Neuroendoscopic Findings of Ventricular Wall in Adult Hemorrhagic Moyamoya Disease: Report of Two Cases

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Moyamoya disease usually manifests as ischemic events in childhood, and as more severe hemorrhagic events, including intraventricular hemorrhage, in adults. Recently, the indication for neuroendoscopic surgery has been extended to cast-formation intraventricular hematomas. However, detailed information about the use of neuroendoscopic surgery for the treatment of intraventricular hemorrhage associated with moyamoya disease has not been reported. We describe two cases of intraventricular hemorrhage with moyamoya disease; one in a 62-year-old and another in a 33-year-old women who both presented with severe neurological symptoms. Cerebral angiography revealed unilateral moyamoya disease. Neuroendoscopic surgery to remove the intraventricular hematoma was performed via bilateral frontal burr holes in both cases. Abnormal findings in the ventricle were observed only in the affected side and the intact side was normal. Specific findings of neuroendoscopic observation were dilated and tortuous vessels, intersection vessels, black-brown macules in the subependyma, and rattan blind-like (Japanese sudare) bleeding vessels. These characteristic neuroendoscopic findings may be useful for the exact diagnosis and treatment of intraventricular hemorrhage associated with moyamoya disease. Endoscopic evacuation of the ventricular hematoma may be important for intracranial pressure control in patients with intraventricular hemorrhage in adult moyamoya disease.

Keywords: moyamoya disease, intraventricular hemorrhage, neuroendoscopy

Introduction

Moyamoya disease usually manifests as ischemic events in childhood, and as more severe hemorrhagic events in adults. Cerebral hemorrhage occurs in only 19% of first attacks in patients with moyamoya disease,[1] but is associated with a poor prognosis, as demonstrated by a post-mortem study that found 16 of 20 adult patients with moyamoya disease died of hemorrhage.[2] Moyamoya disease may cause intracerebral hematoma, subarachnoid hemorrhage, and intraventricular hemorrhage. Intraventricular hemorrhage is particularly common, as observed in 63–81% of cases.[2,3]

The introduction of neuroendoscopic surgery has improved the safety and prognosis of the removal of intraventricular hemorrhage.[4–10] Neuroendoscopic surgery has often been performed to treat hypertensive intracerebral hemorrhage. Recently, the indication for neuroendoscopic surgery has been extended to cast-formation intraventricular hematoma. Neuroendoscopic surgery is effective for the removal of cast-formation intraventricular hematoma caused by ruptured aneurysm with severe neurological symptoms, resulting in improvement in neurological status, resolution of acute hydrocephalus, and shorter ventricular drainage catheter placement times.[4–6] Neuroendoscopic procedures for the evacuation of intraventricular hematoma have been reported in 17 cases, including 1 case of moyamoya disease without particular details.[4] However, detailed information about the use of neuroendoscopic surgery for the treatment of intraventricular hemorrhage associated with moyamoya disease has not been reported.

We describe two cases of adult hemorrhagic type moyamoya disease manifesting as intraventricular hematoma that were treated by neuroendoscopic surgery, and discuss the specific neuroendoscopic observations of this disease, and hematoma removal by neuroendoscopy.

Case Reports

I. Case 1

A 62-year-old woman suffered sudden onset of unconsciousness. Computed tomography (CT) demonstrated a cast-formation intraventricular hematoma and acute obstructive hydrocephalus. The bleeding point was identified as parenchymal hemorrhage in the left para-third ventricle (Fig. 1A). Emergent external ventricular drainage was performed to prevent central herniation. Cerebral angiography revealed moyamoya vessels extending from the left internal carotid artery terminal through the proximal left middle cerebral artery, and absence of the proximal anterior cerebral artery (Fig. 1B, C). Right internal carotid angiography showed no abnormality. No sign corresponding to quasi-moyamoya disease was identified.[11] The diagnosis was unilateral moyamoya disease according to the recommendation for the management of moyamoya disease of the Research Committee on Spontaneous Occlusion of the Circle of Willis.[12]

