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CASE REPORT

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천요추 신경근병증의 임상증상으로 발현된 편측 종아리 근육 가성 비대

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Unilateral Pseudohypertrophy of Calf Muscles as a Clinical Manifestation of Lumbosacral Radiculopathy

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Chronic lumbosacral radiculopathy usually presents with neurogenic muscular atrophy. Hypertrophic changes in accordance with neuropathy are rare. This case report introduces a case of unilateral pseudohypertrophy of calf muscle as a clinical manifestation of chronic lumbosacral radiculopathy due to denervation. The patient presented pseudohypertrophy of muscle resulting from right L5 and S1 radiculopathy due to idiopathic spinal epidural lipomatosis (SEL). Physicians should be aware that muscle hypertrophy, which usually has been regarded as a sign of myopathy, may denote the clinical sign of radiculopathy. Electromyography can be a valuable tool for differential diagnosis in such cases.

Keywords: Muscle denervation, Radiculopathy, Muscular diseases

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Introduction

Radiculopathy commonly presents as pain in dermatomal distribution of involved nerve roots. It is often accompanied by sensory deficit such as paresthesia, hypesthesia, and/or weakness. Neurogenic muscular atrophy is also frequent. However, hypertrophic changes may develop because of denervation in rare cases [1]. Some cases of muscle hypertrophy have been reported for neurogenic causes such as lumbosacral radiculopathy [2-4]. This unusual clinical manifestation most frequently occurs in the calf muscles [2]. The underlying pathophysiologic mechanisms of this neurogenic hypertrophy are unclear, yet some mechanisms have been postulated [2].

This case report introduces a case of unilateral pseudohypertrophy of calf muscle as a clinical manifestation of chronic right L5 and S1 radiculopathy caused by idiopathic spinal epidural lipomatosis (SEL). From our knowledge, no other cases have been reported unilateral pseudohypertrophy of calf muscle after SEL.

Case report

A 65-year-old man (172cm, 82kg, BMI 27.7) visited the rehabilitation medicine department with a chief complaint of swelling of the right lower extremity, which slowly progressed for about three years. The patient had no significant past and family history or underlying diseases. The patient complained of low back pain with limping gait, which began 5 years before the visit. Two years before the visit, the patient experienced subjective weakness of right lower extremity and underwent multiple spinal nerve block procedures including caudal steroid injections. He underwent x-ray imaging of his lumbar spine at the time of the visit for spinal nerve block interventions. He was told that there was a narrowing of the lower lumbar segments which may have caused the mild weakness. It is unknown whether magnetic resonance imaging (MRI) was performed at that time. The symptoms showed wax and wane pattern throughout the multiple trials of nerve blocks, which led him not to take his symptoms seriously. About a year later, the patient noticed the enlargement of his right lower extremity, especially of the calf muscles. A private clinic prescribed the patient with vasoprotectives (Entelon®, vitis vinifera extract) under the diagnosis of dependent edema or lymphedema of undetermined etiology. The prescribed drugs were only helpful for transient resolution of symptoms. He experienced progressive weakness of the right lower extremity, which impeded him from stable walking and tiptoeing. The patient wandered around several hospitals and clinics to find the cause of his enlarged and weakened right leg. The patient refused further evaluation regarding his spine since he presented a new symptom of swollen unilateral leg. Moreover, most of the clinics considered his symptoms to be of vascular origin, because the previous spinal injections did not completely relieve his symptoms. One of the private clinics recommended cardiovascular evaluation at a tertiary hospital; thus, he was referred to our hospital. Cardiovascular evaluation including echocardiography, low-dose chest computed tomography (CT), and ankle-brachial index was performed. The results were all within normal limits. Thyroid function test was performed to exclude hypothyroidism, which can also cause edema of the lower extremity, and the results were normal. Contrast-enhanced CT of the lower extremity artery showed no evidence of significant stenosis or occlusion along the arteries of both lower extremities. However, it revealed the denervation hypertrophy involving the right calf and fatty infiltration in between the muscle fibers of right soleus and gastrocnemius muscles (Fig. 1). After excluding the possible systemic causes of low extremity edema such as hypothyroidism, neoplasm, amyloidosis, or sarcoidosis, the patient was referred from cardiology department to rehabilitation medicine department for further evaluation of possible myopathy.

