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Aims and Scope

Journal of Electrodiagnosis and Neuromuscular Diseases (*J Electrodiagn Neuromuscul Dis*, JEND) is a peer-reviewed journal concerning both normal and abnormal functioning of the muscle, the neuromuscular junction, and the peripheral motor, sensory and autonomic nerves. The journal publishes clinical studies, reviews, and case reports in the fields of electrophysiology, electrodiagnosis, imaging studies including ultrasonography, and management, about neuromuscular diseases. The journal is aimed to provide an open forum for original research in basic science and clinical research that will improve our fundamental understanding and lead to effective treatments of neuromuscular diseases.
JEND is the official journal of the Korean Association of EMG Electrodiagnostic Medicine.

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Editor-in-Chief
Dong Hwee Kim, Korea University, Korea

Editorial Office
Department of Physical Medicine and Rehabilitation, Korea University Ansan Hospital,
123 Jeokgeum-ro, Danwon-gu, Ansan 15355, Korea
Tel: +82-31-412-5330 Fax: +82-31-412-4215 E-mail: editjend@gmail.com

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Prevention and Treatment of Post-Paralytic Synkinesis: Critical Changes Following Severe Facial Palsy

Jin Kim

Department of Otorhinolaryngology, Hallym University Dongtan Sacred Heart Hospital, College of Medicine, Hallym University, Hwaseong, Korea

Facial synkinesis following severe facial palsy has been observed clinically across a wide range of patient groups, from infants to older adults, indicating that it is not limited to any specific demographic. In one of the largest studies on the natural progression of idiopathic peripheral facial nerve palsy, synkinesis was reported in approximately 16% to 55% of patients. The severity of facial synkinesis generally correlates with the severity of the initial facial palsy. Various types of nerve injuries have been categorized based on the extent of damage to the endoneurium, perineurium, and axons. Aberrant responses and ephaptic transmission following facial nerve injury are responsible for the clinical manifestations of facial synkinesis. It is crucial to minimize facial synkinesis in the early stages and to ablate it in the chronic stages to effectively manage this challenging complication following severe facial palsy. We advocate for physical therapy, including neuromuscular retraining during the acute phase, as a viable option for patients at high risk of developing facial synkinesis after a severe episode of Bell's palsy. This approach helps prevent excessive movement (hyperkinesis) and enables patients to recognize and manage unwanted co-contractions (synkinesis). Several treatments targeting the facial nerve and muscles are available, including chemo-denervation, selective myectomy, and selective neurectomy. These interventions aim to address aberrant regeneration and peripheral ephaptic transmission.

Keywords: Facial paralysis; Synkinesis; Physical therapy modalities; Botulinum toxins, type A; Denervation

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Corresponding author:
Jin Kim
Department of Otorhinolaryngology, Hallym University Dongtan Sacred Heart Hospital, College of Medicine, Hallym University, 7 Keunjaebong-gil, Hwaseong 18450, Korea
Tel: +82-31-8086-2836
Fax: +82-31-8086-2874
E-mail: jinsound@gmail.com

Introduction

Facial palsy can lead to significant functional and psychological disorders due to its effects on both static and dynamic facial symmetry [1-3]. In the initial phase of complete paralysis, complications generally stem from muscle weakness in the affected area. Additionally, during partial recovery, patients may experience facial asymmetry and an inability to perform fine movements, which can be linked to prolonged paralysis or the development of post-paralytic synkinesis [4].

Facial muscles that have degenerated or atrophied due to denervation of the facial nerve can undergo re-innervation by axons from non-native muscle groups. This leads to a phenomenon known as synkinesis, which is characterized by unintentional motion in one area of the face during intentional movement in another area [5]. Synkinesis represents one of the most troubling sequelae of facial palsy. Various mechanisms have been suggested to explain synkinesis. These include aberrant regeneration in cases of third-degree nerve injuries as classified by Sunderland [6], ephaptic transmission where neighboring axons at the injury site stimulate each other [7], and hyper-excitability of the facial nucleus in the cerebral motor cortex [8].

Such synkinetic movements, regardless of the underlying mechanism, typically emerge during the neural repair process, 3 to 6 months after injury. They become particularly noticeable during spontaneous facial movements, especially during emotional expressions like involuntary blinking or smiling [9,10]. These observations suggest that nuclear hyper-excitability may significantly contribute to the development of synkinesis, in conjunction with aberrant regeneration. Numerous researchers have reported that the central cortex may also undergo reorganization as a secondary response to peripheral facial palsy [11-13]. The pathophysiology of synkinesis is highly complex and multifactorial.

In a related study, it was observed that synkinesis occurred in approximately 84% of patients when the early degeneration rate exceeded 80% [14,15]. Consequently, even with the early administration of optimal treatments such as steroids or antiviral agents, synkinesis remains an unfortunate inevitability for these patients.

Therefore, it is crucial not only to treat but also to predict and minimize synkinesis at an early stage. Synkinesis typically begins to manifest gradually between 3 and 6 months after the onset of facial palsy. Therefore, during the subacute phase, which spans from 3 to 6 months, it is advisable to engage in neuromuscular retraining therapy (NMRT). This therapy corrects improper muscle movement patterns and suppresses excessive movements, thereby preventing the development of synkinesis [16]. Early NMRT includes educating patients about potential synkinesis before it actually develops. Awareness of likely future facial changes is a crucial component of early NMRT.

Three primary treatment options are available to alleviate existing facial synkinesis: NMRT, botulinum toxin A injections, and selective neurectomy [17,18]. These methods aim to diminish the activity of synkinetic muscles and increase the activity of paralytic muscles, thus enhancing overall facial motility.

However, it is necessary to observe the detailed pattern of synkinetic hemifacial movements before determining appropriate treatment options. Synkinesis is known to manifest in various forms, including oral-ocular, ocular-oral, ocular-chin, ocular-nasal, and platysma synkinesis. These types are categorized based on the two muscle groups involved in each case [19].

In most patients with facial synkinesis, clinicians recommend botulinum toxin type A (BTX-A) injections as the first-line treatment. For those with severe facial synkinesis, selective neurectomy is considered superior [1]. However, numerous studies have shown that combining NMRT with prior BTX-A injections, rather than using BTX-A alone, significantly improves facial synkinesis and asymmetry in cases of chronic facial paralysis. These findings are supported by detailed, accurate, and objective measurement systems [20-22].

Although selective neurectomy is highly effective in eliminating synkinetic movements, it should be approached with caution due to its irreversible nature. The primary objectives of this surgery are to create a natural smile and to eliminate synkinetic movements where feasible. However, outcomes can vary significantly depending on the surgeon's experience and the patient's preferences, which are notably influenced by cultural factors [23]. This procedure can provide satisfactory outcomes in terms of facial tightness and narrowing of the eyelid aperture. For patients seeking to correct limited mouth movement, the approach should be tailored to their specific correction preferences.

Predicting Facial Synkinesis in the Acute Stage of Facial Palsy

Predicting the prognosis of patients with acute facial palsy is crucial. The prognosis for acute patients can be broadly categorized into three outcomes: complete recovery, incomplete recovery with permanent synkinesis and contracture, and no recovery, resulting in a permanently flaccid face [24,25]. When a patient first presents, predicting which of these three patterns will manifest in the coming months or years is a critical step, as it can significantly influence the treatment direction.

In the case of Bell's palsy, which represents the majority of facial palsy cases, about 70% of patients have a favorable prognosis and achieve complete recovery [26]. However, the rest of the patients experience either incomplete recovery or, in some instances, minimal recovery at all.

One of the effective methods for predicting prognosis is the electroneuronography (ENoG) study [27-30]. This study yields the most accurate results when performed 2 to 3 days after the onset of facial palsy, coinciding with the beginning of Wallerian degeneration. It remains valid until 2 to 3 weeks post-onset, which marks the conclusion of Wallerian degeneration. After this period, the accuracy of the measurements may decrease as Schwann cell regeneration begins at the damaged portion of the facial nerve, potentially leading to false-negative or false-positive outcomes. It is important to note that the severity of nerve damage can influence the timing of Wallerian degeneration. In cases of severe nerve damage, Wallerian degeneration may start and end more rapidly, thus shortening the window for obtaining accurate measurements [31,32].

In patients with acute facial palsy, it is crucial to determine which individuals will achieve complete recovery and which will develop synkinesis. This distinction indicates whether more ag-

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gressive treatment is required or if monitoring over time is sufficient. From a pathophysiological standpoint, a tool that can differentiate between facial nerve damage of Sunderland classification grade 3 or higher and less severe damage is essential. The ENoG test meets this requirement.

