A Case of Central Nervous System Lupus Associated with Ruptured Cerebral Berry Aneurysm

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At the age of 29, a woman developed central nervous system manifestations of incontinence, psychosis and a grand mal seizure in February 1982. She was diagnosed as having systemic lupus erythematosus (SLE) based on photosensitivity, oral ulcers and elevated antinuclear and anti-DNA antibodies titers. Three years and one month later the patient had episodes of severe headache and vomiting during the course of maintenance treatment. CT examination of the head revealed blood within subarachnoid cisterns, and a small berry aneurysm was found at the distal portion of the basilar artery by cerebral angiography. The possible role of SLE-associated cerebral vascular changes in the development of this aneurysm is discussed.

Key Words: Incontinence, Psychosis, Grand mal seizure, Basilar artery

A variety of lesions in the wall of small and large blood vessels supplying the central nervous system (CNS) are frequently seen in systemic lupus erythematosus (SLE) involving the CNS (CNS lupus) (1). The incidence of cerebral aneurysms detected at general autopsy is around 5% (2). Although these two diseases are related to blood vessels (1,3), there are few reports of their coexistence (4-6). We report a case of CNS lupus which was complicated by a ruptured cerebral berry aneurysm resulting in a subarachnoid hemorrhage (SAH).

CASE REPORT

A 29-year-old woman with a past history of photosensitivity, and a two-week history of high fever (39°C), weight loss, incontinence, psychosis and a grand mal seizure, was admitted to a hospital emergency ward in February 1982. Laboratory examinations revealed a highly elevated antinuclear antibody (1:160), increased titer of anti-DNA antibody (89.3 U/ml, normal: below 10) and decreased levels of complement (C3: 44.0 mg/ml, normal: 80–200, C4: 5.6 mg/ml, normal: 15–60, CH50: 14.8 U/ml, normal: 31–52). The erythrocyte sedimentation rate (Westergren) was elevated (70 mm/h). However, the findings on cerebrospinal fluid, electroencephalogram, brain CT scans and examinations related to infections were normal. Cerebral angiography was not performed since there were no manifestations related to the complications of cerebral aneurysm.

Under the diagnosis of SLE (7), treatment with prednisolone 60 mg/day orally was started, and resulted in prompt improvement in her condition and laboratory data. The dose of prednisolone was tapered biweekly by 5 mg. Six months later, she was discharged with a maintenance dose of 15 mg/day of prednisolone.

In the following two and a half years, she was treated at the outpatient clinic with 5 to 15 mg/day prednisolone. The dose was tapered or increased depending on the serological data.

The patient suddenly complained of severe headache and vomiting at work in March 1985. Again, she came to the emergency room. No CNS manifestations of SLE were detected. A lumbar tap

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revealed bloody cerebrospinal fluid. CT scans of the brain visualized blood in the subarachnoid cisterns. A small berry aneurysm was seen at the distal portion of the basilar artery by cerebral angiography. No lupus anticoagulant and antiplatelet antibody were detected. Activities of plasminogen, antithrombin III and α₂ antiplasmin were within the normal range. The prothrombine time was 11.2 seconds, the activated partial thromboplastin time was 31.5 sec and the fibrinogen titer was 234 mg/dl.

Four weeks after the rupture of the aneurysm, right frontotemporal craniotomy was performed. The aneurysm at the distal portion of the basilar artery was successfully clipped. However, massive hemorrhage occurred suddenly from the proximal portion of the right internal carotid artery. Due to the bleeding, it was necessary to partially clip the carotid artery in order to make the injured site visible. This manipulation of the artery resulted in a decrease of blood supply and the development of a cerebral vascular lesion.

Left hemiplegia was noted postoperatively. Postoperative brain CT scans disclosed a large infarct in the right frontoparietal region. She is now being treated as an outpatient with a daily prednisolone dose of 10 mg/day, and wears a long leg brace.