Neurologic examination including mental status and cranial nerve examinations showed normal results. Physical examination showed hypertrophy of the right calf muscles with the right calf circumference measuring up to 38 cm, whereas his left calf circumference measured up to 33 cm at resting state although not distinct visually. The circumference of both calves was measured at 10cm below the tibial tuberosity. There was no bulging of the right gastrocnemius muscles at tiptoe position (Fig. 2). There was no pitting edema, color change or skin turgor of his lower

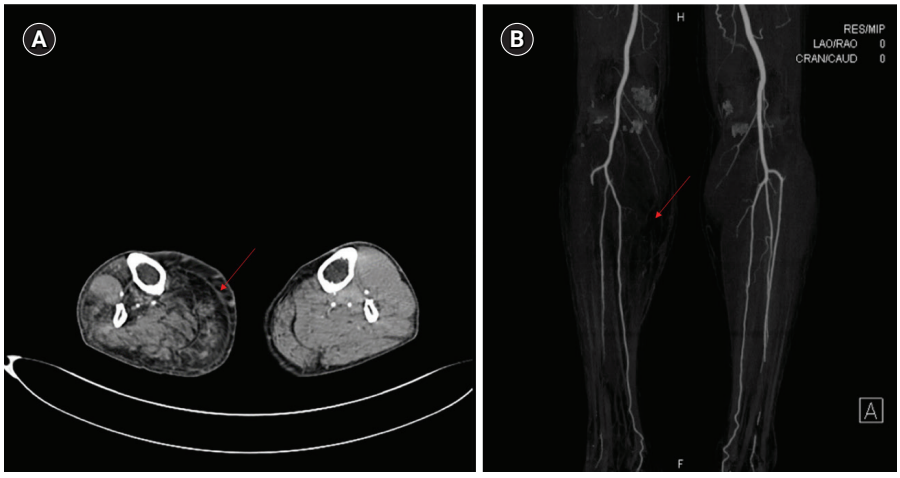

Fig. 1. A CT scan shows denervation hypertrophy of right calf muscle.
There is no evidence of significant stenosis or occlusion along both lower extremity arteries. Fatty infiltration in between muscle fibers of right soleus and gastrocnemius muscles is present, which is suggestive of denervation pseudohypertrophy or infiltrating lipoma.

extremity. Dorsalis pedis pulsation of the patient was intact on bilateral feet. The manual muscle test revealed weakness of right ankle dorsiflexion (4/5), great toe extension (3+/5) and plantarflexion (3/5). Sensory examination showed mild hypesthesia over the sole of the right foot with decreased proprioception of bilateral toes. Serum blood test was performed and muscle enzymes including creatine phosphokinase (CPK), lactate dehydrogenase (LDH) and CPK isoenzymes were all elevated. The CPK level was 601 units per liter(U/L) (normal range: 26-200 U/L) and LDH level was 575 U/L (normal range 250-450 U/L). The CPK isoenzyme pattern can be an indication of muscle damage such as crush injuries, muscle infarction or necrosis, alcoholism, or other muscular disorder including muscular dystrophy, viral myositis, polymyositis, and et cetera. All other laboratory results including inflammatory markers and complements were within normal range. The possibility of myopathy was increased, but the patient denied of muscle pain, history of trauma, travel, heavy exercise or alcohol consumption. Moreover, the patient showed weakness and swelling of the unilateral leg alone.

In order to exclude spinal lesions, the patient was persuaded to undergo MRI of the lumbar spine. The MRI showed an incidental finding of spinal canal lipomatosis extending from L5 to S2 level, mostly abutting the right S1 nerve root, which may be due to obesity or chronic use of steroid injection (Fig. 3).

