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Case report

Amyotrophic lateral sclerosis with a sudden-onset history

Takamichi Kanbayashi [a,*], Takashi Mikata [b], Yuki Hatanaka [a], Masahiro Sonoo [a]

[a] Department of Neurology, Teikyo University School of Medicine, Kaga 2-11-1, Itabashi-ku, Tokyo 1738605, Japan
[b] Department of Neurology, National Hospital Organization National Shimoshizu Hospital, Yotsukaido, Chiba 2840003, Japan

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ABSTRACT

Objective: We report on two patients with amyotrophic lateral sclerosis (ALS) complaining of sudden-onset difficulty in finger elevation.
Case report: A 65-year-old man (the first patient) and a 66-year-old man (the second patient) suddenly became aware of difficulty in finger elevation of one hand. They were not aware of any other symptoms prior to the onset. In the first patient, cerebral infarction at the precentral gyrus was initially suspected. In the second patient, cervical spondylosis was initially suspected, and cervical spine surgery was planned. However, needle EMG revealed widespread neurogenic changes and abundant fasciculation potentials for both patients. Widespread weakness emerged in time and relentlessly progressed, and finally the diagnosis of ALS was made. In both cases, notable weakness in the extensor digitorum (ED) muscle with relatively mild weakness in the other muscles in the affected limb was a characteristic finding. Loss of one motor unit in ED that has already enlarged due to reinnervation must have caused sudden awareness of the weakness.
Significance: Clinicians should recognize the presence of ALS patients with a sudden-onset history because the risk of initial misdiagnosis is high for such patients.

1. Introduction

Amyotrophic lateral sclerosis (ALS) is a chronic progressive neurological disorder, and is naturally believed to develop insidiously. To the best of the authors' knowledge, there are no previous reports of an ALS patient whose complaint was a sudden-onset weakness. We herein report the cases of two ALS patients who noticed a sudden-onset difficulty in finger elevation.

2. Case report

2.1. Patient 1

A previously healthy 65-year-old man suddenly became aware of difficulty in elevating his right index finger when using a computer in his office. He was carefully interviewed, but reported that he did not notice any other symptoms, such as weakness or twitching, prior to the onset of the symptom. He visited an orthopedic clinic, where a cervical spine X-ray and MRI showed no abnormalities. At 16 days after the onset of the symptom, he was referred to our hospital because the symptom failed to improve.

Neurological examinations revealed predominantly distal weakness confined to the right upper limb, which could not be explained by the segmental or peripheral nerve distribution (Table 1). Notably, the extensor digitorum (ED) displayed profound weakness with manual muscle testing (MMT) 2, which was defined as the patient was only able to partially extend the metacarpophalangeal joint. Deep tendon reflexes were normal, and the Babinski sign was indifferent. The other neurological findings were normal. Cerebral infarction at the precentral gyrus was suspected based on the distribution of muscle weakness and sudden-onset of the patient's symptoms. However, brain MRI was normal. Needle electromyography (EMG) at 3 days after admission revealed a definitively reduced recruitment pattern with profuse denervation potentials in ED, and denervation potentials in a number of muscles from C6 to T1 segments. Denervation potentials were not observed in the T1 paraspinal muscle. Nerve conduction studies revealed that the amplitude of the compound muscle action potential of the extensor indicis was reduced; no signs of demyelination were observed in any of the examined nerves.

Based on the presence of widely-distributed neurogenic weakness of acute-onset, the patient was diagnosed with neuralgic amyotrophy without pain. The patient was treated using steroid pulse therapy (methylprednisolone 1000 mg/day for 3 days). However, the weakness progressed, and deep tendon reflexes in the right upper limb became hyperactive. One month later, repeated

* Corresponding author.
E-mail addresses: ta_kanba@yahoo.co.jp (T. Kanbayashi), mikata-neu@umin.ac.jp (T. Mikata), y-hata@med.teikyo-u.ac.jp (Y. Hatanaka), sonoom@med.teikyo-u.ac.jp (M. Sonoo).

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Table 1
The manual muscle testing findings in the present cases.

