Case Report

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A Comprehensive Rehabilitation Approach in a Patient With Serious Neuropsychiatric Systemic Lupus Erythematosus

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Neuropsychiatric systemic lupus erythematosus (NPSLE) involves the central and peripheral nervous system in patients with systemic lupus erythematosus (SLE). It is essential to specify the problems faced by patients with NPSLE because it causes diverse disabilities and impairs quality of life. After performing a comprehensive evaluation, tailored management should be provided for the patient's specific problems. We report here the case of a 30-year-old female with SLE who experienced serious neuropsychiatric symptoms cerebral infarction followed by posterior reversible encephalopathy syndrome and peripheral polyneuropathy. We systemically assessed the patient using the International Classification of Functioning, Disability and Health model as a clinical problem-solving tool and provided comprehensive rehabilitation by focusing on her problems.

Keywords Neuropsychiatric systemic lupus erythematosus (NPSLE), International Classification of Functioning, Disability and Health, Rehabilitation

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease of unknown cause that can affect the skin, joints, nervous system, and/or other organs. Arthritis and skin diseases are the most frequent manifestations of SLE [1], and the main treatments have focused on those symptoms by using medications. Neuropsychiatric systemic lupus erythematosus (NPSLE) can involve the central and/or peripheral nervous system and encompasses a variety of neurological and psychiatric features. Although NPSLE causes diverse disabilities and impairs quality of life, most symptoms and signs are underdiagnosed and undertreated [2]. There are few studies of rehabilitation in patients with NPSLE [3]. We report here a case of NPSLE with severe symptoms. We evaluated

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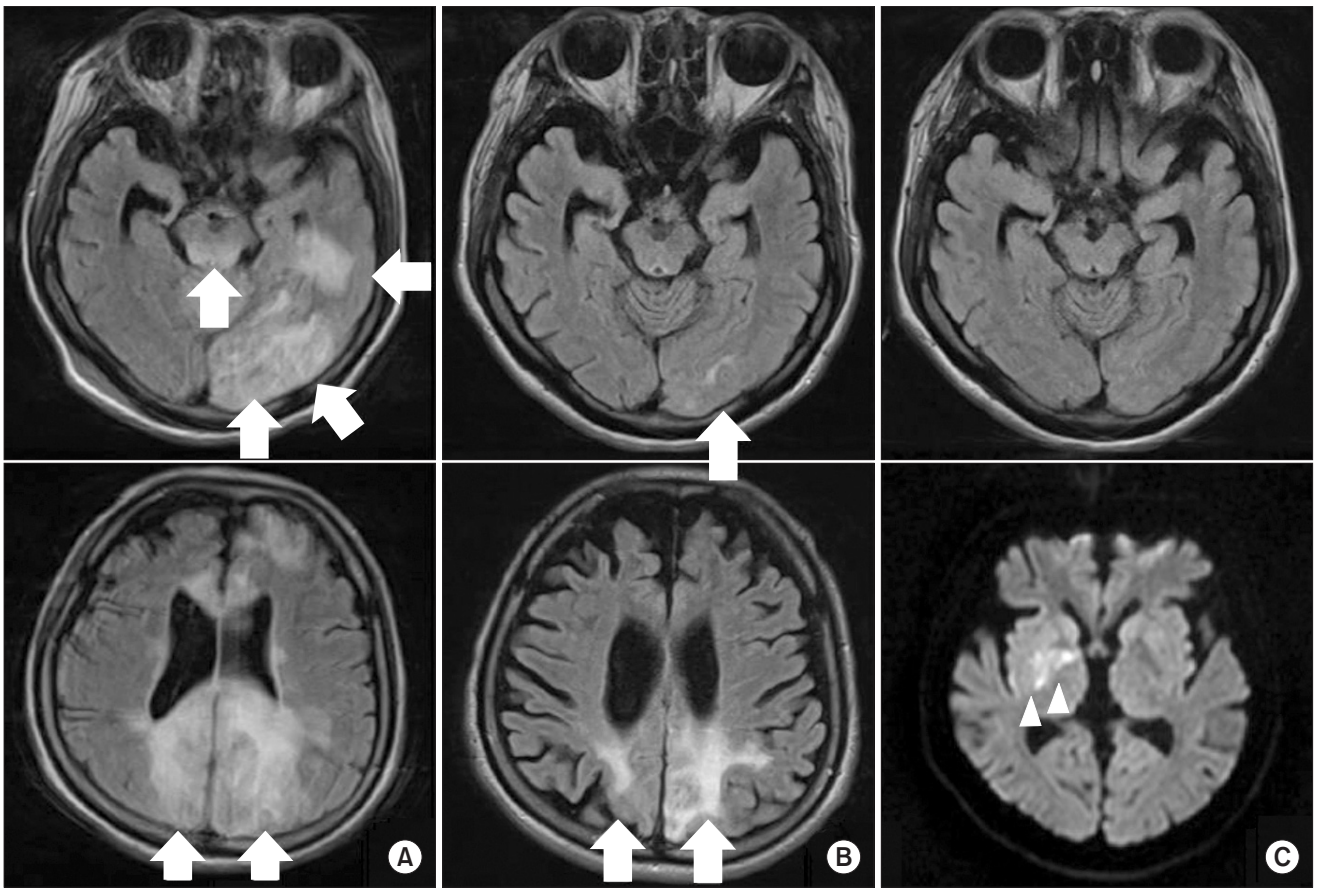

