신경근육재활 및 전기진단

게시일시 및 장소 : 10 월 18 일(금) 13:15-18:00 Room G(3F)

질의응답 일시 및 장소 : 10 월 18 일(금) 15:45-16:30 Room G(3F)

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Benign Monomelic Amyotrophy: a Case Report of 40-Year-Old Korean Male

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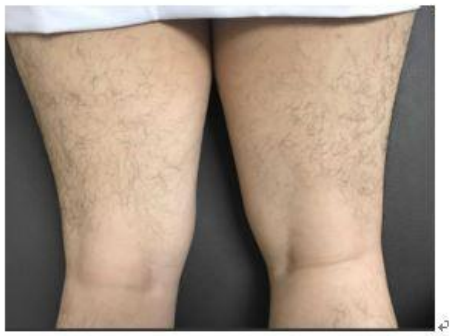
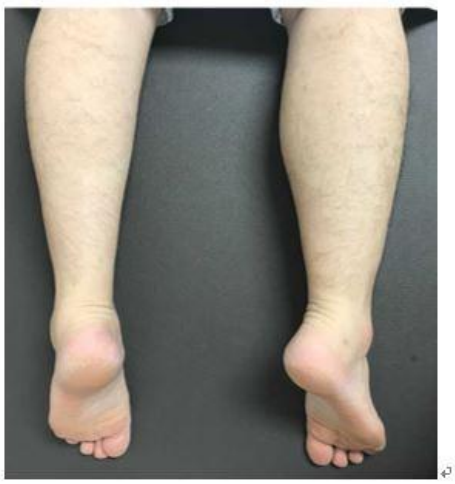
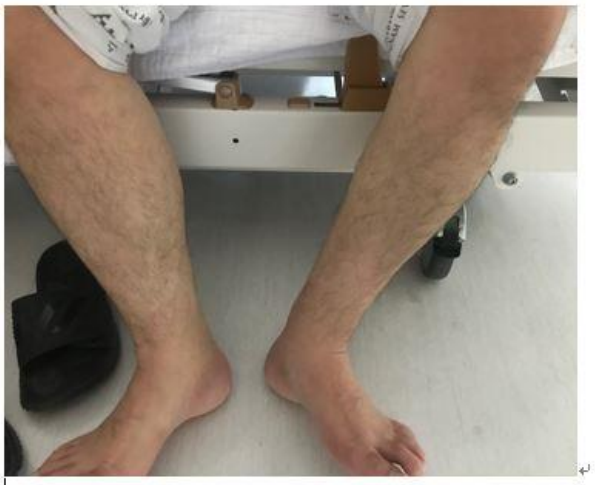
Benign monomelic amyotrophy (BMA) is a rare motor neuron disease in which neurogenic amyotrophy is restricted to either the upper or the lower extremities with sporadic occurrence, insidious onset, and slow progressive course. Numerous cases of unilateral muscular atrophy restricted to the upper or lower limb have been reported from Japan, India, and much less frequently from South Korea. We describe the clinical, electrophysiologic, and magnetic resonance image (MRI) studies of a middle-aged Korean male patient with a late-onset muscular atrophy confined for 3 years in a single lower limb. Case: A 40-year-old male with no significant medical history presented to the outpatient clinic with lower back pain. Upon initial examination, left calf muscle atrophy, and symptoms related to calf muscle weakness (inability to stand on tiptoe or jump) was found. The patient noticed weakness in left calf muscle dating three years ago; however, weakness did not progress over time. There was no history suggestive of poliomyelitis or familiar neuromuscular disease. In addition, he had no previous history of trauma, toxin exposure, or viral infections. According to Medical Research Council (MRC), muscle strength in his left gastrocnemius muscle was 4/5, which differed from his right side significantly. His muscle strength was 5/5 in the remaining lower limb muscles. There was no limitation in range of motion. There were no pathologic upper motor neuron sign at both upper and lower extremities. There was no noticeable defect during the push-off phase in his gait cycle; however, toe walking was limited. Muscle weakness and wasting were localized to the left calf (Figure 1). Lower extremity circumference was measured from the patella margin, which revealed significant difference in circumference at 15cm below the patella margin. Deep tendon reflex was normoreactive and sensory examination did not reveal any abnormalities. Lumbar spine MRI was did not reveal major radiological evidence of compressive lesion of the spinal cord or the nerve root (Figure 2). Patient underwent magnetic resonance imaging of the lower extremity, which revealed marked atrophy and increased signal of left medial gastrocnemius muscle (Figure 3). Nerve conduction study findings were normal without any evidence of motor conduction block or diffuse conduction slowing. Bilateral tibial H-reflex studies showed acceptable findings in both sides. Needle EMG results showed normal, large and long duration polyphasic motor unit on volition in left gastrocnemius. Similar to the previous studies, the reported

patient showed the cardinal features of BMA: 1) insidious onset with arrested course; 2) wasting confined to unilateral limb; and 3) lack of bulbar or sensory symptoms. Diagnosis of BMA is challenging due to its slow progression and lack of biomarkers. With the combination of detailed history taking, MRI of the affected limb, and EMG can lead to accurate diagnosis.







<Lower extremity circumference>

R : +5cm(46cm) +10cm(52cm) +15cm(58cm)
-5cm(40cm) -10cm(39cm) **-15cm(37.5cm)**

L : +5cm(44cm) +10cm(51cm) +15cm(58cm)
-5cm(39cm) -10cm(37cm) **-15cm(33cm)**

Figure 1. Atrophy of the left gastrocnemius muscle in a 40-year-old male. Lower extremity circumference was measured from the patella margin, which revealed significant difference in circumference at 15cm below the patella margin: +15cm (58cm, 58cm), +10cm (52cm, 51cm) +5cm (46cm, 44cm), -5cm (40cm, 39cm), -10cm (39cm, 37cm), -15 (37.5cm, 33cm).

