Persistent Primitive Hypoglossal Artery Associated with Cerebral Aneurysm and Cervical Internal Carotid Artery Stenosis
—Case Report—

Takashi HATAYAMA, Kanji YAMANE, Takeshi SHIMA, Yoshikazu OKADA*, and Masahiro NISHIDA

Department of Neurosurgery, Chugoku Rousai Hospital, Kure, Hiroshima; *Department of Neurosurgery, Tokyo Women’s Medical College, Tokyo

Abstract

A 71-year-old female had vertigo attacks once or twice a day secondary to vertebrobasilar insufficiency. Left carotid angiography revealed persistent primitive hypoglossal artery (PPHA) associated with a large internal carotid artery (ICA) aneurysm and severe stenosis of the ICA. The bilateral vertebral arteries were hypoplastic. The basilar artery was opacified via the PPHA but not via vertebral arteries. Clipping of the aneurysm was performed first because the risk of rupture of the aneurysm was not negligible. One month after clipping, carotid endarterectomy using a T-shaped shunt system was successfully performed. The postoperative course was uneventful and the vertebrobasilar ischemic attacks did not recur. Left carotid angiography demonstrated complete obliteration of the aneurysm and disappearance of the carotid artery stenosis. Low ICA flow (70 ml/min) and low stump pressure of the PPHA (25 mmHg) strongly suggested low perfusion of the posterior circulation. Carotid endarterectomy may be essential for augmentation of the posterior circulation in patients with PPHA associated with ICA stenosis.

Key words: persistent primitive hypoglossal artery, cerebral aneurysm, internal carotid artery stenosis, carotid endarterectomy

Introduction

Persistent primitive hypoglossal artery (PPHA) is one of the rudimentary vessels forming the carotid-basilar anastomoses in the fetus. PPHA is usually observed incidentally at the time of carotid angiography and the estimated incidence is 0.05%. We treated a patient with ischemic attacks of the vertebrobasilar arterial system who had PPHA associated with both cerebral aneurysm and cervical internal carotid artery (ICA) stenosis on the same side.

Case Report

A 71-year-old female had complained of severe vertigo lasting for a few minutes once or twice a day for 2 months before admission. Magnetic resonance angiography indicated a cerebral aneurysm and ICA stenosis, so she was admitted for further evaluation. She had received medication for hypertension and asthma for one year. Her family history was unremarkable.

Neurological and physical examination on admission showed no abnormalities, except vascular bruit at the lateral neck on the left side. Left carotid angiography revealed severe stenosis (81.4%) of the left ICA and an anomalous vessel arising from the cervical ICA at C1–2 levels and anastomosed to the vertebral artery (Fig. 1 left). In addition, there was a large ICA aneurysm on the left side (Fig. 1 right). Right carotid angiography was normal. Vertebral angiography showed the right vertebral artery was hypoplastic and provided blood flow to the posterior inferior cerebellar artery without connection to the basilar artery. The left vertebral artery was not detected. The basilar artery was opacified via the PPHA. The bilateral posterior communicating arte-
ries were not visualized by either carotid or vertebral angiography. A three-dimensional computed tomography (3D-CT) clarified that this anomalous artery passed through the hypoglossal canal (Fig. 2). Based on these findings, the anomalous artery classified as PPHA.

Clipping of the aneurysm was first performed by a pterional approach because the risk of rupture of the aneurysm was not negligible during carotid endarterectomy (CEA). Intraoperatively, the anterior choroidal artery was seen, but the posterior communicating artery could not be identified. The aneurysm was successfully clipped and the anterior choroidal artery was preserved. One month later, CEA for the left ICA stenosis was performed using our T-shaped shunt system, under auditory brainstem response (ABR) and somatosensory evoked potential (SEP) monitoring. The distal end of the shunt was inserted to just proximal to the bifurcation of the ICA and PPHA to secure the blood flow in the ICA and vertebral artery. The position of the distal end of the shunt was checked by intraoperative angiography. The stenotic lesion did not involve the PPHA (Fig. 3). CEA was performed without significant changes in the ABR or SEP.

