Spontaneous Intracranial Internal Carotid Artery Dissection

Report of 10 Patients

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Background: Spontaneous intracranial internal carotid artery (ICA) dissection is an uncommon cause of cerebral infarction, particularly when compared with the dissection of the ICA’s cervical portion. Most reports describe extensive strokes with very high mortality rates.

Objective: To report the clinical and radiological findings of 10 patients with spontaneous intracranial ICA dissection.

Methods: Ten patients (5 women) were included with ages ranging from 15 to 59 years (mean age, 28 years).

Results: Nine patients had a stroke (1 had an associated subarachnoid hemorrhage), whereas 1 patient had only transient ischemic attacks. Severe retro-orbital or temporal headache followed by contralateral hemiparesis was the most common initial clinical symptom. No patient had vascular risk factors or a history of neck or head trauma. Stenosis of the supraclinoid portion of the ICA occurred in 8 patients, with extension to the middle cerebral artery or anterior cerebral artery in 2 patients each. Aneurysm formation in the ipsilateral anterior cerebral artery was seen in 1 patient. Two patients had a total occlusion of the supraclinoid portion of the ICA. All patients did well, with no (n=3), mild (n=4), or moderate (n=3) disability on the Modified Rankin Scale during a 3-month follow-up period.

Conclusions: Spontaneous intracranial ICA dissection can cause ischemic stroke with or without subarachnoid hemorrhage and should be considered in the differential diagnosis of intracranial ICA stenosis or occlusion, especially in young patients. Some patients survive with few or moderate deficits.

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A RTERIAL DISSECTION is now recognized as an important cause of stroke. Most reported dissections involve the extracranial carotid and vertebral arteries. Intracranial arterial dissections are less common, most reported intracranial dissections involve the vertebral and basilar arteries. Vertebrobasilar dissections are known to cause brain infarction, often fatal or disabling, and subarachnoid hemorrhage. Dissecting posterior circulation aneurysms can also cause mass effect on the brainstem and cranial nerves. Intracranial internal carotid artery (ICA) dissections are considered rare and have been reported infrequently. The risk factors, clinical findings, imaging abnormalities, and prognosis in patients with intracranial ICA dissections previously have not been described in detail. We report our experience with 10 patients who had nontraumatic intracranial ICA dissections.

RESULTS

Our study included 10 patients, 5 women and 5 men, with ages ranging from 15 to 59 years (mean age, 28 years). None of the patients had known vascular risk factors or a history of head or neck trauma. However, 3 patients had a history of migraine with aura, 2 patients were in the postpartum period, and 1 patient had fibromuscular dysplasia of the extracranial ICAs that appeared on conventional angiography (Table).

Nine patients experienced a stroke, and 1 patient also had an associated subarachnoid hemorrhage. One patient had only transient ischemic attacks: 2 within a 3-day period (patient 4 in the Table). Severe headache, usually retro-orbital, frontal, and/or temporal, followed by contralateral hemiparesis were the most common initial clinical symptoms (80% of the patients). Neurological signs occurred in most patients (90%) immediately after the headache on-
PATIENTS AND METHODS

Personal files and the New England Medical Center (Boston, Mass) stroke database were searched for patients with the clinical and/or pathologic diagnosis of intracranial ICA dissection. One patient with autopsy-proven intracranial ICA dissection was not included in this series because the case had been previously published.

We reviewed the medical records as well as all of the imaging studies available, including cranial computed tomography and magnetic resonance imaging, magnetic resonance angiography of the head and neck, and conventional cerebral angiography.

Clinical, Radiological, and Outcome Data in 10 Patients With Intracranial Dissection*

