Ischemic chiasmal syndrome and hypopituitarism associated with progressive cerebrovascular occlusive disease.

J Ahmadi, J R Keane, G S McCormick, R J Fallis and C A Miller

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Ischemic Chiasmal Syndrome and Hypopituitarism Associated with Progressive Cerebrovascular Occlusive Disease

Two interesting patients are described with unique clinical and radiographic manifestations of progressive bilateral stenosis-occlusion of the distal internal carotid arteries. Their clinical presentations, ischemic chiasmal syndrome and hypopituitarism, are extremely rare, despite extensive occlusion of the circle of Willis. In one of these patients, the configuration of collateral vessels around the optic chiasm on computed tomography (CT) was so rounded and discrete that it actually had an appearance suggesting a pituitary tumor or an aneurysm. In the second patient, a pseudoaneurysm and a very large intracerebral hematoma with a ring of contrast enhancement were the radiographic manifestations.

Progressive stenosis-occlusion of the distal internal carotid artery is an uncommon variety of cerebrovascular occlusive disorder. When it is associated with an extensive telangiectasia in the region of the basal ganglia, it is often termed moyamoya [1-5]. The occlusive process is usually bilateral and affects primarily the supraclinoid internal carotid artery and its distal bifurcation. Occasionally, the distal end of the basilar artery is also involved. There is a striking female preponderance and predilection for the young [4-7]. The initial clinical symptoms in children are mainly secondary to cerebral ischemia, and predominantly manifest as recurrent transient neurologic deficits. In contrast, intracranial hemorrhage is the most common initial presentation in adults [8]. Ischemic chiasmal syndrome and hypopituitarism are extremely rare, despite extensive occlusion of the circle of Willis. Two interesting patients with unique clinical and radiographic manifestations are the subject of this report.

Case Reports

Case 1

A 40-year-old woman was seen after 2 years of frequent generalized nonthrobrring headaches and 7 months of poor vision in the right eye. She had experienced polyuria for 1 month, cold intolerance for 2 years, and amenorrhea for 20 years. She did not have hoarseness, hair loss, skin changes, weight gain, constipation, memory disturbance, or a change in hand or foot size.

On examination she was alert, cooperative, and obese. Her vital signs and general examination were normal, as were her memory and speech. Visual acuity was 20/40 in the right eye and 20/16 in the left. She missed all of the Hardy-Rand-Rittler color plates with the right eye and nine of 20 with the left. A right afferent pupilary defect was present. Her visual fields showed a complete bitemporal hemianopia, with marked nasal constriction in the right eye and moderate in the left. Fundoscopy revealed bilateral optic disk pallor and diffuse retinal nerve fiber attenuation, more prominent in the right eye. Extraocular movements and efferent pupillary function were intact. The rest of the neurologic examination was normal.
Fig. 1.—Case 1. Collateral vessels around optic chiasm and pituitary stalk. A and B. Postcontrast CT scans. Discrete enhancing lesion in anteromedial part of pituitary fossa and suprasellar cisterns (arrows). C. Metrizamide cisternogram shows corresponding filling defect in suprasellar cisterns. Bilateral carotid occlusion in left frontal (D) and lateral (E) carotid arteriograms. Stenosis of juxtasellar and occlusion of supraclinoid parts of internal carotid, proximal anterior, and middle cerebral arteries. Branches of anterior and middle cerebral arteries are filled via collateral circulation. Some collateral vascular channels are in sellar and suprasellar regions (arrow). F. Right internal carotid artery is completely occluded and thrombosed down to near its origin (arrow).

