Ventricular Hemorrhage at an Early Stage of Moyamoya Disease

—Case Report—

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Abstract

A 19-year-old male presented with intraventricular hemorrhage manifesting as sudden onset of headache. Angiography showed mild stenotic changes in the distal internal carotid artery and proximal anterior cerebral artery only on the right. The anterior choroidal artery and lenticulostriate arteries appeared dilated, and an aneurysm-like shadow was seen in the distal right anterior choroidal artery. He was discharged without treatment. Eighteen months later, he presented with a second intraventricular hemorrhage manifesting as sudden occipitalgia, vomiting, and nausea. He had hyperreflexia of the left extremities and paresthesia of the left upper extremity. Angiography showed marked progression of the vascular abnormalities bilaterally. Moyamoya vessels were also present. He received bilateral encephalo-duro-arterio-myo-synangiosis with good results. Moyamoya disease may cause hemorrhage even at an early stage.

Key words: cerebral angiography, circle of Willis, intraventricular hemorrhage, moyamoya disease

Introduction

Moyamoya disease or syndrome is characterized by the progressive stenosis of the major intracranial arteries associated with the development of abnormal moyamoya vessels. Clinical analyses of patients with moyamoya disease have established some consensus about the process by which the disease progresses and becomes symptomatic.\(^5,7,9,12,17,21\) The primary pathological change is stenosis of the major arteries, which if severe enough may manifest as ischemic symptoms in children because of their high cerebral metabolic demand,\(^1\) followed by the development of abnormal fine collateral (moyamoya) vessels. Overload of these fragile vessels may result in intracerebral or intraventricular hemorrhage due to rupture of these vessels or rupture of microaneurysms formed on these vessels. In addition, changes in the hemodynamics may cause the formation of true aneurysms on the major arteries and result in subarachnoid hemorrhage.\(^6,11,12\) These three types of intracranial hemorrhage tend to occur in adult patients. According to this interpretation of the disease process, intracranial hemorrhages should occur later, i.e. after the severe stenosis has developed in the major arteries. In fact, most patients with moyamoya disease who underwent angiography had severe stenosis or occlusion of the major arteries at the time of hemorrhage.\(^1,3,7,9,11,12,16,20,21,24\)

We describe a 19-year-old patient with moyamoya disease at an early stage who presented with intraventricular hemorrhage when stenotic changes in the major arteries were mild and unilateral.

Case Report

A 19-year-old Japanese male suffered sudden onset of occipital headache while taking an entrance examination for a university. He consulted a physician 2 days later, when computed tomography (CT) revealed an intraventricular hematoma (Fig. 1 left). He was referred to our hospital. No neurological deficit was detected on admission. Angiography
performed 3 days and 1 month after admission showed mild stenotic changes in the distal portion of the internal carotid artery, the proximal portion of the anterior cerebral artery, and around the bifurcation of the middle cerebral artery only on the right side (Fig. 2). The anterior choroidal artery and lenticulostriate arteries appeared somewhat dilated and intensified. The late arterial phase visualized an aneurysm-like shadow in the distal portion of the right anterior choroidal artery. Left carotid and vertebral arteriography revealed no definite abnormality. He had no particular medical history such as sickle cell anemia, neurofibromatosis, tuberculous meningitis, atherosclerosis, or radiation exposure, which may cause abnormal vascular changes. He was discharged without surgical intervention.

Eighteen months later, he experienced a second intraventricular hemorrhage manifesting as sudden occipitalgia, nausea, and vomiting (Fig. 1 right). He had hyperreflexia of the left extremities and paresthesia of the left upper extremity. He developed hydrocephalus 1 week after admission. External ventricular drainage was performed. Angiography showed marked progression of the vascular abnormalities, as the mild stenotic changes previously limited to the right major arteries had progressed further and had become bilateral, which were compatible with moyamoya disease. Abnormal moyamoya vessels were now apparent at the base of the brain and in the terminal portion of the choroidal arteries. The aneurysm-like shadow previously noted in the distal portion of the right anterior choroidal artery had disappeared (Fig. 3). Three months later, bilateral encephalo-duro-arterio-myo-synangiosis was performed. Clumsiness in use of the left hand remained at discharge 2 months after the surgery.

