Intracranial Dissection and Extracranial Hypoplasia of the Internal Carotid Artery

Fabrizio Sallustio, MD, Silvia Di Legge, MD, PhD, Giacomo Koch, MD, PhD, Paolo Stanzione, MD

Dissection of the intracranial portion of the internal carotid artery (ICA) is an uncommon cause of stroke or transient ischemic attack (TIA). Congenital anomalies of the ICA have been rarely associated with cerebral ischemia. However, unilateral ICA hypoplasia can be frequently associated with intracranial arterial anomalies and altered hemodynamics.1 We describe the case of a woman who had sudden onset of right facial pain and transient left hemiparesis. Diffusion-weighted magnetic resonance imaging (MRI) findings were negative for acute ischemia. Magnetic resonance angiography (MRA) and sonography showed hypoplasia of the right extracranial ICA and vertebral artery (VA) and dissection of the intracranial portion of the right ICA and right middle cerebral artery (MCA), with inverted flow of the right anterior cerebral artery (ACA). At 12-month imaging studies, the intracranial hemodynamics appeared normalized, whereas the extracranial findings were unchanged. This case shows an association between hypoplasia, dissection of the ICA, and cerebral ischemia.

Case Report

A 33-year-old right-handed woman was referred to us because of sudden onset of sharp retro-orbital pain and sensorimotor impairment involving the left arm and leg. Symptoms vanished in about 30 minutes, leaving the patient with fatigue and mental slowness. In the previous 4 days, she had been complaining of a right parietal headache. The patient had no classic vascular risk factors, recent infections, or history of drug abuse. Her medical history was unremarkable for migraines, head or neck trauma, heart disease, stroke, or TIA. On admission, the neurologic examination was normal. Sonography was performed using an HD11 system (Philips Medical Systems, Bothell, WA) and a 5- to 7.5-MHz linear array.
probe for extracranial examinations and a 2- to 4-MHz probe for transcranial examinations. Extracranial color-coded sonography showed a diffuse reduction of the vessel diameters of the ICA and VA on the right side compared with the contralateral side, not accompanied by atherosclerotic changes and associated with low flow velocities and increased resistive indices. Transcranial color-coded sonography showed an aliasing effect and high peak systolic velocities (PSVs) in the intracranial portion of the ICA (240 cm/s), the M1 segment of the MCA (252 cm/s), and the posterior communicating artery (217 cm/s) and reversed flow in the ACA. These findings suggested intracranial hemodynamic changes secondary to intracranial dissection of the right ICA with multiple intracranial artery involvement.

The patient underwent 3-T brain MRI and MRA (Achieva 3.0T; Philips Medical Systems). Diffusion-weighted MRI findings were negative for acute infarction. Three-dimensional time-of-flight MRA of the intracranial vessels showed a smaller diameter of both the right ICA up to the siphon and the VA compared with the contralateral side; flame-shaped narrowing of the terminal intracranial portion of the right ICA also involving the M1 segment of the right MCA and the A1 segment of the right ACA was observed. No mural hematoma on axial T1-weighted MRI, malformations, aneurysms of the circle of Willis, abnormal intracranial collateral vessels, or beaded or atherosclerotic changes were detected. Figures 1 and 2 summarize all of the above-described findings. A brain computed tomographic perfusion study showed normal cerebral blood flow and volumes, with no asymmetries between the two hemispheres. Axial T1-weighted MRI showed that the right carotid canal was smaller than the left (Figure 3).

Findings from transesophageal echocardiography, 24-hour electrocardiographic monitoring, and screening for hyperhomocysteinemia and prothrombotic conditions were normal. A thorough workup did not show evidence of vascular abnormalities in other areas. We could not rule out collagenopathies or fibromuscular dysplasia because the patient refused to undergo further screening. She had no recurrent events during admission and began taking antiplatelets (aspirin, 325 mg/d).

