






Peripheral Neuropathy Presenting as Myopathy That was Confused With Inflammatory Myositis

Younghoon KIM¹ , Yunsuek KIM¹ , So-Young JIN² , Hak-Jae ROH³ , Hyun-sook KIM¹ 

¹Department of Internal Medicine, Soonchunhyang University, Seoul, South Korea

²Department of Pathology, Soonchunhyang University, Seoul, South Korea

³Department of Neurology, Soonchunhyang University, Seoul, South Korea

Myopathy accompanying neuropathy is extremely rare. Clinical features alone may be insufficient to distinguish myopathy from neuropathy.¹ Electrodiagnostic studies such as needle electromyography (EMG), nerve conduction studies (NCS), and muscle biopsies are needed to confirm the diagnosis.^{2,3}

A 34-year-old female patient presented with sudden weakness and painful swelling of the left lower extremity. She had also developed multiple bullae that progressed to diffuse swelling (Figure 1a). Laboratory tests resulted in lactate dehydrogenase 881 U/L, aspartate

transaminase 264 U/L, alanine transaminase 102 U/L, creatine kinase 23,535 U/L, myoglobin >3,000 (normal 27-75) ng/UL, and aldolase 122 U/L. Antinuclear, anti-Jo antibody, cytomegalovirus, and Epstein-Barr virus were all negative. Magnetic resonance imaging demonstrated general edema and changes in signal intensity around the posterior segment muscles including the semitendinosus and adductor magnus muscles of the left thigh (Figure 2a, b), but it showed no change in the signal intensity of the left calf muscle (Figure 2c). Initially, there were no specific

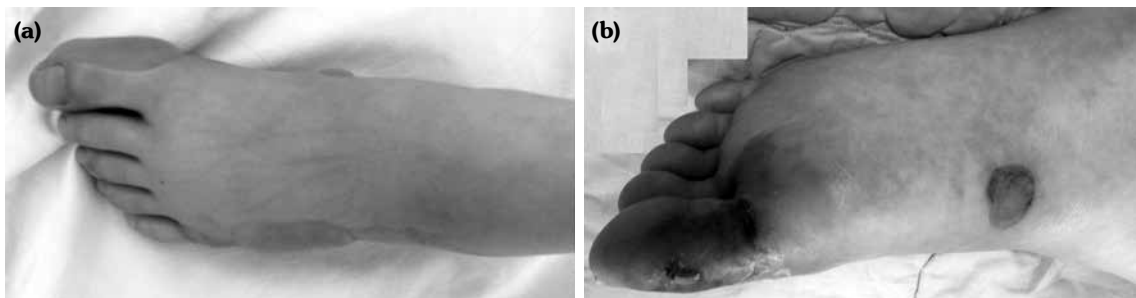


Figure 1. (a) At time of admission, bullae were observed on left foot, with largest measuring 3×4 cm. (b) After one week of glucocorticoid treatment, bullae had revolved almost completely.

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Correspondence: Hyun-sook Kim, MD. Department of Internal Medicine, Soonchunhyang University, 04414 Seoul, South Korea.
Tel: 821062586498 e-mail: healthyra@schmc.ac.kr

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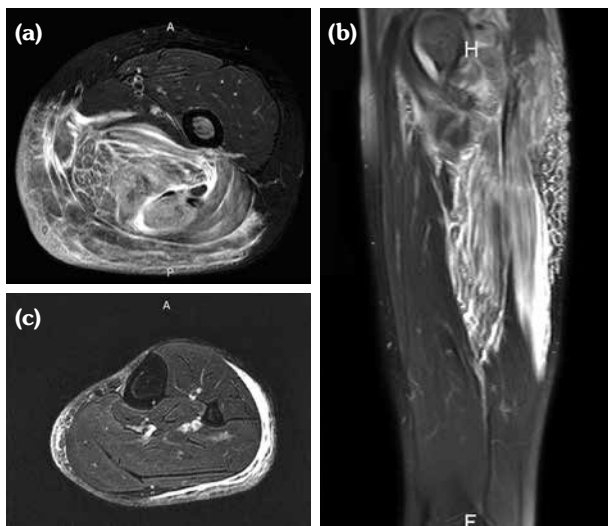


Figure 2. (a, b) T₂-weighted fat-saturated magnetic resonance imaging revealed diffuse high signal intensity in posterior segmental muscles and subcutaneous fat layer of left thigh. (c) T₂-weighted fat-saturated magnetic resonance imaging revealed high signal intensity in subcutaneous fat layer, but not in muscle layer of left calf.

