



Progressive Muscular Atrophy Presenting To PM&R Clinic As Hip Pain and Weakness - A Case Study

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Case Description

A 49 year old female was referred to our PM&R clinic for evaluation of longstanding left hip pain. On further evaluation, the left sided hip pain had begun after she lifted a heavy object and was accompanied by proximal muscle weakness involving both hips, which had gradually worsened over the course of 5-7 years. The patient, who was initially running 25 miles per week, had a gradual functional decline leading to ambulation with walking sticks. Her previous work-up included an unremarkable left hip arthrogram and a lumbar spine MRI, which showed paraspinal atrophy. Outpatient physical therapy had resulted in minimal improvement. Her exam demonstrated proximal bilateral lower extremity weakness as well as distal upper extremity weakness, atrophy and hyporeflexia. This led to referral to the Neuromuscular Clinic, where an EMG was performed. The electrodiagnostic testing suggested a diagnosis of chronic progressive motor neuron disease. Genetic testing was also performed, as Spinal Muscular Atrophy Type 4 and Progressive Muscular Atrophy were the initial differentials.

Results

Needle EMG testing was notable for widespread denervation and chronic reinnervation across all sampled muscles in the cervical and lumbosacral body segments, with changes of chronic reinnervation also seen in the thoracic paraspinals at T6.

Needle EMG Data

Muscle	Side	Spontaneous Activity			Motor Unit Morphology			Interference Pattern	
		Insertional Activity	Fibs/Pos. Waves	Fascicls	Duration	Amplitude	Phases	Activation	Recruitment
Deltoid	R	Normal	0	0	+1	+1	Normal	Normal	-1
Flexor Carpi Radialis	R	Increase	+1	0	+2	+3	Normal	Normal	-4
Extensor Digitorum Communis	R	Increase	+1	0	+2	+2	Normal	Normal	-4
First Dorsal Interosseous	R	Increase	+2	0	+2	+2	Normal	Normal	-3
Thoracic paraspinal	R	Normal	0	0	+2	+2	Normal	Normal	-2
Tensor Fasciae Latae	R	Increase	+2	0	+3	+3	Normal	Normal	-3
Vastus Medius	R	Increase	+2	0	+1	+2	Normal	Normal	-4
Tibialis Anterior	R	Increase	+2	0	+2	+2	+2	Normal	-4
Medial Gastrocnemius	R	Increase	+2	0	+1	+3	+1	Normal	-3

Motor nerve conduction studies of the right ulnar and tibial nerves were normal. Right median CMAP amplitude was mildly reduced. The sensory nerve conduction studies of the right median, ulnar, medial antebrachial cutaneous, and sural nerves were normal.

Discussion

Often, referrals for joint pain directed to PM&R result in the diagnosis of a musculoskeletal condition. Neurological conditions, however, should be kept in the differential diagnosis. In this case, the patient was suspected to have a neuromuscular disease due to the pattern, onset and progression of weakness, absence of upper motor neuron signs, as well as first dorsal interossei and thenar atrophy on exam. Electrodiagnostic testing confirmed this clinical suspicion, demonstrating findings consistent with a chronic progressive motor neuron disease. The patient's genetic testing showed variables of unknown significance, and she was diagnosed with Progressive Muscular Atrophy.

Conclusion

Physiatrists should keep neuromuscular conditions in the differential diagnosis when referred for evaluation of joint pain with associated muscle weakness.