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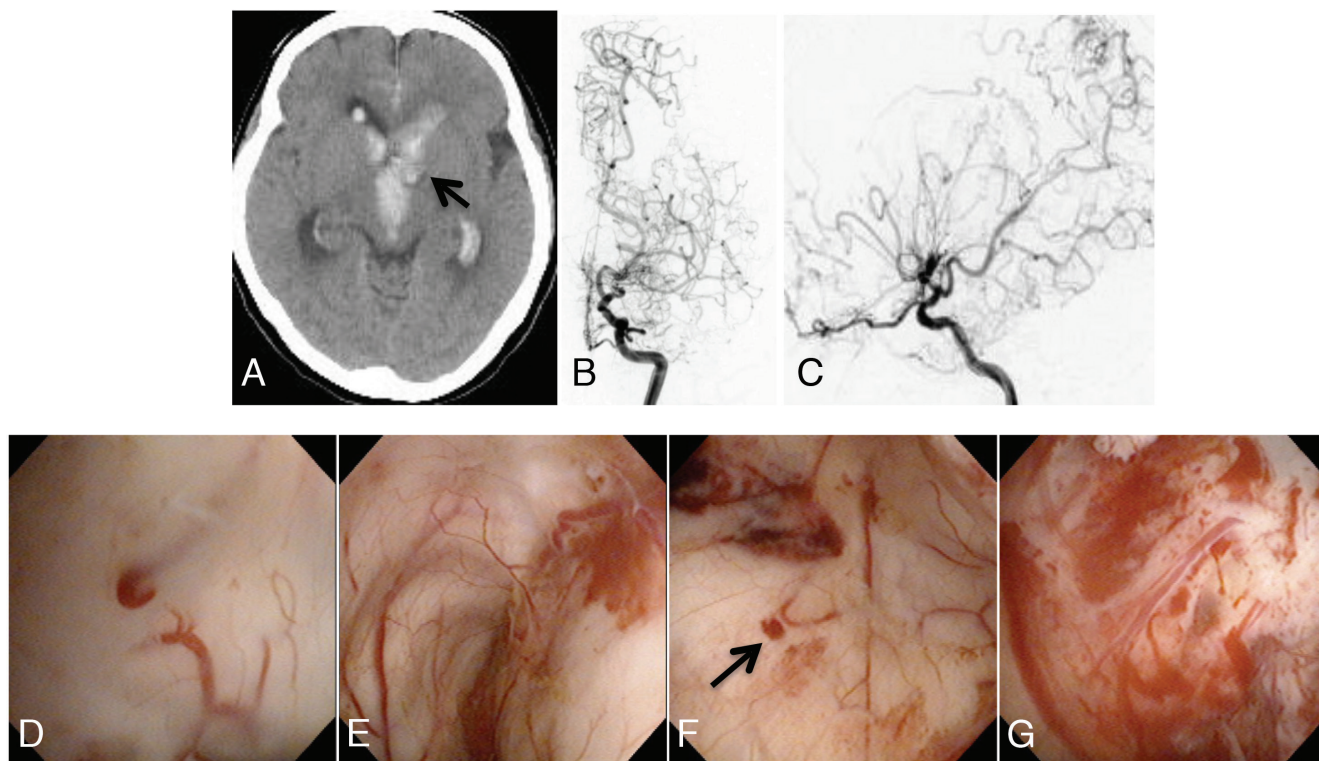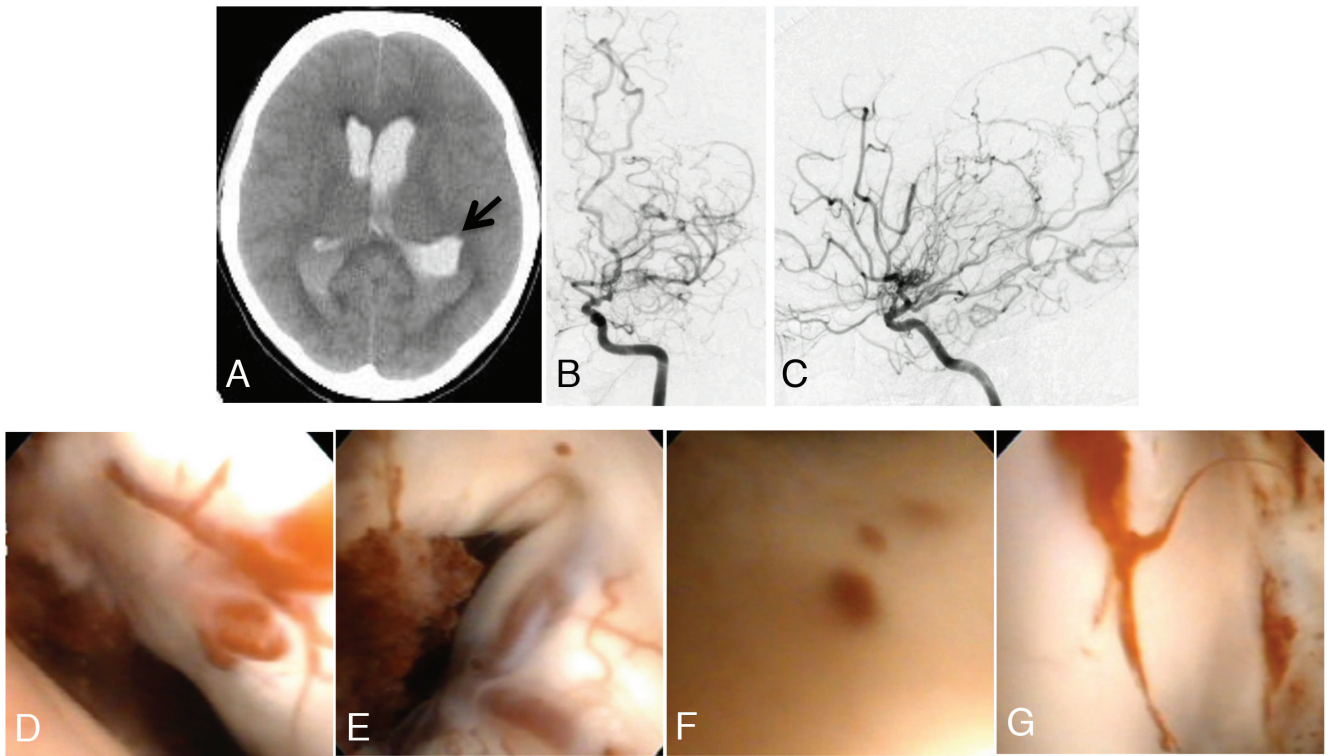
Fig. 1 Case 1. A: Computed tomography scan taken at onset showing bleeding from the left para-third ventricle (black arrow). B: Left internal carotid angiogram demonstrating moyamoya vessels extending from the internal carotid artery terminal, with stenosis of the middle cerebral artery and absence of the anterior cerebral artery. C: Lateral view of left internal carotid angiogram. D: Left lateral ventricle body. Subependymal tortuous vessels are exposed in the ventricle. E: Left lateral ventricle body. Abnormal small vessels are tortuous and dilated, with subependymal hemorrhage. F: Left lateral ventricle body. Black macules are present (black arrow). G: Left lateral side of the third ventricle. Exposed rattan blind-like vessels are bleeding. Copious subependymal hemorrhage is present.