Fig. 1. Sequential brain magnetic resonance images. A T2-fluid attenuation inversion recovery image shows multiple high-signal-intensity lesions in the left temporal and frontal lobes, both occipital lobes, and the midbrain (A). Fifteen days later, the previously noted lesions were markedly resolved (B). Although the previously noted lesions were completely resolved, acute right basal ganglia infarction (arrowheads, C) was seen on brain diffusion magnetic resonance imaging 1 week later.

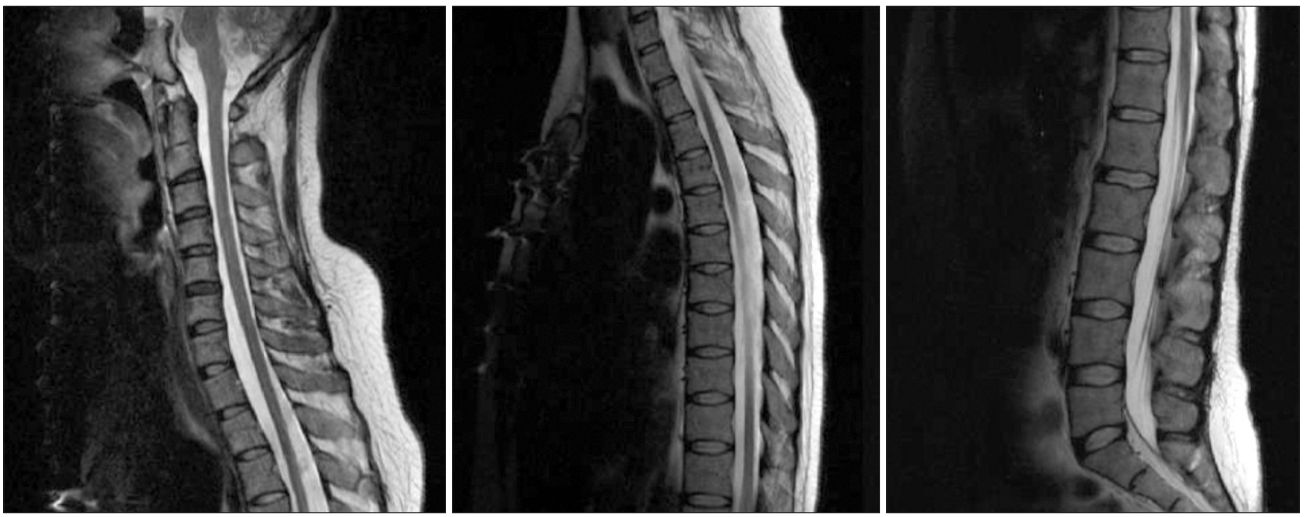

Fig. 2. Whole-spine magnetic resonance image. The cross-sectional area of the whole spinal cord was markedly thin on the sagittal T2-weighted image.

multiple facets of the patient using the International Classification of Functioning, Disability and Health (ICF) model, and provided comprehensive rehabilitation management.