In the following month, the patient remained asymptomatic, and the neurologic examination was normal. Sonography showed unmodified extracranial vessel diameters with a substantial reduction in flow velocities in the intracranial vessels on transcranial color-coded imaging, although the PSV was still above the normal limit, and the flow in the ACA was antegrade, suggesting a hemodynamic adjustment. Follow-up MRA, MRI, and extracranial and transcranial color-coded sonographic studies were repeated 6 and 12 months after the initial study. Intracranial MRA showed an upgraded luminal caliper with no filling defects. Sonography confirmed the presence of smaller diameters of the right ICA and VA and normal flow velocities in the previously stenosed intracranial vessels, with antegrade flow in the right ACA (Figures 4 and 5).

Discussion

In the case described, both the clinical presentation and combined MRA and sonographic findings suggested intracranial ICA dissection involving the MCA and ACA. Follow-up MRA and sonographic findings, coupled with MRI evidence of carotid canal hypoplasia, ruled out extracranial ICA dissection and allowed diagno-
sis of a congenital arterial anomaly. To the best of our knowledge, such an association has not been reported previously. Up to now, there have been no established criteria for diagnosis of carotid hypoplasia because of the small number of reported cases. Magnetic resonance imaging findings of diffuse narrowing of the ICA might suggest an acquired etiology, such as dissection or atherosclerosis. However, in all of the cases investigated by color duplex sonography (CDS), a uniform narrowing of the luminal and external vessel diameters not associated with wall thickening and flow disturbances should suggest ICA hypoplasia. Although the combined use of CDS and MRA allows a diagnosis of ICA hypoplasia, diagnostic confirmation should be obtained by depiction of a small carotid canal on computed tomography or MRI of the skull base.

Spontaneous intracranial ICA dissection, in particular in the terminal segment of the artery, is a rare cause of stroke or TIA. Dissection more often occurs at the level of the carotid knee just beyond the ophthalmic artery and also involves the ipsilateral MCA and ACA. Flame-shaped narrowing of the arterial lumen is a frequent angiographic appearance of dissection. High diagnostic accuracy in detecting dissection can be achieved by duplex sonography, which allows noninvasive evaluation of tissue, flow, and hemodynamics of extracranial and intracranial areas. A recent study by Benninger et al., examining the accuracy of CDS in detecting ICA dissection, suggests that normal sonographic findings in the cervical ICA allow reliable exclusion of ICA dissection in patients with a first ischemic event in the territory of the ICA because of their high sensitivity and negative predictive values. However, due to false-positive findings on CDS, a diagnosis of ICA dissection may require MRI and MRA confirmation. In particular, a hyperintense signal on T1-weighted MRI is considered highly suggestive of intramural hematoma in the dissected vessel, although this feature may be absent in more than 50% of patients. In our patient, intracranial ICA dissection was suggested by the absence of atherosclerotic changes throughout the extracranial and intracranial vessels, increased PSVs in the intracranial portion of the ICA, M1 segment of the MCA, and posterior communicating artery, and reversed flow in the ACA and supported by the progressive normalization of these hemodynamic parameters and MRA findings on follow-up studies.

Hypoplasia of the ICA is a rare condition of uncertain etiology, and its frequent association with anomalies in the circle of Willis suggests a congenital origin. Although our report does not provide any proof of a causal relationship, alterations in the hypoplasic arterial wall could be a plausible explanation for the association with dissection and cerebrovascular events, although the exact mechanism of the latter remains unclear. In this regard, a recent review...
of published series and a personal series of patients with aplasia or hypoplasia of the ICA showed a high prevalence of intracranial saccular aneurysms, particularly among patients older than 30 years, suggesting an acquired condition over a developmental one.1 This study might suggest optimization of stroke prevention through careful control of classic vascular risk factors in patients with arterial hypoplasia, a condition that seems to be not as benign as previously considered.

Our patient also had possible hypoplasia of the right VA. Congenital variations and asymmetry of the VAs are not infrequent (the left one being the dominant artery in 50% of the population), and they are often regarded as normal variants. However, VA hypoplasia has been recently recognized as a possible predisposing factor for posterior circulation strokes, presumably of embolic origin in half of the cases.11 We propose that headaches and neurologic symptoms in a patient with arterial hypoplasia should initiate a thorough investigation for arterial dissection and other intracranial abnormalities.

References