Caudate Hemorrhage With Moyamoya-like Vasculopathy From Atherosclerotic Disease

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Background and Purpose: Caudate hemorrhage usually results from hypertension, rupture of arteriovenous malformation or aneurysm, or rarely, moyamoya disease. Moyamoya-like changes related to severe atherosclerotic occlusive disease, usually causing ischemic stroke, have been reported.

Case Description: A 51-year-old normotensive patient was admitted with headache due to a left caudate hematoma with ventricular extension. There was a history of smoking, leg claudication, elevated cholesterol, and coronary artery disease. Angiography demonstrated complete extracranial carotid occlusion on the left and atherosclerotic stenosis at the bifurcation on the right, with supraophthalmic occlusion distally. At the base of the brain, bilateral moyamoya-like vessels, presumed to be secondary to atherosclerotic occlusion, were evident, but neither aneurysm nor arteriovenous malformation was present. Cerebral blood flow and transcranial Doppler studies indicated severely impaired cerebral perfusion that improved after bilateral extracranial-to-intracranial bypass surgery.

Conclusions: Atherosclerotic occlusive carotid disease with moyamoya-like changes may be a rare cause of caudate hemorrhage. A decrease in moyamoya vessels with bypass surgery may reduce the risk of recurrent hemorrhage.

KEY WORDS • carotid artery diseases • cerebral hemorrhage • moyamoya disease

Caudate hemorrhage is an uncommon stroke syndrome, accounting for 3–7% of all primary intracerebral hematomas.1-3 Hypertension appears to be the most important risk factor,1,2,4,5 although arteriovenous malformations (AVMs) or aneurysms are also potential causes.2,5 Rarely, caudate hemorrhage has occurred in the setting of occlusive disease of the intracranial internal carotid artery (ICA) and the circle of Willis, associated with an abnormal network of collateral vessels at the base of the brain or with moyamoya disease.6-7 Vascular changes similar to those in typical moyamoya disease have been associated with atherosclerosis, usually presenting with cerebral infarcts, and less often, with subarachnoid and ventricular hemorrhage.8-11 We report the case of an unusual patient with atherosclerotic risk factors and bilateral carotid occlusions with moyamoya-like vessels who presented with a caudate hemorrhage. Extracranial-to-intracranial (EC-IC) bypass surgery was performed to reduce the risk of recurrent hemorrhage and improve cerebral hypoperfusion. This patient illustrates the theory that one cause of caudate hemorrhage is moyamoya-like changes in the setting of presumed atherosclerotic occlusive disease.

Case Report

A 51-year-old non-hypertensive white man experienced the sudden onset of severe headache, nausea, and vomiting without focal symptoms. There was a history of long-standing cigarette smoking and mild hypercholesterolemia. Intermittent claudication had been recently diagnosed. Initial neurological examination was normal except for minimal nuchal rigidity. Vomiting occurred repeatedly in the first hours after onset, whereas headache persisted over a few days.

Computed tomographic scan on the following day revealed a left caudate hemorrhage with extension into the lateral, third, and fourth ventricles (Figure 1). Selective angiography demonstrated delayed filling of the right ICA caused by a high-grade stenosis at its origin and occlusion of its supraophthalmic portion (Figure 2, left panel). A moyamoya-like vascular network was seen at the base of the brain around the supraclinoid ICA, with collateral flow through the ethmoidal arteries supplied by the ophthalmic artery (Figure 3, left panel). The left ICA was occluded at its origin without filling of the intracranial supraophthalmic segment, probably caused by absent collateral flow from the ophthalmic artery, which did not opacify (Figure 2, right panel). Maxillary branches supplied small ethmoidal collaterals (Figure 3, right panel). The vertebral, basilar, and posterior cerebral arteries were all normal. The posterior communicating arteries supplied the anterior circulation. However, the left posterior communicating artery was stenotic at its ostium in the ICA.
FIGURE 1. Ventricular hemorrhage emanating from head of left caudate nucleus on initial computed tomographic scan.

Leptomeningeal collaterals from the posterior to the middle and anterior cerebral arteries were also present. There was no evidence of aneurysm or AVM. Duplex Doppler studies were consistent with the extracranial angiographic findings. Three-dimensional transcranial Doppler sonography demonstrated abnormally low flow velocity in both middle cerebral arteries (mean velocities: 29 cm/sec, right; 34 cm/sec, left) and confirmed collateral supply to the anterior circulation through the posterior communicating arteries. Flow velocity was markedly increased at the junction of the left posterior communicating artery and ICA, indicating local stenosis. Cerebral blood flow studies using the xenon-133 inhalation method showed severely reduced resting flow bilaterally with an initial slope index of 34.5 m⁻¹ for the right hemisphere and 34.0 m⁻¹ for the left, although hypercapnic vasoreactivity was normal (right, 3.3%/mm Hg; left, 4.1%/mm Hg). A right EC-IC bypass was performed 20 days after symptom onset, followed by another bypass on the left side 32 days later. On follow-up examination 9 months after the second bypass operation, the patient was neurologically asymptomatic. He had suffered a small myocardial infarction 2 weeks after discharge from the hospital. Transcranial Doppler confirmed patency of both anastomoses and retrograde flow in both middle cerebral arteries with a higher mean flow velocity on the right (75 cm/sec) than on the left (42 cm/sec). Cerebral blood flow was increased bilaterally (right, 38.2 m⁻¹; left, 39.4 m⁻¹), and hypercapnic vasoreactivity remained normal (2.1%/mm Hg bilaterally).

