Received: 5 May 2018 | Revised: 11 September 2018 | Accepted: 17 September 2018

DOI: 10.1002/jgf2.213

CASE REPORT

Journal of General and Family Medicine Open Access WILEY

A case of amyotrophic lateral sclerosis which was diagnosed with progressive dysphagia and muscle atrophy

Tadashi Komata MD, PhD

Department of Rehabilitation, Niigata Rehabilitation Hospital, Niigata, Japan

Correspondence
Tadashi Komata, Department of Rehabilitation, Niigata Rehabilitation Hospital, Niigata, Japan.
Email: tdskmt5@yahoo.co.jp

Abstract

A 79-year-old man presented to our outpatient clinic with symptoms of dysphagia, dysarthria, and muscle atrophy of the trunk and upper extremities. These symptoms were gradually progressive, and he had lost substantial weight—20 kg in 2 years. One month later, he was admitted due to dehydration and received tube feeding. The presence of "split hand" suggested amyotrophic lateral sclerosis (ALS). Finally, the patient was diagnosed with ALS by two neurologists. When elderly patients present with progressive dysphagia and muscle atrophy, especially with "split hands," ALS should be included as a differential diagnosis.

KEYWORDS

amyotrophic lateral sclerosis, dysarthria, dysphagia, muscle atrophy, split hand, weight loss

1 | INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a common and well–recognized motor neuron disease that involves degeneration of both upper and lower motor neurons, leading to progressive muscular paralysis. Death typically occurs within 1-5 years after the onset of the disease.[1] Here, we describe an elderly patient who exhibited "split hand"[2,3] with progressive dysphagia and muscle atrophy, which suggested a diagnosis of ALS.

2 | CASE REPORT

A 79-year-old man developed dysarthria, dysphagia, and weakness of the hands 1 year before admission, which gradually progressed. Therefore, 6 months later, he was referred to a neurosurgical hospital. A definitive diagnosis was not confirmed even after brain computed tomography (CT). Five months later, he presented to our outpatient clinic. One month postpresentation, he was admitted to our hospital owing to appetite loss and dehydration. He had lost substantial weight (20 kg; starting weight: 75 kg; final weight: 55 kg; body height: 158 cm) in 2 years. He had no family history of neurodegenerative disease.

On admission, he was alert (Glasgow Coma Scale score: 15). We observed atrophy and fasciculation of the tongue; dysphagia; and dysarthria. Other cranial nerves were normal. Muscle atrophy was observed (Table 1). The first dorsal interosseous muscles and thenar eminence muscles showed "split hand"[2,3] signs bilaterally (Figure 1). Gripping power (kg) was 0/0. MMT findings were as follows: deltoid (3/3), biceps (4+/4+), triceps (4+/4+), wrist ext. (3−/3−), wrist flex. (3+/3+), opponens pollicis (3/3), iliopsoas (4+/4+), quadriceps (4+/4+), hamstrings (4+/4+), tibialis anterior (5/5), and gastrocnemius (5/5). The muscle tone was flaccid in all four extremities. Neither sensory impairments nor cerebellar ataxias were observed. Deep tendon reflexes were as follows: upper extremities (2+/2+) and lower extremities (3+/3+). Abnormal reflexes/fasciculation details are shown in Table 1. No bladder or rectal disturbances, nor extrapyramidal disorders were observed. He could walk using a U-shaped walker. All other physical examination results were unremarkable.

The differential diagnoses were ALS, brain-stem tumor, cervical spondylosis, multiple sclerosis, chronic inflammatory demyelinating

TABLE 1 Summary of the clinical signs on admission

	Lower motor neuron signs	Upper motor neuron signs
Brain stem	Muscle weakness and atrophy of the facial muscle and tongue Fasciculation of the tongue	Jaw jerk reflex (hyper) Snout reflex (+)
Cervical cord	Muscle weakness and atrophy of the neck and upper extremities and hands Fasciculation of upper extremities	Not apparent
Thoracic cord	Muscle weakness and atrophy of the upper and middle trunk	Not apparent
Lumbosacral cord	Muscle weakness and atrophy of the lower trunk and lower extremities Fasciculation of lower extremities	Babinski reflex (+)

polyneuropathy, progressive muscular atrophy, myasthenia gravis, and myopathy.

Precontrast brain CT and cervical X-ray film excluded other intracranial or cervical abnormalities. Electrophysiological examinations of the bilateral median and ulnar nerves revealed mild reduction in amplitude in motor nerve velocity tests; no abnormality was recorded in sensory nerve velocity tests. Brain MRI was not performed because he had a DDD pacemaker. Lumbar puncture was not executed because he had lumbar spondylosis. Hence, the aforementioned differential diagnoses could not be completely excluded.

The final diagnosis was made by two neurologists based on the Awaji algorithm, combined with the El Escorial criteria, which are considerably useful for early ALS diagnosis.[4] The diagnostic grade was "probable."[4]

Regarding rehabilitation, because there is no effective training for symptoms of ALS, we modified his daily environment.[1,5] For 1 month after admission, his muscle weakness progressed noticeably. He could not walk, despite staff assistance. He experienced breathing difficulty and received frequent intratracheal aspiration. Two months after admission, he was transferred to a center for intractable neural disease. Three months after admission, he died of acute hypercapnic respiratory failure.

