Multiple Cerebral Aneurysms Associated with Aortitis Syndrome

—Case Report—

Shinjitsu NISHIMURA, Michiyasu SUZUKI, Kazuo MIZOI, and Takashi YOSHIMOTO

Division of Neurosurgery, Institute of Brain Diseases, Tohoku University School of Medicine, Sendai

Abstract

A 48-year-old female presented with four aneurysms in the anterior half of the circle of Willis associated with aortitis syndrome. All the aneurysms were successfully clipped. In general, intracranial hemodynamic change, due to stenosis or occlusion of carotid arteries, is considered to initiate aneurysm growth in the vertebrobasilar system in this syndrome, but renal hypertension was probably involved in our patient. Careful screening for multiple aneurysms, even in the anterior circulation, should be undertaken in patients with aortitis syndrome who present with an aneurysm.

Key words: cerebral aneurysm, aortitis syndrome, multiplicity, renal hypertension, autopsy

Introduction

Aortitis syndrome is caused by stenosis or obstruction of the aorta and its major branches due to an acquired, chronic, nonspecific inflammation. Associated brain lesions include cerebral aneurysms, middle cerebral artery (MCA) or internal carotid artery (ICA) occlusion due to emboli from intraneurysmal thrombus of the aorta, and cerebral hemorrhage due to renal hypertension. Several cases of intracranial aneurysms associated with aortitis syndrome have been reported recently. However, the mechanism of aneurysm growth is not well understood. The incidence of this association is also uncertain.

We describe a 48-year-old female with aortitis syndrome and multiple aneurysms in the anterior half of the circle of Willis, and discuss the mechanism of association and the incidence of aortitis syndrome associated with cerebral aneurysms.

Case Report

A 48-year-old female presented with persistent severe headache. Her father and brother had histories of hypertension. Previously, she experienced right retinal hemorrhage and was diagnosed as hypertensive in 1979. Aortitis syndrome was diagnosed based on elevated erythrocyte sedimentation rate, positive C-reactive protein and rheumatoid factor, an abdominal aortic aneurysm, and renal hypertension in 1982. One year later she underwent a radical operation for the abdominal aortic aneurysm. Postoperative aortography showed slight stenosis of the distal portion of the origin of the left subclavian artery, but no stenosis or obstruction of the main trunk leading rostrally. She experienced sudden onset of headache and a similar incident 2 days later in 1989. Since the headache improved gradually, she did not seek medical help.

She again experienced severe headache, and was admitted to our department when the headache persisted in October, 1992. She had no neurological abnormalities and her blood pressure was maintained within the normal range (132/76 mmHg) by medication. Laboratory tests revealed no abnormalities, and no acute worsening of the aortitis syndrome was indicated. Computed tomography (CT) revealed an
aneurysm and an old cerebral hemorrhage (Fig. 1). Right carotid angiography revealed a large aneurysm at the anterior communicating artery (AComA) complex, and left carotid angiography showed an aneurysm at the bifurcation of the ICA (Fig. 2). Right vertebral angiography demonstrated no abnormality.

The AComA and the ICA bifurcation aneurysms, and two more small aneurysms of the right MCA discovered during operation were clipped on December 17, 1992 (Fig. 3). The postoperative course was uneventful, and she was discharged without neurological deficits.

Discussion

The 17 previously reported patients with aneurysms associated with aortitis syndrome harbored a total of 27 aneurysms (Table 1). The incidence of aneurysms located on the vertebrobasilar system (11/27, 40.7%) was considerably higher than the natural incidence shown by studies of all intracranial aneurysms (5.4%, 4.0%). Multiplicity is also characteristic of cerebral aneurysms associated with aortitis syndrome. Seven of the 17 patients (41.2%) had multiple aneurysms, a considerably higher incidence than found in large series of aneurysm patients (between 7.7% and 19.0%).

The aneurysms probably develop due to hemodynamic stress caused by changes in blood circulation because an autopsy study (Shimabukuro et al., personal communication) has indicated ab...
sence of vascular inflammation of the cerebral aneurysm or parent artery in aortitis syndrome patients. Since the vertebrobasilar system is less affected than the carotid system in this syndrome, dilatation of the open vertebral artery and increased blood flow result in hemodynamic stress which promotes aneurysm formation in the vertebrobasilar system.\(^3,6,7,10,11,13,17,18,20\) Recently, several patients have been described with an aneurysm of the AComA.\(^1\) These patients demonstrated unilateral hypoplasia of the A\(_1\) portion of the anterior cerebral artery and stenosis or obstruction of the ipsilateral common carotid artery, which would lead to severe hemodynamic stress in the AComA and development of aneurysms.\(^7\) This is consistent with the suggestion that hemodynamic abnormalities of the anterior half of the circle of Willis are often found associated with AComA aneurysms.\(^8,9\) However, a four-vessel study in our patient did not reveal stenosis or obstruction of the main trunk arteries by the aortitis syndrome, so the development of the aneurysms was probably due to general hemodynamic stress in the brain caused by the renal hypertension.

The actual incidence of aortitis syndrome associated with cerebral aneurysm is still unknown. An autopsy study of 26 aortitis syndrome patients found only one (3.8%) with cerebral aneurysms,\(^1\) and none were found in another study of 77 patients.\(^16\) Review of published autopsies in Japan (Annual of the pathological autopsy cases in Japan by the Japanese Society of Pathology) has found a total of 133 aortitis syndrome cases in the past decade, including five patients with aneurysms (3.8%). The natural occurrence of aneurysm is between 0.2% and 9%,\(^5\) so the incidence of aortitis syndrome associated with aneurysm is within that range.

Various mechanisms for the development of aneurysms associated with aortitis syndrome have been proposed, but further work is required to clarify whether aortitis syndrome is directly responsible for the development of cerebral aneurysms. Although the coincidence of these lesions is rare, our present case emphasizes that care should be taken to screen for multiple aneurysms, even in the anterior circulation, once an aneurysm is revealed by angiography.

References


Neurou Med Chir (Tokyo) 34, December, 1994


Address reprint requests to: M. Suzuki, M.D., Department of Neurosurgery, Iwate Medical University, 19-1 Uchimaru, Morioka 020, Japan.