CASE REPORT

A 30-year-old female was diagnosed with SLE at 12 years of age. At around 27 years of age, she experienced several instances of seizures and began taking antiepileptic drugs. On brain magnetic resonance imaging (MRI), diffuse cortical atrophic changes and an old infarct lesion in the left superior parietal cortex were detected. Three years later, she had fever, clonic seizure and loss of consciousness. On T2-fluid attenuation inversion recovery (FLAIR) imaging, a high-intensity signal change was detected in the left temporal and frontal lobes, both occipital lobes, and the midbrain (Fig. 1A). She was also diagnosed with antiphospholipid syndrome. After 15 days of treatment, which included intravenous methylprednisolone, immunoglobulin and anti-epileptic drugs, she regained consciousness and her motor weakness was

Table 1. ICF codes of the patient's symptoms and their management

ICF code	Description	Management
b7304	Power of muscles of all limbs	Passive and active range of motion exercise, strengthening exercise
d410	Changing basic body position	Rolling over training, sitting balance training, tilt table standing training
d415	Maintaining a body position	NMES applied to all extremities
		Supportive care: trochanteric roll, foot plate applied
b140	Attention function	COMCOG program 10 times per week, 30 minutes per time
b144	Memory function	Verbal memory, visual memory, auditory continuous performance training
b1520	Appropriateness of emotion	Atypical antipsychotic and antidepressant medications
b1521	Regulation of emotion	Psychological therapy
		Check socioeconomic status and provide support
		Participate in occupational therapy group
s1110	Structure of the cortical lobes	Antiepileptic medication
s198	Structure of the nervous system	Anticonvulsant medication (pregabalin) for neuropathic pain
b2800	Generalized pain	Application of TENS to the pain site (at a constant frequency of 80 Hz) 30 minutes per day
b415	Blood vessel functions	Warfarin with regular monitoring of INR
		Target INR: 2–3
b4200	Increased blood pressure	Calcium-channel blocker
		Blood pressure monitoring
b430	Hematological system functions	Ferrous sulfate and oral steroids
		Continuous follow-up laboratory tests
b810	Protective functions of the skin	Antibiotic ointment and a moisturizer cream
b820	Repair functions of the skin	Instructed patient to avoid ultraviolet rays
		Pressure ulcer care with low-powered laser therapy
d530	Toileting	Hygiene for the inguinal area after urination and defecation
		Adequate water intake
		Oral antibiotics if the symptomatic urinary tract infection occurred
		Education about proper position during urination or defecation
b2153	Functions of lachrymal glands	Eye-drops and ointment for dry eye

ICF, International Classification of Functioning, Disability and Health; NMES, neuromuscular electrical stimulation; COMCOG, computer-aided cognitive rehabilitation training system (MaxMedica Inc.); TENS, transcutaneous electrical nerve stimulation; INR, international normalized ratio.

improved. Follow-up brain MRI showed improvement of the previous lesions, which were suggestive of posterior reversible encephalopathy syndrome (PRES) (Fig. 1B). As she had fever and a urinary tract infection (UTI), the infectious condition following immunosuppressant therapy with cyclosporine was thought to be the cause of PRES. At her first admission to the Department of Rehabilitation Medicine, consistently with the dramatic neurological recovery commonly observed in patients with PRES, she showed mild functional and cognitive impairment. One week after the start of rehabilitation, weakness of the left extremities and fever were noted. Brain MRI showed acute infarction in the right basal ganglia (Fig. 1C). Acute cerebral infarction was considered related to SLE activation due to UTI. One month after treatment in the Department of Rheumatology, she was returned to the Department of Rehabilitation Medicine. We performed an electrodiagnostic study and whole spinal cord MRI to rule out other superimposed diseases, such as neuropathy, transverse myelitis, and myelopathy. The cross-sectional area of the spinal cord from the cervical through the conus medullaris was markedly thin on sagittal images, and no other significant abnormal findings were evident (Fig. 2). The electrodiagnostic study showed severe sensory peripheral polyneuropathy involving the lower extremities.