	Case 1		Case 2	
	MMT[1] (right/left)	1 year later	MMT (right/left)	6 months later
Deltoid	5/5	3/4	4/5	3/5
Infraspinatus	4/5	4/5		
Serratus anterior	5/5		5/5	
Biceps brachii	5/5	3/4	5/5	4/5
Triceps brachii	5/5	3/4	4/5	4/4
Wrist extensor	4/5	1/4	3/5	2/4
Wrist flexor	4/5	2/5	4/5	4/5
Extensor digitorum	2/5		2/5	0/4
Extensor pollicis brevis	4/5		4/5	
Extensor pollicis longus	4/5		4/5	
Flexor pollicis longus	4/5		4/5	2/5
Flexor digitorum profundus (index)	4/5		5/5	4/4
Flexor digitorum profundus (little)	4/5		4/5	4/4
Abductor pollicis brevis	4/5		5/5	4/4
First dorsal interossei	4/5		4/5	3/4
Abductor digit minimi	4/5		4/5	2/4

[1] Manual muscle testing.

EMG revealed fasciculation potentials in the biceps brachii and triceps brachii muscles. Finally, a diagnosis of ALS was made based on the progressive clinical course and the needle EMG findings. One year later, the weakness showed further progression with left upper limb involvement (Table 1).

2.2. Patient 2

A previously healthy 66-year-old man suddenly became aware of difficulty in elevating his right middle finger on one day. Despite being asked repeatedly, the patient reported that he was not aware of any other symptoms prior to this complaint. The weakness remained unchanged for three months, although he came to notice widespread muscle twitching in all four limbs and his trunk. He visited an orthopedic clinic four months after the onset of symptoms. Cervical MRI revealed spinal canal stenosis from C3 to C7. Cervical spine surgery was planned, although he was referred to us to rule out ALS.

Neurological examinations revealed mild weakness (MMT 4) in the right upper limb muscles from myotomes C5 to C8, and marked ED weakness (MMT 2; Table 1). Deep tendon reflexes of his left upper limbs were slightly hyperactive. The other neurological findings were normal. The tentative diagnosis after neurological examinations was cervical spondylotic amyotrophy (CSA). However, needle EMG revealed definite neurogenic changes in the flexor digitorum superficialis muscle belonging to the T1 myotome (Chiba et al., 2015), which was not clinically affected. Abundant fasciculation potentials were observed in the triceps and the trapezius muscles. ALS was strongly suspected based on the needle EMG findings. We therefore suggested that the orthopedists should refrain from surgery.

Six months later, the patient was again referred for EMG. Weakness in the right upper limb had worsened, and the left upper limb had become slightly weak (Table 1). However, the patient's lower limb muscle strength was normal and no upper motor neuron signs were observed. Finally, we diagnosed the patient with brachial amyotrophic diplegia-type ALS (Katz et al., 1999).

3. Discussion

These two patients complained of sudden-onset difficulty in elevating a finger. Despite a careful interview, they did not notice any symptoms prior to the onset of this symptom. Marked weakness of the ED with relatively mild weakness of the other muscles in the affected limb was a characteristic finding in both cases.

It is unlikely that the disease process of ALS actually began suddenly. The truth was that the weakness was just perceived suddenly, although the motor unit loss must have begun insidiously prior to the awareness of the weakness. We hypothesize that the patients were unaware of their symptoms because finger extensor weakness has little influence on daily life until an obvious limitation in the range of motion occurs and the ability in opening the hand from a fist is impaired. Prior to clinical onset, substantial motor neuron loss and subsequent reinnervation may well have already occurred, resulting in the increased innervation ratio of the remaining motor units. Then, the loss of a single motor neuron may have resulted in a considerable loss of power, crossing the threshold for efficient finger elevation, which caused the patients to perceive a "sudden-onset" difficulty in finger elevation.

The relatively mild weakness in the muscles other than ED, including small hand muscles, may also have contributed to the perception of isolated ED weakness. Such a distribution of weakness is rather atypical for ALS. Commonly, the weakness of an ALS patient is prominent in small hand muscles, especially those on the radial side, resulting in the split-hand syndrome (Wilbourn, 2000), which was not even observed in the present patients (Table 1). A sudden-onset presentation may be a feature of such a rather rare type of ALS showing ED weakness as the initial main symptom.

In such cases, the risk of an initial misdiagnosis is high. In fact, stroke was initially suspected in the first patient, and the patient was subsequently misdiagnosed with neuralgic amyotrophy. Sudden-onset weakness in the upper-limb may also be confused with CSA that often presents with acute-onset weakness (Sonoo, 2016). This was the case in the second patient and the cervical spine surgery was actually planned before the referral to us. Inappropriate neck surgery is hazardous for patients with ALS (Pinto et al., 2014), and therefore it is important to recognize the existence of ALS with a sudden-onset history.

Conflict of interest

No conflict of interest.

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