Numerous studies have reported degeneration rates in ENoG tests that correlate with the development of synkinesis. Typically, a degeneration rate ranging from 60% to 80% is considered indicative of the potential for synkinesis [33-36].

Based on the author's experience, when the degeneration rate on acute ENoG tests ranges from 60% to 80%, mild synkinesis may occur. This condition is often so subtle that it is either barely noticeable or, when noticed, does not significantly interfere with daily activities. Therefore, aggressive treatment is typically not pursued in these cases. Conversely, when the degeneration rate exceeds 80%, synkinesis can significantly disrupt daily life, necessitating more aggressive treatment [16]. An accelerated and severe Wallerian degeneration, surpassing 80% as observed on ENoG and coupled with facial function inferior to House-Brackmann grade IV, is expected to correlate with widespread aberrant regeneration following Bell's palsy. The rates of degeneration were independently quantified for each of the five branches of the facial nerve, spanning from the forehead to the neck. Evaluating each of these branches separately can provide insights into the rate of neural injury. This approach could potentially offer invaluable information regarding the specific patterns of synkinetic movements that might develop in the future within each region [16].

Minimizing Facial Synkinesis through Early-Stage Neuromuscular Retraining Therapy

Synkinesis in patients with Bell's palsy can be minimized through early intervention with physical therapy, including NMRT, before the onset of synkinesis [16]. The timing of NMRT is crucial. Historically, NMRT has primarily been employed to address already developed synkinetic mass-like movements [37]. This therapy focuses on correcting neuromuscular abnormalities by enhancing muscle coordination, rather than increasing muscle strength.

Early NMRT involves informing patients about potential synkinesis before its onset. This awareness of anticipated facial changes is a crucial aspect that sets NMRT apart from conventional approaches. Clinicians guide patients to minimize excessive movements in areas such as the eyelids, mouth corners, forehead, nostrils, neck, and any other regions where the patient observes slight involuntary movements. The therapy encompasses soft tissue mobilization and selective stretching to strengthen the paralyzed muscles. The use of any electrical stimulation that causes mass-like twitching or movement is strictly prohibited to avoid negative impacts on synkinesis and improper muscle contractions [38-40]. It is advisable to consult a rehabilitation medicine specialist for guidance on the appropriate types and frequencies of electrical therapy for facial palsy rehabilitation.

At the onset of physical therapy, each patient is informed about the potential for synkinesis. Understanding the possibility of future facial changes is a crucial aspect of early NMRT. The therapist explains various types of synkinesis to the patients, including oral-ocular/ocular-oral, chin-oral/chin-ocular, nasal-oral/ocular-nasal, and platysmal synkinesis, which may develop 5 to 6 months after the onset of palsy. The therapist demonstrates how to prevent excessive movements in the eyelids, mouth corners, forehead, nostrils, and neck. Additionally, if a patient reports any involuntary movements in other facial areas, they are instructed on techniques to minimize these movements. In their daily routines, patients are encouraged to practice small, slow, and symmetrical facial movements to establish new movement patterns.

After informing the patients that synkinesis involves uncoordinated, inappropriate movements, the therapist explains that the main purpose of NMRT is to learn new patterns to minimize synkinesis (Table 1).

NMRT is commonly prescribed for facial palsy patients who already exhibit synkinesis. However, initiating NMRT early, as soon as movement returns to the affected side after recovery from Bell's palsy, can provide insights into the potential for synkinesis and awareness of possible future facial changes.

However, it is essential to comprehend that all efforts to minimize synkinetic movements are predicated on the assumption that critical interventions are performed during the acute phase. These interventions include the timely administration of steroids or antiviral agents and, if necessary, the implementation of facial nerve decompression surgery. These measures are crucial during the acute phase and establish the groundwork for all subsequent efforts.

Two Treatment Options: Chemo-Denervation Combined with Neuromuscular Retraining Therapy and Surgical Denervation

NMRT normalizes resting muscle tone and enhances facial expressions by inhibiting synkinetic movements [41]. During NMRT, patients engage in subtle facial exercises to enhance facial symmetry [42]. Since facial muscles lack sensory receptors,

Table 1. Early Neuromuscular Retraining Therapy to Minimize Synkinesis

Relaxation of any restriction in dynamic motion caused by simultaneous contraction of an opposing muscle, and the control of potentially aberrant gross motor activity
Explanation of the current abnormal movement pattern of the patient's face
Prohibition of nonspecific maximum-effort exercises aiming to strengthen muscles, regardless of the inhibition or coordination of abnormal patterns. Gross facial movements should not be performed. The therapist focuses on the development of small motor units that could be precisely refined to yield complex, subtle movements.
Education on the sensory information imparted by facial movements, to enhance symmetrical adaptation and learning.
Every effort should be made to develop the facial muscles responsible for expressions that convey happiness.
Motivation is maximized by individualized instruction and active patient participation in facial musculature control.
Neuromuscular retraining therapy is not "delivered" to the patient; rather, the patient performs the therapy because the psychosocial and educational aspects of rehabilitation are very important and are thus prioritized.

NMRT aids in increasing self-awareness of facial posture and movements. A systematic review demonstrated that biofeedback with electromyography is an effective treatment for synkinesis and asymmetry following facial palsy [43]. Similarly, biofeedback utilizing a mirror has proven to be an effective treatment strategy [17]. Individualized facial neuromuscular re-education has shown greater effectiveness in improving facial symmetry in patients with Bell's palsy compared to conventional treatments, such as electrical stimulation, gross facial expression exercises, and massage [44].

Although NMRT is the cornerstone treatment for synkinesis, BTX-A has shown superior efficacy in reducing facial hyper-contractions during both spontaneous and rehabilitation-induced recovery of facial movements. Therefore, the effectiveness of BTX-A may be enhanced by integrating NMRT [20,45,46].

The combination of NMRT and BTX-A, specifically targeted at the affected synkinetic regions, has proven to be more effective than using NMRT alone [20]. A previous review indicated that NMRT and BTX-A work synergistically to treat synkinesis [41]. Our earlier research confirmed that combining NMRT and BTX-A is advantageous for patients with mild facial synkinesis localized to the oral-ocular region who also have a high score on the Sunnybrook Facial Grading System [19].

In most patients with facial synkinesis, the initial treatment typically involves BTX-A injections and/or NMRT. However, for those with severe facial synkinesis that does not respond to BTX-A and/or NMRT, selective neurectomy may be considered as an alternative [47-49]. The primary goals of selective neurectomy are to create a natural smile and eliminate synkinetic movements. It is important to note that surgical outcomes may vary based on the surgeon's experience and the patient's preferences.

Selective neurectomy is effective in eliminating synkinetic movements, but it requires careful execution as it is an irreversible surgical procedure. The most important consideration is identifying which branch of the facial nerve should be removed. Performing excessive neurectomy can compromise facial expression, while insufficient removal may result in persistent synkinetic movements. Furthermore, to prevent recurrence due to axonal sprouting or regeneration post-surgery, the neurectomy site is reinforced by applying hemoclips in two to three layers at the proximal and distal cut ends of the nerve branches [23].

Surgical intervention can yield gratifying results in alleviating facial tightness and abnormal eyelid constriction. However, for individuals seeking to improve restricted mouth mobility, the approach should be customized based on the patient's preference for correction. While the vertical tilt of the mouth corner can see substantial improvement, the enhancement of horizontal angles may be less than optimal.

Conclusion

It is well established that the most critical factor for patients with acute facial palsy is the timely administration of steroids and antiviral agents. However, patients with relatively severe facial palsy, as indicated by an ENoG degenerative rate exceeding 80% or a House-Brackmann grade higher than IV, are likely to develop synkinesis despite appropriate initial therapy. Therefore, it is crucial to suppress the intensification of synkinesis and provide re-education through early NMRT in patients predicted to develop synkinesis. In cases where synkinesis has already strongly formed, it is recommended to initiate treatment with a combination of BTX-A and NMRT. If synkinesis is so severe that it significantly impacts daily life, or if there is no significant improvement with BTX-A or NMRT, selective neurectomy can be considered as a viable surgical alternative.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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ORCID

Jin Kim, https://orcid.org/0000-0001-6634-7750

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CASE REPORT

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A Rare Case of Radiculoplexopathy Induced by Herpes Zoster

Dong Chan Kim, Jihye Park

Department of Rehabilitation Medicine, Eunpyeong St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea

This case report discusses a rare instance of herpes zoster infection presenting with radiculoplexopathy, exhibiting both preganglionic and postganglionic features. A 62-year-old male patient experienced sudden right upper limb weakness following a recent herpes zoster diagnosis. A nerve conduction study, electromyography, and magnetic resonance imaging revealed a right brachial plexopathy involving the upper trunk and posterior cord, accompanied by lesions in the middle to lower cervical roots. Even after several months, the patient's recovery remained limited, indicating the significance of this case as a reference for clinical progression in instances of both preganglionic and postganglionic neuropathy. This case emphasizes the role of electrophysiological evaluation in zoster-associated paresis to predict disease progression and improve patient management.

