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Stiff Person Syndrome With an Evidence of Polymyositis Secondary to Sustained Muscle Contraction

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Introduction

Stiff person syndrome (SPS) is a rare neuroimmunological disorder characterized by progressive muscular rigidity and spasms that affect axial and limb muscles. There have been a few reports that patients with SPS had evidences of polymyositis (PM). There have been no clear explanations about the characteristics of PM in SPS. We report a case of SPS with an evidence of PM secondary to sustained muscle contraction that partially responded to immunomodulatory agent.

Case

A 36-year-old woman presented with a 1-year history of progressive rigidity and pain in her proximal upper and lower limbs. Her medical history revealed idiopathic chronic kidney disease diagnosed 3 years ago and depression for 15 years. Physical examination revealed normal deep tendon reflexes and absence of motor and sensory impairment. Sustained contraction of the trapezius, biceps, triceps, quadriceps, and hamstring muscles during rest was prominent (Fig. 1A). Laboratory results revealed elevated creatinine kinase (CK; 828 U/L) levels, although autoantibody tests, including anti-GAD antibody (0.23 U/mL), rheumatoid factor, antithyroglobulin, and antinuclear antibody levels, were all negative. Electromyography revealed continuous motor unit activity in the agonist and antagonist muscles despite attempts by the patient to relax the muscles (Fig 2). There was no evidence of fibrillation potentials or positive sharp waves on EMG during the silent period. Brain MRI was normal. Limb MRI revealed bilaterally increased signal intensity in the biceps, triceps, rectus femoris, semitendinosus, and biceps femoris muscles (Fig. 3A and B). PET showed increased uptake within the same muscles and did not reveal evidence of malignancy (Fig. 3C). Based on the clinical feature, MRI, PET and electromyography results, SPS was presumptively diagnosed. However, elevated CK level and signal change in the biceps and rectus femoris muscles on MRI were not fully explained by the diagnosis of SPS. Thus, muscle biopsy on the right rectus femoris was performed. Pathology revealed pathologic signs of inflammatory myopathy (Fig. 3D). Since administration of oral diazepam and steroid exhibited no effect, we attempted to administer intravenous immunoglobulin (IVIG). After IVIG therapy, CK levels were normalized, although sometimes it is not fully. Despite serial IVIG treatment, her stiffness, hypertrophy, and limb and axial musculature pain have gradually worsened over the previous four years (Fig. 1B and C).

Discussion

SPS diagnosis is challenging and requires a high degree of suspicion. SPS is a rare neuroimmunological disorder in which an evidence of PM can be seen. PM characteristics

or pathophysiology in SPS have not been fully explained. We describe a patient diagnosed with SPS associated with PM secondary to sustained muscle contraction.

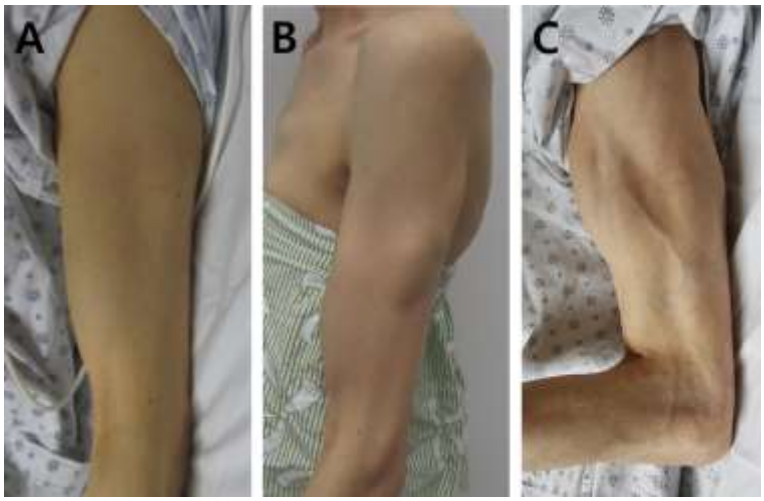

Fig 1. Inspection of the left arm. (A) The patient's left biceps and triceps during the first visit. (B) Hypertrophic change in the biceps and triceps at 1 year after diagnosis. (C) Hypertrophic change aggravated at 4 years after diagnosis

