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Ruptured Cerebral Aneurysm in patient with Systemic Lupus Erytematosus

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Abstract The authors present a case of ruptured cerebral aneurysm in a patient with systemic lupus erythematosus. This is the rare case in which a radical operation was performed on cerebral aneurysm associated with systemic lupus erythematosus. We discuss a saccular aneurysm formation in systemic lupus erythematosus.

Key Words: Ruptured cerebral aneurysm, Fusiform aneurysm, Angiitis, Fibrinoid necrosis, Systemic lupus erythematosus

Introduction

In neuropathological study, subarachnoid homorrhage is relatively common in systemic lupus erythematosus (SLE)[1]. However, there are no reports in which a radical operation on confirmed cerebral aneurysm in SLE was performed.

We here describe a patient with established SLE in whom a saccular aneurysm at the junction of the right pericallosal artery and the callosomarginal artery was clipped with good result following subarachoid hemorrhage, and discuss the pathogenesis of cerebral aneurysm in an SLE patient.

Case Report

On November 4th 1983, a 47-year-old woman was admitted to the internal medicine ward at Yamaguchi University Hospital after onset of headache, vomiting and consciousness disturbance.

Computerized tomography (CT) scans taken at the internal medicine department showed a subarachnoid hemorrhage, so she was immediately transferred to our ward.

She had a past history of subarachnoid hemorrhage 7 years previously and a cerebral angiogram performed at that time showed a 1 mm diameter sized aneurysm at the junction of the pericallosal artery and the callosomarginal artery with wall irregularity of involved vessels which may be attributable to angiitis (Fig. 1). She was medically treated at that time because she was known to have had SLE for the past 5 years. She had been treated with prednisolone 20mg at every exacerbation of her exanthem, arthralgia and myalgia.

On admission, her blood pressure was 170/100 mmhg and her pulse rate 70/min. and regular. She appaeared cushingoid and in moderate discomfort from headache. She was in a drowsy state.

Ophthalmoscopic examination revealed

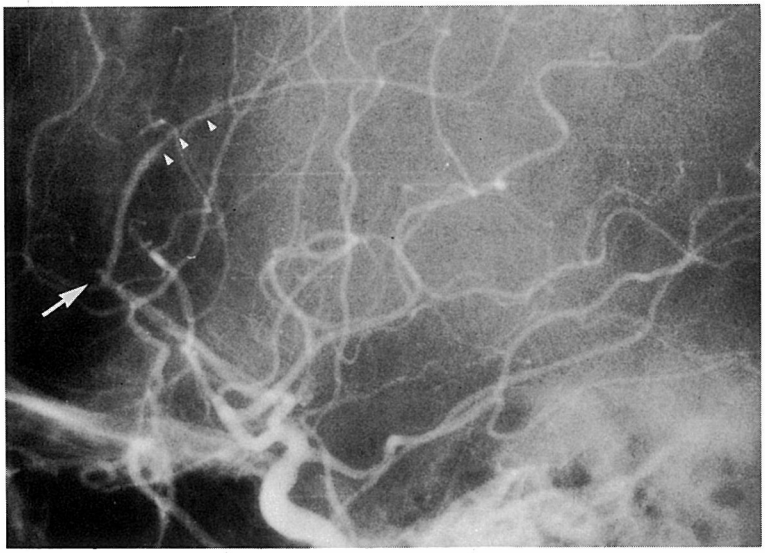

Fig. 1 Right carotid artery angiogram, lateral view, showing a very tiny aneurysm (arrow) at the junction of the pericallosal artery and the callosomarginal artery with wall irregularity (small arrowheads).