Keywords: Herpes zoster; Brachial plexus neuropathies; Radiculopathy

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Corresponding author:
Jihye Park
Department of Rehabilitation Medicine, Eunpyeong St. Mary's Hospital, College of Medicine, The Catholic University of Korea, 1021 Tongil-ro, Eunpyeong-gu, Seoul 03312, Korea
Tel: +82-2-2030-2875
Fax: +82-2-2030-2876
E-mail: sophia@catholic.ac.kr

Introduction

Herpes zoster is caused by varicella-zoster virus (VZV), which colonizes the dorsal root ganglion after infection and reactivates when the patient is in immunocompromised state. The most common neurological symptom after a herpes zoster infection is neuropathic pain, which is known as postherpetic neuralgia; however, it can occasionally lead to muscle weakness or paralysis. Previous research has identified plexopathy as the most common cause of limb paresis associated with herpes zoster, with less frequent instances of radiculopathy and mononeuropathy [1]. This report details a unique case of a patient exhibiting both brachial plexopathy and cervical radiculopathy due to herpes zoster infection, thereby demonstrating a combination of preganglionic and postganglionic manifestations.

Case Report

A 62-year-old man presented to a neurosurgery outpatient clinic, reporting sudden-onset right upper limb weakness that had occurred 2 months ago. One week before the onset of weakness, he had been diagnosed with herpes zoster at a local clinic and was subsequently prescribed antiviral medication and topical ointment. His medical history included hypertension and hyperlipidemia.

A neurological examination of the patient's right upper limb, conducted using the Medical Research Council (MRC) scale, revealed muscle weakness graded as 1/5 in shoulder flexion, 2/5 in elbow flexion, and 4/5 in wrist extension. The patient reported paresthesia in his right thumb and index finger, with pin-pricking pain rated as 4 on a numerical rating scale. Initially, eschars were observed on the right lateral upper arm and lateral forearm, which were fully healed 2 months after the onset. To determine the cause of the right upper limb paresis, a comprehensive diagnostic plan was formulated, including nerve conduction study (NCS), electromyography (EMG), computed tomography (CT) of the cervical spine, and magnetic resonance imaging (MRI) of the brachial plexus.

NCS and EMG were performed 2 months following the onset of weakness. Sensory NCS showed reduced amplitudes of sensory nerve action potentials in right lateral antebrachial cutaneous and radial nerves (Table 1). Motor NCS showed reduced amplitudes of compound muscle action potentials in right radial nerve recording at the extensor indicis proprius muscle, right axillary nerve recording at the deltoid muscle, and right musculocutaneous nerve recording at the biceps brachii muscle.

Needle EMG showed abnormal spontaneous activities at rest in the right C4-T2 paraspinalis, supraspinatus, infraspinatus, teres major, deltoid, biceps, extensor digitorum communis, and flexor carpi radialis muscles (Table 2). However, the right extensor indicis proprius muscle showed normal findings. Single interference patterns were shown on maximal volition in the right biceps muscle, while no motor unit action potentials were observed in the right supraspinatus, infraspinatus, teres major, and deltoid muscles. Consequently, the electrodiagnostic findings were consistent with a diagnosis of right brachial plexopathy, predominantly affecting the upper trunk and the posterior cord, and appeared to be associated with lesions in the middle to lower cervical roots.

After performing NCS and EMG, a radiologic evaluation was executed. Cervical spine CT showed mild bulging discs at C3-4 and C5-6, but without root compression. Brachial plexus MRI

Table 1. Results of Nerve Conduction Studies

Study	Nerve	Latency (ms)	Amplitude	Velocity (m/sec)
Sensory nerve conduction (R/L)	Median	2.4/2.6	18.1/11.5	
	Ulnar	2.5/2.6	11.6/13.7	
	Radial	1.6/1.6	4.1*/15.0	
	MABC	1.2/1.4	9.9/9.4	
	LABC	1.5/1.6	5.8*/17.9	
Motor nerve conduction (R/L)	Median: wrist-elbow	3.4/3.4	6.3/7.7	56/60
	Ulnar: wrist-elbow	2.8/2.7	6.3/9.5	59/59
	Radial: forearm-spiral groove	2.5/2.2	1.9*/3.5	61/65
	Axillary: Erb's point	3.8/3.1	0.1*/12.0	
	MCN: Erb's point	4.6/4.6	0.4*/7.0	

Amplitudes were measured in millivolts for motor nerves and microvolts for sensory nerves.
R, right; L, left; MABC, medial antebrachial cutaneous; LABC, lateral antebrachial cutaneous; MCN, musculocutaneous.
*Abnormal findings are represented with asterisks; an abnormal finding was defined by a greater than 50% reduction of amplitude or 30% delay of latency compared to the unaffected side, or no response of sensory nerve action potential and compound motor action potential.

Table 2. Results of Needle Electromyography

Muscle	Spontaneous activities	MUAP	Recruitment pattern	Interference pattern
C4-T2 PSP L	Nml			
C4-6 PSP R	Fib 1+, PSW 1+			
C6-T2 PSP R	Fib 1+, PSW 2+			
SSP R	Fib 2+, PSW 3+			No MUAP
ISP R	Fib 2+, PSW 3+			No MUAP
RM R	Nml	Nml	Nml	Nml
TM R	Fib 1+, PSW 1+			No MUAP
SA R	Nml	Nml	Nml	Nml
Deltoid R	Fib 1+, PSW 2+			No MUAP
Biceps R	Fib 2+, PSW 2+	Nml		Single
Triceps R	Nml	Nml	Nml	Reduced
EDC R	PSW 1+	Long, polyphasic	Nml	Nml
EIP R	Nml	Nml	Nml	Nml
FCR R	PSW 1+	Nml	Nml	Reduced
FCU R	Nml	Nml	Nml	Nml
FDI R	Nml	Nml	Nml	Nml
APB R	Nml	Nml	Nml	Nml

MUAP, motor unit action potential; PSP, paraspinalis; L, left; Nml, normal; R, right; Fib, fibrillation potential; PSW, positive sharp wave; SSP, supraspinatus; ISP, infraspinatus; RM, rhomboid major; TM, teres major; SA, serratus anterior; EDC, extensor digitorum communis; EIP, extensor indicis proprius; FCR, flexor carpi radialis; FCU, flexor carpi ulnaris; FDI, first dorsal interosseous; APB, abductor pollicis brevis.

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showed edema and enhancement of the right C5 and C6 nerve roots, as well as edema of the upper trunk and the posterior cord of the right brachial plexus, compared to the left side (Fig. 1), corresponding to the localization of lesions indicated in NCS and EMG.

Following an electrophysiological diagnosis of right radiculoplexopathy, the patient was prescribed gabapentin and amitriptyline to relieve sensory symptoms, and he underwent rehabilitation for his right arm, which included strengthening exercises and electrical stimulation therapy targeting shoulder flexors and abductors. Four months after the onset of weakness, he revisited the outpatient clinic for evaluation. At this time, his right upper limb muscle strength exhibited a slight improvement in shoulder flexion with the MRC scale of 2/5, but the MRC scale for elbow flexion remained unchanged at 2/5. The paresthesia in the right thumb persisted. Seven months after the onset of weakness, the patient showed no significant improvement in muscle strength of the right upper limb.

Written informed consent by the patient was waived due to a retrospective nature of our study.

Discussion

Approximately 5% to 30% of patients with herpes zoster may develop motor weakness, usually within the first 2 weeks following the skin eruption [2]. Notably, motor nerve involvement may not always align with the same dermatome of the rash [3]. Li and Feng [4] have identified age, pain severity, and involvement of C6 or C7 nerve roots as potential risk factors for zoster paresis. Conversely, Tang et al. [5] have argued that age and pain severity are not significant factors, instead suggesting that upper limb involvement and comorbidities are more critical risk factors.

Zoster-induced radiculoplexopathy is rare, with only a few cases reported in the literature [1,6]. Liu et al. [1] conducted retrospective review of 1,393 patients infected with herpes zoster and identified eight cases of zoster-associated paresis. Among these, two cases categorized as radiculopathy exhibited favorable prognoses, achieving full motor recovery within 3 months without treatment, while the other two cases categorized as plexopathies and three cases categorized as radiculoplexopathies demonstrated poor prognoses, varying from partial recovery after 1 year to no motor recovery observed even after 2 years [1].