Neuroendoscopic surgery to remove the intraventricular hematoma was performed on the 12th hospital day, as her neurological symptom had not improved and intraventricular hematoma was insufficiently washed out. Prolonged extraventricular drainage was predicted. Neuroendoscopy used a fiberscope (VEF type V, Olympus, Tokyo) and peel-off introducer sheaths (17.5 Fr, Medikit, Tokyo), which were introduced into the bilateral frontal burr holes. Hematoma was removed from the third ventricle and then the aqueduct was opened. The intact right lateral ventricle had a white, smooth, and glossy ependymal surface. In contrast, the left lateral ventricle contained tortuous, abnormally dilated subependymal vessels, exposed in the ventricle and affecting the entire left lateral ventricle body (Fig. 1D, E). Black-brown macules were present in the lateral ventricle body (Fig. 1F). The ependyma was pale on the affected side. The vulnerable blood vessels drooped like a rattan blind in the anterior part of the lateral ventricle, which contained parenchymal hemorrhage (Fig. 1G). No such findings were observed in the anterior horn of the lateral ventricle and anterior part of the third ventricle. Third ventriculostomy was then performed. Hydrocephalus did not recur after removal of the drainage catheter. One week after surgery, computed tomography (CT) showed cerebral infarction in the bilateral frontal lobes.

