Case Report

Recovery Phase Rehabilitation for Atypical Amyotrophic Lateral Sclerosis: A Case with Lumbar Vertebral Fracture Complications

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Abstract: Amyotrophic lateral sclerosis is a rapidly progressive disorder involving degeneration of motor neurons. Excessive exercise impact can actually decrease muscle strength. We would like to report our success with a patient in whom home-based convalescence had been difficult due to a lumbar vertebral fracture. She was able to return home after appropriate adjustments. The patient was a 64-year-old woman who became aware of diminishing muscle strength in her extremities 12 years previously and was diagnosed with amyotrophic lateral sclerosis. She had fallen at home and suffered a lumbar vertebral fracture. After a month of inpatient therapy at another hospital, she was transferred to our institution. She tired easily and could not perform muscle strength training, so her program was adjusted so that walking, which was not as exhausting for her, was the focus of the training. Gradually, her fatigue began to lessen, and she was able to undergo muscle strength training. Ultimately, she became independent enough to conduct her activities at home, allowing her to be discharged home. Even in cases such as ours where the patient initially finds the training too challenging, implementing alternative exercises such as the use of repetitive low impact training can lead to improvements in a patient's activities of daily living.

Key Words: Amyotrophic lateral sclerosis, Lumbar vertebral fracture, Rehabilitation.

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Introduction

Amyotrophic lateral sclerosis (ALS) is a refractory disease of unknown cause where both upper and lower motor neurons degenerate, leading to progressive muscle weakness. Motor neuron degeneration has been reported to halve the number of motor units in just half a year[1], leading to progressive limb muscle weakness and bulbar paralysis. By approximately 3 years after onset, many patients develop walking difficulties and dysphasia, and ultimately, many of the patients die of respiratory failure. Elisabetta et al. reported that the 1-year/5-year/10-year survival rates in 483 cases of ALS were 76.2%/23.4%/11.8%, respectively[2], and that there are slowly progressing cases where survival beyond 10 years is possible. Activities of daily living (ADL) are compromised as a result of ALS making home convalescence difficult, so rehabilitation must be carried out in an inpatient setting. However, excessive exercise loads during rehabilitation can actually lead to overwork weakness and may even hasten the progression of ALS[3]. In addition, the primary disease may progress during hospitalization so that eventually, instead of improved ADL functioning, the patient can no longer be discharged home. We would like to report our experience with a patient who had atypical slowly-progressive ALS that was complicated by a lumbar vertebral fracture in whom we instituted recovery stage rehabilitation. By adjusting her exercise treatment appropriately, overwork weakness was avoided and the patient was discharged home.

Case Report

A 64-year-old woman

Past medical history

Breast cancer

Family history

Her younger brother was diagnosed with ALS.

Social background

She lives with her husband and eldest son. Before the injury, she was able to walk for 20 meters indoors using a wheeled walker or by holding on to walls, while outdoors she was able to walk for about 100 m with her husband's help. As her symptoms progressed, she often fell even inside her home. In terms of ADLs, she required assistance in shampooing, bathing, and dressing, and her husband took over the housework. She was designated as requiring Long-Term care level 2, and although her home had been renovated for her needs, there were still areas with level differences but with no railings, such as the entranceway, and neither the toilet nor bathroom had railings.

History of present illness

The patient became aware of diminishing muscle strength in her extremities in 2002, and was diagnosed with ALS by a neurologist. Because her disease progressed slowly, she received electrophysiological testing in order to rule out other diagnoses.

On EMG, giant spikes were observed for the tongue and for the upper and lower extremities

broadly, and only some fibrillation potentials were observed in contrast. These findings were interpreted as a disorder of anterior horn cells, and it meant that there were lower motor neuron disorders in her upper and lower extremities. For tendon reflexes, other than a reduced lower jaw reflex and patellar tendon reflex hyperactivity, deep tendon reflexes at other sites were normal. Babinski reflex was negative. Although the lower motor neuron disorder was found by EMG, upper motor neuron disorder findings were limited to physical findings. According to the revised El Escorial criteria[4], she was diagnosed clinically with possible ALS. As disease progression was slower than the typical case, hers was considered to be a case of atypical ALS. Lung function tests were conducted at the same time: FVC was 1.96l, %VC was 71.3%, and she exhibited a restrictive disease pattern.

