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Case Report

Neuralgic Amyotrophy with Isolated Ulnar Nerve Involvement: A Case Report

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Neuralgic amyotrophy involving the lower trunk of the brachial plexus, particularly the ulnar nerve, is exceedingly rare. Here, we report a 41-year-old female who presented with acute right shoulder pain and paresis of the right hand. An electrodiagnostic study showed patterns compatible with acute axonopathy of the right ulnar nerve. However, other electrophysiologic, laboratory, and imaging studies failed to identify a clear cause of the neuropathy. After receiving five weeks of systemic steroid and five months of rehabilitation program, pain and grip strength gradually improved. This is a rare case of neuralgic amyotrophy with isolated ulnar nerve involvement confirmed by an extensive diagnostic workup. (Rehabil Pract Sci 2023; 2023(1): 29 - 34)

Key Words: neuralgic amyotrophy, ulnar nerve, electrodiagnosis

INTRODUCTION

Neuralgic amyotrophy (NA) is an acute peripheral neuropathy characterized by severe pain in the upper extremity followed by prominent multifocal weakness.[1] The most commonly affected nerve is the long thoracic nerve, followed by the suprascapular and anterior interosseous nerves.[2] Even in cases with the most typical presentations, recognizing this condition is often challenging[3] resulting in misdiagnosis and inadequate treatment.[2] In patients with NA, involvement of the lower trunk of the brachial plexus, particularly the ulnar nerve, is unusual.[4,5] Here, we report a rare case of NA with isolated ulnar nerve involvement confirmed by comprehensive neurophysiologic, imaging, and laboratory workup.

CASE REPORT

A 41-year-old female presented with acute right shoulder pain (8/10 on the visual analog scale) for ten days that extended to the lateral side of the right upper arm, forearm, lateral hand, and the first three digits after waking up from a nap. The pain was persistent and unresponsive to over-the-counter pain relievers, together with profound weakness of the hand, preventing her from performing normal daily activities. A history of common cold about 2 weeks before symptom onset was also reported. Accompanying symptoms included numbness over the right medial forearm, medial hand, and the fourth

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and fifth digits. Decreased grip strength and weakness with the abduction of the fifth digit were also noted. Physical examination revealed decreased muscle strength of the right hand, graded 2+/5 on the Medical Research Council scale for finger flexors of the fourth and fifth digits, abductors of the fifth digit and the flexor carpi ulnaris muscle. Sensory testing revealed diminished sensation to both light touch and pin prick over the right medial forearm, medial hand, and the fourth and fifth digits. Muscle stretch reflexes of the biceps brachii, triceps brachii, and brachioradialis muscles were normal and symmetric. The skin color, thickness and temperature were symmetric in both upper limbs. No increased hair, nail growth or hyperhidrosis over the affected limb were present. There was no observable abnormality of shoulder movements or shortness of breath. An electrodiagnostic (EDX) study (Table 1 and Table 2) was performed on the tenth day, with findings suggestive of acute right ulnar neuropathy with axonopathy. The results of electrophysiologic and imaging studies such as magnetic resonance imaging (MRI) of the head and cervical spine, computed tomography (CT) of the chest, and motor evoked potential of four limbs by trans cranial magnetic stimulation were unremarkable. The results of metabolic and autoimmune panel workup including levels of serum vitamin B12, folate, anti-nuclear antibody, rheumatoid factor and thyroid function tests were all within normal limits. Screening tests for the human immunodeficiency virus were also negative. High-resolution sonography showed no mass-like lesion in the right upper extremity or focal swelling in the right ulnar nerve from the axilla to the wrist. The patient was given a course of corticosteroids that included intravenous methylprednisolone 40 mg twice daily for five days, followed by four weeks of oral prednisolone 25 mg twice daily. Analgesic drugs with pregabalin (75mg/cap) 1 cap daily and tramadol/acetaminophen (37.5/325mg/tab) 1 tab four times a day were also given. Rehabilitation programs consisting of motor control training, hand function training, scapular stabilization exercises, periscapular musculature endurance training as well as training for daily living activities were provided. The EDX study was repeated on the sixth week (Table 1 and Table 2), with similar nerve conduction study findings as to the first, but with positive sharp waves and fibrillation potentials in multiple ulnar-innervated muscles on electromyography, indicating subacute right ulnar neuropathy with axonopathy. Pain and weakness gradually improved over the following weeks. During the last follow-up clinical visit five months after symptom onset, the patient's muscle strength in the affected muscles returned to normal, and she was able to perform all daily living activities independently. Trivial and vague discomfort in the right shoulder was the only residual symptom reported by the patient.

