Cerebral Hemiatrophy in Systemic Lupus Erythematosus: Report of a Case

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An 18-year-old woman with systemic lupus erythematosus developed neuropsychiatric disorders, including aseptic meningoencephalitis, organic brain syndrome and seizure. A series of computed axial tomography scans revealed the progression of marked atrophy of the right cerebral hemisphere for a period of 3 years without occlusion or stenosis of large vessels on cerebral angiography. I-123 IMP single photon emission computed tomography disclosed a markedly decreased uptake of I-123 IMP in the right cerebral hemisphere, and also in the left cerebellar hemisphere (crossed cerebellar diaschisis), which disappeared within 2 years.

Key words: Computed axial tomography, Central nervous system, Crossed cerebellar diaschisis

Central nervous system (CNS) involvement in systemic lupus erythematosus (SLE) (CNS-lupus) has been reported to occur relatively frequently as one of the most serious complications of this SLE (1, 2). However, the pathogenesis of CNS disorders still remains unclear in contrast to lupus nephritis (1, 2). Although there have been some reports on the deposition of immunoglobulins and complement components in the choroid plexus and on the presence of microinfarction and vasculitis, these phenomena can not always explain the CNS disease activity or the variability of the neuropsychiatric manifestations (1, 2).

Regarding the diagnosis of CNS-lupus, some investigators have reported the efficacy of computed axial tomography (CAT) scans (3, 4), and magnetic resonance imaging (5). Thus, many patients with CNS-lupus have been reported to have cerebral atrophy as typified by sulcal enlargement with or without ventricular dilatation (3, 4). However, it has also been reported that corticosteroids may exacerbate the cerebral atrophy (6). Moreover, the atrophy seen on CAT scan does not fully explain the pathophysiologic basis for the myriad of CNS manifestations, such as psychiatric disorders, organic brain syndrome and seizures (2), in SLE patients.

In the present report, we describe an SLE patient who developed organic brain syndrome and seizures. The CAT scan of the patient showed marked atrophy of the right cerebral hemisphere without arterial occlusion or stenosis on cerebral angiography.

CASE REPORT

An 18-year-old Japanese woman had suffered from severe proteinuria, arthralgia and facial erythema since June 1981. She developed aseptic meningoencephalitis in July 1983. Repeated CAT scan was normal at this time. She was diagnosed as SLE by positive ANA test (peripheral pattern at a titer of 1:10 and speckled pattern at a titer of 1:160) and positive lupus band test. In September 1983, in addition to leukocytopenia and polyarthritis, she

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Fig. 1a. EEG on October 27, 1983 showing diffuse slow waking activity with occasional high voltage delta activity in the left parietal and occipital regions.

Fig. 1b. EEG on October 27, 1983 showing no significant voltage differences nor deficit of the normal components on either side.

Fig. 1c. EEG on December 13, 1984 showing positive spikes in the right temporal, central, parietal and occipital areas.

Fig. 1d. EEG on August 4, 1986 showing predominant alpha waking activity of 9–10 Hz with occasional theta activity. Voltage differences between the right and left leads are remarkable.
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also showed organic brain syndrome characterized by mild mental retardation and disturbances of perception and intellectual function. Both anti-Sm and anti-RNP antibodies were positive by gel immunodiffusion techniques. Total hemolytic complement (CH50) was 19.5 U/ml (normal: 29.3–43.9 U/ml). At this time, EEG showed diffuse slow waking activity with occasional high voltage delta activity in the left parietal and occipital areas with the normal components on both sides (Fig. 1a and 1b). All cerebrospinal fluid (CSF) IgG, IgA and IgM indices, which are considered to reflect intrathecal immunoglobulin (Ig) production, were elevated. She recovered from these conditions along with the decrease in the CSF Ig indices after oral administration of prednisolone 30 mg daily was started.

In May 1984, she began to complain of numbness and weakness of the left upper extremity. In July 1984, CSF IgM and IgG indices were found to be elevated. In December 1984, she developed a generalized convulsion originating from the left upper limb. At this time, EEG revealed positive spikes in the right temporal, central, parietal and occipital regions (Fig. 1c), and both CSF IgM and IgG indices were also elevated. Phenobarbital was started at a dose of 60 mg daily, and convulsion was well controlled, although spikes in the right hemisphere continued to appear on EEG. Although the systemic disease activity was rather well controlled with oral prednisolone 10–20 mg daily, CSF IgM and IgG indices were found to be elevated in November 1985. In February 1986, she began to complain of mild vertigo and dizziness. At this time, spikes were not observed on EEG. In July 23, 1986, after phenobarbital was discontinued on July 10, convulsions developed. CAT scan at this time showed marked right cerebral hemiatrophy (Fig. 2a). She was admitted to the University of Tokyo Hospital for further evaluation and treatment.

Physical examination on admission revealed a blood pressure of 116/72 mmHg, pulse rate of 84 beats/min, and finger and facial erythema. She was alert and well-oriented. Her mental function was normal. Neurological examination revealed mild left hemiparesis and minimal left cerebellar sign. Urinalysis was normal; her complete blood cell count demonstrated hemoglobin 12.2 g/dl, white blood cell count $5.3 \times 10^3$/mm$^3$ with a mild left shift and platelet count $22.3 \times 10^4$/mm$^3$. The renal function test and the liver function test were normal. The test for C-reactive protein was negative, and erythrocyte sedimentation rate (Westergren) was 14 mm/h. Both anti-ds DNA IgG antibody and anti-RNP antibody were positive by enzymeimmunoassay and by immunodiffusion technique, respectively. CH50 was

Fig. 2a. CAT scan on July 10, 1986 showing marked atrophy of the right cerebral hemisphere.

