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Case Report

The Use of Korean Medicine to Treat Patients with Spinobulbar Muscular Atrophy, Kennedy's Disease - A Case Study

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Key Words
herbal medicine, Kennedy's disease, Korean medical treatment, pharmacopuncture, spinobulbar muscular atrophy

Abstract

Objectives: Studies involving patients with spinobulbar muscular atrophy (SBMA), which is often referred to as Kennedy's disease, similar to those involving patients with progressive muscular disease (PMD), are rare. This paper reports a case study involving the use of Korean medicine to treat a patient with SBMA.

Methods: We treated a patient with SBMA with unique symptoms by using various kinds of pharmacopuncture and herbal medicines for about two and a half years. After the treatment had ended, we evaluated the patient's conditions and the side effects of the treatment.

Results: After treatment, the patient's symptoms were stabilized, and the patient suffered no abnormalities or side effects. No special changes in condition were noted during treatment period, and the patient was very satisfied with his response to treatment.

Conclusion: Existing treatments have some considerable after effects and are difficult to apply in domestic clinics. In this regard, our findings should open possibilities for new clinical guidelines. Nevertheless, the limitations associated with this case study should be resolved, and more studies need to be conducted.

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1. Introduction

X-linked spinobulbar muscular atrophy, also known as Kennedy's disease, is a neurodegenerative disease caused by an expansion of a polymorphic CAG tandem-repeat in the androgen receptor gene. It presents as weakness and wasting of bulbar, facial, and proximal limb muscles owing to loss of anterior horn cells in the brain and the spinal cord. The age at onset is between 20-and 40 years. It follows a slowly progressive course, and the patient has normal or minimally- reduced life span [1].

Although several therapeutic strategies have been proposed, currently no effective therapy exists to arrest or delay disease progression [2]. Two case reports on the treatment of progressive muscular disease (PMD) by using Korean traditional medicine can be found in the literature [3, 4]. According to these papers, significant improvements in muscle function were achieved by using Korean traditional medicine to treat patients with PMD.

However, studies involving patients with Kennedy's disease, similar to those involving patients with PMD, are,

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to the best of the authors knowledge, nonexistent. For that reason, we present the case of a patient with X-linked spinobulbar muscular atrophy who was treated using Korean traditional medicine.

2. Case study

The patient was a 42-year-old male, whose chief complaints were proximal muscle weakness since the summer of 2010 and dysarthria since December 2012. When he visited our clinic, he stated the he had been weaker and less flexible than others. His past medical history was non-specific, and his family's medical history showed that his mother suffered from head tremors and his father suffered from gastric cancer. His current medications were tanamin (80 mg, one tablet once a day) and gliatilin (400 mg, one tablet twice a day).

The patient had been a heavy drinker since his 20 s, and since that time his hands had been trembling. He drank two bottles of soju five days a week for two and a half years when he was in his 30 s. After that, he drank two bottles of soju about twice a week. At that time, he weighed as much as 83 kg and his breasts were large. Later, he lost weight (70 kg), but his breasts were still large.

From the summer of 2010, he has had difficulty running, climbing stairs, and walking for a long time, and he has had increasing weakness in his arms and legs. In addition, he has had difficulty holding up his hands while washing his hair because of arm weakness.

He exercised two hours a day, four to five times a week, but he felt that he was losing strength. He had easily climbed three flights of stairs until 2012, but now climbing above the first floor was difficult. From December 2012, he has experienced hoarseness, and he has felt an intermittent pulling sensation in his left fascial muscle. He has had mildly slurred speech and a nasal voice because of facial and tongue palsy.

A few years ago, his finger started to shake unconsciously. When he tried to sleep, he felt muscle fasciculation intermittently.

In recent years, his weight has been relatively stable, but the muscles of both limbs have started to sag. He has experienced an atrophied tongue (Fig. 1), gynecomastia (Fig. 2), and sexual disorders such as atrophied testis (Fig. 3), hypospermia, and erectile dysfunction. Deoxyribonucleic acid (DNA) testing for Kennedy's disease confirmed the diagnosis and revealed an expansion of 47 CAG repeats (normal being < 33).

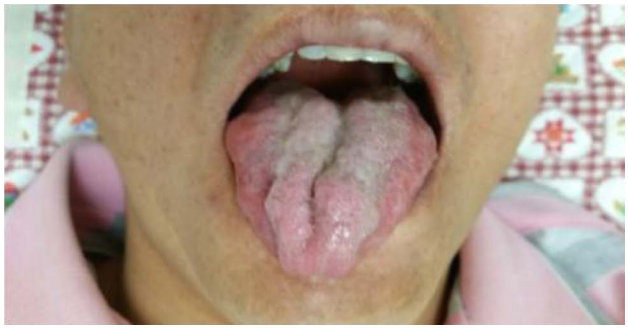

Figure 1 Atrophied tongue with valley used with the patient's permission.

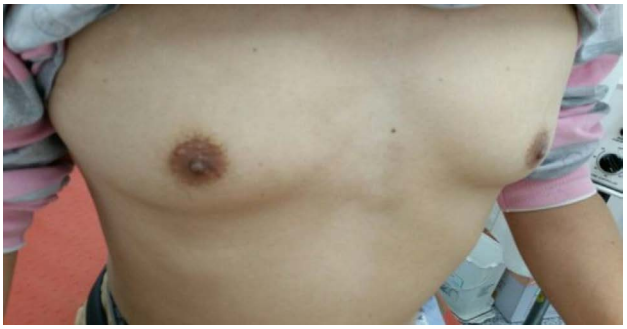

Figure 2 Gynecomastia used with the patient's permission.