DISCUSSION

Neuralgic amyotrophy, also known as idiopathic brachial plexus neuropathy,[2] Parsonage—Turner syndrome,[3] brachial plexus neuritis,[3] or brachial plexitis,[1] is a frequently underappreciated acute peripheral neuropathy. The incidence rate has been reported to be 1 per 1000 per year,[6] much higher than previously believed. The classic clinical picture is a patient who wakes up with new-onset severe shoulder or upper arm pain that usually exceeds 7/10 on a numeric rating scale.[2,3] In about half of the patients, antecedent events including viral or bacterial infections, trauma, or surgery can be identified days or a few weeks prior to the onset of symptoms.[1] Hours to days after symptom onset, prominent motor weakness develops and usually involves the long thoracic, anterior interosseous, and the suprascapular nerves.[2] Sensory symptoms frequently coexist and most commonly affect the axillary, lateral antebrachial cutaneous, and the superficial radial nerves.[2,3]

Patients with atypical presentations of NA account for less than a third of all NA cases.[2] In one study of 246 patients,[5] lower trunk-predominant NA comprised only 4.17% of the patient population. In another study of 281 cases, Ferrante and Wilbourn[4] reported a low rate of ulnar nerve involvement at 1.13% across all lesion distributions. We report an extremely rare case of NA with isolated ulnar nerve involvement demonstrated by extensive neurophysiologic, imaging, and laboratory workup. Despite the advances of neuroimaging modalities such as high-resolution ultrasound and magnetic resonance neurography, NA is still considered a clinical diagnosis and a diagnosis of exclusion. Detailed history taking, thorough physical examination, and a comprehensive

workup should be conducted to rule out other conditions that might mimic NA clinically.[2,3]

The presence of a preceding event, the rapid onset of symptoms, severe shoulder pain, and failure to fit the signs and symptoms into a classic neurological localization paradigm all matched the typical presentation of NA in our case. The only exception to this classic clinical pattern was the weakness of the ulnar-innervated muscles which is rare in NA.[4] The differential diagnosis included cervical radiculopathy, neurogenic thoracic outlet syndrome, lesions of the central nervous system, Pancoast tumor causing compression of the lower brachial plexus, infectious peripheral neuropathy, and other secondary causes of ulnar neuropathy. By using extensive electrodiagnostic study, appropriate imaging and laboratory examinations, these possible diagnoses were ruled out.

Electrodiagnostically, the patient initially presented with acute axonopathy of the right ulnar nerve, sparing other major nerves in the right upper extremity. No pick-up of the ulnar sensory conduction study of the affected side indicated that the lesion is at or distal to the dorsal root ganglion, which excluded the possibility of a C8 or T1 radiculopathy. This is further supported by normal results of the C8 and T1 paraspinal muscles sampled on electromyography. Normal parameters of the median motor conduction study recording the abductor pollicis brevis muscle and the medial ante brachial cutaneous sensory study, together with sparing of C8- or T1-innervated non-ulnar muscles on electromyography including the extensor digitorum communis, extensor indicus proprius, abductor pollicis brevis, and the flexor digitorum profundus muscles make the diagnosis of lower trunk/medial cord brachial plexopathy and neurogenic thoracic outlet syndrome much less likely. The second-EDX study revealed chronological evolution of the nerve lesion, with the appearance of fibrillation potentials and positive sharp waves in ulnar-innervated muscles. This pattern of axonopathy on EDX is consistent with NA,[1,8] as nine out of ten patients with NA in one study demonstrated evidence of axonopathy on electrodiagnostic study.[8]

