

## A Case of SLE Presenting Stroke-Like Symptoms

Koji Ishitsuka<sup>1,2</sup>, Tetsuro Ago<sup>2</sup>, Kenji Fukuda<sup>1,2</sup>, Yoshihisa Fukushima<sup>1,2</sup>, Seiji Gotoh<sup>1,2</sup>, Tomohiro Yubi<sup>1,2</sup>, Masahiro Kamouchi<sup>2</sup> and Takanari Kitazono<sup>2</sup>

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### Abstract

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We report a 50-year-old woman with systemic lupus erythematosus (SLE) who developed stroke-like symptoms, including disturbance of consciousness and severe left hemiparesis. Despite the stroke-like symptoms, MR imaging showed quite atypical findings for stroke; broad high-intensity areas in the right frontoparietal lobes on diffusion-weighted images were shown mainly as iso-intensity areas on the ADC map, without any findings of stenosis of the cerebral large arteries. The cerebral blood flow in these areas was significantly decreased, as evaluated by single photon emission computed tomography. The present case suggests that small vessel vasculopathies localized in the unilateral hemisphere could cause “stroke-like” symptoms in SLE.

**Key words:** SLE, NPSLE, stroke, small vessel vasculopathy, MRI, SPECT

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### Introduction

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Systemic lupus erythematosus (SLE) is a systemic autoimmune disease, in which various organs are involved (1). Nervous system involvement, described as neuropsychiatric syndromes of SLE (NPSLE), is very common and is reported to occur in 25-60% patients with SLE (1, 2). According to the American College of Rheumatology Nomenclature (2), NPSLE is divided broadly into 2 groups, central and peripheral nervous system NPSLE; each group is further classified into various disease types. “Cerebrovascular disease (=stroke) type” is one of the major central nervous system NPSLE: although any type of stroke can occur in SLE patients, medium-to-large size cerebral vessels are usually affected by either atherosclerosis or embolism, and antiphospholipid antibodies are often present in these cases (3, 4). In contrast, other NPSLE, including confusion, seizure, cognitive dysfunction, mood disorder, and psychosis (2), is often caused by diffuse vasculopathies of small size cerebral vessels, such as vasculitis and immunocomplex deposition (5). The diffuse small vessel vasculopathies usually occur symmetrically in the bilateral hemisphere (5). These psychiatric symptoms may be caused by mild but diffuse cerebral hy-

poperefusion, which does not result in irreversible focal ischemic damage, *i.e.* infarction. Despite its high incidence, psychiatric type NPSLE is difficult to diagnose due to the wide variety of neuropsychiatric manifestations and the absence of golden standard modalities. It has been thought that single photon emission computed tomography (SPECT) is the most sensitive modality to detect psychiatric type NPSLE, although it has poor specificity (6).

In the present report, we describe an interesting case with NPSLE presenting “stroke-like” symptoms, in which its pathogenesis was considered to be small vessel vasculopathies occurring in the unilateral hemisphere.

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### Case Report

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We describe a 50-year-old woman presenting stroke-like symptoms during the course of SLE. The patient was diagnosed with SLE at age 47. She started taking prednisolone at that age, and had done well on a low dose of prednisolone (5 mg per day) for 3 years. There were no physical changes before the onset of the following neurological symptoms.

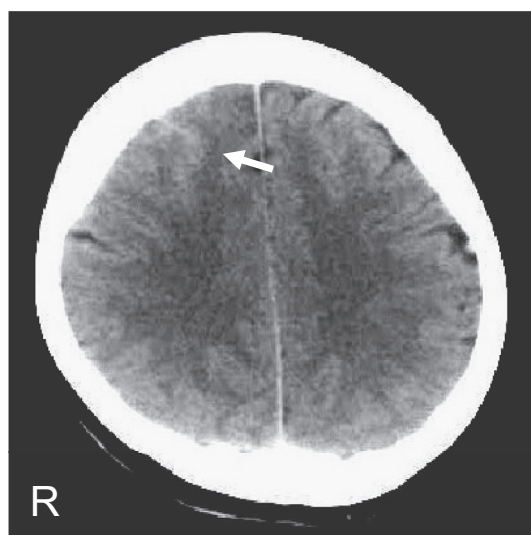
When she woke up one morning, she noticed left hemiparesis and was brought to our hospital by ambulance. Sei-

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<sup>1</sup>Division of Cerebrovascular Disorders, St. Mary's Hospital, Japan and <sup>2</sup>Department of Medicine and Clinical Science, Graduate School of Medical Sciences, Kyushu University, Japan

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Correspondence to Dr. Tetsuro Ago, agou@intmed2.med.kyushu-u.ac.jp



**Figure 1.** CT on admission shows a linear high-density area along a cerebral sulcus in the right frontal lobe.

zure was not found during the course. On admission, her consciousness was slightly drowsy with mild disorientation. Her blood pressure was 172/88 mmHg. Heart rate was 65 beats per minute and regular. Her body temperature was 35.9°C.

