Progressive Vascular Changes in Moyamoya Syndrome

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SUMMARY Progressive vascular changes are described in an adult with moyamoya syndrome and similar cases are reviewed. Angiographic findings support the concept of a progressive acquired stenosis of the internal carotid artery as the primary abnormality in this syndrome.

DEMONSTRATION of early stenosis of the internal carotid bifurcation with subsequent development of the angiographic findings characteristic of the syndrome of multiple progressive intracranial arterial occlusions, or moyamoya syndrome, has been infrequently reported. A patient is described here in whom such a sequence was observed with a review of previously reported patients with similar progressive changes.

Patient Report

A 29-year-old woman was admitted to the University of Iowa Hospital in September 1975, one day after sudden onset of severe headache and confusion. She had had recurrent generalized seizures from age 13 to 21 and frequent headaches preceded by visual blurring for the past eight years. Arterial hypertension was found after her only pregnancy at age 23.

On examination she was drowsy but easily arousable. Her blood pressure was 155/100 and pulse 68/min. Cardiovascular examination was normal. There were no cervical or cranial bruits. Funduscopes, visual fields and extra-ocular movements were normal. Her neck was stiff. There was a mild left hemiparesis with left-sided hyperreflexia and bilateral withdrawal on plantar stimulation.

The cerebrospinal fluid was bloody with xanthochromic supernatant. It contained 140,000 red blood cells/mm³, 700 white blood cells/mm³, 55mg% of glucose and 250 mg% of protein.

Cerebral angiography demonstrated stenosis of the right internal carotid artery bifurcation and the initial segment of the right anterior cerebral artery. Proximally the horizontal segment of the right middle cerebral artery was not visualized, but further distally its trunk and bifurcation were opacified, the contrast reached it through prominent lenticulostriate vessels (fig. 1). The left carotid and right vertebral angiograms were normal. Distal right middle cerebral artery branches filled through leptomeningeal anastomosis from the right posterior cerebral artery. No aneurysms were visualized.

The patient was treated by dexamethasone, epsilon aminocaproic acid (EACA) intravenously, alpha-methyl-dopa, hydrochlorothiazide and phenytoin.

On the ninth hospital day she became less respon-
artery was supplied (fig. 2). Irregular stenosis of the bifurcation of the left internal carotid artery was now present. Both anterior cerebrials and the left middle cerebral artery filled from the left carotid. Transdural anastomosis was evident on the right.

A right superficial temporal-middle cerebral artery anastomosis was performed to improve right hemisphere perfusion. The patient has remained asymptomatic following the bypass.

Discussion

Including this report, progressive vascular changes have been demonstrated by angiography in 15 patients with the moyamoya syndrome (table). The interval between the angiograms demonstrating the changes ranged from 7 to 84 months with a mean of 27 months. As in the present patient, stenosis of the supraclinoid internal carotid artery and proximal segments of the middle and anterior cerebral arteries were the earliest demonstrable abnormalities, with development of prominent lenticulostriate vessels and leptomeningeal and transdural anastomosis as the ICA stenosis became more pronounced or occlusion occurred. In this patient and the children reported by Handa and Handa, a progressive decrease in the diameter of the cervical carotid artery was also noted in the follow up angiograms.

Although the number of patients with documented progression is small when compared to the total number of reported patients with moyamoya syndrome, the finding of progressive changes in 53% (15/28), where angiography was repeated several months to years after the initial vascular event, suggests that this is a much more common feature than previously considered.

The angiographic changes described favor the concept of a progressive acquired occlusion of the supraclinoid internal carotid artery as the pathogenic mechanism in this syndrome. Because major tributaries of the circle of Willis are involved early in the disease, perfusion of the brain is maintained through the development of collateral channels represented by the lenticulostriate vessels and leptomeningeal and transdural anastomosis. The progressive narrowing of the internal carotid artery proximal to the occlusion and in its cervical portion observed by Handa and Handa and in this patient, is probably the result of decreased blood flow through the artery rather than true "hypoplasia" as previously suggested by several authors.

<table>
<thead>
<tr>
<th>Author</th>
<th>Age and Sex</th>
<th>Interval between Angiograms (months)</th>
<th>Clinical Course</th>
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<tbody>
<tr>
<td>Adams et al.</td>
<td>35 F</td>
<td>12</td>
<td>Recurrent alternating hemiparesis and vertigo</td>
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<tr>
<td>Handa &amp; Handa</td>
<td>8 F, 3 M</td>
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<td></td>
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<td>Iivanainen et al.</td>
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<tr>
<td>Kudo</td>
<td>4 M</td>
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<td>Nishimoto &amp; Takeuchi</td>
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<td></td>
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<td>52 F</td>
<td>23</td>
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<tr>
<td>Present author</td>
<td>29 F</td>
<td>26</td>
<td>Asymptomatic</td>
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References


Primitive Hypoglossal Artery and Carotid Endarterectomy

JOE A. PINKERTON, JR., M.D., KENDRICK C. DAVIDSON, M.D., AND BLAINE Z. HIBBARD, M.D.

SUMMARY A patient with transient ischemic symptoms in the carotid and vertebrobasilar distribution is reported. His arteriogram demonstrated a persistent primitive hypoglossal artery which in part may explain the clinical picture. The embryology, radiology, clinical manifestations, and surgical considerations of this rare anomaly are discussed.

An arch aortogram with bilateral selective biplane carotid evaluation was performed (fig. 1). A persistent hypoglossal artery was found arising from the left internal carotid artery and providing blood flow to the basilar artery and all its major branches except the posterior inferior cerebellar artery (figs. 2, 3). An atherosclerotic plaque with severe stenosis and ulceration was demonstrated just above the bulb of the left internal carotid artery and proximal to the origin of the hypoglossal artery. The right carotid arterial system was normal and both vertebral arteries, while hypoplastic, provided flow to the posterior inferior cerebellar arteries with no connection to the basilar artery.

The patient had a left carotid endarterectomy using continuous electroencephalographic monitoring. The left internal carotid mean arterial stump pressure at operation was 25 mm Hg and a temporary inlying shunt was used during carotid occlusion. The patient had no complications and was discharged on the third postoperative day.

Embryology

The embryologic development of the carotid and vertebrobasilar systems has been described by Padget based on his extensive dissections of the 4-5 mm human embryo. In the human embryo the carotid arteries arise from the third paired aortic arches. The
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