Discussion

To our knowledge, there have been no previous reports of caudate hemorrhage attributed to atherosclerotic moyamoya-like changes. A moyamoya-like pattern due to atherosclerosis is a rare observation. In a review of the literature in 1985, Sato et al. found eight reported cases of ventricular hemorrhage, four of which involved the carotid artery. We are aware of only one other report of a patient with moyamoya-like vessels due to severe extracranial atherosclerotic carotid

FIGURE 2. Lateral view of selective carotid angiograms. Left panel: High-grade stenosis of right internal carotid artery (ICA) at bifurcation, with adaptive narrowing of vessel lumen distal to stenosis. Intracranial occlusion is immediately beyond origin of ophthalmic artery. Right panel: Complete extracranial occlusion of left ICA.
disease, in whom ventricular hemorrhage occurred after carotid endarterectomy.

The angiographic findings in our patient met the criteria for typical moyamoya disease:1,13 1) The process was bilateral, involving the supraophthalmic segment of the ICA on one side. Because of the absence of an ophthalmic collateral, it can be assumed that the primary site of the contralateral ICA occlusion was also intracranial, with retrograde thrombosis to its cervical bifurcation. 2) Moyamoya-type vessels at the base of the brain were present, with dilated lenticulostriate arteries and ethmoidal collaterals. 3) The posterior circulation was not involved. However, we infer that atherosclerotic disease was the most likely cause of the occlusions because our patient had generalized atherosclerosis (leg claudication and coronary heart disease), with evidence of atherosclerotic disease at the carotid bifurcation on the right side. Extracranial carotid disease in classical moyamoya disease must be considered extremely rare.7 In a study of 13 patients with moyamoya disease, only one patient had an occlusion of the extracranial carotid artery; however, the occlusion was distal to the bifurcation, possibly a result of progressive secondary narrowing.

Chronic hypertension that produces pathological changes of small penetrating arteries is probably the most frequent cause of caudate hemorrhage.12,25 Carotid or anterior cerebral artery aneurysm and AVM are other possible sources of bleeding.2,5 In one study,3 3% of 315 patients with hypertensive intracerebral hemorrhages and 6% of 70 patients with AVMs presented with caudate hematomas. In our normotensive patient, the ventricular hemorrhage emanating from the head of the caudate nucleus was most likely caused by rupture of small, penetrating, moyamoya-like vessels because there was no angiographic evidence of aneurysm or AVM.

This mechanism has been postulated as the cause of hemorrhages in patients with typical moyamoya disease. Suzuki13 suggested that the formation of "pseudo-aneurysms" of small, penetrating arteries due to chronic ischemia of the white matter adjacent to the lateral ventricle is a frequent cause of intraventricular bleeding in moyamoya patients. Among 60 computed tomography–documented hematomas in patients with classic moyamoya disease,6,7 19 (32%) were primary intraventricular hemorrhages, while 14 (23%) each were medial ganglionic (i.e., caudate and thalamus) and lateral ganglionic (i.e., putaminal and capsular). Thirteen other hematomas (23%) had a variable distribution, with predominant involvement of the temporal lobe (n=7). The reported angiographic data are too limited to determine whether medial and lateral hemorrhages have different patterns of intracranial artery occlusion and collaterals.7

By contrast, only a few examples of intracranial hemorrhage due to moyamoya-changes related to atherosclerotic disease have been reported. Among five reported cases, the exact location of the hemorrhage was not known in three because computed tomography was not available.11,14 One patient had ventricular hemorrhage without obvious parenchymatous involvement,9 and another had a temporal lobe hematoma.15

The preserved hypercapnic vasoreactivity in our patient may reflect differences in the underlying vascular disease process between typical moyamoya disease and moyamoya-like vasculopathy associated with atherosclerosis. Findings consistent with those of our case were observed in a study using the 133Xe inhalation technique.16 In that study, hypercapnic vasoreactivity was markedly lower in patients with moyamoya disease than in those with atherosclerotic bilateral carotid occlusions, although resting blood flow was lower in the latter group.

We considered this patient to have a high risk of both recurrent hemorrhage due to moyamoya-like vasculopathy and ischemic stroke from insufficient collateral
supply to the anterior circulation. As in typical moyamoya disease, EC-IC bypass surgery appeared to be the most rational therapy for this condition.

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