II. Case 2

A 33-year-old parous woman suffered sudden onset of unconsciousness. Her son had been diagnosed with moyamoya disease at age 4 years. CT showed a cast-formation intraventricular hematoma and acute obstructive hydrocephalus. The bleeding point was identified as parenchymal hemorrhage at the left pulvinar (Fig. 2A). Ventricular catheter was installed immediately. Cerebral angiography revealed moyamoya vessels extending from the left internal carotid artery terminal through the proximal left middle and anterior cerebral arteries (Fig. 2B). Right internal carotid angiography showed no abnormality. Brush-like capillary vessels were aggregated at the pulvinar (Fig. 2C). Unilateral moyamoya disease was diagnosed.

Neuroendoscopic hematoma evacuation was performed on the 6th hospital day, as her neurological condition had not improved and intraventricular hematoma was insufficiently washed out. The procedure was similar to that for Case 1. Third ventriculostomy was also then performed. Intact right lateral ventricle ependyma had a white, smooth, and glossy surface. Subependymal vessels were tortuous and abnormally dilated, mainly in the left lateral ventricle body (Fig. 2D), but without obvious exposure in the ventricle. Vessels across the ventricle were close to the foramen of Monro (Fig. 2E). Many black-brown macules were observed in the

Fig. 2 Case 2. A: Computed tomography scan taken at onset showing bleeding from the pulvinar of the forward left ventricle trigone (black arrow). B: Left internal carotid angiogram demonstrating moyamoya vessels extending from the internal carotid artery terminal, with stenosis of the middle cerebral artery and absence of the anterior cerebral artery. C: Brush-like arteries branched from the anterior choroidal artery at the origin of the parenchymal hemorrhage. D: Left lateral ventricle body. Subependymal vessels are dilated and tortuous. E: Left lateral ventricle body. Intersecting vessels are present on the side of the lateral ventricle. F: Left lateral ventricle body. Black macules are seen. G: Left lateral side of the third ventricle. Exposed rattan blind-like vessels are the origin of venous bleeding.

lateral ventricle body (Fig. 2F). Exposed vessels from the medial wall of lateral ventricle body near the bleeding point were the source of the venous bleeding (Fig. 2G). CT did not demonstrate intraventricular hematoma on the day after surgery. One week after surgery, CT showed cerebral infarction in the cingulate gyrus.

Discussion

Late-onset cerebral infarction after intraventricular hemorrhage occurred in both our patients. Infarction in peripheral vascular areas in the subacute phase has been reported in hemorrhagic type adult moyamoya disease.[1,13–16] Including our cases, 10 patients developed infarction during the 7th through 16th days after hemorrhage. Continuous high intracranial pressure (ICP) due to massive hemorrhage and obstructive hydrocephalus induces insufficient collateral flow resulting in irreversible ischemic damage.[13–15] Therefore, ICP must be controlled adequately after intraventricular hemorrhage to prevent late-onset cerebral infarction.[13–15] Intraventricular hematoma may also induce late-onset vasospasm.[16,17] The pathology of late-onset infarction is unclear, but may be indirectly related to ICP change caused by hydrocephalus and hematoma after intraventricular hemorrhage. Neuroendoscopic evacuation of the ventricular hematoma may be effective to resolve these problems.

The techniques and instruments for neuroendoscopic evacuation of intraventricular hematoma are still under development.[5,8,10] Therefore, we initially performed emergent extraventricular drainage in our two patients with intraventricular hematoma associated with moyamoya disease. However, hematoma evacuation was subsequently performed safely with a fiberscope. The intraventricular hematoma was easily evacuated under negative pressure. The presumed bleeding point was covered by solid hematoma with destroyed parenchymal brain tissue and choroid plexus, so aggressive evacuation was not performed. Evacuation of hematoma lodged in the septum pellucidum caused venous bleeding, which spontaneously ceased after continuous irrigation with artificial cerebrospinal fluid (CSF) for about 5 minutes.