When she was returned to the Department of Rehabilitation Medicine, we specified her clinical problems multiphasically using the ICF (Table 1). After addressing the full spectrum of problems that she had, we comprehensively and rapidly planned how to care for and provide a tailored rehabilitation program to the patient.

Regarding the lower extremity weakness and poor functional level, physical therapy including tilt table standing, trunk muscle-strengthening training and sitting-balance training were administered. To investigate cognitive dysfunction, neurocognitive testing was performed using the Korean version of the Consortium to Establish a Registry for Alzheimer's Disease Assessment Packet (CERAD-K), and a computerized neuropsychological test was also performed. These showed remarkable impairment in the praxis recall, word list recall, and trail making tests (Table 2). She underwent a therapeutic program using a computer-aided cognitive rehabilitation training system (COMCOG; MaxMedica Inc., Seoul, Korea), which included memory function training and attention training components. As she experienced severe cerebral infarction after UTI, we aggressively controlled her condition to prevent development any further infections as confounding factors. To manage the tingling sensation and neuropathic pain in both of her lower extremities, we prescribed the anticonvulsant, pregabalin. She also experienced psychological problems, including depression and avolition, for which we prescribed anti-depressants and referred her to a psychologist and social work team.

Table 2. Results of the Korean version of the Consortium to Establish a Registry for Alzheimer's Disease Assessment Packet (CERAD-K)

Measurement	Patient's score /max points	Normal reference (age 60–74 yr)
Verbal fluency	11/-	15.3±3.4
Boston naming	11/15	11.3±2.4
Word list memory	18/30	18.3±3.1
Constructional praxis	10/11	10.4±1.0
Word list recall	7/10	6.4±1.8
Word list recognition	10/10	9.3±1.2
Constructional praxis recall	2/11	7.2±2.6
Trail making A & B	Not testable	-

Using the ICF code, our multidisciplinary rehabilitation team could determine the appropriate therapeutic approach to each problem. For example, several interventions were chosen to treat the target intervention "d530 toileting". A physician prescribed antibiotics to prevent UTI, a physical therapist instructed the patient and her parents about proper position during urination or defecation and nurses checked her water intake. This approach allowed us to rapidly achieve the goal. Thus, we believe that use of the ICF code facilitates precise and rapid communication among rehabilitation team members.

After completing the comprehensive rehabilitation program, the patient showed favorable improvements in multiple respects (Table 3).

DISCUSSION

It is difficult to detect multiple problems in patients with SLE because of the disease's variable activity and severity [1]. We reported a patient with NPSLE with severe involvement of both the central and peripheral nervous

Table 3. Changes in the patient's function and problems before and after rehabilitation therapy

ICF code	Description	Evaluation after PRES	At re-admission to the rehabilitation unit after infarction	At discharge after rehabilitation therapy
b7304	Power of muscles of all limbs	Both upper and lower extremities (MRC): all IV	Right upper extremity (MRC): I	Right upper extremity (MRC): III
			All the rest: 0	All the rest: I
d410	Changing basic body position	Walk independently on an even surface with a walker	Bedridden with no head or neck control	Side rolling to left side
d415	Maintaining a body position		Tolerate tilt table at 50º	Sit up alone in wheelchair with good head and neck control
				Tolerate tilt table at 70º
b140	Attention function	Advanced Grade 2 in COMCOG	Beginner Grade 3 in COMCOG	Intermediate Grade 4 in COMCOG
b144	Memory function	Advanced Grade 1 in COMCOG	Beginner Grade 2 in COMCOG	Intermediate Grade 1 in COMCOG
b1520	Appropriateness of emotion	PHQ-9: 10	PHQ-9: 23	PHQ-9: 14
b1521	Regulation of emotion			
s1110	Structure of the cortical lobes	Seizure (–)	Seizure (–)	Seizure (–)
s198	Structure of the nervous system	VAS: 3	VAS: 6	VAS: 1–2
b2800	Generalized pain			
b415	Blood vessel functions	INR 2–3	INR 2–3	INR 2–3
b4200	Increased blood pressure	130/90 mmHg	120/80 mmHg	110/80 mmHg
b430	Hematological system functions	Anemia	Anemia	Mild anemia
		Low platelet	Low platelet	Low platelet
b810	Protective functions of the skin	-	Coccyx sore 4×5 cm	Coccyx sore 1×1 cm
b820	Repair functions of the skin		Stage III	Stage I
d530	Toileting	Minimal assist	Total assist	Maximal assist
		Upper urinary tract infection	Cystitis	Cystitis
b2153	Functions of lachrymal glands	Dry eye (+)	Dry eye (+)	Dry eye (+)

