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Case Report

Brief Exercise as a Diagnostic Aid in a Case of Lambert Eaton Myasthenic Syndrome

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Abstract

Background: Lambert-Eaton myasthenic syndrome (LEMS) is an autoimmune disease, affecting the presynaptic neuronal transmission.

Objectives: To represent a case evaluated and confirmed of Lambert Eaton Myasthenic Syndrome.

Methods: 35year old lady with proximal muscle weakness was evaluated with detailed NCS and RNS.

Results: Routine RNS showed decremental response, NCS showed very low CMAPs with marked potentiation post short exercise. Thus incremental response in a patient with pure motor, proximal weakness, areflexia confirmed the diagnosis of LEMS. Her paraneoplastic workup was negative.

Discussion: Non-paraneoplastic LEMS may present with lower limb predominant proximal weakness with subtle autonomic symptoms especially in young females. Bedside clinical examination for pupillary reflexes and postexercise potentiation of tendon reflexes along with increment in CMAP amplitudes aids in confirmation of the diagnosis.

Keywords: LEMS; Potentiation; Non-Paraneoplastic; Presynaptic Neuromuscular Disorders; RNS

Abbreviations

CMAP - Compound Muscle Action Potential, SNAP - Sensory Nerve Action Potential, RNS- repititive nerve stimulation, APB- Abductor Pollicis Brevis

Introduction

Lambert-Eaton myasthenic syndrome is a presynaptic neuromuscular disorder commonly associated with carcinoma lung (PLEMS). Non-Paraneoplastic (NP)- LEMS is seen in younger females with good treatment responsiveness and better prognosis.

Case Summary

A 35years non-diabetic lady, presented with painful cramps in the calf muscles since 8 months. Three months later she developed progressive proximal weakness with difficulty climbing stairs. She had persistent malaise which slowed her household work requiring to rest in between her work. Symptomatic medications gave no relief. She had no buckling or stiffness of limbs, paraesthesias, oculo-bulbar symptoms or bowel and bladder disturbances.

There was progressive change in her voice over next 5 months later becoming low pitched despite rest with exertional breathless-

ness. On enquiring she reported frequent dryness of mouth since 8 months. There were no associated rheumatological symptoms.

She had normal systemic evaluation with normal cognition. Pupils were both 4mm sluggishly reacting to light. Muscle bulk and tone were normal without any fasciculations. Proximal muscle weakness was noted predominantly in lower limbs with modified medical research council grade 4 power while grade 4+ in both upper limbs with mild truncal and neck flexor weakness. There was generalized areflexia with flexor plantar responses. Sensory and cerebellar evaluation was normal. She could walk unassisted with a waddling gait.

Electrophysiology revealed very low CMAP amplitudes with normal velocities, latencies and normal SNAPs. A 3 Hz RNS in right median showed decremental response of 33.8% (Figure 1B). She could not tolerate rapid RNS (30Hz). Post brief (10sec) exercise CMAPs showed significant incremental responses (Figure 1A) confirming the diagnosis of LEMS.

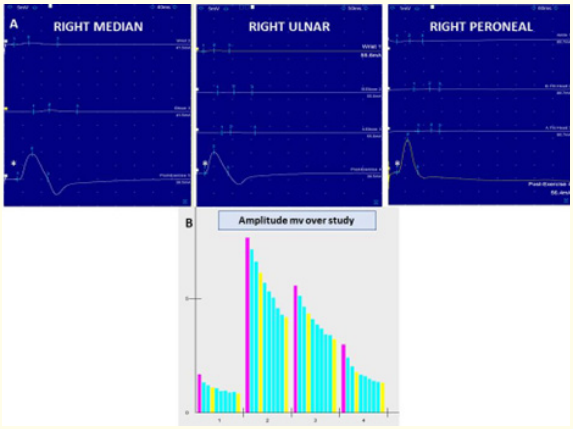

Figure 1: A) Nerve conduction testing at proximal and distal sites at rest (the upper traces) showed very low amplitudes for right median and ulnar nerves with almost non-recordable trace for right peroneal nerve and marked post short exercise (10sec) potentiation (the last trace in each recording *). B) Graphical representation of CMAP amplitudes of R. median nerve slow RNS at 3hz recorded at APB at rest (1), immediately after brief exercise (2), 30seconds after the brief exercise (3) and at 1minute (4). There was decremental response at 3 Hz repetitive stimulation but significant increment in the baseline amplitudes was noted after brief exercise.

Her autoimmune profile showed positive Anti-RNP antibodies. Metabolic work up (TFTs, vitamin D3, electrolytes and blood gas analysis) and malignancy screening were negative. Serum VGCC antibodies could not be tested.

She was started on 20mg Dalfampridine, 40mg of Prednisolone and 100mg of Azathioprine per day. At three months she reported marked improvement in her gait and voice with resolution of dryness of mouth.

Discussion

LEMS is an auto-immune disorder of neuro-muscular junction (NMJ). Paraneoplastic - PLEMS has median age of onset at 60 years and relative male preponderance [1]. Non-paraneoplastic - NP-LEMS accounts for 30-50% of all cases, has bimodal presentation at 4th decade with female preponderance and 6th decade and relatively equal sex distribution [2]. In NP-LEMS, a mean interval between the onset of symptoms and diagnosis is longer than in PLEMS cases. Both are characterized by antibody mediated inhibition of P/Q-type voltage gated calcium channels (VGCC) responsible for calcium influx at pre-synaptic nerve terminals of NMJ and ganglionic cholinergic receptors.