Our present cases demonstrated some specific findings during endoscopic evacuation of intraventricular hemorrhage in adult moyamoya disease: dilated and tortuous vessels (Figs. 1D, 2D), intersecting vessels (Figs. 1E, 2E), black-brown macules in the subependyma (Figs. 1F, 2F), and rattan blind-like bleeding vessels (Figs. 1G, 2G). Dilated subependymal arteries lacking internal elastic lamina were observed in 12 of 20 previous autopsy cases. Our neuroendoscopic findings of dilated subependymal arteries were consistent with the previous autopsy report in Japan.[2] Artery and vein

intersections were similar to the changes of the ocular fundus induced by hypertension. Such small vessels may have developed under local hemodynamic hypertension. Subependymal black-brown macules were considered to be hemosiderin-like deposits originating from subependymal microbleeding. These macules were suspected to represent previous subclinical subependymal hemorrhage. The rattan blind-like bleeding vessels were all subependymal vessels, and may have appeared after destruction of the ventricle wall by the primary intraventricular hemorrhage. The bleeding could be easily stopped by irrigation with artificial CSF. Fibrinoid necrosis and microaneurysm have not been reported in hypertensive parenchymal hemorrhage associated with adult moyamoya disease,[2] so intraventricular hemorrhage in adult moyamoya disease may have a different pathology from arteriosclerosis. In fact, the neuroendoscopic appearance of the ventricle ependyma near the hemorrhagic origin was intact in hypertensive parenchymal hemorrhage. The ventricular anatomic structures were easily identified in a series of intraventricular hematoma excluding moyamoya disease.[7] The present cases of intraventricular vascular abnormality associated with moyamoya disease had almost the same findings as the previous postmortem study.[2]

The main clinical manifestation of moyamoya disease is main trunk stenosis or occlusion, but dilation of the perforators, anterior choroidal artery (AchA), and posterior communicating artery (PcomA) also occur.[18,19] Dilation and branch extension of the AchA and PcomA are predictors for adult hemorrhagic type moyamoya disease, and may be observed in young patients, but are rare in ischemic type moyamoya disease, as the dilated AchA and PcomA have branches that act as collateral vessels.[18] The autopsy study also observed perforator dilation more commonly in young patients.[19] Additional sclerotic change in the dilated artery will lead to local lateral projection and microaneurysm formation.[19] The AchA and PcomA acting as collateral vessels developed with dilation changes and the requirement for ischemic tolerance. However, sclerotic change progresses with aging and hemodynamic stress, so patients with adult moyamoya disease tend to suffer bleeding from dilated arteries. The presence of dilated arteries may be correlated with intraventricular hemorrhage[13] and parenchymal microbleeding.[20]

Both our patients had unilateral moyamoya disease. The present abnormal findings were observed only in the ventricle of the affected side, whereas normal findings were observed in the intact side. Our present neuroendoscopic findings showed that vessels under hemodynamic load underwent dilation changes in the ventricle of the diseased side. Such vessels increased in the subependyma, and were fragile because of the absence of internal elastic lamina. Such fragile vessels could induce hemorrhage by disruption of the ependyma which is less strong than the parenchyma. In contrast, the ventricle had almost normal appearance outside the AchA perfusion area, including the anterior horn of the lateral ventricle and the floor of the third ventricle. The anterior horn of the lateral ventricle without choroid plexus appeared normal even on the diseased side. These findings suggest that little hemodynamic change occurred outside the

area of AchA perfusion. Therefore, basic techniques such as anterior puncture and third ventriculostomy could be performed safely in the present cases.

Conclusion

Neuroendoscopic observation of our two cases of adult hemorrhagic type moyamoya disease manifesting as intraventricular hematoma found dilated and tortuous vessels, intersecting vessels, black-brown macules in the subependyma, and rattan blind-like bleeding vessels on the affected ventricle walls, which are specific findings of collateral vessels of moyamoya disease.

Conflicts of Interest Disclosure

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