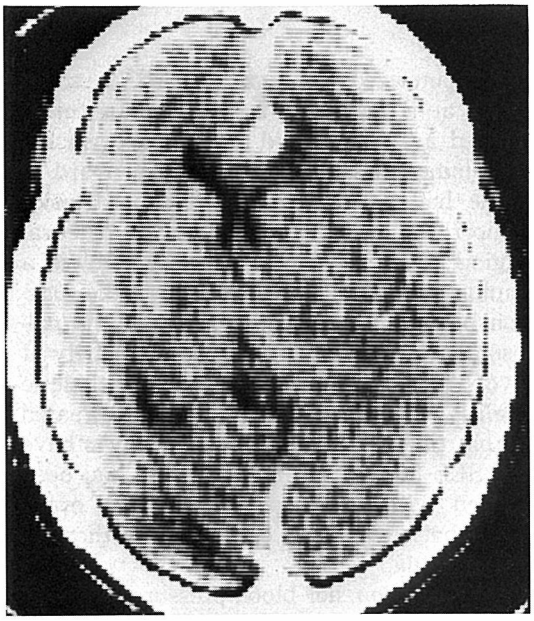

Fig. 2 A CT scan without contrast-medium enhancement showing high-density lesion in the interhemispheric fissure and right medial frontal lobe with ventricular deformity.

severe arteriolar narrowing and retinal bleeding. The neck was stiff. Cardiac rhythm was regular without murmur. There was no motor weakness and no sensory disturbance but Babinski sign was noted at the right side.

Lumbar puncture revealed grossly bloody spinal fluid. The CT scans of the brain showed high-density area in the subarachnoid space, especially in the anterior interhemispheric fissure and the right medial frontal lobe (Fig. 2).

Laboratory tests on admission revealed normal values except leucocytosis.

A four-vessel angiogram performed via the femoral route showed a saccular aneurysm at the junction of the pericallosal artery and the callosomarginal artery which had increased in size to about 4 mm in diameter (Fig. 3 A, B).

The patient's consciousness level markedly improved after intravenously administered glycerol, so she was operated on November 10th, 1983.

Operation.

Under general anesthesia, the patient was placed in the supine position with fixation of the head in a three-point headrest. A coronal

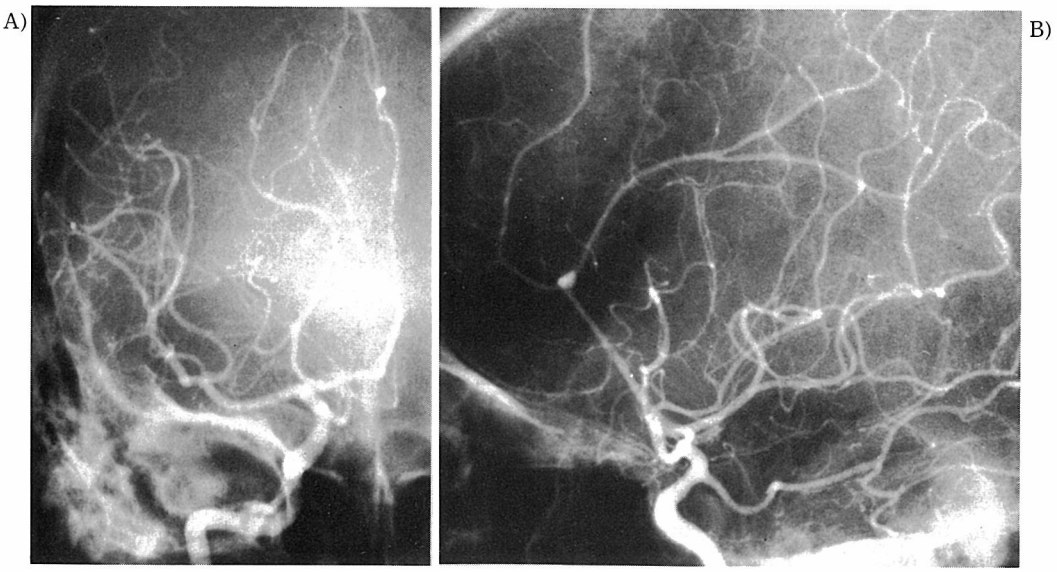

Fig. 3 Right carotid artery angiogram, anterior-posterior view (A) and lateral view (B), showing a saccular aneurysm at the junction of the pericallosal artery and the callosomarginal artery which had increased in size.