Neuromuscular ultrasound plays an important role in the evaluation of NA. It helps exclude compressive peripheral neuropathies which is the one of the most common differential diagnosis of NA.[1] Occasionally, nerve swelling with complete or incomplete construction on ultrasound can be seen.[7] These findings, in an appropriate clinical context, are not only highly indicative of NA but also help guide the treatment.[1,7] Patients with nerve constriction are found to be unresponsive to corticosteroid and should be best treated with surgical neurolysis with detorsion.[7] Conservative treatment with systemic corticosteroid, adequate analgesia and comprehensive rehabilitation is sufficient in our case, in whom ultrasound did not reveal constriction or other structural abnormality.

NA has been generally considered to have good prognosis. [1,2,5,8] In one study, six out of ten patients (60%) with NA have their motor power completely recovered six months after disease onset.[8] The mean grades of muscle power improvement were 2.4 ± 0.6 to 4.5 ± 0.7 at 7.8 ± 3.8 months of follow-up. [8] In another study of 246 cases with NA, more than half of the patients stated to have at least 50% overall recovery after 6-12 months. Van Eijk also found that the majority of patients with NA have prominent recovery of strength in their paretic muscles over the course of 6-18 months.[2] As NA is axonal loss in nature and reinnervation progresses at 1mm/day for nerves with severe axonal loss, Van Eijk believed course of several months for recovery is reasonable. [2] Based on the current literature, it is reasonable that our case achieved a nearly full recovery from paralysis, especially considering her early use of systemic corticosteroid, negative sonographic findings, and early initiation of comprehensive rehabilitation programs.

A short course of systemic corticosteroid is considered the treatment of choice for NA in the absence of constriction of nerve.[3,7] When administered within one month of symptoms onset, it has been proven to shorten the duration of pain and achieve a higher recovery rate at 1 year.[3] Multidisciplinary rehabilitation programs have been reported to help patients regain strength in paretic muscles, restore occupational capability and performance in daily living activities as well as reduce persistent pain and fatigue.[2] The patient in our case received a comprehensive rehabilitation program that focused on hand function training, resulting in a good recovery five months after symptom onset.

Table 1. Nerve conduction studies

Initial study at day 10									
Nerve stimulated	Stimulation site	Recording site	Onset latency (ms)		Amplitude Motor (mV) Sensory (μV)		Velocity (m/s)	
			R	L	R	L	R	L
Motor studies								
Median	Wrist	APB	3.1	3.0	6.7	8.2	61	58
	Antecubital fossa		7.0	6.9	6.7	8.0		
Ulnar	Wrist	ADM	NP*	2.7	NP*	5.9		
	Below elbow		NP*	6.1	NP*	5.8	NP*	59
	Above elbow		NP*	8.0	NP*	5.3	NP*	53
Radial	Forearm	EIP	2.3	2.3	2.5	3.0		
	Elbow		5.1	NT	2.5	NT	59	NT
	Spiral groove		6.5	NT	2.2	NT	69	NT
Axillary	Erb's point	Deltoid	3.4	3.3	4.0	5.9		
Musculocutaneous	Erb's point	Biceps brachii	4.1	3.8	7.0	6.6		
Sensory studies								
Median	Palm	3rd digit	1.9	1.9	54.8	68.2	37	37
	Wrist		3.3	3.2	48.1	61.9	50	54
Ulnar	Wrist	5th digit	NP*	3.1	NP*	28.5	NP	45
Dorsal ulnar cutaneous	Ulnar wrist	Dorsal 4th web space	NP*	2.5	NP*	13.3	NP	52
Radial	Forearm	Snuffbox	2.4	2.4	37.8	39.0	45	47
Lateral antebrachial cutaneous	Elbow	Lateral forearm	2.5	2.4	25.2	39.0	48	50
Medial antebrachial cutaneous	Elbow	Medial forearm	2.4	2.2	23.1	26.3	50	55
Follow-up study at week 6								
Nerve stimulated	Stimulation site	Recording site	Onset latency (ms)		Amplitude Motor (mV) Sensory (μV)		Velocity (m/s)	
			R	L	R	L	R	L
Motor studies								
Median	Wrist	APB	3.0	2.9	5.8	6.4	59	61
	Antecubital fossa		7.2	6.9	5.7	5.9		
Ulnar	Wrist	ADM	2.7	2.6	0.4*	7.6		
	Below elbow		5.6	5.9	0.4*	7.3	54	59
	Above elbow		8.1	8.2	0.5*	6.3	52	59
Radial	Forearm	EIP	2.3	2.2	2.9	2.3		
	Elbow		5.0	4.3	2.5	2.0	61	67
Sensory studies								
Median	Wrist	3rd digit	3.2	3.2	46.2	49.8	58	59
Ulnar	Wrist	5th digit	NP*	3.0	NP*	26.2	NP*	45
Radial	Forearm	Snuffbox	2.5	2.4	33.2	38.0	46	48