Jones et al. [6] reviewed 49 cases of zoster-induced paresis, providing a comparative analysis of preganglionic and postganglionic lesions localized by electrophysiological evaluation. The findings revealed that postganglionic lesions were associated with a greater severity of weakness than preganglionic lesions [6]. The mean minimum duration of weakness was 166.3 days in preganglionic lesions and 210.3 days in postganglionic lesions [6].

Castro et al. [7] reviewed 19 cases of zoster-induced paresis, in

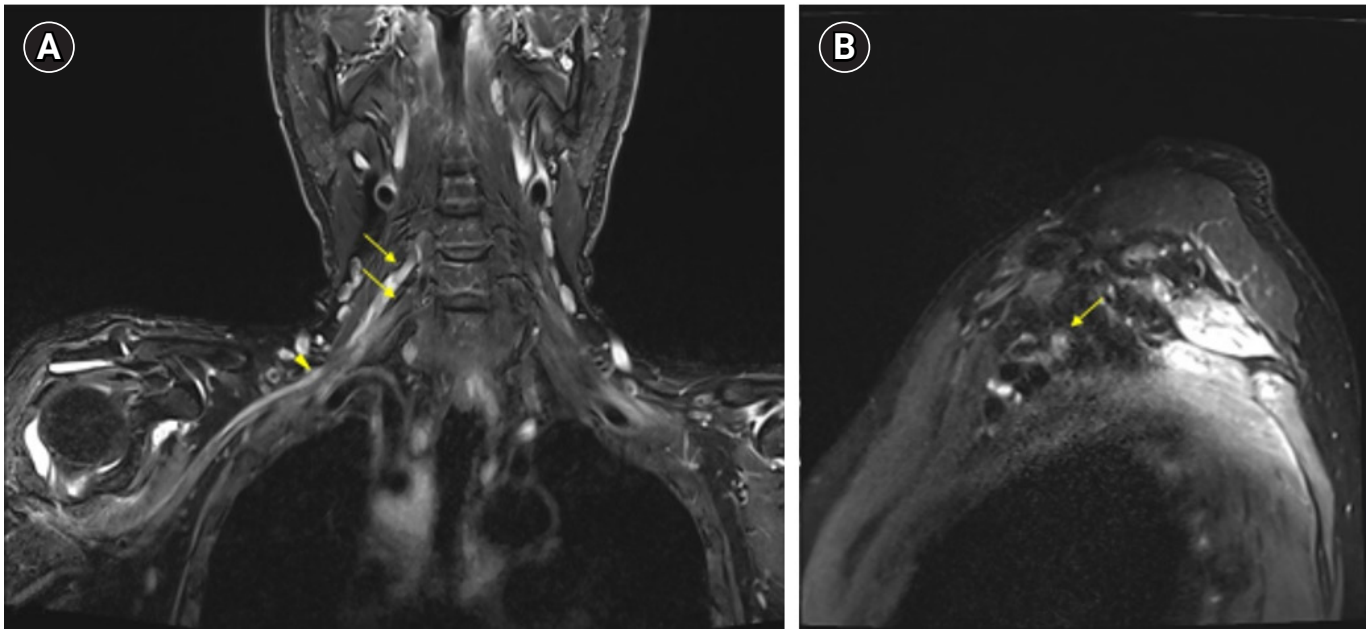

Fig. 1. Contrast-enhanced coronal T2-weighted brachial plexus magnetic resonance imaging. (A) Edema and enhancement of right C5 and C6 nerve roots (arrows) and edema of the upper trunk of the brachial plexus (arrowhead). (B) Edema at the right posterior cord level of the brachial plexus (arrow).

which six patients reported partial recovery, while other patients reported full recovery. Excluding two cases of abdomen involvement, the case reports reporting partial recovery exhibited reduced amplitude or a failure to evoke the sensory nerve action potential, implying postganglionic involvement. The longest time to evaluate partial recovery in a postganglionic lesion case was 2 years. Consequently, zoster-induced paresis tends to exhibit more severe muscle weakness and a poorer prognosis in instances involving postganglionic lesions than preganglionic lesions.

This tendency is evident not only in cases of zoster infection but also in neuropathies with other causes. Radiculopathy, which is defined as compression or irritation of nerve roots from the spine, generally has a more favorable prognosis than plexopathy. Previous studies have shown that functional outcomes in patients with radiculopathy, including chronic conditions, improve with appropriate treatment [8]. In contrast, plexopathy, which is defined as damage to brachial or lumbosacral plexuses, requires more caution. The etiology is variable, potentially including trauma, neoplasm, or radiation; therefore, its pluralistic nature requires a more detailed history, along with more complex imaging, and electrodiagnostic studies [9]. Extensive nerve involvement complicates the diagnosis, leading to delayed decision-making regarding treatment.

In this case, initial clinical presentation suggested radiculopathy; however, subsequent findings revealed additional postganglionic involvement. Although the patient exhibited a 1-grade improvement in shoulder flexion on the MRC scale 4 months post-onset, no further changes in the MRC scale were observed 7 months after onset. This outcome aligns with previous research documenting that a poor prognosis is associated with postganglionic involvement [1].

While there is a lack of studies directly comparing instances of zoster-induced radiculoplexopathy and plexopathy in isolation, it is presumed that concurrent preganglionic and postganglionic involvement suggests a poor prognosis. This assumption is based on the pathophysiology of VZV, which spreads from the dorsal root ganglion to distal nerves [10]. Further investigation is warranted to comprehensively compare concurrent and singular lesions in zoster-induced paresis, considering the limitation of a restricted number of available cases.

The current study highlights a rare case of zoster-induced radiculoplexopathy, emphasizing its long-term progression. Differentiating the lesion locations in zoster-induced neuropathy can assist clinicians in predicting patient outcomes. Therefore, we recommend that clinicians conduct NCS and EMG in patients presenting with zoster-induced paresis to better anticipate disease progression.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

ORCID

Dong Chan Kim, https://orcid.org/0009-0007-7808-8259
Jihye Park, https://orcid.org/0000-0002-9764-128X

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CASE REPORT

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Compression Neuropathy Caused by Pelvic Lymphocele after Laparoscopic Surgical Staging

Dong Jin Chae, Jong Bum Park, Mi Jin Hong, Jungyun Kim, Cho E. Sim, Seung-Eun Kim, Yung Jin Lee

Department of Rehabilitation Medicine, Konyang University Hospital, Konyang University College of Medicine, Daejeon, Korea

Lymphocele is a complication of pelvic surgery that infrequently leads to compressive neuropathy. We present a case of compressive obturator neuropathy resulting from lymphocele development after pelvic surgery. Electrodiagnostic studies revealed severe axonal disruption in the left obturator nerve, which is associated with poor functional recovery. This case underscores the role of electrodiagnostic testing in the diagnosis and rehabilitation of patients experiencing lower limb weakness following gynecological pelvic surgery.

Keywords: Lymphocele; Obturator nerve; Nerve compression syndromes

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Corresponding author:
Yung Jin Lee
Department of Rehabilitation Medicine, Konyang University College of Medicine, 158 Gwanjeodong-ro, Seo-gu, Daejeon 35365, Korea
Tel: +82-42-600-2181
Fax: +82-42-612-2034
E-mail: eutravel@kyuh.ac.kr

Introduction

Lymphocele, defined as a collection of lymphatic fluid encased by thin fibrous walls [1], is a known complication following pelvic surgery, with an incidence rate between 25% and 61% [2]. While it is typically asymptomatic, symptoms can manifest in about 3% to 6% of patients, leading to a range of complications [1]. A limited number of reports have described lower extremity weakness resulting from compressive neuropathy after pelvic surgery.

Previous studies have reported that the incidence of neuropathy following gynecological pelvic surgery ranges from 1.9% to 3.2% [3,4]. Lower extremity weakness after prolonged pelvic surgery is often caused by neuropathy, primarily due to sustained nerve compression. This compression can result from incorrect patient positioning or the prolonged or inappropriate use of surgical retractors, and it can meaningfully affect patient functioning and quality of life by causing lower limb impairment [4]. Additionally, neuropathy may arise from other causes, such as intraoperative nerve damage or hematoma.

This report details the case of a patient who developed obturator neuropathy caused by pelvic lymphocele following laparoscopic staging surgery for ovarian cancer. This condition was diagnosed with an electrodiagnostic examination.