<table>
<thead>
<tr>
<th>Patient/Sex/ Age, y</th>
<th>Clinical Symptoms</th>
<th>MRI</th>
<th>MRA/Cerebral Angiography</th>
<th>Clinical Outcome at 3 mo†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/28</td>
<td>Right retro-orbital pain followed by fluctuating left hemiparesis (3 mo post partum)</td>
<td>Right IC infarct</td>
<td>Stenosis of the supraclinoid portion of the right ICA and MCA</td>
<td>2</td>
</tr>
<tr>
<td>2/F/38</td>
<td>Right temporal and retro-orbital headache followed by fluctuating left hemiparesis; history of migraine</td>
<td>Right IC infarct</td>
<td>Stenosis of the supraclinoid portion of the right ICA and MCA</td>
<td>2</td>
</tr>
<tr>
<td>3/F/26</td>
<td>Frontal headache with vomiting followed by decreased level of consciousness and left hemiparesis (3 mo post partum)</td>
<td>Right caudate, right MCA, and left ACA infarcts</td>
<td>Stenosis of the supraclinoid portion of the right ICA and ACA; aneurysm formation in the right A2 segment; hypoplastic left A1 segment</td>
<td>3</td>
</tr>
<tr>
<td>4/F/35</td>
<td>Left temporal headache followed 3-5 d later by 2 spells of right-hand numbness; history of migraine</td>
<td>Normal</td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>0</td>
</tr>
<tr>
<td>5/M/20</td>
<td>Right retro-orbital headache followed by transient left-arm clumsiness and slurred speech; 10 d later, acute onset of slurred speech, left hemiparesis, and neglect</td>
<td>Right LS infarct</td>
<td>Occlusion of the supraclinoid portion of the right ICA; fibromuscular dysplasia in the cervical portion of both ICAs</td>
<td>3</td>
</tr>
<tr>
<td>6/F/35</td>
<td>Left-sided headache followed by transient paresis of the right arm; 6 d later, transient episode of expressive aphasia; 2 wk later, right hemiplegia and dysarthria</td>
<td>Left corona radiata infarct</td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>2</td>
</tr>
<tr>
<td>7/M/22</td>
<td>Right temporal headache followed by acute left hemiplegia</td>
<td>Right caudate and IC infarcts</td>
<td>Stenosis of the supraclinoid portion of the right ICA with thrombosis of the right A1 segment</td>
<td>3</td>
</tr>
<tr>
<td>8/M/59</td>
<td>Left-sided headache followed by expressive aphasia</td>
<td>Left frontal infarct</td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>0</td>
</tr>
<tr>
<td>9/M/15</td>
<td>Right-sided headache followed 3 d later by left hemiplegia</td>
<td>Right LS infarct</td>
<td>Occlusion of the supraclinoid portion of the right ICA</td>
<td>1</td>
</tr>
<tr>
<td>10/M/25</td>
<td>Left occipital/temporal headache followed by transient right-sided weakness; 2 h later, right hemiparesis and aphasia; history of migraine</td>
<td>Left LS infarct</td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>0</td>
</tr>
</tbody>
</table>

*MRI indicates magnetic resonance imaging; MRA, magnetic resonance angiography; IC, internal capsule; ICA, internal carotid artery; MCA, middle cerebral artery; ACA, anterior cerebral artery; and LS, lenticulostriate.
†Modified Rankin Scale score.

Six patients were immediately treated with anticoagulants, and 3 other patients were given antiplatelet agents. No patient had a hemorrhagic complication. The remaining patient with intracranial ICA dissection complicated by subarachnoid hemorrhage was initially treated with steroids because the presumed diagnosis was vasculitis. Three months later, the patient was switched to antiplatelet treatment. Serial neuroimaging studies performed in 9 patients showed partial or complete recanalization in 6 of them.

All patients did well, with no (n=3), mild (n=4), or moderate (n=3) disability on the Modified Rankin Scale during a 3-month follow-up period.

Spontaneous dissecting aneurysms of the intracranial carotid system are uncommon in adults, especially when...
Figure 1. Patient 1: A, T2-weighted magnetic resonance imaging scan shows a right basal ganglia infarct (arrow). B, Magnetic resonance angiography of the head demonstrates a filling defect (arrowhead) in the distal portion of the right internal carotid artery (ICA) and proximal middle cerebral artery (MCA) stem. Cerebral angiography shows the presence of double lumen in the right MCA stem (C, arrow) and a narrowing of the supraclinoid portion of the right ICA (D, arrow).

Figure 2. Patient 3: Left image, Magnetic resonance imaging scan of the head shows both a right middle cerebral artery (left arrow) and left anterior cerebral artery (right arrow) stroke. Cerebral angiography demonstrates an irregular aneurysm in the right A2 segment (middle image, arrow) with proximal narrowing of the right A1 segment and supraclinoid portion of the right ICA (right image, arrow).
Compared with those of the vertebrobasilar system. Fewer than 100 cases have been reported in the English-language literature. Our case material includes 10 patients, most of them younger than 40 years.

Intracranial ICA dissection has been associated with fibromuscular dysplasia, cystic medial necrosis, intimal fibroelastic aberrations, and atherosclerosis. However, no microscopic pathologic changes are usually detected in these patients. Only 1 of our patients was found to have changes suggestive of fibromuscular dysplasia in both extracranial ICAs during conventional angiography. None of them had evidence of hypertension, diabetes, hyperlipidemia, cardiovascular disease, coagulation abnormalities, or systemic disorders or a history of head or neck trauma.