It begins with a proximal weakness, especially of the legs often precipitated with exertion and heat [3]. Muscle pain or cramps are rarely reported in 5% of patients [4]. Post-exercise facilitation due to calcium accumulation in the nerve terminal and increased acetylcholine release in the synaptic gap is a characteristic feature. As in this case marked increase in the CMAP amplitudes immediately after brief exercise verifies the same and may circumvent the need of rapid RNS for diagnosis. Hyporeflexia improving after brief exercise was reported in 43.7% [5].

Widespread autonomic dysfunction noted in about 80% cases [3]. may precede the development of muscle weakness by years. Features such as dry mouth, constipation, and impotence are the presenting symptoms in 3-6%. Pupillary dysfunction in the form of sluggish pupillary reactions may serve as important diagnostic aid.

Conclusion

Non-paraneoplastic LEMS may present with lower limb predominant proximal weakness with subtle autonomic symptoms especially in young females.

Nerve/Sites	Muscle	Latency ms	Amplitude mV	Rel Amp %	Segments	Distance mm	Lat Diff ms	Velocity m/s	RelVel %
R Median -APB									
Wrist	APB	2.15	0.8		Wrist - APB	80			
Elbow	APB	6.29	0.6		Elbow - Wrist	242	3.96	61	100
After 10sec exercise	APB	2.94	9.5	1087					
R Ulnar- ADM									
Wrist	ADM	2.17	0.3		Wrist - ADM	80			
B. Elbow	ADM	5.98	0.3		B. Elbow - Wrist	223	3.81	58	100
A. Elbow	ADM	7.15	0.2		A. Elbow - B. Elbow	80	1.17	69	117
After 10sec exercise	ADM	2.63	7.9	2533	Axilla - A. Elbow		-4.52		
R Peroneal-EDB									
Ankle	EDB	3.48	0.1		Ankle - EDB	80			
B. Fib Head	EDB	8.98	0.1		B. Fib Head - Ankle	353	5.50	64	100
A. Fib Head	EDB	10.35	0.1		A. Fib Head - B. Fib Head	80	1.38	58	90.7
After 10sec exercise Ankle	EDB	3.46	2.5	2400	at rest - A. Fib Head		-6.81		
L Peroneal- EDB									
Ankle	EDB	NR	NR	NR	Ankle - EDB	80			
B. Fib Head	EDB	NR	NR	NR	B. Fib Head - Ankle		NR		
A. Fib Head	EDB	NR	NR	NR	A. Fib Head - B. Fib Head		NR		
After 10 sec exercise Ankle	EDB	3.52	2.8	2700					
R Tibial- AH									
Ankle	AH	NR	NR	NR	Ankle-AH	80			
Knee	AH	NR	NR	NR	Knee- Ankle		NR		
After 10sec exercise Ankle	AH	4.9	3.5	3400					
L Tibial- AH									
Ankle	AH	5.92	0.3	100	Ankle- AH	80			
Knee	AH	12.65	0.2	126	Knee- Ankle	421	6.73	63	100
After 10sec exercise Ankle	AH	5.43	3.8	1166					

Table 1A: Shows the details of nerve conduction study. Right median, ulnar, peroneal and left tibial nerves show very low CMAP amplitudes while left peroneal and right tibial almost non-recordable CMAPs. After brief 10seconds exercise however all the sampled sites showed significant incremental response as shown in bold.

Nerve / Sites	Rec. Site	Onset Lat	Peak Lat	NP Amp	Segments	Distance	Velocity
		ms	ms	µV		mm	m/s
L Sural - (Antidromic)							
Calf	Ankle	2.06	2.77	15.3	Calf - Ankle	140	68
R Sural - (Antidromic)							
Calf	Ankle	2.06	2.88	13.5	Calf - Ankle	140	68
R Superficial peroneal - Ankle							
Lat leg	Ankle	2.65	3.46	10.8	Lat leg - Ankle	140	53
L Superficial peroneal - Ankle							
Lat leg	Ankle	2.48	3.31	12.5	Lat leg - Ankle	140	56
R Median - Orthodromic (Dig II, Mid palm)							
Dig II	Wrist	1.90	2.48	39.0	Dig II - Wrist	130	69
R Ulnar - Orthodromic, (Dig V, Mid palm)							
Dig V	Wrist	1.69	2.21	25.0	Dig V - Wrist	110	65

Table 1B: Shows sensory nerve conduction study with all normal parameters. (CMAP: Compound Muscle Action Potential, SNAP: Sensory Nerve Action Potential, APB: Abductor Pollicis Brevis, ADM: Abductor Digiti Minimi, EDB: Extensor Digitorum Brevis, AH: Abductor Hallucis).

Bedside clinical examination for pupillary reflexes and postexercise potentiation of tendon reflexes along with increment in CMAP amplitudes aids in confirmation of the diagnosis.

Learning point for the clinicians

- LEMS should be suspected in pure motor, progressive hypoareflexic proximal lower limb predominant weakness especially in young women irrespective of risk for malignancy.
- Subtle autonomic features like tonic sluggishly reacting pupil, dryness of mouth are important clinical clues.
- Post-exercise facilitation of reflexes and marked elevation CMAP amplitudes aid in diagnosis.

Conflicts of Interest

None declared.

Consent of Patient

Written informed consent taken.

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