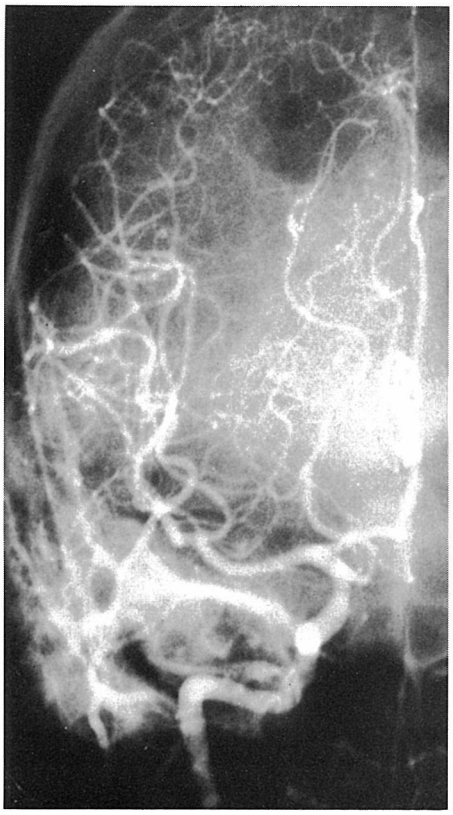

Fig. 4 Postoperative right carotid angiogram, anterior-posterior view showing complete obliteration of the aneurysm.

incision was made and the bone-flap was formed across the midline and extended posterioly to just in front of the coronal suture. The aneurysm was approached by midline exposure between the medial frontal lobe and falx using a microscope. A hematoma was noticed in the interhemispheric fissure and right cingulate gyrus. After evacuation of the hematoma, a partially thrombosed aneurysm with bleb was found at the junction of pericallosal artery and the callosomarginal artery. The aneurysm was clipped using two Sugita's clips.

The postoperative course was uneventful and a postoperative angiogram showed complete obliteration of the aneurysm (Fig. 4).

Discussion

Subarachnoid hemorrhage is a relatively common neurological complication of SLE but it is usually secondary to intracerebral hemorrhage[2]. Ellis and Verity[1] have reported a 30% incidence of subarachnoid hemorrhage in a neuropathological study of 57 patients with SLE. In all cases except one, the subarachnoid hemorrhage was accompanied by intracerebral hemorrhage or

diffuse petechial microhemorrhage, especially throughout the subcortical white matter. They reported that subarachnoid hemorrhage may have a close relationship to angiitis[3~5].

Harvey et al[2] reported a cerebral hemorrhage secondary to the rupture of a small aneurysm. They noted evidence of arteritis seen at necropsy, but did not describe the aneurysm histologically.

In SLE, however, there are a few reports in which cerebral aneurysm was shown angiographically following subarachnoid hemorrhage. Ferris[6] demonstrated multiple fusiform aneurysms at the bifurcation of the cerebral artery in SLE.

Taveras and Wood[7] also reported multiple fusiform aneurysms at sites of cerebral arterial bifurcation in SLE. Kelley et al[8] reported a patient with established SLE in whom a subarachnoid hemorrhage developed due to the rupture of a fusifrom aneuryam at the left posterior communicating artery, and at necropsy there was extensive transmural and lesser perivascular inflammation of the vessels at and around the site of rupture with fragmentation of the elastica and marked focal subintimal proliferation.

Ferris[6] reported that at autopsy, focal fibrinoid necrosis and elastic tissue disruption of the involved vessels were found. He theorized that fibrinoid necrosis commonly seen in SLE may be responsible for the aneurysm formation by producing local weakness in the walls of small vessels.

In the present case, a cerebral angiogram showed a saccular aneurysm which is different from that in reported SLE. In the operation, we could not obtain a specimen from the aneurysm and involved vessels. However, our patient has a history of subarachnoid hemorrhage 7 years previously, and a cerebral angiogram performed at that time showed a small aneurysm with wall irregularity of involved vessels which may have been attributable to angiitis. Generally, the histopathological features in SLE with involvement of the central nervous system are focal fibrinoid necrosis and elastic tissue disruption of the involved cerebral vessels[3~5].

From our angiographic findings and from the histopathological features in SLE, the authors presume that the focal fibrinoid necrosis and elastic tissue disruption of involved cerebral vessels may be responsible for a saccular aneurysm formation.

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