx – axonal deg and amyloid deposition within fascicles and around endoneurial microvessels
- Light chain amyloidosis may be the cause of a multiple mononeuropathy pattern in the absence of a more diffuse peripheral neuropathy
- Primary amyloidosis results from overproduction of monoclonal Ig light chains (λ in 2/3 patients) and subsequent deposition of light chains in vulnerable tissues including nerves
- Systemic complaints, including fatigue and LOW, common

MRI cervical spine + brachial plexus



Acta Neurol Scand. 1997 May;95(5):319-20.

Acute brachial plexus neuropathy as a presenting sign of peripheral nervous system involvement in paraproteinaemia.

Martinelli P, Macrì S, Scaglione C, Stumpo M, Poppi M.

- ABPN as a presenting sign of paraproteinemic neuropathy
- NCS/EMG – focal injury of the upper trunk of BP + mild, diffuse sensorimotor neuropathy
- IgG λ paraprotein
- BMAT – low grade plasmacytoma
- Sural N Bx – demyelinating neuropathy with IgG deposits in Schwann cells; c/w immunomediated mechanism
- ? Coincidental onset

J Neurol Sci. 2010 Apr 15;291(1-2):89-91. doi: 10.1016/j.jns.2010.01.018. Epub 2010 Feb 10.

Light chain deposition in peripheral nerve as a cause of mononeuritis multiplex in Waldenström's macroglobulinaemia.

Luigetti M¹, Frisullo G, Laurenti L, Conte A, Madia F, Profice P, Batocchi AP, Montano N, Tarnani M,
Tonali PA, Sabatelli M.

- WM is a form of monoclonal IgM gammopathy a/w B-cell lymphoplasmacytic lymphoma
- Mononeuritis multiplex previously reported
- Nerve biopsy demonstrates light chain depositions which subverts the normal architecture of the endoneurium and epineurium resulting in fascicular hyalinosis and epineurial artery disruption
- Ig deposition is one of several mechanisms of nerve damage in IgM-related neuropathy