Case Report

A 52-year-old woman was referred to the Department of Rehabilitation Medicine due to weakness in her left leg and gait disturbance that had persisted for 2 weeks following pelvic surgery. She had been diagnosed with ovarian cancer and had undergone staging laparoscopy, which included total laparoscopic hysterectomy, bilateral salpingo-oophorectomy, pelvic lymph node dissection, and omentectomy. Four days after surgery, she began experiencing difficulty lifting her left leg, inguinal stiffness, and discomfort when walking.

Physical examination revealed muscle strength of Medical Research Council (MRC) grade 2 in the left hip flexor and adductor muscles and grade 3 in the knee extensor muscles. All muscles of the right lower limb were normal. The patient also report-

ed hypoesthesia in the left medial thigh. Deep tendon reflexes, including knee and ankle jerks, were normoactive.

Needle electromyography (EMG) (Table 1) and nerve conduction studies (NCS) (Table 2) were performed 2 weeks after the onset of symptoms. NCS of the femoral nerve displayed a typical response. However, EMG of the left adductor longus muscle demonstrated abnormal spontaneous activity and decreased voluntary recruitment of motor unit action potentials (MUAPs), indicative of left obturator neuropathy.

In addition to electrodiagnostic examination, the patient underwent pelvic magnetic resonance imaging (MRI) (Fig. 1). This imaging revealed a lobulated cystic lesion of approximately 3.2 × 3.2 × 4.2 cm at the left pelvic wall, suggestive of lymphocele. The potential for associated obturator nerve injury could not be excluded. Instead of opting for drainage, conservative treatment (including rehabilitation therapy) was considered for the patient, given the possibility of spontaneous absorption.

Thus, we conducted a comprehensive rehabilitation therapy

Table 1. Needle Electromyography

Side	Muscle	ASA		Motor unit potentials			Interfer.
		Fibs	PSW	Polyphasia	Amplitude	Duration	
Left	Adductor longus	–	+	–	N	N	STP
	Iliopsoas	–	–	–	N	N	Normal
	Vastus lateralis	–	–	–	N	N	Normal
	Gluteus medius	–	–	–	N	N	Normal
	Gluteus maximus	–	–	–	N	N	Normal
	Biceps femoris	–	–	–	N	N	Normal
	Tibialis anterior	–	–	–	N	N	Normal
	Gastrocnemius	–	–	–	N	N	Normal
	L3 PSP	–	–				
	L4 PSP	–	–				
	L5 PSP	–	–				
	S1 PSP	–	–				

ASA, abnormal spontaneous activity; Fibs, fibrillation potentials; PSW, positive sharp wave; Interfer., interference pattern; N, normal; STP, single to partial.

Table 2. Nerve Conduction Studies

	Stimulation	Latency (ms)*	Amplitude	CV (m/sec)
Sensory nerve				
Rt. superficial peroneal (foot)	Calf	3.23	19	53.8
Rt. sural (lat. malleolus)	Calf	3.39	15.3	52.7
Rt. lat. femoral cutaneous	Thigh	2.19	16.8	81.2
Lt. superficial peroneal (foot)	Calf	3.33	20.4	52.7
Lt. sural (lat. malleolus)	Calf	3.39	20.1	49.8
Lt. lat. femoral cutaneous	Thigh	2.19	16.8	68.1
Motor nerve				
Rt. common peroneal (EDB)	Ankle	3.13	4.3	
	Fibular head	8.39	4.3	47.5
	Knee	10	4	52.6
Rt. tibial (AH)	Ankle	3.07	27.2	
	Knee	9.53	21.9	48.8
Rt. femoral (VM)	Inguinal canal	3.23	10.4	
Lt. common peroneal (EDB)	Ankle	2.81	3.7	
	Fibular head	8.18	3.7	48.5
	Knee	9.43	3.6	64
Lt. tibial (AH)	Ankle	2.97	22	
	Knee	9.64	21.9	49.5
Lt. femoral (VM)	Inguinal canal	3.65	8.5	

Amplitudes are measured in microvolts (μV, sensory) and millivolts (mV, motor).
CV, conduction velocity; Rt., right; lat., lateral; Lt., left; EDB, extensor digitorum brevis; AH, abductor hallucis; VM, vastus medialis.
*Sensory nerve: peak latency; motor nerve: onset latency.

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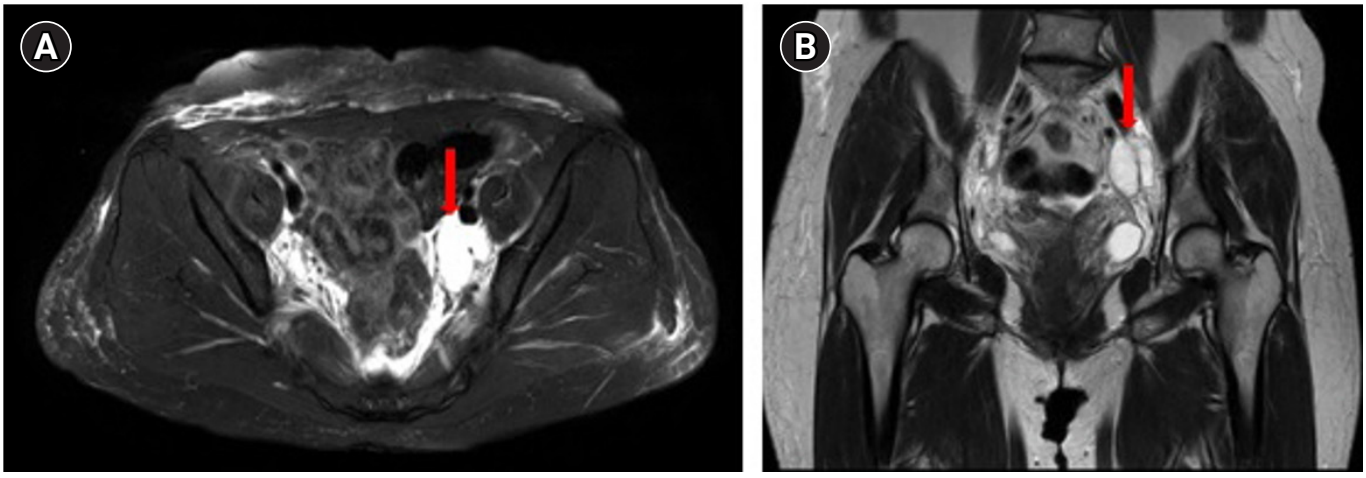

Fig. 1. Axial and coronal views of pelvic magnetic resonance imaging show a 3.2 × 3.2 × 4.2 cm lobulated cystic lesion, suggestive of lymphocele (indicated by the arrows), at the left pelvic wall. (A) Suggestive lymphocele from axial view and (B) suggestive lymphocele from coronal view.

program that included hip range-of-motion exercises, progressive resistance exercises, gait training, and neuromuscular electrical stimulation.

Electrodiagnostic evaluation, performed 3 months later (Tables 3, 4), revealed an improvement in the MRC scale from 2 to 4. Increased recruitment of MUAPs was confirmed on needle EMG examination. Four months later, improvements were noted on abdominopelvic computed tomography (CT) (Fig. 2). Although complete recovery had not been achieved, the improving trend led us to continue conservative treatment, including rehabilitation therapy, while monitoring progress.

Written informed consent was obtained from the patient. This study was approved by the Institutional Review Board of Konyang University Hospital Hospital (IRB no: 2023-02-020-001).

Discussion

One notable complication of pelvic surgery is the formation of lymphoceles, which are collections of lymphatic fluid surrounded by thin fibrous walls. Pelvic lymphoceles can develop after surgery when lymphatic vessels are damaged due to truncation or improper ligation [1]. The incidence of symptomatic lymphocele (3% to 6%) is lower than that of the asymptomatic condition (13% to 20%). Symptoms primarily result from pressure on adjacent anatomical structures, leading to pain, swelling, numbness, or deep venous thrombosis [1]. Treatment is indicated based on the size or symptoms of lymphocele, and various methods have been described, including needle aspiration, percutaneous catheter drainage with or without sclerotherapy, and surgical drainage [5]. In a previous study, conservative management, including mechanical compression and a low-fat diet, was shown to be effective against swelling [6]. Consequently, we employed simple physical therapy and range-of-motion exercises to increase lymphatic flow, reduce swelling, and improve MRC grade.

The obturator nerve arises from the anterior division of the L2, L3, and L4 spinal nerve roots. It descends within the psoas major muscle, emerges from its medial border, and runs along the lateral wall of the lesser pelvis. The nerve then enters the thigh via the obturator canal, where its anterior branch innervates the muscles of the medial thigh. Obturator mononeuropathy can lead to adductor weakness and cutaneous sensory deficits in the medial thigh [7].