Nerve conduction studies (NCS) and Electromyography (EMG) were performed for differential diagnosis of myopathy and neuropathy. The NCS showed slightly decreased amplitudes of sensory nerve action potential (SNAP)s in bilateral superficial peroneal and sural nerves (Table 1). Motor nerve action potential (CMAP)s showed no definite right to left discrepancy. The EMG depicted abnormal spontaneous activities in the right tibialis anterior, peroneus longus, gastrocnemius, soleus, and bilateral lower lumbar paraspinal muscles (Table 2). Polyphasic configuration of motor unit action potentials (MUAP) on minimal volition in right gluteus maximus, peroneus longus and soleus muscles were observed. There was no definite electrodiagnostic evidence of myopathy. Quantitative EMG was performed on the right vastus medialis, tibialis anterior and left tibialis anterior muscles. Turns and amplitude patterns were within reference limits and showed neuropathic patterns. Right peroneus longus, gastrocnemius, soleus, abductor hallucis and extensor hallucis longus muscles depicted discrete to reduced interferential patterns on maximal volition. Complex repetitive discharges were seen in right lower lumbar paraspinal, soleus and abductor hallucis muscles. Decreased insertional activities were obtained in the right soleus muscle. Above EMG/NCS showed MUAPs of neuropathic pattern, in which profuse denervation potentials were

Table 1. Nerve Conduction Study Result

Sensory Nerve Conduction Studies					
Nerve/Sites	Onset Lat ms	Peak Lat ms	Amp μV	Vel. m/s	Dist. cm
L SURAL - Lat Mal					
Calf	2.70	3.75	7.7	51.9	14
R SURAL - Lat Mal					
Calf	2.60	3.65	7.7	53.8	14
R SUP PERONEAL - Ankle					
Lat Leg	2.75	3.70	8.0	50.9	14
L SUP PERONEAL - Ankle					
Lat Leg	2.85	3.75	7.3	49.1	14

Motor Nerve Conduction Studies						
Nerve/Sites	Onset Lat ms	Peak Lat. ms	Amp. mV	Dur. ms	Dist. cm	Vel. m/s
R COMM PERONEAL - (EDB)						
Ankle	2.85	5.05	5.0	3.90		
Fib Head	9.85	13.25	4.5	4.90	33	47.1
L COMM PERONEAL - (EDB)						
Ankle	3.10	5.30	4.3	4.50		
Fib Head	9.80	13.10	3.8	5.90	33	49.3
R TIBIAL (KNEE) - AH						
Ankle	2.95	6.70	5.9	4.85		
Popliteal	11.85	16.55	2.8	6.70	43	48.3
L TIBIAL (KNEE) - AH						
Ankle	3.35	6.55	4.6	4.40		
popliteal	12.85	15.80	2.8	6.00	43	45.3
R COMM PERONEAL - Tib Ant						
Fib Head	3.30	9.05	2.9	8.90		
Knee	4.40	9.95	2.8	8.70	6	54.5
L COMM PERONEAL - Tib Ant						
Fib Head	2.15	5.20	4.9	12.40		
Knee	3.35	7.20	4.9	12.25	6	50.0

EDB: extensor digitorum brevis, AH: abductor hallucis, Tib Ant: tibialis anterior

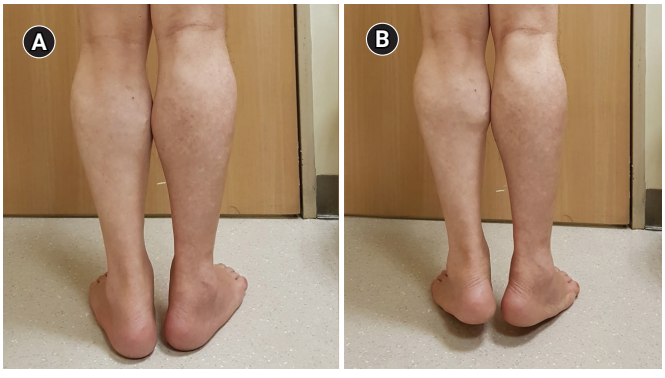

Fig. 2. Clinical photos of the patient's calf muscles.
Physical examination shows (A) hypertrophy of the right calf muscles compared to the left side. The right calf diameter measured up to 38 cm and his left calf diameter measured up to 33 cm at resting state. (B) Patient of tiptoe position: There is no bulging of the right gastrocnemius muscles.