In September 2012, she fell while adjusting a built-in fan and injured herself at home. Severe dorsal lumbar pain made it difficult for her to move her body, and she was taken to an emergency hospital. She was diagnosed with an L1 vertebral fracture and was hospitalized for treatment. Although conservative treatment with a hard corset was instituted, she could no longer be discharged home due to further decreases in ADL function, so in October of the same year, she was transferred to our hospital in order to undergo inpatient rehabilitation training.

Status on admission to our institution

No apparent impairment of mental function was noted. Mild dysarthria, as well as tongue atrophy and fasciculations, was noted. Signs of limb muscular atrophy and muscular weakness were present with a 2/5 score on a manual muscle test (MMT) of the shoulder joint muscles and 3/5 score in all other muscles tested in the upper limb. In the lower limbs, the hip girdle muscle was 2/5, the knee extensor was 4/5, and plantar dorsiflexion was 2/5 (Table 1). Grip strengths were: right-5.1kgf; left-＜5kgf. There was hyperreflexia of the patellar tendon. There was no observation of sensory impairment, bladder or bowel incontinence, eye movement disorder, or pressure ulcers. Upper motor neuron signs and lower motor neuron signs were present, and the patient met the diagnostic criteria for possible ALS. There were hardly any complaints of lumbar pain, and she could move to her wheelchair with assistance.

Table 1. MMT at admission and at discharge

		admission		discharge	
		Right	Left	Right	Left
shoulder	flexion	2	2	2	3
	abduction	2	2	2	3
elbow	flexion	3	3	3	3
	extension	3	3	3	3
hip	flexion	3	3	3	3
	extension	3	3	2	2
	abduction	2	2	3	3
knee	flexion	4	4	4	4
	extension	4	4	4	4
ankle	dorsiflexion	2	2	3	3
	plantarflexion	2	2	2	2

MMT: manual muscle test

SpO2 at rest on room air was 95%. Her scores on the Simple Test for Evaluating Hand Function (STEF) were 71 points on the right and 85 points on the left (standard cutoff points at 60 years of age: 88 points). At admission, her functional independence measures (FIM) were all decreased with the exception of sphincter control and self-care, while the motor score subtotal was 61 points and cognitive subtotal was 35 points. In the lumbar spine x-ray lateral view at the time of admission, 34% collapse was noted in the anterior column of L1. Callus was already formed, and her course of treatment was uncomplicated (Fig. 1). Due to her mild dysphagia, we used a thickener in administering a video fluoroscopic swallowing study (VFSS). She exhibited slight defects in chewing, transport, and bolus formation, but an induced delay of the swallowing reflex was not observed. Laryngeal elevation was mildly defective, and mild pharyngeal residue was also detected. By adding a thickening agent to the barium meal solution we assured there was no aspiration.

Issues

This patient's potential issues were as follows.

Functional disorders included: (1) muscle weakness (2) decreased endurance; and (3) finger dexterity impairment.

Decreased functioning was noted in: (1) difficulty walking; and (2) decreased ADL ability.

Social disadvantages were: (1) home environment; and (2) home convalescence difficulties.

Treatment protocol

Physiotherapy included muscle strength training and walking training, while occupational therapy focused on ADL training. While taking care to avoid muscle fatigue due to overuse, muscle strength was checked on the day after training to adjust training intensity accordingly. Treatment goals were to walk indoors with modified independence, and perform all ADL (other than bathing) with modified independence so that she could be discharged home.

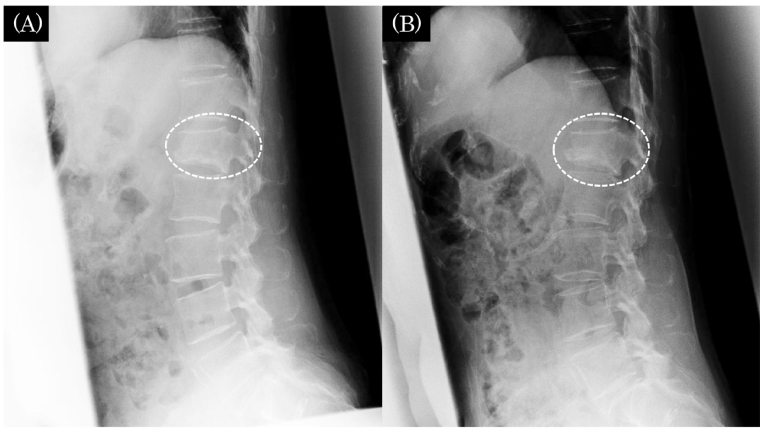

Fig. 1. Radiography of lumbar spine.
(A) A compression fracture of the 1st lumbar vertebra was seen on admission.
(B) Callus formation at the fracture site was seen at discharge.