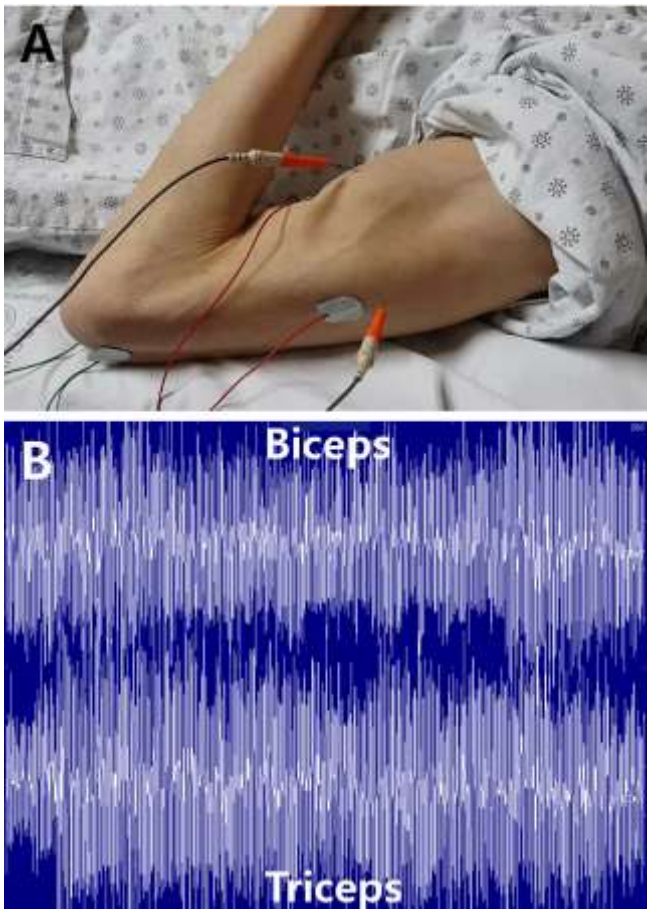

Fig 2. Electrophysiologic study. (A) Dual channel electromyography recorded with needle electrodes of the left biceps and triceps. (B) During rest, electromyography revealed continuous motor unit activity simultaneously in both the biceps and triceps (sensitivity of 200 μV per division and sweep speed of 200 ms per division)

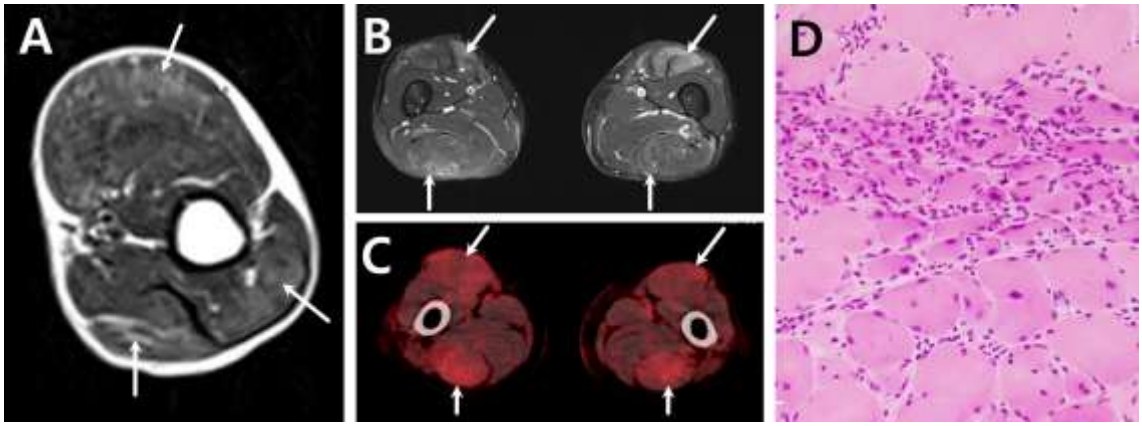

Fig 3. Results of imaging studies and biopsy. (A) Axial T2-weighted image on left upper arm MRI revealed signal changes in the biceps as well as the long and lateral head of the triceps (arrows). (B) Axial T2-weighted fat-suppressed turbo spin echo image on thigh MRI revealed increased signal intensity in the rectus femoris, semitendinosus, and biceps femoris muscles (arrows). (C) Positron emission tomography revealed increased uptake in the rectus femoris, semitendinosus, and biceps femoris muscles (arrows). (D) Biopsy of the right rectus femoris revealed markedly increased fiber size variation (20–100 μm), necrotic and regenerating fiber, internal nuclei, and inflammatory cell infiltration.