This report presents a case of obturator mononeuropathy caused by lymphocele formation after pelvic surgery. According to prior research, neuropathy following pelvic surgery occurs in about 1.9% to 3.2% of cases [3,4]. Obturator mononeuropathy is relatively rare due to the nerve's protected location within the pelvis and medial thigh [8]. Previous studies have estimated the incidence of neuropathy after pelvic surgery to be around 2%, with the primary causes being improper patient positioning and retractor usage. Other nerves frequently affected in such cases include the ilioinguinal/iliohypogastric nerve, genitofemoral nerve, femoral nerve, and lumbosacral nerve plexus [9].

We diagnosed obturator mononeuropathy using electrodiagnostic evaluation. To differentiate it from conditions such as peripheral neuropathy or lumbar plexopathy, we performed needle EMG on the adductor muscle groups. The obturator nerve descends through the fibers of the psoas major, complicating nerve conduction testing. For an accurate diagnosis of obturator nerve injury, it is necessary to examine both the adductor magnus mus-

Table 3. Needle Electromyography after 3 Months

Side	Muscle	ASA		Motor unit potentials			Interfer.
		Fibs	PSW	Polyphasia	Amplitude	Duration	
Left	Adductor longus	+	+	–	N	N	Partial
	Iliopsoas	–	–	–	N	N	Normal
	Vastus lateralis	–	–	–	N	N	Normal
	Gluteus medius	–	–	–	N	N	Normal
	Gluteus maximus	–	–	–	N	N	Normal
	Biceps femoris	–	–	–	N	N	Normal
	Tibialis anterior	–	–	–	N	N	Normal
	Gastrocnemius	–	–	–	N	N	Normal
	L3 PSP	–	–				
	L4 PSP	–	–				
	L5 PSP	–	–				
	S1 PSP	–	–				

ASA, abnormal spontaneous activity; Fibs, fibrillation potentials; PSW, positive sharp wave; Interfer., interference pattern; N, normal.

Table 4. Nerve Conduction Studies after 3 Months

	Stimulation	Latency (ms)*	Amplitude	CV (m/sec)
Sensory nerve				
Lt. superficial peroneal (foot)	Calf	3.23	15.8	54.9
Lt. sural (lat. malleolus)	Calf	3.49	23.7	51.7
Lt. lat. femoral cutaneous	Thigh	2.5	15.2	67.8
Motor nerve				
Lt. common peroneal (EDB)	Ankle	3.07	3.6	
	Fibular head	8.54	3.4	48.5
	Knee	10.1	3.4	51.2
Lt. tibial (AH)	Ankle	2.86	21	
	Knee	10	17.8	46.2
Lt. femoral (VM)	Inguinal canal	3.13	6.2	

Amplitudes are measured in microvolts (μV, sensory) and millivolts (mV, motor).
CV, conduction velocity; Lt., left; lat., lateral; EDB, extensor digitorum brevis; AH, abductor hallucis; VM, vastus medialis.
*Sensory nerve: peak latency; motor nerve: onset latency.

cle, which is innervated by the posterior branch, and one of the muscles innervated by the anterior branch—either the adductor longus, brevis, or gracilis. However, one limitation of this case was that only the adductor longus muscle was examined. The test results strongly suggested axonotmesis of the obturator nerve, which was consistent with the symptoms exhibited by the patient.

The prognosis for acute onset compressive neuropathy is generally favorable, according to a limited number of studies. A case series suggested that most patients with acute onset recovered well following conservative treatment, regardless of cause or the severity of EMG findings [8]. For asymptomatic lymphocele, spontaneous regression is commonly observed, and the condition can be monitored over time; however, this process may take several months [10]. In the case presented here, the patient did not achieve full recovery despite conservative management of symptomatic lymphocele, even though follow-up pelvic CT indicated that the lymphocele had resolved. This raises the question of whether early intervention or aspiration surgery might have led to complete recovery. Additionally, determining the optimal timing for invasive intervention, based on the condition's progression over time, could improve the prognosis in future cases.

Postoperative nerve compression syndrome has various causes, including hematoma and direct nerve injury. In the present case, imaging and NCS identified lymphocele as the cause of compression neuropathy. Lymphocele is a condition commonly discussed in the contexts of cancer and lymphatic rehabilitation, but it is relatively rare in the field of physical medicine and rehabilitation. Although most cases are asymptomatic and managed conservatively, persistent symptoms may necessitate interventions such as percutaneous catheter drainage or surgical removal, with success rates of 79% to 82% in previous research [10]. This case report de-

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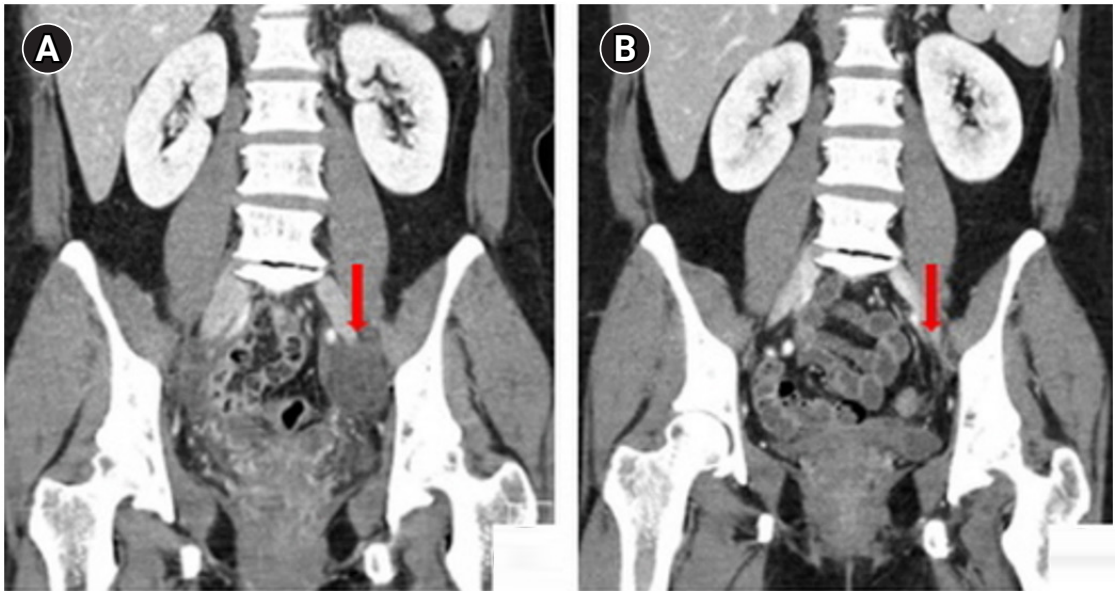

Fig. 2. Abdominopelvic computed tomography scans. (A) Lymphocele (indicated by the arrow) at diagnosis. (B) After 4 months, we could confirm that almost the entire lymphocele (arrow) had been naturally absorbed.

scribes a patient with obturator nerve entrapment syndrome caused by lymphocele, who demonstrated functional improvement through conservative management and rehabilitation. The report also includes a review of the literature on lymphocele.

In conclusion, our findings emphasize the importance of considering lymphocele in patients who present with lower limb weakness following pelvic surgery. Various evaluations, including electrodiagnostic testing and MRI, should be combined to obtain an accurate diagnosis and establish an appropriate rehabilitation plan that facilitates a successful return to daily activities.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

ORCID

Dong Jin Chae, https://orcid.org/0000-0003-4258-5572
Jong Bum Park, https://orcid.org/0000-0002-0087-845X
Mi Jin Hong, https://orcid.org/0000-0002-4170-0762
Jungyun Kim, https://orcid.org/0000-0003-3439-9075
Cho E. Sim, https://orcid.org/0009-0008-2842-5330
Seung-Eun Kim, https://orcid.org/0009-0001-0019-4154
Yung Jin Lee, https://orcid.org/0000-0003-4479-9136

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CASE REPORT

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Diagnosis of *ADSSL1* Mutation-Induced Myopathy through Electrophysiology and Genetic Tools

Dong Jin Chae, Yung Jin Lee, Mi Jin Hong, Cho E. Sim, Seung-Eun Kim, Jong Bum Park

Department of Rehabilitation Medicine, Konyang University Hospital, Konyang University College of Medicine, Daejeon, Korea

Mutations in the adenylosuccinate synthase 1 (*ADSSL1*) gene, resulting in adenylosuccinate synthase deficiency, are a rare genetic anomaly characterized by muscular weakness, elevated serum creatine kinase levels, and pathological muscle findings. However, these clinical symptoms are similar to those observed in many other myopathies, increasing the risk of misdiagnosis. In an era of rapidly expanding genetic knowledge, the authors sought to verify the diagnostic utility of electromyography for genetic disorders. Through combined electrophysiological and genetic studies, a patient initially thought to have Becker's muscular dystrophy was conclusively diagnosed with *ADSSL1* mutagenic myopathy. This case underscores the importance of re-evaluating diseases that do not follow the typical clinical progression of traditional myopathies, especially in light of recent diagnostic advancements.