*Indicates abnormal data based on our reference values.

APB, abductor pollicis brevis; ADM, abductor digiti minimi; EIP, extensor indicus proprius; NP, no-pickup; NT, not tested.

Table 2. Electromyography for the right upper limb

Muscle	Spontaneous activity			Voluntary motor unit action potentials				
	Insertional activity	Fibrillation potentials	Positive sharp waves	Amplitude	Duration	Polyphasia	Recruitment	Interference pattern
Initial study at day 10								
Deltoid	NL	0	0	NL	NL	NL	NL	Full
Biceps	NL	0	0	NL	NL	NL	NL	Full
PT	NL	0	0	NL	NL	NL	NL	Full
EDC	NL	0	0	NL	NL	NL	NL	Full
EIP	NL	0	0	NL	NL	NL	NL	Full
APB	NL	0	0	NL	NL	NL	NL	Full
FDP 4, 5	Increased	0	0	NL	NL	NL	Discrete	Incomplete
FCU	Increased	0	0	NL	NL	NL	Discrete	Incomplete
ADM	Increased	0	0	NL	NL	NL	Discrete	Incomplete
FDI	Increased	0	0	NL	NL	NL	Discrete	Incomplete
C7 Parasp	NL	0	0	NL	NL	NL	NL	>80%
C8 Parasp	NL	0	0	NL	NL	NL	NL	>80%
T1 Parasp	NL	0	0	NL	NL	NL	NL	>80%
Follow-up study at week 6								
EIP	NL	0	0	NL	NL	NL	NL	Full
APB	NL	0	0	NL	NL	NL	NL	Full
FDP 4, 5	Increased	3+	2+	NL	NL	NL	Discrete	Incomplete
FCU	Increased	3+	2+	NL	NL	NL	Discrete	Incomplete
ADM	Increased	3+	3+	NL	NL	NL	Discrete	Incomplete
FDI	Increased	3+	3+	NL	NL	NL	Discrete	Incomplete
C7 Parasp	NL	0	0	NL	NL	NL	NL	Full
C8 Parasp	NL	0	0	NL	NL	NL	NL	Full
T1 Parasp	NL	0	0	NL	NL	NL	NL	Full

NL, normal; PT, pronator teres; EDC, extensor digitorum communis; EIP, extensor indicus proprius; APB, abductor pollicis brevis; FDP, flexor digitorum profundus; FCU, flexor carpi ulnaris; ADM, abductor digiti minimi; FDI, first dorsal interosseous; Parasp, paraspinal muscle.

CONCLUSION

Neuralgic amyotrophy is a condition that should not be overlooked when evaluating patients with acute upper limb pain or acute neuropathy of the upper extremities. In cases with atypical presentations, ancillary tests may be useful in ruling out other pathologies. Appropriate medications and comprehensive rehabilitation programs should be provided as they optimize patient outcomes.

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