Her erythrocyte sedimentation rate was 46 mm/hr. The antinuclear antigen determination was normal. Fasting cholesterol and triglyceride levels were 237 mg/dl and 80 mg/dl, respectively. The serum T4 was 2.2 μg/ml (normal, 4.6–11.0 μg/ml); T3 was 0.81 μg/ml (normal, 0.1–1.0 μg/ml); and thyroid-stimulating hormone (TSH) was 1.8 IU (normal, 0.5–5 IU). The follicle-stimulating hormone (FSH) (normal, 20 mIU/ml) was less than 1.51 mIU/ml. An 8 a.m. cortisol level was 1.0 μg/dl (normal, 15–25 μg/dl). The serum prolactin was 15.2 μg/ml (normal, 9–20 μg/ml), and the random serum growth hormone was 0.6 μg/ml (normal, 0.3–7.5 μg/ml).

Plain skull films and polytomograms showed a normal sella turcica. Precontrast computed tomography (CT) revealed a slightly hyperdense (40 H) abnormality in the anteromedial part of the suprasellar cistern and pituitary fossa. This lesion enhanced (67 H) after intravenous injection of contrast medium (41 g I). CT metrizamide cisternography demonstrated a corresponding filling defect in the suprasellar cisterns (figs. 1A–1C). Bilateral carotid and vertebral angiography showed narrowing of the juxtasellar and occlusion of the supraclinoid parts of the left internal carotid artery, involving the proximal middle and anterior cerebral arteries. There were some collateral vascular channels in the sellar and suprasellar regions (figs. 1D–1F). The right internal carotid artery was completely occluded. There were leptomeningeal collaterals and retrograde blood flow from the vertebrobasilar circulation into branches of the middle and anterior cerebral arteries.

Repeated examinations over the subsequent 4 years demonstrated fluctuating visual function, with overall mild improvement in acuity, color plate visualization, and visual fields. Repeated contrast CT showed the lesion to have remained essentially unchanged. The patient was given thyroid and corticosteroid replacement therapy.

Four years after her initial presentation, thyrotropin- and gonadotropin-releasing hormone stimulation was performed. TSH remained less than 1.0 IU/ml at 0, 20, 30, 60, and 90 min. Similarly, the FSH did not rise above 1.5 mIU/ml. She remained amenorrheic.

Case 2

A 34-year-old man was admitted with the diagnosis of terminal multiple sclerosis. He was the product of a difficult delivery and was mentally retarded.

At age 29, he developed a rigid neck, unsteady gait, drooling, and a change in speech. He recovered partly, and later the same year
Fig. 2.—Case 2. A, Postcontrast CT scan shows very large hematoma in right cerebral hemisphere with peripheral rim of contrast enhancement, indicating that at least intracerebral hematoma is several weeks old. Lateral (B and C) and slightly oblique frontal (D and E) angiograms. Aneurysm (arrows) arising from inferior frontal branch, which apparently originates near anterior communicating artery and was filled during both right and left carotid injections. Right middle cerebral artery was completely occluded, with stenotic lesions in distal internal carotid artery and in both anterior and left middle cerebral arteries.

was admitted to another hospital, with left hemiparesis and right gaze preference. His vision was poor, and he was able to recognize coins with each eye only at distances of 8 cm or less. A nuclear brain scan and echoencephalogram were normal. Diffuse slowing was seen on electroencephalography. Cerebrospinal fluid examination was normal, aside from findings of 9 monocytes/3 ml and slightly increased immunoglobulins of 13 mg/dl. A diagnosis of multiple sclerosis was made; steroid treatment was begun, and the patient improved.

At age 32, similar symptoms reappeared and a course of steroid therapy provided only minimal benefit. He remained dysarthric, unable to walk without assistance, and unable to swallow solid foods. At age 34, he became mute and barely able to move. He was admitted to the LAC-USC Medical Center unresponsive but with normal vital signs. Bilateral optic atrophy and severe spastic quadriaparesis were present, more pronounced on the left side.