Keywords: ADSSL1; Mutation; Diagnostic errors; Electromyography; Genetic diseases

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Corresponding author:
Jong Bum Park
Department of Rehabilitation Medicine, Konyang University Hospital, Konyang University College of Medicine, 158 Gwanjeodong-ro, Seo-gu, Daejeon 35365, Korea
Tel: +82-42-612-2180
Fax: +82-42-612-2034
E-mail: jbocean@kyuh.ac.kr

Introduction

Mutations in adenylosuccinate synthase 1 (*ADSSL1*) can cause myopathy, which is characterized by symptoms that typically begin in adolescence. These symptoms include primarily distal muscle involvement, mild facial muscle weakness, and a slight increase in serum creatine kinase (CK) levels. This form of distal myopathy was first identified in Korea through a study using exome sequencing conducted in 2015 [1]. Globally, there are fewer than 200 known cases [2].

Distal myopathies represent a diverse group of rare, progressive hereditary muscle disorders that predominantly affect the muscles of the distal limbs [1]. To date, researchers have identified more than 16 genes responsible for distal myopathy, with the majority linked to autosomal dominant (AD) inheritance patterns [1]. Recently, the *ADSSL1* gene has been implicated in this condition. ADSSL1 encodes the muscle-specific enzyme adenylosuccinate synthetase like 1, which is crucial for purine nucleotide interconversion. This enzyme catalyzes the initial step in the de novo synthesis of adenosine [3].

Here, we describe a case of *ADSSL1* mutagenic myopathy that was misdiagnosed and initially treated as Becker muscular dystrophy (BMD). Furthermore, we discuss the importance of re-evaluating diseases that deviate from the typical clinical course of traditional myopathies, in light of recent advances in diagnostic techniques.

Case Report

A 39-year-old man presented at the Department of Rehabilitation Medicine for an evaluation of progressive symptoms, which included balance issues during standing and walking, as well as weakness in the distal extremities. At the age of 20, he sought treatment at another tertiary hospital for facial weakness and poor physical performance, notably difficulty in running. He had no issues with swallowing, pulmonary function, or cardiac

health. A physical examination, conducted using the Medical Research Council (MRC) score, revealed a muscle strength grade of approximately 3 to 4 in both the upper and lower extremities.

Laboratory findings from 2002 indicated hyperCKemia, with a CK level of 1,707 IU/L (reference range, 0 to 170). A muscle biopsy was conducted, and hematoxylin-eosin staining showed increased nuclei internalization, hypertrophic fibers, degenerating fibers, myophagocytosis, and fiber splitting. Immunohistochemistry revealed dystrophin expression along the sarcolemma. The dystrophin gene deletion test showed no exon deletions. Due to the limitations of genetic testing at the time, it was not possible to identify other genetic disorders that could present with these muscular symptoms. The presence of dystrophin, distinguishing the condition from Duchenne type, led to a presumptive diagnosis of BMD [4]. In his late 20s, as his symptoms worsened, including difficulties in grasping and pinching with his hands, he found it challenging to perform everyday activities.

At the time of the patient's visit to our hospital, his cognitive function and other medical conditions remained unchanged. Bilateral shoulder abduction, elbow flexion, and wrist extension had a score of 3/3; bilateral finger flexion and extension scored 2/2; bilateral hip flexion and knee extension had a score of 3/3; and bilateral ankle dorsiflexion and plantar flexion scored 2/2, according to the MRC scale. The deep tendon reflexes in the bilateral ankle jerks were hypoactive. He continued to have no issues with swallowing, pulmonary function, or cardiac health, and his family history was unremarkable (Fig. 1).

Despite the initial diagnosis, the clinical course and current neurological state suggested that a presumptive diagnosis of BMD was regarded as inappropriate. The primarily affected muscles exhibited distal differences, which contrasted with the typical manifestations of BMD. Consequently, we conducted nerve conduction studies and electromyography (EMG), including quantitative EMG. The nerve conduction study was normal, except for a reduced amplitude of the compound motor action potential in the bilateral extensor digitorum brevis muscles (Tables 1, 2). The EMG findings indicated myopathic patterns predominantly in the distal limbs (Tables 1, 2, Fig. 2).

Based on the patient's family history, facial weakness, prominent distal muscle atrophy, and the identification of myopathic patterns in the distal muscles on EMG, which were compatible with distal myopathy, we requested genetic testing. Diagnostic exome sequencing revealed pathogenic variants of *ADSSL1*—specifically. c.910G > A and c.1048 del mutations, with minor allele frequencies of 0.0034% and 0.0081%, respectively, in the human population (Table 3). In light of the diagnosis of *ADSSL1* mutagenic myopathy, we provided genetic counseling for myopathy of autosomal recessive (AR) inheritance, which is different from AD or sex-linked inheritance.

The requirement for written informed consent was waived because it was impractical to obtain, refusal was unlikely, and the risk to research subjects was extremely low. The study received approval from the Institutional Review Board of Konyang University Hospital (IRB no: 2023-03-004-004).

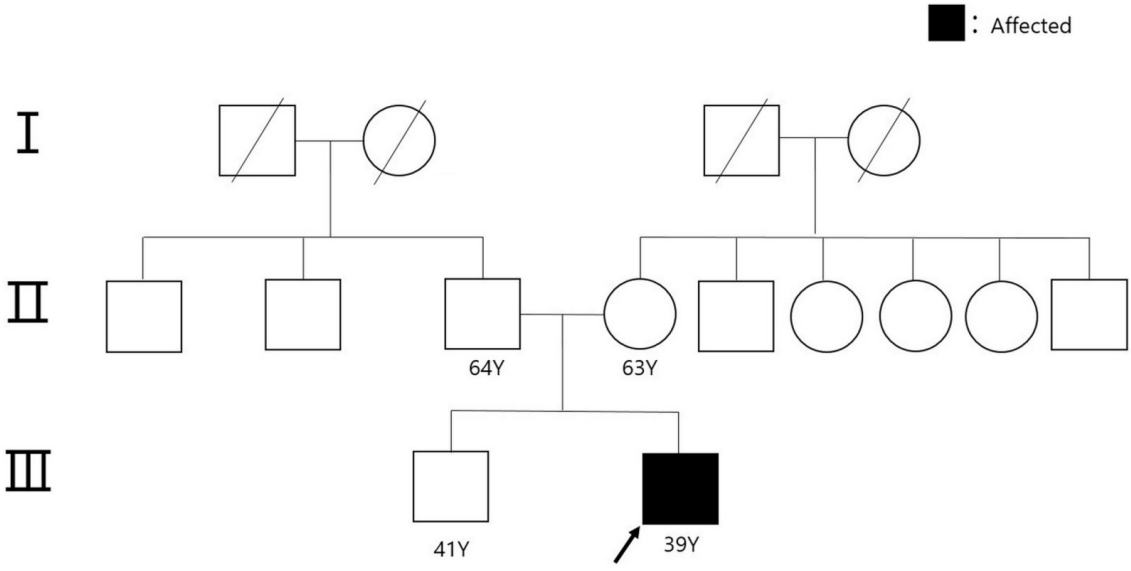

Fig. 1. Pedigree of a patient with adenylosuccinate synthase 1 (*ADSSL1*) gene mutagenic myopathy. The arrow indicates the proband (square: male; circle: female; filled: affected; unfilled: unaffected).

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Table 1. Nerve Conduction Studies

	Stimulation	Latency (ms)	Amplitude	CV (m/sec)
Sensory nerve				
Rt. superficial peroneal	Calf	3.28	6.4	49
Rt. sural (lat. malleolus)	Calf	4.11	22.4	44.9
Lt. superficial peroneal	Calf	3.54	6.7	40.6
Lt. sural (lat. malleolus)	Calf	4.06	15	41.1
Motor nerve				
Rt. common peroneal (EDB)	Ankle	5	0.8*	
	Fibular head	12.29	0.7*	46.6
	Knee	13.49	0.7*	50.1
Rt. tibial (AH)	Ankle	5.52	8.8	
	Knee	13.33	7.3	49.9
Lt. common peroneal (EDB)	Ankle	4.58	0.8*	
	Fibular head	13.02	0.7*	45.6
	Knee	9.43	0.7*	50.1
Lt. tibial (AH)	Ankle	5.42	5	
	Knee	13.49	5	47.1

Amplitudes are measured in microvolts (μV, sensory) and milivolts (mV, motor).
CV, conduction velocity; Rt., right; lat., lateral; Lt., left; EDB, extensor digitorum brevis; AH, abductor halluces.
*Abnormal findings on nerve conduction studies.

