Multiple Saccular Cerebral Aneurysms associated with Systemic Lupus Erythematosus

—Case Report—

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Abstract

A rare case of multiple saccular cerebral aneurysms in a patient with systemic lupus erythematosus (SLE) is presented. The aneurysms were located at the junction of the left internal carotid artery (ICA) and the posterior communicating artery (PComA), at the anterior communicating artery, at the middle cerebral artery, at the junction of the right ICA and the PComA, and at the top of the basilar artery. All the aneurysms, except for the one on the basilar artery, which was unruptured, were successfully clipped. Histopathological examination of one resected aneurysmal dome confirmed the diagnosis of transmural arteritis secondary to SLE. Postoperatively, she died of a massive hemorrhage from a rectal ulcer. The etiology and prognosis of cerebral aneurysms in SLE are discussed.

Key words: multiple aneurysms, saccular aneurysm, transmural arteritis, systemic lupus erythematosus

Introduction

Although several cases of fusiform aneurysms in patients with systemic lupus erythematosus (SLE) have been reported, few cases of multiple saccular aneurysms associated with SLE have been previously reported. We describe such a case.

Case Report

A 52-year-old female was admitted to our department with severe headache, nausea, and vomiting of sudden onset. She had noticed a butterfly rash on her face 10 years prior to this hospitalization, and had soon thereafter developed fatigability. She had been admitted to our hospital 2 months prior to the attack. The diagnosis of SLE had been made on the basis of clinical symptoms, blood examination, and a renal biopsy, which had revealed mild, diffuse, proliferative and patchy membranous changes in the glomeruli. Two weeks prior to the attack she had been started on 40 mg/day of prednisolone. She had no history of hypertension.

On admission, she was alert but somewhat disoriented. No focal signs were present. Blood pressure was within the normal range. Routine blood examinations were negative except for an increased erythrocyte sedimentation rate. Computed tomography (CT) disclosed a subarachnoid hemorrhage, which was particularly severe in the left Sylvian fissure (Fig. 1). Four-vessel cerebral angiography revealed aneurysms at the junction of the left internal carotid artery (ICA) and the posterior communicating artery (PComA) (Fig. 2A), at the anterior communicating artery (AComA), at the right middle cerebral artery (MCA) (Fig. 2B), at the junction of the right ICA and the PComA (Fig. 2C), and at the top of the basilar artery (Fig. 2D). All these aneurysms were saccular.

Because the CT findings strongly suggested that
the aneurysm at the junction of the left ICA and the PComA was the one that had ruptured, this aneurysm was approached first, on the day of admission. It had in fact ruptured and was successfully clipped via the left pterional approach. Blood clots in the surrounding cistern were removed, and a large piece of the aneurysmal dome was resected for histopathological examination.

Microscopic examination of the resected specimen showed destruction of the normal structure of the vessel wall and diffuse, extensive infiltration by inflammatory cells through the vessel wall (Fig. 3). The diagnosis was transmural arteritis.

The postoperative course was uneventful until the fourth day, when she developed right hemiparesis and disturbance of consciousness. These events were thought to be attributable to angiospasm. Although she remained hemiparetic, her consciousness improved over the next 10 days.

A second operation was performed 51 days after the subarachnoid hemorrhage. All aneurysms except the one at the top of the basilar artery were uneventfully clipped via the right pterional approach. The basilar artery aneurysm was not clipped because the procedure could not be accomplished without clipping the perforating arteries as well. None of the aneurysms ruptured during surgery.

Although her postoperative recovery was excellent, she developed massive rectal bleeding on the second postoperative day. Endoscopy revealed a large ulcer on the posterior wall of the rectum. Because conservative treatment was not effective, the rectum was resected. However, she developed disseminated intravascular coagulation (DIC) and died on the 14th postoperative day. An autopsy was not performed.

Fig. 1 A CT scan demonstrating a subarachnoid hemorrhage, which is especially severe in the left Sylvian fissure.

Fig. 2 A: Left carotid angiogram disclosing an aneurysm (arrow) at the junction of the ICA and PComA. B: Right carotid angiogram, anteroposterior view, revealing an aneurysm on the AComA (arrow) and another on the MCA (double arrow). C: Right carotid angiogram, lateral view, demonstrating an aneurysm (arrow) at the junction of the right ICA and PComA. D: Left vertebral angiogram showing an aneurysm (arrow) at the basilar artery top.

Fig. 3 Photomicrograph showing destruction of the normal structure of the arterial wall and diffuse infiltration by inflammatory cells through the arterial wall. HE stain, × 400.

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Discussion

Although the incidence of subarachnoid hemorrhage ranges from 15.3 to 30% in autopsied SLE patients, the true incidence of cerebral aneurysms associated with SLE is unknown. Although cerebral angiography has disclosed multiple stenosis and fusiform aneurysms in some patients with SLE, few cases of multiple saccular aneurysms have been reported.

Cerebral arteritis, which almost always involves primarily the small arteries, is considered to be the most probable cause of aneurysmal formation in patients with SLE. Kelley et al. described one case in which transmural arteritis was histologically proved to be the cause of a fusiform aneurysm of the PComA. Ferris suggested that fibrinoid necrosis, a common histological finding in SLE, may be responsible for aneurysmal formation, since it produces local weakness in the walls of small arteries. In our case, transmural arteritis of medium to large arteries may have been the cause of aneurysmal formation, although involvement of medium and large arteries is uncommon in SLE.

Three instances of mortality have been reported in patients with SLE and cerebral aneurysms. Two patients died as a result of aneurysmal rupture and the third of cardiac insufficiency. Our patient died of massive bleeding from a rectal ulcer. Although the number of cases is too small to draw a conclusion, the prognosis for cerebral aneurysms associated with SLE seems to be poor.

References


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