CT of the head revealed a very large hematoma occupying most of the right frontal and parietal lobes. Its anterior part was hyperdense, surrounded by a peripheral isodense rim that enhanced after intravenous injection of contrast medium. There was marked compression of the right lateral ventricle, with displacement of the midline structures to the left, and hydrocephalus due to compression of the foramen of Monro and transfalcial and uncal herniation (fig. 2A). A cerebral angiogram revealed occlusion of the right middle cerebral artery, with stenotic lesions in the right anterior cerebral artery and in the left middle and anterior cerebral arteries. Collateral blood flow to the anterior circulation was provided by leptomeningeal and periphthalmic anastomoses and channels from the posterior circulation. There was an aneurysm on an inferior frontal branch arising in the vicinity of the anterior communicating artery (figs. 2B–2E).

The patient died 8 months later. A postmortem angiogram was obtained that demonstrated a fine vascular rete arising from the right internal carotid artery. However, the postmortem angiogram did not reveal an aneurysm.

At autopsy, the vessels at the base of the brain showed striking diffuse circumferential thickening of the walls of the middle and anterior cerebral arteries and the supraclinoid part of the internal
carotid artery. There were no aneurysms, vascular malformations, or artheromatous plaques. The posterior circulation, including the basilar and vertebral arteries, showed normal vascular structures. The cerebral hemispheres were asymmetric, with marked enlargement and subfalcine herniation of the right frontal lobe. There was also a mild degree of transtentorial herniation of the uncus bilaterally.

Gross examination of the brain after fixation with 10% phosphate-buffered formalin revealed widespread hemorrhage within the lateral ventricles, extending from the white matter of the right frontal lobe. There was also subarachnoid extension of the hemorrhage into the posterior fossa and overlying the cerebellar hemispheres. A 5-mm-diameter circumscribed area of old cystic infarction was present within the left basal ganglia.

Microscopically, cross sections of the vessels revealed marked diffuse fibrosis and thickening of the intima. The internal elastic membrane was fragmented in some regions, as revealed in aldehyde fuchsin-stained sections (figs. 3A and 3B). The media showed moderate proliferation of the connective tissue component. There were no inflammatory infiltrates, amyloid, fibrin, or lipid degeneration or cholesterol deposits within the vessel walls. Several smaller vessels also showed luminal occlusion and recanalization by a fine meshwork of endothelial cells. The vascular lesions did not appear to be the result of an acute or chronic inflammatory process. There was pallor and some demyelination of the optic chiasm (figs. 3C and 3D). Post-mortem examination of the brain showed no evidence of multiple sclerosis.

Discussion

Moyamoya disease is a rare and unique cerebrovascular occlusive disorder of unknown etiology with three distinctive angiographic features [1-13]:

1. Bilateral stenotic and occlusive changes affecting the distal internal carotid bifurcation and proximal parts of the main trunk of the middle and anterior cerebral arteries. Branches of the middle and anterior cerebral arteries distal to the stenotic segments remain normal (therefore being available for possible bypass surgery).

2. Leptomeningeal and transdural anastomoses ("rete mirabile") derived from the middle meningeal and superficial temporal arteries. With involvement of the ophthalmic arteries, cerebral perfusion becomes totally dependent on collaterals from the external carotid and vertebral arteries.

3. Development of an extensive parenchymal collateral network in the region of the basal ganglia. The angiographic appearance of this telangiectasis has been likened to a puff of smoke ("moyamoya" in Japanese). The telangiectatic channels that serve as collateral routes (supplied by the striate vessels and choroidal arteries) increase with the evolution of the stenotic process and are more pronounced when the occlusive process begins earlier in childhood and progresses slowly. However, further progression of the occlusive process may ultimately compromise the origin of the striate arteries and decrease the collateral network.

The optic chiasm and pituitary gland are uniquely situated to receive blood supply from multiple arteries of the circle of Willis [14-21]. Branches arising from the internal carotid and anterior cerebral arteries supply each side of the lateral chiasm. The inferomedial chiasm is perfused predominantly by the right and left superior hypophyseal arteries that arise from the medial aspect of the internal carotid at the level of the origin of the ophthalmic artery. The posteroinferior chiasm is mainly supplied by perforating twigs arising from the posterior communicating arteries. The superior surface of the chiasm receives multiple fine branches from the distal internal carotid, anterior cerebral, and anterior communicating arteries. The superior hypophyseal artery as well as the anastomotic plexus surrounding the infundibular stalk nourishes the pituitary gland. This perinfundibular anastomosis is supplied by the ipsilateral and contralateral connecting branches of the internal carotid, anterior cerebral, posterior communicating, and superior hypophyseal arteries. There is generally a common blood supply to the pituitary gland and optic chiasm.