Table 2. Needle Electromyography

Side	Muscle	ASA		Motor unit potentials			Recruitment	Interfer.
		Fibs	PSW	Poly	AMP	Duration		
Left	Tibialis anterior	+	–	–	Small	Short	Early	Complete
	Rectus femoris	–	+	+	Small	Short	Normal	Complete
	Gastrocnemius	–	+	+	Small	Short	Early	Complete
	Vastus lateralis	–	–	–	Normal	Normal	Normal	Complete
Right	Tibialis anterior	–	–	–	Small	Short	Early	Complete
	Rectus femoris	–	–	–	Small	Short	Normal	Complete
	Gastrocnemius	–	–	–	Small	Short	Early	Complete
	Vastus lateralis	–	–	+	Normal	Normal	Normal	Complete

ASA, abnormal spontaneous activity; Fibs, fibrillation potentials; PSW, positive sharp wave; Poly, polyphasia; AMP, amplitude; Interfer., interference pattern.

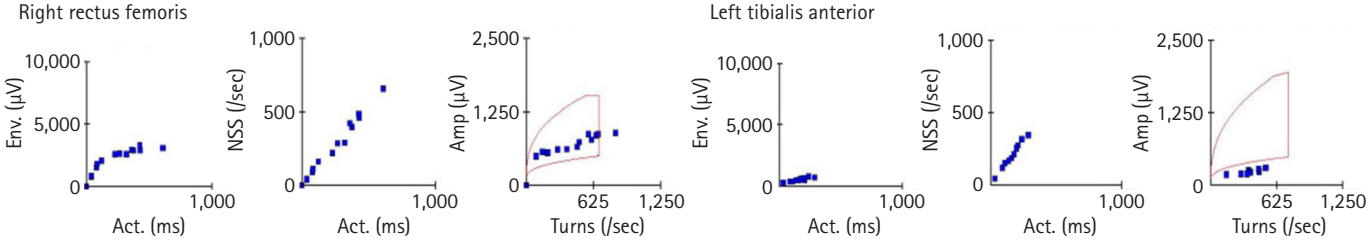

Fig. 2. Quantitative electromyography findings. For the tibialis anterior muscle, the values of turns/amplitude (Amp) data were shifted toward the lower quadrant and outside the zone of normal cloud (red line), showing high turns and small amplitudes, which indicate myopathic patterns. For the rectus femoris muscle, it shows results within the zone of the normal cloud. Env., envelope; Act., activity; NSS, number of small segments.

Discussion

Distal myopathy is a genetic disorder characterized by the progressive loss of muscle tissue. In contrast to proximal dystrophies, which mainly involve defects in sarcolemmal proteins, distal muscular dystrophies are most often caused by mutations in

Table 3. Diagnostic Exome Sequencing Test

Gene	DNA change	Predicted AA change	Zygosity	OMIM disease	Inherit	Class
ADSS1	c.910G>A	p.Asp304Asn	Het	MD5	AR	PV
ADSS1	c.1048del	p.Ile350SerfsTer25	Het	MD5	AR	PV

Reference sequencing: NM_199165.2(ADSS1).
AA, amino acid; OMIM, Online Mendelian Inheritance in Man; *ADSS1*, adenylosuccinate synthase 1; Het, heterozygous; MD5, myopathy distal 5; AR, autosomal recessive; PV, pathogenic variant.

genes that encode sarcomeric proteins [5]. It remains unclear why these specific genetic defects predominantly affect distal muscles. These mutations alter the structural and functional integrity of the sarcomere [5]. Further research is essential to investigate potential therapeutic strategies.

ADSSL1 mutagenic myopathy is an AR distal myopathy, predominantly reported in Asians [6], with an estimated prevalence of less than 0.1 per 1,000,000 population and a currently reported number of cases ranging between 100 and 200 [2]. *ADSSL1*, which is located on chromosome 14q32.33, encodes the ADSSL1 protein, a muscle-specific enzyme crucial for energy metabolism in skeletal muscles, particularly in the muscle and heart [7]. Defects in ADSSL1 disrupt the purine nucleotide cycle, interfering with normal muscle function and leading to progressive muscle weakness and myopathy. Similar to other metabolic and mitochondrial diseases affecting the muscular system, the severity and progression of the disease vary among patients. However, it typically presents in adolescence during periods of rapid growth, when structural stress from stretching across growing bones and demands on protein synthesis and cellular energy are at their peak [2]. From the standpoint of physiatrists, there is a need for more accuracy regarding the methods, intensity, and frequency of rehabilitation therapy for patients with distal myopathy.

Before the widespread adoption of genetic testing in diagnostics, primary tools for diagnosing muscular diseases included blood tests, EMG, and biopsies. These methods helped identify affected areas and differentiate between various myopathies, such as inflammatory myopathy. To diagnose *ADSSL1* mutations, exome sequencing is essential. However, until recently, challenges in genetic testing have led to reports of muscular diseases being misdiagnosed as BMD in cases diagnosed several years ago [8,9].

Previous studies have indicated that both muscle biopsy and EMG are viable methods for diagnosing neuromuscular diseases. EMG, which is widely accessible, provides advanced quantitative assessments, boasts higher sensitivity, and can be applied to various affected areas. However, its major limitation is that it lacks specificity and offers no insights into pathogenesis. In contrast, muscle biopsy, particularly with the advent of electron microscopy, has become highly sensitive but remains significantly invasive [10]. The recent surge in genetic testing has lessened the reliance on these invasive biopsies, which are not only costly but also time-consuming. Nonetheless, the implementation of genetic testing requires careful consideration of both ethical issues and expenses.

An incorrect diagnosis can delay appropriate treatment and potentially cause fatal harm to patients. It is crucial to raise awareness about conditions that are frequently misdiagnosed. Previous studies have shown that patients with myopathy, who were misdiagnosed with BMD, did not undergo genetic or EMG testing at the time of their diagnosis [8,9]. Clinical courses and subsequent reevaluations, which included genetic and EMG testing, have led to the identification of new genetic diseases [8,9]. Therefore, a meticulous evaluation that includes comprehensive history taking, physical examinations, EMG, and genetic testing is beneficial for accurate long-term diagnoses of genetic disorders.

As for *ADSSL1* mutagenic myopathy, effective treatments have yet to be developed. This underscores the necessity for future research, similar to other genetic diseases where treatments are still not established.

In conclusion, accurate diagnoses are essential for managing progressive diseases, such as neuromuscular disorders, to effectively guide treatment and rehabilitation strategies. This case underscores the significance of an accurate diagnosis by correcting an initial presumptive diagnosis of BMD through EMG and genetic testing, thereby reinforcing the diagnostic utility of EMG for future applications.

We provided rehabilitative treatment focused on the distal muscles to enhance daily functioning and to educate patients about their condition and the associated inheritance risks. Periodic reassessment of long-term conditions, especially genetic diseases, helps differentiate similar conditions, improve patient education, refine treatment planning, and enhance overall future quality of life.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

ORCID

Dong Jin Chae, https://orcid.org/0000-0003-4258-5572
Yung Jin Lee, https://orcid.org/0000-0003-4479-9136
Mi Jin Hong, https://orcid.org/0000-0002-4170-0762
Cho E. Sim, https://orcid.org/0009-0008-2842-5330
Seung-Eun Kim, https://orcid.org/0009-0001-0019-4154
Jong Bum Park, https://orcid.org/0000-0002-0087-845X

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Instructions for Authors

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Refrain from an excessive review of historical studies, textbook facts, or irrelevant references. Interpret the results with respect to the objective of the study, and describe differences with previous studies and significant findings, which lead to the deduction of the conclusion. Accentuate newly obtained observations from the study and include significant limitations of the study. Do not repeat the results in detail or other information that is given in the Introduction or the Results section.

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Acknowledgment

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4) Online resource

National Library of Medicine: Fact sheet: AIDS information resources [Internet]. Bethesda: National Library of Medicine; 2003 [cited 2007 Mar 26]. Available from: http://www.nlm.nih.gov/pubs/factsheets/aidsinfs.html.

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