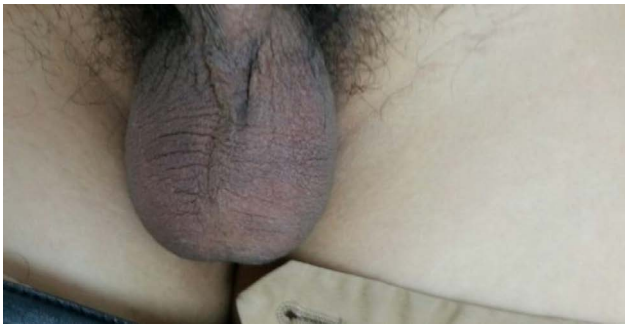

Figure 3 Atrophied testes used with the patient's permission.

A neurologic examination, and the results were normal, his mental status was alert. He had tongue fasciculation, but other cranial nerve functions were normal. The results from examinations of his upper and lower extremities are shown in Tables 1, 2, respectively. The results for the deep tendon reflex (DTR, deep tendon reflex) are shown in Table 3.

The patient's squatting, hopping, toe gait and heel gait were intact, as was his sensory touch, pinprick test. Finger to nose

Table 1 Results from examination of the upper extremities

Part	Motion	Right	Left
Neck	Flexion	GV	-
	Extension	GV	-
Shoulder	Abduction	GIV^{+}	GIV^{+}
	Abduction	GV^{-}	GV^{-}
Elbow	Flexion	GIV^{+}	GIV^{+}
	Extension	GIV^{+}	GIV^{+}
Wrist	Flexion	GV^{-}	GV^{-}
	Extension	GV^{-}	GV^{-}
Thumb	Abduction	GV^{-}	GV^{-}
	Adduction	GV^{-}	GV^{-}
Hand Grip	-	GV^{-}	GV^{-}

Table 2 Results from examination of the lower extremities

Part	Motion	Right	Left
Hip	Abduction	GV^-	GV^-
	Abduction	GV^-	GV^-
Knee	Flexion	GV	GV
	Extension	GV	GV
Ankle	Plantarflexion	GV	GV
	Dorsiflexion	GV	GV
Great toe	Plantarflexion	GV	GV
	Dorsiflexion	GV	GV
Hand Grip	-	GV^-	GV^-

Table 3 Results for the deep tendon reflex

Kinds of DTRs	Right	Left
Biceps	+	+
Brachioradialis	+	+
Triceps	+	+
Knee jerk	+	++
Ankle jerk	+	+

DTR, deep tendon reflex.

movement, rapid alternating movement, and heel to shin movement, which are cerebellar functions, were all intact, his tandem gait was without sway. His hands had atrophied, and he had gynecomastia.

The nerve conduction study (both limbs) showed low sensory nerve action potentials (SNAPs) over the F-W and the W-E segments on the left median and ulnar nerves, as well as low SNAPs on the bilateral sural and superficial peroneal nerves. F-waves were demonstrated on neither the bilateral median and left ulnar nerve nor the right ulnar nerve. H-reflexes were demonstrated bilaterally.

Electromyography (EMG, left limb, thoracic vertebrae part) revealed moderate to severe denervation potential with reduced interference pattern in the left abductor pollicis brevis muscle, moderate denervation potential with reduced interference pattern in the left extensor digiti communis muscle, and mild denervation in the right T10 paraspinal muscle. The EMG findings for the left tibialis anterior muscle and the left T10 paraspinal muscle were normal.

From October 29, 2013, to April 25, 2016, the patient was treated with 0.2 cc of Scolopendrid pharmacopuncture (Korean Pharmacopuncture Institutem, Korea) injected at each of the following acupoints: Joksamni (ST36), Susamni (LI10), Hapgok (LI4), Pungbu (GV16), and Pungji (GB20). From October 29 to November 9, 2013, he was prescribed a concoction of *Exsiccata lacca sinica* 10 g, *Semen strychni* 2 g, *Rhizoma smilacis* 10 g, *Flos lonicerae* 10 g, *Herba protulacae* 10 g, *Herba oldenlandiae diffusae* 10 g, *Radix polygalae* 2 g, and *Galla rhois* 10 g administered 30 minutes after eating meal (3 times a day). From October 29 to November 29, 2013, '*Aconibal Tab*', and '*Lactic acid bacteria*' were administered three times a day. After more than a year of outpatient treatment, the conditions and the symptoms of the patient remained unchanged.

3. Discussion

X-linked spinobulbar muscular atrophy (Kennedy's disease) affects muscles and motor neurons, manifesting as weakness and wasting of bulbar, facial, and proximal limb muscles due to loss of anterior horn cells in the brain and spinal cord [5]. As in this case, patients typically present in middle age with muscle cramps, bulbar weakness or leg weakness [6]. Muscle weakness, wasting, and fasciculations are generally slowly progressive, with some patients becoming wheelchair-dependent 20 to 30 years into the course of the disease [7]. Interestingly, in one large series of patients, nearly 100% of the patients showed abnormalities in their sensory responses on nerve conduction studies [6].

Unfortunately, no effective therapy for this condition is available; a recently concluded trial of dutasteride was unable to demonstrate any statistically significant effect on the patient's muscle strength versus those receiving a placebo [8].

Our patient received outpatient treatment, total of 44 times, from October 29, 2013, to April 25, 2016. No progression of the disease and no special change in condition were noted during treatment period. The patient was very satisfied with his response to treatment.

The limitations of this study include small sample size (one patient), uncontrolled design, and a dubitable before and after comparison. Because existing treatments would cause several considerable side effects and are difficult to apply in domestic clinics, we expect our findings, despite the limitations of the study, to lead possibly to new clinical guidelines.

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Conflict of interest

The authors declare that there are no conflicts of interest.

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