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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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.9 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>Left LS infarct</td>
<td></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 IC infarct</td>
<td></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>Right caudate, right MCA, and left ACA infarcts</td>
<td></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 corona radiata infarct</td>
<td></td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>0</td>
</tr>
<tr>
<td>5/M/20</td>
<td>Right LS infarct</td>
<td></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 LS infarct</td>
<td></td>
<td>Stenosis of the supraclinoid portion of the left ICA</td>
<td>2</td>
</tr>
<tr>
<td>7/M/22</td>
<td>Right caudate and IC infarcts</td>
<td></td>
<td>Stenosis of the supraclinoid portion of the right ICA with thrombosis of the right A1 segment</td>
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<tr>
<td>8/M/59</td>
<td>Left frontal infarct</td>
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<td>Stenosis of the supraclinoid portion of the left ICA</td>
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<tr>
<td>9/M/15</td>
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<td>Occlusion of the supraclinoid portion of the right ICA</td>
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<tr>
<td>10/M/25</td>
<td>Left LS infarct</td>
<td></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.

Comment

Spontaneous dissecting aneurysms of the intracranial carotid system are uncommon in adults, especially when...
Figure 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).
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 suprachinoid 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 has 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 suprachinoid ICA stenosis or occlusion, especially when no other stroke risk factors are identified.

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Author contributions: Study concept and design (Drs Chaves and Caplan); acquisition of data (Drs Chaves, Estol, Esnaola, Gorson, O'Donoghue, de Witt, and Caplan); analysis and interpretation of data (Drs Chaves and Caplan); drafting of the manuscript (Dr Chaves); critical

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REFERENCES