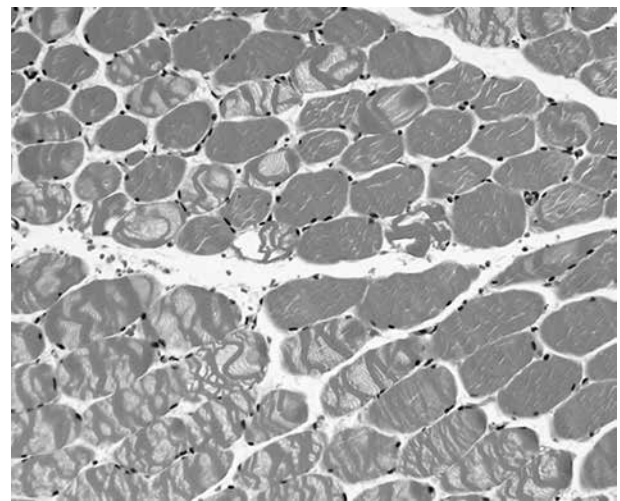


Figure 3. Histologically, left semitendinosus muscle did not change in overall contour or size of muscle fibers, nor was there any central movement of nucleus. Varying degrees of vesicle degeneration and local loss of striation appeared in myocyte cytoplasm. There was no size diversity, mass atrophy, or fat infiltration, and no interstitial lymphocyte infiltration (H-E×200).

findings of peripheral neuropathy on EMG or NCS. Despite improvement of bullae, edema, and returning to normal range of muscle enzyme after the glucocorticoid pulse therapy (methylprednisolone 250 mg/day for three days) (Figure 1b), the histological findings of the left semitendinosus muscle were nonspecific without interstitial lymphocyte infiltration (Figure 3). Although the patient could extend the left great toe, the left foot plantar flexion and dorsiflexion

had not recovered completely. Ultimately, repeated EMG and NCS revealed decreased motor unit activation in the muscles innervated by the left peroneal and tibial nerves. The NCS revealed decreased amplitude, compared with the right side, and suggested a neurogenic condition (Tables 1 and 2). Seven months later, axonal regeneration and the motor unit activation were improved in the same muscles on EMG and NCS (Tables 1 and 2).

Table 1. There were no signs suggestive of myopathy or neuropathy on needle electromyography at the time of admission. However, three weeks after admission, the needle electromyography findings revealed decreased motor unit activation in the muscles innervated by the peroneal and tibial nerves (the external hallucis longus, tibialis anterior, tibialis posterior, and peroneus longus muscles). After treatment, axonal degeneration and regeneration were observed and the motor unit activation was improved in the same muscles.

Muscle (left)	3 weeks after admission			9 weeks after admission			7 months after admission		
	ASA	MUAP	IP	ASA	MUAP	IP	ASA	MUAP	IP
External hallucis longus	-	-	Z	+	PPP	S-P	-	Normal	P-C
Tibialis anterior	-	-	Z	+	PPP	P	+	PPP	P-C
Tibialis posterior	-	-	Z	+	Normal	P-C	-	Normal	P
Peroneus longus	-	-	Z	+	PPP	P	-	PPP	P-C
Gastrocnemius (medial)	-	PPP	S-P	+	PPP	P	-	Normal	C
Biceps femoris short	-	PPP	C	++	PPP	C	-	Normal	C
Semitendinosus	-	Normal	C	-	Normal	C	-	Normal	C
Semimembranosus	-	PPP	C	-	Normal	C	-	Normal	C
Biceps femoris long	-	Normal	C	++	Normal	C	-	Normal	C

ASA: Abnormal spontaneous activity; MUAP: Motor unit action potential; IP: Interference pattern; Z: Zero; PPP: Polyphasic pattern; S: Single; P: Partial; C: Complete.

Table 2. Nerve conduction study findings revealed decreased amplitude of the left peroneal and tibial nerves, compared with the right side. These findings suggested axonal loss of the left peroneal and tibial nerves. After treatment, the amplitudes of the left peroneal and tibial nerve were increased and the degree of axonal loss was improved

	3 weeks after admission	9 weeks after admission	7 months after admission
Left deep peroneal nerve-EDB	No response	No response	0.5 mV
Left deep peroneal nerve-TA	3.6 mV (51% of right side)	4.3 mV	5.5 mV
Left tibial nerve-AH	4.1 mV (23% of right side)	2.7 mV	7.3 mV

EDB: Extensor digitorum brevis; TA: Tibialis anterior; AH: Abductor hallucis.

Myopathy and neuropathy are generally considered to be different, although concomitant myopathy and neuropathy are rarely reported, called neuromyositis.⁴ EMG and NCS are useful tools for identifying neuromyositis.⁵ Muscle and nerve biopsies may also be helpful for diagnosing neuromyositis, but further studies are needed to establish the typical histologic findings. Myopathy is rarely accompanied by bullae. If the skin cells are damaged by various causes and their structural function is affected, the cells separate from each other and fluids build up between the separated cells, causing bullae.^{6,7} In our case, a similar mechanism was suggested that the rapid progression of myopathy accompanied by neuropathy caused cutaneous edema and dermoepidermal disruption, resulting in bullae.

Although EMG and NCS are useful tests for distinguishing myopathy and neuropathy, the interpretation of the results may vary depending on the time of the tests. Typically, NCS and EMG reveal pathological findings beginning a few days after the onset of symptoms, but they may appear normal for up to three weeks. Therefore, if unexplained abnormal sensation and disability occur, it is appropriate to repeat EMG and NCS.

Declaration of conflicting interests

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