Neurologically, gaze palsy to the left side and left hemiparesis with dominance of the lower limb (Manual Muscle Testing: upper limb 2/5, lower limb 1/5) were observed. Sensory disturbance was not observed. Pathological reflex including Babinski's reflex on the left side was positive. These neurological findings seemed to be consistent with those of "stroke" injuring the right hemisphere including the fronto-parietal cortex.

Peripheral blood cell counts and coagulation studies were normal. Biochemical studies were also normal except for a mild elevation of blood sugar (134 mg/dL). CRP (0.2 mg/dL) and ESR (8 mm/hr) were normal. Immunologically, specific autoantibodies, including anti-phospholipid antibodies, anti-double strand DNA antibody, and anti-ribosomal P antibody, were negative, although anti-nuclear antibody was slightly positive (80-fold). Cerebrospinal fluid examination was normal (cell count 1/μL, protein 26 mg/dL, IgG index 0.56, and MBP <40 pg/mL) except that interleukin 6 was significantly elevated (57.4 pg/mL).

Head computed tomography (CT) on admission showed a slight linear high-density area along a cerebral sulcus in the right frontal lobe (Fig. 1). Although conventional magnetic resonance images (MRI), T<sub>1</sub>-weighted images (T<sub>1</sub>WI) (Fig. 2A) and T<sub>2</sub>WI (Fig. 2B), showed a slight abnormality in the right frontal lobe on both day 0 and day 3, diffusion-weighted images (DWI) detected high-intensities in much broader areas of the right fronto-parietal lobes (Fig. 2C). Unexpectedly, these lesions were shown mainly as iso-intensity areas, with a small low-intensity area, on apparent diffusion coefficient map (Fig. 2D). In addition, T<sub>2</sub>\*-weighted images showed a low-intensity area along a cere-

bral sulcus in the right frontal lobe (Fig. 2E). These findings indicated that the right hemispheric lesion consisted mainly of vasogenic edema, with small ischemic change and micro-hemorrhage in the subarachnoid space. Neither MR angiography of the cerebral arteries nor ultrasound sonography of the cervical arteries showed abnormality (data not shown). We further examined cerebral blood flow (CBF) using <sup>123</sup>I-IMP SPECT, and found a significant decrease in the CBF of the right hemisphere, especially in the fronto-parietal lobes (Fig. 3A). Taken together, we diagnosed this patient with NPSLE, in which inflammation and vasogenic edema at the level of small size vessels in the right hemisphere caused the "stroke-like" symptoms.

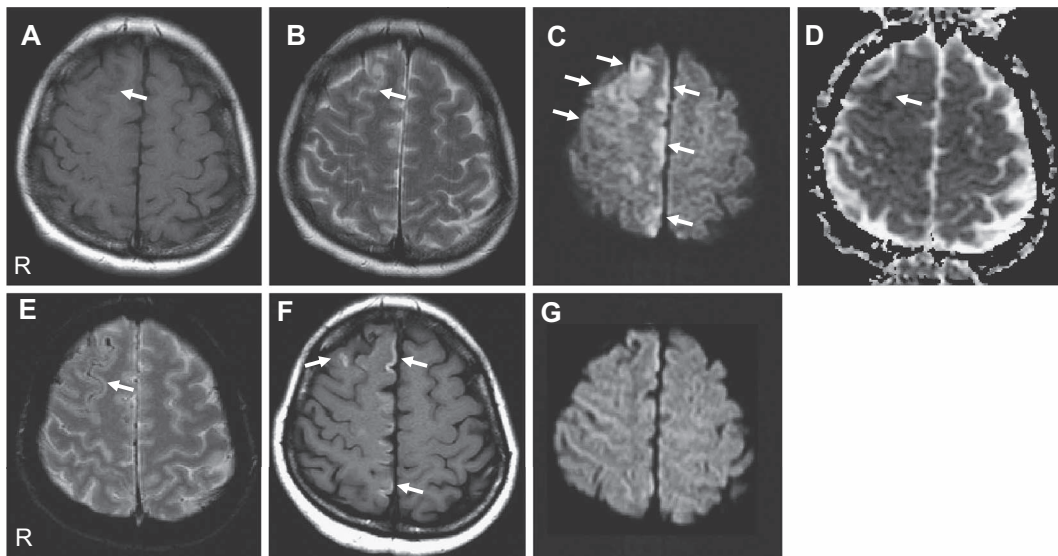
We started an anticoagulation therapy with heparin while continuing steroid therapy. These treatments drastically improved the disturbance of consciousness on the following day, and markedly improved the left hemiparesis, with mild weakness of the lower extremity, in one month. The subsequent MRI on day 30 showed that the high-intensity areas found on admission (Fig. 2C) completely disappeared on DWI (Fig. 2G) with residual cortical high-intensity areas in the right frontal lobe on T<sub>1</sub>WI (Fig. 2F). The cerebral blood flow in the right hemisphere was also improved significantly in the subsequent SPECT (Fig. 3B).