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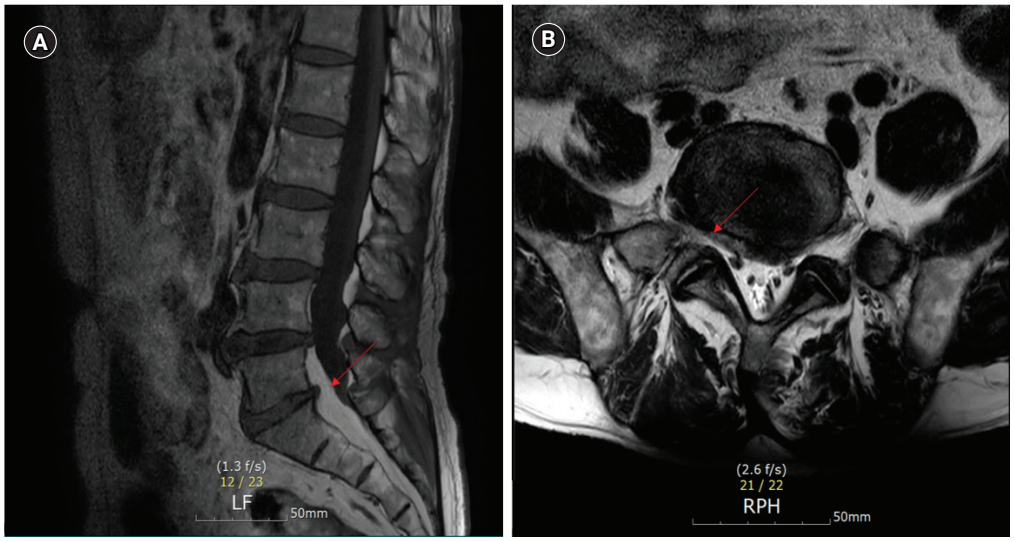

Fig. 3. MRI of the lumbar spine of spinal canal lipomatosis: A) Sagittal T1-weighted-image shows a bright signal intensity posterior epidural mass extending from L5 to S2 level, which is suggestive of spinal lipomatosis B) axial T2-weighted-image shows the epidural lipomatosis abutting the right S1 nerve root.

Table 2. Needle Electromyography Study Result

	Needle Electromyography								
	Spontaneous					MUAP			Interferential
	IA	Fib	PSW	Fasc	CRD	Amp	Dur.	PPP	pattern
B. Cerv(C4-T1) PSP	N	None	None	None	None				
R. LUMB PSP (U)	N	None	None	None	None				
R. LUMB PSP (M)	N	None	None	None	None				
R. LUMB PSP (L)	N	None	1+	None	CRD				
L. LUMB PSP (U)	N	None	None	None	None				
L. LUMB PSP (M)	N	None	None	None	None				
L. LUMB PSP (L)	N	None	1+	None	None				
R. GLUTEUS MAX	N	None	None	None	None	7m V	N	Poly	N
R. GLUTEUS MED	N	None	None	None	None	7m V	N	N	N
R. VAST MEDIALIS	N	None	None	None	None	N	N	N	N
R. RECT FEMORIS	N	None	None	None	None	N	N	N	N
R. ADD MAGNUS	N	None	None	None	None	N	N	N	N
R. ILIOPSOAS	N	None	None	None	None	N	N	N	N
R. BIC FEM (L HEAD)	N	None	None	None	None	N	N	N	N
R. BIC FEM (S HEAD)	N	None	None	None	None	N	N	N	N
R. TIB ANTERIOR	N	None	1+	None	None	N	N	N	N
R. PERON LONGUS	N	None	1+	None	None	N	N	Poly	Discrete
R. GASTROCN (MED)	N	None	1+	None	None	N	N	N	Discrete
R. SOLEUS	Dec	None	1+	None	CRD	N	N	Poly	Discrete
R. ABD HALLUCIS	N	None	None	None	CRD	N	N	N	Reduced
R. EXT HALL LONG	N	None	None	None	None	N	N	N	Reduced
L. GLUTEUS MAX	N	None	None	None	None	N	N	N	N
L. GLUTEUS MED	N	None	None	None	None	N	N	N	N
L. ILIOPSOAS	N	None	None	None	None	N	N	N	N
L. VAST MEDIALIS	N	None	None	None	None	N	N	N	N
L. TIB ANTERIOR	N	None	None	None	None	N	N	N	N
L. GASTROCN (MED)	N	None	None	None	None	N	N	N	N
L. SOLEUS	N	None	None	None	None	N	N	N	N

observed in the sampled L5 and S1 innervated muscles. The findings of EMG/NCS correlated with the findings on MRI.