The stump pressure of the ICA, measured under cross-clamping of the common carotid artery and the PPHA, was 20 mmHg. The stump pressure of the PPHA, measured under cross-clamping of the common carotid artery and the ICA, was 25 mmHg. Electromagnetic flowmetry showed blood flow in the ICA at proximal to the second bifurcation was 70 ml/min before and 240 ml/min after CEA. Her postoperative course was uneventful and the vertebrobasilar ischemic attacks did not recur. Postoperative left carotid angiography demonstrated complete obliteration of the aneurysm and disappearance of the stenotic ICA lesion (Fig. 4).

Discussion

The criteria for diagnosis of PPHA are that the artery arises from the cervical ICA at the levels of C-1 to C-3, the artery passes through the hypoglossal canal to the posterior cranial fossa, the basilar artery is filled only by the distal part of the junction with the anastomosis, and angiography indicates deficiency or absence of the posterior communicating artery. Our case satisfied these criteria. In ad-
condition, 3D-CT demonstrated that the artery passed through the hypoglossal canal which confirmed the diagnosis.

Patients with PPHA may have various associated lesions. Like other persistent primitive arteries, the main coexisting lesions are cerebral aneurysm and occlusive cerebrovascular disease. The clinical features of 134 patients with PPHA associated with intra- or extracranial lesions showed that cerebral aneurysms were the most frequent association (26.9%), followed by occlusive cerebrovascular disease (20.9%), brain tumors (9.7%), and arteriovenous malformation (3.0%). These high incidences may be explained by the contribution of congenital factors. However, the coexistence of both cerebral aneurysm and ipsilateral ICA stenosis with PPHA is extremely rare. The clinical manifestations of patients with PPHA can be classified into three types: No symptoms and incidental detection by angiography; symptoms caused by associated lesions such as cerebral aneurysms, cerebral arteriovenous malformation, or brain tumor; and symptoms caused by ischemia in the posterior circulation.

Circulatory effects of the PPHA: A higher incidence of posterior fossa ischemic attacks may be associated with persistent carotid-basilar anastomoses than previously thought. The vertebral arteries in patients with persistent carotid-basilar anastomosis tend to be hypoplastic or aplastic as in our case. The posterior circulation is maintained mainly by the persistent carotid-basilar anastomosis. Under such specific circulatory conditions, ICA stenosis may easily cause ischemia in the vertebrobasilar system. In our patient, low flow (70 ml/min) in the ICA proximal to the second bifurcation and low stump pressure of the PPHA (25 mmHg) strongly indicated low perfusion in the posterior circulation, which could be a cause of vertigo, though thromboembolism due to migration of thrombus at the additional bifurcation or thrombus at the atheromatous plaque through the PPHA could not be denied.

CEA in patients with PPHA: Five prior patients including ours with PPHA associated with the ICA stenosis presented with ischemic attacks of the vertebrobasilar arterial system. Therefore, CEA for prevention of further ischemic attacks in the posterior circulation is a reasonable treatment. In three of the five cases, the stenotic lesions were limited to the ICA, so CEA was performed only on the ICA. In the other two patients, CEA was performed on both the PPHA and the ICA because stenotic lesions involved the PPHA as well. Cross-clamping of the ICA can cause ischemia in both the distal area to the ICA and in the basilar artery territory, so we used the T-shaped shunt system which can shorten the occlusion time of the PPHA and the ICA. In addition, intraoperative angiography could be easily performed through the side arm of the shunt. If the stenotic lesion extends to the PPHA, a special shunt system, the distal end...
of which is divided into two tubes, would be beneficial for CEA of both the PPHA and the ICA. Intraoperative monitoring by ABR and SEP is helpful for assessing the circulatory condition during CEA.

**Order of the operations:** The order in which the procedures were performed is an important point. The ICA stenosis was apparently the cause of ischemia in the vertebrobasilar system, so CEA was planned at first, but the risk of rupture of the aneurysm during CEA was not negligible. Therefore, the aneurysm was clipped before CEA. Intraoperative flow measurement demonstrated that the ICA flow remarkably increased to 240 ml/min after CEA. This great increase in ICA flow could have reinforced hemodynamic stress on the aneurysm, and resulting in possible rupture if CEA had been performed first.

**References**


Address reprint requests to: T. Hatayama, M.D., Department of Neurosurgery, Chugoku Rousai Hospital, 1–5–1 Hirotagaya, Kure, Hiroshima 737–0193, Japan.