The relationship between migraine and intracranial ICA dissection is unclear. Sinclair reported a fatal case in which a 27-year-old patient with a long-standing history of migraine developed a spontaneous MCA dissection during a migrainous attack. The author posited that local vascular edema during the migraine contributed to the development of the intracranial artery dissection. Although 3 of our patients with intracranial ICA dissection had a long-standing history of migraine, all of them stated that the headaches preceding the onset of their ischemic symptoms were more severe than their usual migraines and involved predominantly the retro-orbital, frontal, and/or temporal regions. These locations are frequent sites of referred pain described during stimulation of the distal portion of the ICA and MCA.

Similar to the cases described in the literature, the neurological signs in most of our patients followed almost immediately after the headache’s onset. This clinical manifestation is different from that usually seen in patients with extracranial ICA dissection, in which initial symptoms may precede the stroke by several days. Also, whereas patients with extracranial ICA dissection may initially experience headaches, Horner syndrome, or pulsatile tinnitus without cerebral ischemia, patients with intracranial ICA dissection almost invariably have brain ischemia and cerebral infarcts.

Fluctuation of neurological signs during the first 2 weeks after symptom onset was common in our patients, occurring in 50% of the cases. Cerebral hypoperfusion was probably the mechanism of many of these events, in contrast to distal embolism, which is thought to be the most important mechanism of cerebral ischemia in patients with extracranial ICA dissection.

The typical angiographic findings of intracranial ICA dissection are similar to those observed with dissection of its extracranial portion. String sign, double lumen, irregular scalloped stenosis, and vessel occlusion are usually seen when the dissection involves the subintimal and intramedial layers, and aneurysm formation typically occurs when the subadventitial layer is affected. The most common intracranial site for aneurysm is the supraclinoid portion of the ICA with occasional extension into the MCA and/or ACA. All of our patients had involvement of the supraclinoid portion of the ICA (8 with stenosis and 2 with occlusion), with extension to the MCA or ACA in 2 patients each. Aneurysm formation involving the ipsilateral A2 segment was present in 1 patient who developed a subarachnoid hemorrhage.

Subarachnoid hemorrhage and aneurysm formation with mass effect are common complications in patients with intracranial ICA dissections but rare in patients with dissection of the extracranial vessels. The presence of thinner medial and adventitial layers and the lack of a well-developed external elastic lamina in the intracranial arteries have been implicated as the main factors causing subarachnoid hemorrhage in these patients. Most of the cases reported to date have involved the vertebrobasilar arteries. With a few reports describing subarachnoid hemorrhage in patients with dissection of the ACA or MCA. The reason for this discrepancy is not known.

Occasionally, patients with intracranial ICA dissections have been misdiagnosed as having vasculitis and are treated with steroids. This happened in 1 of our patients (patient 3). Among all types of vasculitis with central nervous system involvement, the only one known to affect the distal portions of the ICAs is giant cell arteritis. In most of these patients, only the petrous and cavernous segments of the ICA are affected, with no involvement of its supraclinoid portion. Giant cell arteritis generally affects a much older population and is associated with an increased erythrocyte sedimentation rate.

The treatment of patients with intracranial ICA dissection is controversial. The development of hemorrhagic transformation and progression of the dissection during treatment with heparin have prompted speculation that anticoagulation may be harmful in these patients. On the other hand, spontaneous progression in patients who were not given anticoagulants has supported the opposite argument. Six of our patients received immediate anticoagulant therapy with no hemorrhagic transformation or progression of the dissection. We had no complications in the patients treated with antiplatelet agents. Even though our sample was too small to allow any conclusions, the immediate administration of either type of treatment seems to be relatively safe.

In contrast to most of the literature, in which massive stroke has been the rule with a 75% mortality rate, all 10 of our patients with intracranial ICA dissection did well, with no to moderate disability during a 3-month follow-up period. The discrepancy with prior studies probably represents a bias in the population described; cases diagnosed during autopsy have dominated the literature. In more recent reports, the outcome of these patients has been significantly better than previously described, probably because of a higher index of suspicion and improved diagnostic methods allowing antemortem diagnosis.

In conclusion, we believe that intracranial ICA dissection can have a good prognosis and should be considered in the differential diagnosis of supraclinoid ICA stenosis or occlusion, especially when no other stroke risk factors are identified.

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revision of the manuscript for important intellectual content (Drs Chaves, Estol, Esnaola, Gorson, O'Donoghue, de Witt, and Caplan); administrative, technical, and material support (Drs Estol, Esnaola, and Gorson); study supervision (Drs Chaves, Estol, O'Donoghue, de Witt, and Caplan).

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REFERENCES