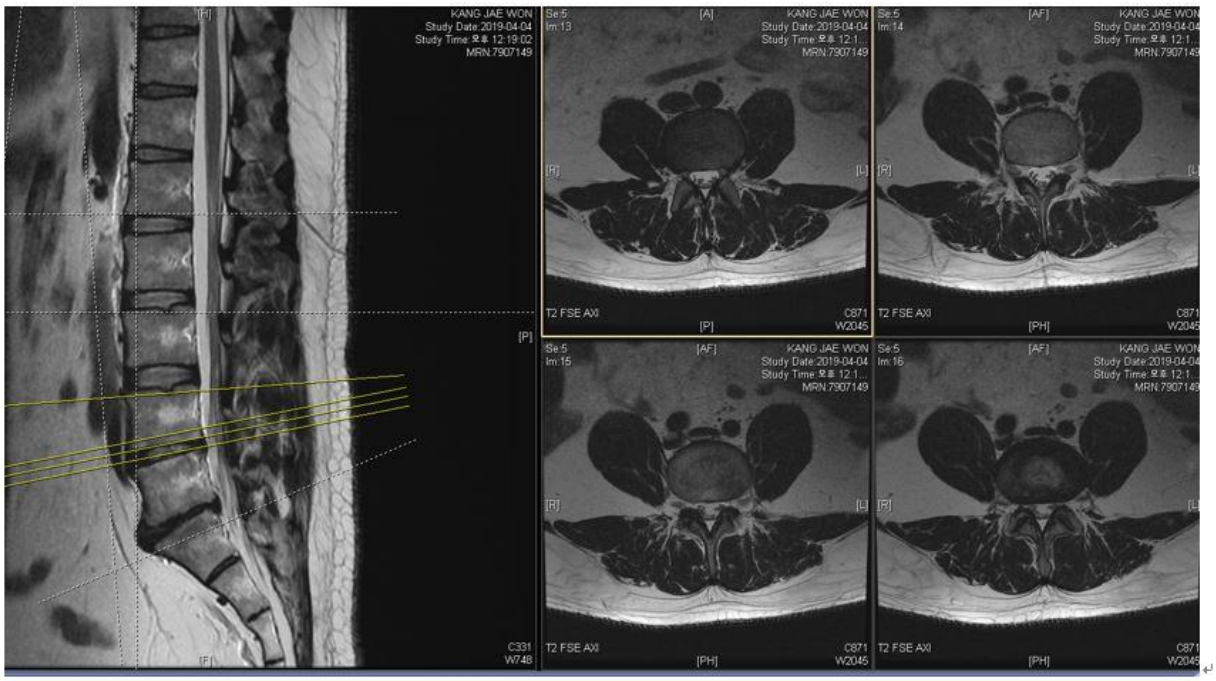


Figure 2. Magnetic resonance imaging of the spine. No major radiological evidence of compressive lesion of the spinal cord or the nerve root

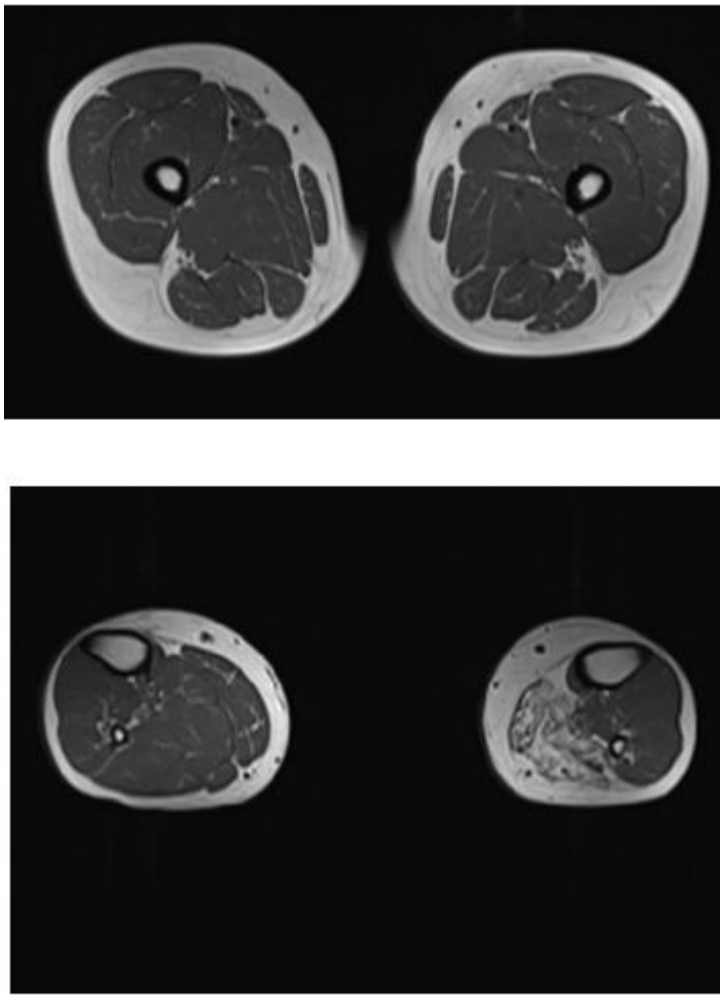


Figure 3. Axial images of lower extremity MRI. T2 weighted-images revealed marked atrophy and increased signal of left medial gastrocnemius muscle.