Course of Treatment

It was necessary for our patient to continue wearing the brace for her lumbar compression fracture. Initially, just 50 repetitions of standing and sitting exercises led to residual fatigue. Due to this residual fatigue and complaints of shortness of breath, muscle strength training could not be carried out the following day. She was able to perform walking training with few complaints. In order to maintain a certain degree of training, the treatment protocol was changed so that rehabilitation training would focus on walking exercises. After about 3 weeks, the patient's resilience improved and with fewer complaints of shortness of breath, she became able to perform muscle strength training with 0.5 kg weights doing 2 sets of 10 repetitions each. There was no progression of her lower back pain or collapsed vertebra via simple x-ray. Because bone union was observed, the corset was discontinued three months after her initial injury. Her complaints of shortness of breath disappeared, and it was possible to initiate an aggressive muscle strength training program. Ultimately, muscle strength measurements of left shoulder joint flexion and abduction, bilateral hip joint abduction, and right ankle dorsiflexion improved (Table 1). Grip strength increased from 5.1 kgf to 6.7 kgf in the right hand but the left hand remained at less than 5 kgf for the duration of her hospital stay. She eventually could walk alone for 100 m using a walker wheeler, and with help from her husband, she could even walk for 50 m with a T-cane. STEF deteriorated from 72 points to 56 points on the right and 85 points to 64 points on the left, in detailed mention evaluation items using small things, such as a pin mainly decreased. But there was no impact on her ADL performance. FIM motor items included grooming, dressing, toileting, transfers, locomotion, and stair climbing, while her total score improved from 61 to 95 points (Table 2). Her home environment was also improved by placing handrails at the entranceway, toilet, and bathroom, and having achieved modified independence in ADL function at home, she was discharged home on Day 86 of hospitalization.

Table 2. Motor FIM Scores

	admission	discharge
eating	6	6
grooming	7	7
bathing	1	4
upper dressing	4	5
lower dressing	4	5
toileting	5	7
bladder management	7	7
bowel management	7	7
bed/chair transfer	5	6
toilettransfer	5	6
tub/shower transfer	1	4
walk	5	6
stairs	4	5
total motor FIM score	61	75

FIM: functional independence measure

Discussion

Increased muscle weakness related to overuse can occur in ALS as well as in the post-polio syndrome, and this results from excessive loading in muscles with reduced nerve function due to lost collateral branches within nerve fibers[5)6)]. By attempting to increase baseline muscle strength through strength training, it is possible to exacerbate the nerve degeneration[6)]. ADL functions may, however, improve if muscle strengthening exercises are implemented while taking care to avoid overwork weakness[3)7)]. Recent reports have found exercise therapy can improve functioning and delay disease progression, thus alleviating the burden of needed caretaking[8)], and more papers have reported implementing outpatient exercise therapy[9)10)]. Unfortunately, there is no consensus on the optimal exercise intensity, clinicians debate whether exercise therapy should be enforced or avoided, and the efficacy of exercise has not yet been established. If the disease progresses rapidly, not only is there no hope of improving activities of daily living (ADL) performance, but even if the patient is hospitalized long-term for rehabilitation, the patient's ADL function may deteriorate during that time due to ALS progression, making it difficult for the patient to be discharged home. Our patient had atypical slowly-progressive ALS that was complicated by a lumbar compression fracture, and primary disease progression during hospitalization was limited. By careful adjustment of exercise therapy, it was possible to achieve improvements in ADL function which allowed this patient to be discharged home.

In lumbar vertebral fractures, reports suggest that compared to inpatient therapy, outpatient therapy may decrease the risk of compromising ADL function[11)]. Therefore in hospitalized patients, it is necessary to initiate early mobilization and aggressive exercise to minimize decreases in ADL due to disuse.