Follow-up cerebral angiography, CT, magnetic resonance imaging, and cerebral blood flow study with single photon emission CT were carried out for 5 years. Good collateral blood flow through the surgical anastomoses and cessation of progression of occlusive changes and basal moyamoya formation were confirmed at 2 years following the surgery. The aneurysm-like shadow in the distal right anterior choroidal artery found on the first admission was no longer visualized.

**Discussion**

In the present case, initial angiography did not provide a diagnosis of moyamoya disease as the cause of intraventricular hemorrhage, since the stenotic changes in the major arteries were only on the right side and not very prominent. However, the dilation of the perforating arteries and the anterior choroidal artery, and the terminal microaneurysm were also abnormal findings, which retrospectively suggested that the patient had been in an early stage of moyamoya disease. Abnormal moyamoya vessels were now apparent at the base of the brain and in the terminal portion of the choroidal arteries. The aneurysm-like shadow previously noted in the distal portion of the right anterior choroidal artery had disappeared (Fig. 3). Three months later, bilateral encephalo-duro-arterio-myo-synangiosis was performed. Clumsiness in use of the left hand remained at discharge 2 months after the surgery. The diagnostic criteria established by the Japanese Ministry of Health and Welfare propose that patients with unilateral angiographic changes should be categorized as having "probable"
moyamoya disease. Recently, 23 patients including 17 children with "probable" moyamoya disease have developed bilateral involvement, 22 of whom presented with symptoms other than intracranial hemorrhage such as transient ischemic attack, infarction, or convulsion. In contrast, our patient was a young adult who presented with intracranial hemorrhage.

Intracranial hemorrhages may occur in moyamoya disease by the following process. Chronic ischemia due to stenotic changes in the major arteries triggers the development of two types of collateral vessels: the collateral pathway within the intracranial arteries, the basal moyamoya vessels, and the choroidal arteries; and the collateral pathway originating from the external carotid arteries, the "ethmoidal moya" and "vault moya." The latter type is seen only in children. Overload of the former type of collaterals in adults may result in formation of true aneurysms in the major arteries, which may rupture and cause subarachnoid hemorrhage. Review of the involvement of hemodynamic changes in the formation of true aneurysms showed that aneurysms were frequent in the basilar artery in patients with "bilateral" moyamoya disease and frequent in the anterior communicating artery in patients with "unilateral" moyamoya disease.

These hypotheses for the origin of intracranial hemorrhage require that the stenotic changes in the major arteries are severe enough to stimulate the development of the collateral vessels. However, in the present case, the stenotic changes in the major arteries were not very prominent. The severest change was narrowing of the anterior cerebral artery to less than 30% of the normal diameter, resulting in a blood flow reduction of less than 10%. The severity of stenotic changes required to trigger the formation of the collateral vessels is unknown but the dilatation of the perforating arteries and the anterior choroidal artery in this patient appeared to be disproportionately great compared with the stenotic changes. The collateral vessels apparently developed in parallel with the stenotic changes, suggesting that the ischemic trigger is not continuous but a transient repetitive process, so that the development of abnormal fine vessels is not a result of ischemia but a primary pathological process associated with the occlusion of the circle of Willis.

The present case shows that intracranial hemorrhage may occur at an early stage of moyamoya disease. It has been well documented that adult patients with moyamoya disease tend to present with hemorrhages. However, we cannot conclude that this disease does not cause hemorrhage before the completion of the typical angiographic changes unless the timing of hemorrhages associated with the various angiographic stages is analyzed in a large number of patients. Follow-up study with serial angiography is necessary in patients with intracranial hemorrhage of unknown origin since moyamoya disease might be an underlying cause of the hemorrhage.

**References**


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