Compression is the usual cause of the chiasmal syndrome. Tumors are the most common source of pressure on the chiasm [22-25], but aneurysms [26, 27], ectatic carotid or anterior cerebral arteries [28-30], arachnoiditis [31-35], a dilated third ventricle [36], and a transtentorial herniation are other unusual etiologies. Rare, noncompressive causes of bitemporal hemianopia include trauma, multiple sclerosis, and ischemia [34]. Ischemic damage to the anterior visual pathways occurs predominantly at two sites: (1) the inner retina, in association with central retinal artery occlusion, and (2) the retrolaminar optic nerve, with multiple posterior ciliary artery occlusions. Chiasmal ischemia in the absence of associated

Fig. 3.—Case 2. A, Cross section through carotid artery reveals markedly thickened intima and fragmented internal elastic membranes (arrows) (×45). B, At high magnification, smaller artery adjacent to internal carotid artery with proliferation of intima and recanalization of occluded lumen (×45). C, Section of optic chiasm contains fibers with intact myelin. D, Midportion of chiasm adjacent to perforating vessels shows focal patches of myelin loss (Luxol fast blue ×10).
lesions (such as tumors, ectatic vessels, meningitis, or arachnoiditis) is exceedingly rare. The few well documented cases have been described in association with temporal arteritis [37, 38].

Intracranial carotid occlusion is a rare but recognized cause of hypopituitarism [39, 40]. Ischemic infarction of the pituitary would require occlusion of multiple branches of the arteries supplying the pituitary gland. Pituitary hypofunction may be explained by thrombosis of these small branches, such as those demonstrated histologically in our case 2.

Few vascular diseases produce such extensive disease of the circle of Willis without cortical infarction. In our patients, collateral vessels from the external carotid artery and the posterior circulation must have provided adequate cerebral perfusion. Such widespread collateralization is often found in patients with moyamoya disease.

CT findings in moyamoya disease have been described [41–45]. Significant abnormalities noted were multiple low-density lesions simulating those of multiple infarcts, ventricular dilatation, and cerebral atrophy. On occasion, the abnormal avascular network in the region of the suprasellar cisterns and the basal ganglia has been demonstrated by higher doses of intravenous contrast medium. McCormick and Schochet [46] demonstrated extensive collateral vascular channels around the optic chiasm on postmortem examination in a patient with moyamoya disease (fig. 4). Abnormal enhancement in the sella and suprasellar cisterns in our case 1 represents similar extensive collateralization. This case is of further interest in that the configuration of the enhancing lesion was so rounded and discrete that it actually had an appearance suggesting a pituitary adenoma or aneurysm (or other mass lesion).

In case 2 an aneurysm of a distal part of the frontopolar artery was demonstrated angiographically. The peripheral location of this aneurysm and the failure to find an aneurysm postmortem either angiographically or at autopsy suggested that a pseudoaneurysm may have been present. True aneurysms as well as pseudoaneurysms have been reported occasionally in patients with moyamoya disease [47–59]. Pseudoaneurysm is thought to have developed at the site of hemorrhage from a small artery. In our case 2, presumably the blood had penetrated the adjacent encephalomalacic ischemic brain tissue and produced a huge hematoma. A ring of contrast enhancement seen on the CT scan indicates that the intracerebral hematoma was in a resolving state [60, 61]. Although a large space-occupying lesion with resultant herniation could have resulted in chiasmal compression and ischemia, the visual deficit in our patient had long preceded the intracerebral hematoma.

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