In this case, in addition to the lumbar vertebral fracture, the patient had already suffered a decrease in ADL performance due to ALS itself before the injury, making home recuperation difficult and requiring hospitalization during the acute phase. In addition to the mild decrease in respiratory function, as exemplified originally by 95% SpO_2 at rest, it was necessary for our patient to wear a brace for the treatment of her lumbar vertebral fracture. She complained of shortness of breath after even light exercise. This is a slowly progressing case where the patient was already more than 10 years past ALS onset. That is the reason why the contribution of ALS to ADL decline at the time the patient was transferred to our rehabilitation hospital was considered minor. Decreases in ADL functioning that had been noted when the patient was transferred to our hospital were primarily the result of inactivity during hospitalization in an acute care institution for the fracture. Furthermore, learned nonuse, due to inactivity and decreased resilience[12)13)], led to disuse which compromised movements that should have been possible.

Exercise therapy for ALS includes moderate resistance training, and there are reports that improved muscle strength and endurance can be achieved with such moderate resistance training[9)10)], and so initially the patient had been scheduled for muscle strength training which included resistance training. Nevertheless, restriction of thoracic movement due to the patient's corset resulted in shortness of breath, which prevented us from completing our schedule of muscle strength training. Because the admission x-ray showed mild callus formation and collapse, pain was not a limiting factor for rehabilitation, and we were able to remove the brace at an early stage. For that reason there was an original deterioration of respiratory function, however we believed it possible to increase the amount

of training. Thus, the plan was changed to one with repeated walking exercise that the patient found less tiring and which led to improved endurance. As a result, it was now possible for high frequency muscle strength training to have an impact. Functional impairment and disability were improved by training, and our patient was able to be discharged home within the 90-day deadline of her admission to our convalescent rehabilitation facility.

We did not note any signs of overwork, such as fatigue, muscle pain, or soreness after exercise. STEF deteriorated in this period, in contrast to improved function, but we did not consider rehabilitation or disease progression as a cause. We consider this may be from 'learned-nonuse' in the hand. Usually, ALS rehabilitation targets large joints[14], such as the shoulder, knee, or ankle, and adaptive equipment is applied for hand impairment. Hand exercises might have been insufficient in our case. We mainly used exercise hours for muscle training, as we had greater concern for ADL.

Even if the patient had developed learned nonuse due to atypical ALS complicated by lumbar compression fractures, if the ALS progression is slow, with repeated low impact training, it was thought that it would be possible to break free from the vicious cycle of learned nonuse and to improve ADL performance.

Conflicts of Interest: None exist

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〈和文抄録〉

腰椎圧迫骨折を合併した非典型的筋萎縮性側索硬化症に回復期リハビリテーションを行った1例

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筋萎縮性側索硬化症は発症後平均約3年で呼吸不全から死に至る，急速進行性の運動ニューロン変性疾患である．自宅療養の継続にリハビリテーションは有効とされるが，過剰な運動負荷は筋力の低下を招くことも知られており，疾患の進行に配慮した訓練内容の設定が必要である．また入院を余儀なくされると，疾患自体の進行により退院困難になることがある．腰椎圧迫骨折の受傷により入院し退院が困難であったが，リハビリテーションの工夫により自宅退院が可能になった1例を報告する．64歳女性．12年前に四肢の筋力低下を自覚し，筋萎縮性側索硬化症と診断され，日常生活では夫の介助を要していた．自宅で転倒して腰椎圧迫骨折を受傷し，他院で1ヵ月間入院加療をうけた後，当院に転院した．疲労感のため筋力増強訓練が行えず，低負荷の歩行訓練を中心に行うよう訓練内容を変更した．次第に疲労感は軽減し筋力増強訓練が可能になり，最終的に動作が自立したため，自宅への退院が可能になった．本症例のように訓練に困難をともなう筋萎縮性側索硬化症の症例であっても，低負荷訓練を高頻度に行うなど訓練内容の工夫で，日常生活活動が改善することがあり，積極的な介入が必要と考えた．

キーワード：筋萎縮性側索硬化症，腰椎圧迫骨折，リハビリテーション．