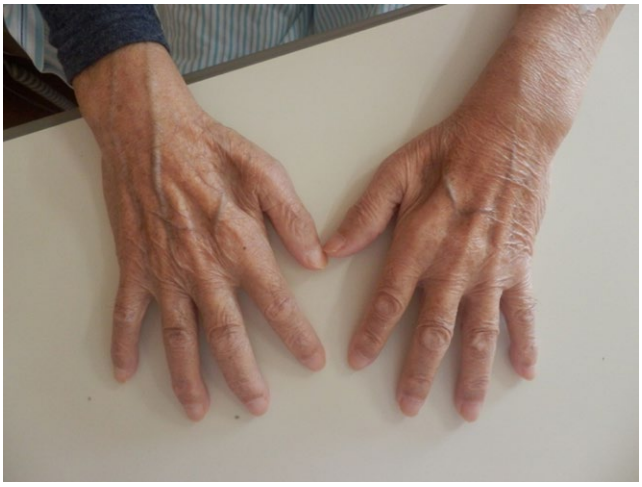

FIGURE 1 Picture of dorsal side of both hands showing atrophy especially of the first dorsal interosseous muscles and thenar eminence muscles

3 | DISCUSSION

To definitively diagnose ALS, initial diagnosis should be confirmed by a neurologist. However, it is important for the attending physician to collect relevant medical information before consultation with the neurologist.[1,6] In this case, positive signs were apparent and progressive: Negative signs were consistent with ALS. Therefore, when ALS is suspected, diagnosis may not be difficult. An important consideration is the need for suspicion of this disease at an earlier stage of its progressive clinical course. Chio suggested that the diagnostic process for ALS may be excessively prolonged: The median time from onset to diagnosis was 12-17 months.[7] The reasons included the presence of other diseases, misinterpretation of examination findings, and lack of familiarity with ALS.[7] In our case, "lack of familiarity" was thought to have delayed the diagnosis. However, the suspicion of ALS was greatly enhanced owing to the observed "split hand" sign, which is a highly specific ALS symptom.[2,3]

Murphy et al[8] reported that ALS incidence rates steadily increased by 3% per year over the 22 years (from 1985 to 2006). They confirmed older age, male sex, and bulbar onset as adverse prognostic factors.[8] Furthermore, Tanaka et al[9] suggested that patients with late-onset ALS showed more rapid disease progression than those with early-onset ALS using the progression rate. Our case is "late-onset ALS with bulbar onset," which showed rapid disease progression.

In summary, when physicians encounter elderly patients with progressive dysphagia and muscle atrophy, they should consider that those patients may have neurodegenerative diseases: Thus, they should examine whole–body condition and function, including muscle mass and strength.

ACKNOWLEDGEMENTS

I especially thank all the nursing and rehabilitation staff for their devoted and capable nursing care and rehabilitation. I greatly appreciate Dr. Keiko Tanaka and Dr. Toru Imamura for the diagnosis

of this patient. I also appreciate Dr. Nao Sanada, Dr. Seiko Ishikawa, Dr. Tatsuya Kikuchi, and Dr. Yoko Sakimura for discussion regarding rehabilitation. I greatly appreciate Dr. Tatsuya Sato for providing hospital care after transferring to Agano Hospital.

CONFLICT OF INTERESTS

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

REFERENCES

1. Francis K, Bach JR, DeLisa JA. Evaluation and rehabilitation of patients with adult motor neuron disease. Arch Phys Med Rehabil. 1999;80(8):951–63.
2. Wilbourn AJ. The split hand syndrome. Muscle Nerve. 2000;23:138.
3. Kuwabara S, Mizobuchi K, Ogawara K, Hattori T. Dissociated small hand muscle involvement in amyotrophic lateral sclerosis detected by motor unit number estimates. Muscle Nerve. 1999;22:870–3.
4. Carvalho MD, Swash M. Awaji diagnostic algorithm increases sensitivity of El Escorial criteria for ALS diagnosis. Amyotroph Lateral Scler. 2009;10(1):53–7.
5. Strand EA, Miller RM, Yorkston KM, Hillel AD. Management of oral-pharyngeal dysphagia symptoms in amyotrophic lateral sclerosis. Dysphagia. 1996;11(2):129–39.
6. Paganoni S, Karam C, Joyce N, Bedlack R, Carter GT. Comprehensive rehabilitative care across the spectrum of amyotrophic lateral sclerosis. NeuroRehabilitation. 2015;37(1):53–68.
7. Chiò A. Update on ISI survey: Europe, North America and South America. Amyotroph Lateral Scler Other Motor Neuron Disord. 2000;1(Suppl 1):S9–11.
8. Murphy M, Quinn S, Young J, Parkin P, Taylor B. Increasing incidence of ALS in Canterbury, New Zealand: 22-year study. Neurology. 2008;71(23):1889–95.
9. Tanaka Y, Yoshikura N, Harada N, et al. Late-onset patients with sporadic amyotrophic lateral sclerosis in Japan have a higher progression rate of ALSFERS-R at the time of diagnosis. Intern Med. 2012;51:579–84.

How to cite this article: Komata T. A Case of amyotrophic lateral sclerosis which was diagnosed with progressive dysphagia and muscle atrophy. *J Gen Fam Med*. 2019;20:25–27. https://doi.org/10.1002/jgf2.213