Fig. 2b. Cerebral angiography on September 3, 1986. No stenosis or occlusion was demonstrated in the right internal carotid artery or the right middle cerebral artery.
29.0 U/ml. CSF examination showed 0.3 mononuclear cells/mm³ and 31 mg protein/dl. CSF IgG, IgA and IgM indices were 1.11 (normal 0.28–0.79), 0.50 (0.11–0.39) and 0.11 (0.003–0.074), respectively. EEG (Fig. 1d) revealed predominant alpha waking activity of 9–10 Hz with occasional theta burst and reduced voltage in the right hemisphere regions. Right carotid artery angiography revealed no stenosis or occlusion (Fig. 2b). However, N-isopropyl I-123 p-iodoamphetamine single photon emission computed tomography (I-123 IMP SPECT) (7) demonstrated a markedly decreased uptake of I-123 IMP in the right frontal, temporal and parietal cortices and in the right thalamus (Fig. 3a) and a slightly decreased uptake in the left cerebellar hemisphere, presumably by crossed cerebellar diaschisis (remote effects of supratentorial injury) as first described by Baron et al (8) (Fig. 3b). The I-123 IMP SPECT performed in April 1989 showed normal cerebellar blood flow, indicating the disappearance of the crossed cerebellar diaschisis.

DISCUSSION

Our patient showed a series of neurological manifestations, including aseptic meningoencephalitis, organic brain syndrome and focal motor seizure of the Jacksonian type. Since all CSF IgM, IgA and IgG indices were elevated in 1983 when she showed organic brain syndrome and in 1986 when she was admitted to our hospital, the CNS complications were considered to be caused by active SLE (8–10). Actually, the organic brain syndrome observed in 1983 was ameliorated along with the decrease in CSF Ig indices by increasing the dose of steroid (data not shown).

The CAT scan performed in July 1986 showed marked atrophy of the right cerebral hemisphere. However, repeated CAT scans in 1983 had all been normal when the patient developed aseptic meningoencephalitis and organic brain syndrome. Moreover, the patient had no history of perinatal accidents, postictal disorders, developmental abnormality or episodes of paralysis in childhood. Therefore, the right cerebral atrophy of the patient was considered to develop between 1983 and 1986. Since she began to complain of numbness and weakness of the left upper extremity in May 1984 and CSF Ig indices were increased in July 1984, the involvement of the right hemisphere most likely started around this time. Although the systemic disease activity of SLE had been fairly well controlled, she developed Jacksonian type convulsions originating from the upper left extremity with the elevation of CSF Ig indices in December 1984, when EEG showed spikes in the right hemisphere. This is not surprising since the CNS disease activity does not always parallel the systemic disease activity in CNS lupus patients (9–12). In fact, although administration of phenobarbital prevented the attack of seizure and her systemic disease activity of SLE was fairly well controlled after December 1984, CSF Ig indices were...
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still elevated in November 1985. Moreover, after phenobarbital was discontinued in July 1986, the patient again developed convulsions. Therefore, it is suggested that the CNS disease activity had not been well controlled after December 1984 and that the right hemiatrophy of the patient was most likely caused by active SLE.

A variety of abnormalities on CAT scan may be observed in patients with SLE, including generalized and perisulcal atrophy, infarcts, intracerebral hemorrhages, and basal ganglia calcifications (1-4, 6, 13). Among these, generalized and perisulcal atrophy have been the most commonly observed, although the pathological significance is still controversial. Thus, the etiologic possibilities of cerebral atrophy in SLE are too numerous. For example, corticosteroids (6) or anti-neuronal antibodies (14) can result in cerebral atrophy by affecting neurons. Considering that cerebral atrophy was restricted to the right side in our patient, it seems unlikely that anti-neuronal antibodies or corticosteroids were responsible for the atrophy in our patient. Occlusion of the right internal carotid artery or the middle cerebral artery has been observed in SLE patients with lupus anti-coagulant (1). Moreover, brain infarcts due to deep venous thrombosis in SLE has been reported (15). However, cerebral angiography revealed no occlusion or stenosis of the right internal carotid artery or the middle cerebral artery or the venous system, and lupus anti-coagulant was not demonstrated in our patient. Interestingly, I-123 IMP SPECT showed a marked decrease in the uptake of I-123 IMP in the right frontal, temporal and parietal cortices, indicating decreased blood flows and a decreased number of vital neurons in these areas (7). Thus, the presence of the right cerebral hemiatrophy is considered to result in a marked decrease of I-123 IMP uptake.

The patient complained of vertigo and dizziness since January 1986, and neurological examination on admission revealed mild left cerebellar signs. The left cerebellar signs appeared to be caused by a decrease in blood flow in the left cerebellar hemisphere through the crossed cerebellar diaschisis (7, 8) as demonstrated by I-123 IMP SPECT. The crossed cerebellar diaschisis was no longer observable in April 1989, which is consistent with previous findings that the crossed cerebellar diaschisis might occur only temporarily after damage of the contralateral cerebral hemisphere (7, 8).

In summary, we report a patient with right cerebral hemiatrophy who had a prolonged history of CNS lupus with elevated CSF Ig indices. The pathogenesis could not be explained either by the occlusion of large vessels or by the presence of lupus anticoagulant, and it is therefore postulated that in this patient, who had a history of proceeding aseptic focal meningoencephalitis, the hemiatrophy was the result of active CNS lupus.

REFERENCES