It was concluded that the patient had pseudohypertrophy of the right lower extremity due to chronic L5 and S1 radiculopathy. The patient was referred to the department of neurosurgery for a second opinion for the prognostic value of operative management. Conservative care was recommended by the neurosurgeon due to the slow progressive nature of the symptoms. Conservative management including administration of non-steroid anti-inflammatory medications, analgesics, and physical therapy was started. Despite the subtle but progressive improvements of his pain and weakness, the patient desired rather rapid improvements of his symptoms. The patient was referred to the pain clinic of the anesthesiology department and underwent neuroplasty of right L5 and S1 nerve roots by using the epidural catheter. The patient's pain and hypesthesia improved after about two months of strengthening exercise, physical therapy modalities, and neuroplasty. The patient no longer had limping gait and began to work out in the gym with gradual improvements of his weakness.

Discussion

This report highlights a case of pseudohypertrophy as a clinical manifestation of lumbosacral radiculopathy. The patient presented muscle pseudohypertrophy resulting from a neurogenic cause, which was L5 and S1 radiculopathy in this case.

Pathologic muscle hypertrophy is known to be most commonly associated with myopathic disorders. Myopathic muscular hypertrophies, such as muscular dystrophy, hypothyroidism induced Hoffman syndrome, and inflammatory myopathies, usually manifest in diffuse and symmetric pattern unlike the muscular hypertrophy of neuropathy origin, which is not focused on muscles of dermatomal distribution [5]. Moreover, myopathic muscle hypertrophy shows elevation of the muscular enzymes, but the neurogenic cause cannot be ruled out solely based on CPK levels [2]. Analyses of motor unit potentials via EMG are crucial in the diagnosis of myopathy [6], but MRI of suspected muscle lesion and corresponding muscle biopsy may be required in some cases.

Neurogenic denervation pseudohypertrophy usually manifests with a painless, gradual onset of swelling of the limb without paresis. The affected muscles are usually innervated by the same nerve roots of specific myotome [5]. Moreover, pseudohypertrophic muscles are frequently paradoxically weak. The underlying pathophysiological mechanism of neurogenic muscle pseudohypertrophy remains unclear. However, it is suggested that the mechanism may involve compensatory hypertrophy of remaining healthy muscles, sprouting and synapsing of remaining axons with denervated fibers, which results in abnormal spontaneous electrical activity of denervated muscle fibers [7]. In cases of unilateral hypertrophied muscles, EMG frequently demonstrates complex repetitive discharges (CRD). This may be due to the overuse of partially denervated muscle fibers and it may be the result of a neurogenic process [8]. Therefore, a diagnosis of neurogenic muscle hypertrophy secondary to radiculopathy was made, and EMG played a crucial role in differential diagnosis.

Unlike previously published cases, the merit of this case report lies in the fact that it reports a patient with symptomatic swelling of the unilateral leg which resulted from SEL. Although not rare, SEL is usually underestimated because it is usually regarded as a benign radiologic finding. Despite that it is believed to be mainly associated with obesity and endocrinopathies, exogenous steroid injection is an important risk factor of SEL as seen in the present case [9]. Identifying the possible cause of SEL of the present case is beyond the scope of this report. However, it is important for clinicians to recognize that patients with SEL, which is usually asymptomatic, can present with symptoms resulting from nerve or spinal cord compression.

The treatment guidelines of neurogenic muscle hypertrophy have not yet been established. Treatment can either be surgical or conservative. Oral corticosteroids with prednisone or prednisolone at a dose of 15-45 mg per day has been reported for its effectiveness in cases of unilateral calf hypertrophy induced by radiculopathy with combined focal myositis [3]. Early detection is critical in deciding the proper treatment option and necessary for preventing deleterious effects of progressive muscle pseudohypertrophy.

In conclusion, this case addresses the clinicians to be alert for the possibility of neurogenic conditions when dealing with unilateral hypertrophy of muscles. Thorough history taking with the use of NCS/EMG has an important role in revealing the underlying pathology involving spinal nerve roots. Physicians must be aware of the variability in muscle response to denervation for prevention of unnecessary delays for proper diagnosis.

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