**DISCUSSION**

The incidence of cerebral berry aneurysms increases with age (2, 8). Autopsy findings support the acquired rather than the congenital theory of the pathogenesis of cerebral berry aneurysms (3). On the other hand, there are biochemical (9) and clinical data (10) suggesting the congenital origin of such aneurysms.

A number of non-traumatic acquired conditions including vasculitis are associated with cerebral berry aneurysms (3, 10). In the literature, several cases of vasculitis or connective tissue diseases have been described as complications of cerebral berry aneurysms (10). The relationship of these aneurysms to the connective tissue diseases may be fortuitous, since reported cases having the above two conditions are scarce. However, typical vasculitic lesions or atypical forms of vascular changes in connective tissue diseases possibly induce a weakening of the

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**Fig. 1.** Frontal projection of vertebral arteriogram showing a small berry aneurysm (arrow) at the distal portion of basilar artery or at the bifurcation of the posterior communicating artery.

**Fig. 2.** CT scan of the brain. Blood in the basal cisterns of the subarachnoid space.
supporting connective tissue as well as the adjacent muscle coat in the wall of cerebral artery, predisposing an aneurysm formation during a long course of such diseases.

In CNS lupus, the typical vasculitic findings, invariably seen in cases of lupus nephritis, may be less common than previously supposed (1). Instead, vascular damage, including perivascular infiltration of lymphocytes, vascular hyalinization, and endothelial proliferation, is found in over 60% of the cases of CNS lupus (1). In the present case, easy bruisability prevented us from performing a biopsy during the aneurysm clipping operation for histological examination of the aneurysmal wall. Berry aneurysms are prone to rupture. Most ruptured aneurysms measure 5–10 mm in diameter (11). The initial process of rupture may involve aneurysmal dilatation.

Although SAH is the most frequent complication (not manifestation) of CNS lupus outbreaking in as many as 30% of active cases (1), cerebral aneurysms are undetected in these hemorrhaged cases even during autopsy examinations (1). Kelley et al. (4) reported a case, having a 5-year history of lupus nephritis and a 3-week history of CNS lupus, which was complicated with cerebral fusiform aneurysm found at autopsy. Angitis was detected at the site of the aneurysm. Fody et al. (5) noted another case of active CNS lupus of 12 days duration combined with aneurysms of spinal artery. Similarly, a postmortem study demonstrated these aneurysms to be involved in vasculitis. Hashimoto et al. (6) presented an additional case of SLE with a 3-year history of lupus nephritis and a 1-year-8-month history of CNS lupus associated with multiple cerebral fusiform aneurysms. In this case, aneurysms were confirmed radiologically and vasculitis in the small arteries of the CNS was found at autopsy. The present patient seems to be the 4th case of CNS lupus on steroid within 3 years associated with cerebral berry aneurysm. Different from the above 3 cases, she is alive as an outpatient. Five cases of lupus nephritis associated with cerebral aneurysms were reported (4, 6 12-14). Also, there are several cases of SLE (12, 14-21) with manifestations apparently associated with cerebral aneurysms. In one (18) of these cases, vasculitis in the aneurysmal wall was noted at autopsy.

The present case was reported elsewhere from a neurosurgical point (15) in which nothing was noted on the clinical pictures and course of the CNS lupus. A typical vascular disorder called cerebral berry aneurysm was revealed following a definite period (3 years) of development of another collagen vascular disease of CNS (named CNS lupus). These apparent, sequentially related clinico-pathological (CNS lupus and cerebral aneurysm) changes are supposed to be interesting from the stand point of the acquired theory in the pathogenesis of cerebral berry aneurysms (3). In addition berry aneurysm, found in the present case, is the most common form of cerebral aneurysms (3). On the other hand, two previously reported cases of CNS lupus (4, 6) were combined with cerebral fusiform aneurysms, which are thought to be arteriosclerotic in origin (3). In the other case of CNS lupus (5), an unidentified shaped aneurysm was found microscopically in the spinal artery.

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