## Discussion

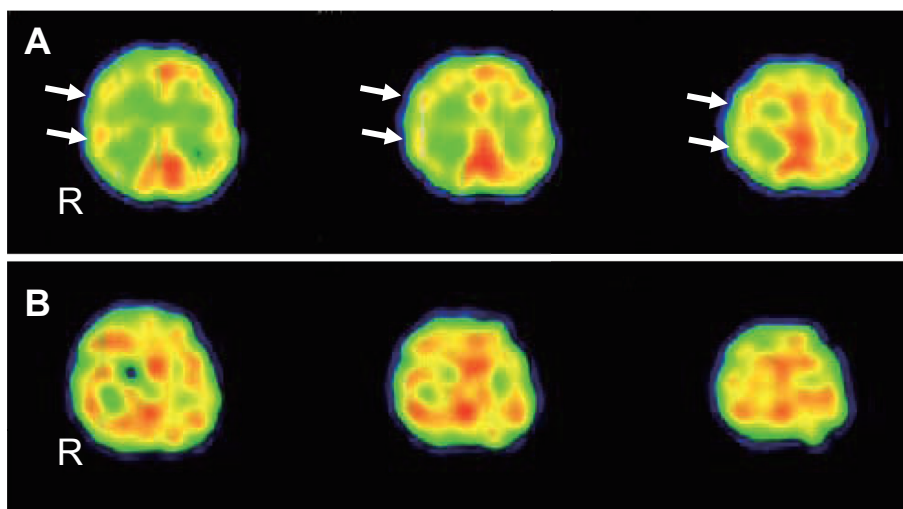
In this report, we describe a patient who presented sudden stroke-like symptoms during the course of SLE. There are two interesting points in the present case.

First, in SLE patients, various types of stroke can occur, mainly due to the presence of antiphospholipid antibodies. Although this patient presented stroke-like symptoms, neither coagulation abnormalities nor stenotic lesions in the cerebral and cervical arteries were found. Moreover, patterns of brain damage were not compatible with those of any types of stroke. Judging from the findings of SPECT, an acute decrease in CBF appeared to occur diffusely in the right hemisphere. Thus, we speculate that small vessel vasculopathies occurred broadly in the right hemisphere, and among them, particularly severe ones may have caused a focal neuronal ischemia leading to the left hemiparesis. This speculation is well consistent with the MRI findings (Fig. 2C & D). We could not find any autoimmune abnormalities suggesting exacerbation of SLE disease activities, such as anti-double strand DNA antibody and anti-ribosomal P antibody. It is indeed reported that there are no clear relationships between NPSLE and SLE disease activities (7). In the present case, since the anticoagulation therapy without reinforcement of immunotherapies significantly improved the neurological symptoms, disturbance of microcirculation caused by small vessel vasculopathies, regardless of SLE disease activities, may have been the major pathophysiology of this case.

In this context, we cannot completely exclude the possibility that the hypoperfusion in the right hemisphere may



**Figure 2.** MRI on day 3 shows a slightly low-intensity area on T<sub>1</sub>WI (A, arrow) and a high-intensity area on T<sub>2</sub>WI (B, arrow) in the right frontal lobe. (C) DWI detects high-intensities in broader areas of the right frontal and parietal lobes (arrow). (D) ADC map shows iso-intensity areas with a small low-intensity area (arrow) in the right frontal lobe. (E) T<sub>2</sub>\*-weighted imaging shows a low-intensity area along the cerebral sulcus in the right frontal lobe (arrow). The subsequent MRI on day 30 shows almost normal study on DWI (G) with residual cortical high-intensity areas on T<sub>1</sub>WI (F).



**Figure 3.** (A) <sup>123</sup>I-IMP SPECT at day 8 shows a significant decrease in CBF in the right hemisphere, especially in the fronto-parietal lobes. (B) The CBF was much improved on the subsequent SPECT on day 89.

have been attributable to post-ictal neuronal suppression leading to Todd's paresis. We think, however, that the following points do not positively support the post-ictal mechanisms in this case. 1) no occurrence of convulsion was found throughout the course, 2) focal epileptiform discharges were not found in electroencephalogram during hospitalization, and 3) the left hemiparesis, especially of the lower extremity, lasted for more than one month, while post-ictal Todd's paresis usually lasts for no more than 48 hours. At any rate, the present case suggests that diffuse small vessel vasculopathies, without any occlusion of the medium-to-large size arteries, can cause stroke-like symp-

toms in SLE patients.

Second, consistent with previous reports about psychiatric-type NPSLE (6), SPECT was much more sensitive to detect the lesions of the present case than CT and conventional MRI. In the present case, DWI was another sensitive method to detect the lesions of NPSLE (Fig. 2C & G). In addition, the findings of SPECT and DWI similarly reflected the disease states of the present case. Since the specificity of SPECT for NPSLE is low albeit its high sensitivity (6), a combination of SPECT and DWI may be useful to evaluate NPSLE.

In conclusion, physicians should note that small vessel

vasculopathies can occur in the unilateral hemisphere, thereby causing “stroke-like” symptoms in SLE patients, although they usually occur in the bilateral hemisphere and are often related to psychiatric symptoms. In such cases, a combination with SPECT and DWI would be helpful for diagnosis and observation of the disease states.

**The authors state that they have no Conflict of Interest (COI).**

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