ICF, International Classification of Functioning, Disability and Health; PRES, posterior reversible encephalopathy syndrome; MRC, Medical Research Council; COMCOG, computer-aided cognitive rehabilitation training system (MaxMedica Inc.); PHQ-9, Patient Health Questionnaire-9; VAS, visual analog scale; INR, international normalized ratio.

system. To our knowledge, this is the first study to evaluate a patient's functional impairment and disability in detail using the ICF model, and attempt to manage the identified issues to improve the patient's quality of life.

As NPSLE causes diverse disabilities and impairs the quality of life, management should be tailored to each patient. The first step of management in patients with NPSLE is to identify aggravating factors, such as hypertension, infection, and metabolic abnormalities. Particularly, infection control is important because an infection can increase SLE disease activity, enabling circulating antibodies to attack blood vessels and increase the risk of cerebral infarction [4]. In progressive cases, immunosuppressive agents and anticoagulation medication can be used for events related to a prothrombotic vascular injury. The next step is to provide symptomatic therapy and tailored, comprehensive rehabilitation.

In clinical practice, the ICF enables precise description of the patient's functional state; illustrates the patient's experience of functioning and the relationship between the rehabilitation goals and appropriate intervention targets; and provides an overview of the resources required to improve specific aspects of human functioning, and the changes in functional state following rehabilitative intervention. This also supports a common understanding of the patient's functioning and communication among rehabilitation team members [5].

Aringer et al. [6] designed ICF core sets for patients with SLE to adequately describe the typical spectrum of problems in functioning. In our case, this core set did not cover all of the problems. For example, our patient had pancytopenia, neuropathic pain, and susceptibility to infection. Thus, modifications specific to each individual patient are required when using the ICF.

Many signs and symptoms of NPSLE can pose a significant diagnostic and therapeutic challenge to physicians and rehabilitation team members. We confirmed the patient's functional status and comorbidities with this team approach using the ICF and designed appropriate interventions to achieve her goals. Thus, we believe that use of the ICF facilitates team communication and enables provision of comprehensive rehabilitation programs.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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REFERENCES

1. Cervera R, Khamashta MA, Font J, Sebastiani GD, Gil A, Lavilla P, et al. Systemic lupus erythematosus: clinical and immunologic patterns of disease expression in a cohort of 1,000 patients. The European Working Party on Systemic Lupus Erythematosus. Medicine (Baltimore) 1993;72:113-24.
2. Hanly JG. Diagnosis and management of neuropsychiatric SLE. Nat Rev Rheumatol 2014;10:338-47.
3. Lee IH, Ryu YU. Physical therapy combined with corticosteroid intervention for systemic lupus erythematosus with central nervous system involvement: a case report. J Phys Ther Sci 2014;26:1839-41.
4. Nencini P, Baruffi MC, Abbate R, Massai G, Amaducci L, Inzitari D. Lupus anticoagulant and anticardiolipin antibodies in young adults with cerebral ischemia. Stroke 1992;23:189-93.
5. Rauch A, Cieza A, Stucki G. How to apply the International Classification of Functioning, Disability and Health (ICF) for rehabilitation management in clinical practice. Eur J Phys Rehabil Med 2008;44:329-42.
6. Aringer M, Stamm TA, Pisetsky DS, Yarboro CH, Cieza A, Smolen JS, et al. ICF core sets: how to specify impairment and function in systemic lupus erythematosus. Lupus 2006;15:248-53.