Extravasation from Anterior Choroidal Artery in a Child with Moyamoya Disease
—Case Report—

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Abstract
A rare association of moyamoya disease with intra- and periventricular hemorrhage in an 8-year-old girl is presented. Angiography showed that the cause of hemorrhage was extravasation from the dilated anterior choroidal artery. Ventriculoperitoneal shunt and encephalomyosynangiosis have prevented clinical or neurological deficits for over 6 years.

Key words: moyamoya disease, intraventricular hemorrhage, extravasation, anterior choroidal artery, encephalomyosynangiosis

Introduction
Intracranial hemorrhage is the most common initial manifestation of moyamoya disease in adults. However, this is rarely the case in children. Among the reported cases of moyamoya disease in children, there are only four cases of intraventricular hemorrhage (IVH) and two autopsy cases of subarachnoid hemorrhage or intracerebral hemorrhage. The advent of computed tomographic (CT) scanning has shown that most moyamoya disease cases present with intracerebral hemorrhage or IVH.

We present a child case of IVH demonstrated by CT scanning. Cerebral angiography showed that the cause of hemorrhage was bleeding from the anterior choroidal artery associated with moyamoya disease. The reasons for the low frequency of IVH in childhood moyamoya disease are discussed.

Case Report
An 8-year-old girl suffered a sudden onset of severe headache with vomiting and subsequently lost consciousness on April 8, 1982. She was referred to the Sannocho Hospital after pediatric consultation. Her brother was wheelchair-bound after a stroke associated with moyamoya disease.

On admission, she was stuporous with neck stiffness, right hemiparesis, and hyper-reflexia. There was bilateral Babinski sign. CT scans revealed high-density areas filling all the ventricles and extending to the left periventricular region (Fig. 1). Emergency cerebral angiography revealed narrowing of the anterior cerebral arteries, bilateral basal moyamoya vessels, and marked development of the left anterior choroidal artery, the bilateral lenticulostriate arteries and posterior cerebral arteries. Left carotid angiogram demonstrated extravasation of the contrast medium from the peripheral portion of the dilated...
anterior choroidal artery (Fig. 2). No aneurysm was detected. CT scanning after the angiography disclosed pooling of contrast medium in the posterior horn of the left lateral ventricle. The diagnosis was moyamoya disease associated with IVH and periventricular hemorrhage. The angiographic findings showed the early stage of moyamoya disease according to Suzuki's classification 12,11; stage 3 on the left and stage 2 on the right.

Her level of consciousness gradually improved, but she was mute. Follow-up CT scans revealed marked hydrocephalus. A ventriculoperitoneal shunt and encephalomyosynangiosis (EMS) were established on the left on April 26. She became responsive to verbal commands on April 30, and could speak a few words despite dysphasia 3 days later. The dysphasia and right hemiparesis gradually resolved. She was discharged and walked home on May 29, 1982. She was subsequently doing well and enjoying school without neurological deficit.

On August 16, 1983, follow-up angiography demonstrated newly developed anastomotic vessels between the EMS and cortical arteries. The left anterior choroidal artery dilatation was resolved. A stenosis at the middle cerebral artery origin and basal moyamoya had developed on the right. Right EMS was established to prevent ischemic stroke on August 23, 1983. In the subsequent 6-year follow-up, she has never experienced ischemic stroke, hemorrhage, mental deterioration, or physical disability.

**Discussion**

In childhood moyamoya disease, the most common manifestation is ischemic stroke caused by progressive occlusion of the circle of Willis.\(^8,12,13\) Angiography and histology show that occlusions and stenoses occur in the circle of Willis and also in the cerebral peripheral arteries.\(^4,16\) Chronic ischemia probably induces anastomotic vessels to develop as collateral channels.

Suzuki and Takaku\(^12\) named the abnormal net-like vessels “moyamoya.” Moyamoya vessels may be basal moyamoya, ethmoidial moyamoya, and vault moyamoya.\(^12\) Additionally, the anterior or posterior choroidal artery may establish a dominant anastomosis, rupture of which causes IVH. However, among 50 child patients with moyamoya disease in the last 10 years at our hospitals, IVH lead to the diagnosis in only three cases including the present one. In these cases, angiography demonstrated the early stage of moyamoya disease and the development of anterior choroidal arteries. The four reported cases of moyamoya with IVH\(^1,2,14\) demonstrated neither aneurysmal rupture nor angiographic extravasation. Three of these four cases were apparently in the early stage of moyamoya disease with an enlarged anterior choroidal artery as in our IVH cases.\(^1,14\) Marked development of the posterior choroidal artery occurred in one case.\(^2\)

Our case is the first moyamoya patient in childhood to demonstrate extravasation from the dilated peripheral anterior choroidal artery without associated aneurysm. Yamashita et al.\(^16\) reported a higher frequency of dilated perforating arteries in young patients, but only one case of intracerebral hematoma.\(^16\) Therefore, we consider that although the dilated perforating arteries are not fragile, the anastomotic vessels of the choroidal arteries are easily broken by the hemodynamic stress in childhood moyamoya disease, especially where a choroidal artery has developed in the early stage. The cause of adult intracranial hemorrhage associated with moyamoya disease has been shown to be extravasation or pseudoaneurysm of the peripheral choroidal arteries.\(^3,5,10,11,15\) Dilated choroidal arteries are therefore not sufficiently resistant to hemodynamic stress in both childhood and adult moyamoya disease. However, only one child case of SAH due to aneurysmal rupture has been reported,\(^10\) whereas cerebral aneurysmal rupture frequently occurs in adults in the major arteries and also in the peripheral or perforating arteries.\(^2,6,7,16,17\)

Surgical treatment, such as external carotid-internal carotid bypass and EMS, has been used to treat...
moyamoya disease. Such preventive surgery probably decreases the frequency of hemorrhage because the extent of moyamoya vessels and choroidal artery dilatation decreases postoperatively, as seen in our case. We are undertaking a long-term follow-up study to evaluate such preventive surgery for both ischemic stroke and hemorrhage associated with moyamoya disease. In our case, EMS effectively prevented stroke, hemorrhage, or mental deterioration during a period of 6 years.

Acknowledgment

The authors wish to thank Professor Ryuichi Tanaka, Department of Neurosurgery, Brain Research Institute, Niigata University, for his valuable comments.

References


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