Long-term Follow-up of Occlusive Cervical Carotid Dissection

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We retrospectively studied 19 cases of occlusive cervical carotid dissection encountered at our hospital between 1974 and 1984 and followed for 5–13 (mean 8.2) years to assess the long-term prognosis of the disease. Five patients had transient ischemic attacks, seven had minor stroke, six had major stroke, and one had epileptic seizures. Angiography demonstrated the typical string sign in 17 cases, a double lumen with occlusion in one, and multiple scalloped narrowings with distal occlusion in the other. Three patients died within 1 month and three remain severely disabled (overall mortality and major morbidity 32%), five have permanent deficits, and seven are neurologically intact; the remaining patient was lost to follow-up. Five patients were treated surgically (two had extracranial-intracranial bypass and three had cervical carotid exploration), and the other 14 were treated medically. The overall rate of reopening was 47% with eight of 10 patients demonstrating recanalization on control angiography and another patient demonstrating recanalization at surgery. These nine patients remain clinically stable on follow-up evaluations. However, vascular abnormalities in the healed arteries were notable and include kinking, fibromuscular dysplasia, dissecting aneurysms, intracranial occlusion, and a residual mural defect.

Spontaneous dissection of the cervical carotid artery is an increasingly recognized vascular disorder with a rather benign prognosis. Although the pathogenesis, angiographic picture, and treatment of this disorder have recently been discussed in several papers, little is known about its long-term course. The angiographic spectrum of carotid dissection ranges from luminal stenosis to frank occlusion and may include dissecting aneurysms, double lumen, and intimal flaps. In many reports complete occlusion constitutes a rather uncommon presentation of the disease, accounting for 18% of the carotid dissections in the series of Houser et al; however, this presentation constituted 60% of our 42 patients during 15 years (1974–1988), probably because for many years a great number of patients with carotid occlusion were referred for extracranial-intracranial anastomosis. We retrospectively studied 19 patients with occlusive dissection of the cervical carotid artery admitted to our hospital during 1974–1984 who were followed for 5–13 years.

Subjects and Methods

We reviewed the medical records and angiograms of patients with a diagnosis of carotid occlusion following spontaneous dissection between 1974 and 1984. All but two patients showed the string sign on angiograms (Table 1), which is considered highly suggestive of a carotid dissection. We excluded patients with other conditions known to cause an angiographic string sign, particularly atherosclerotic pseudo-occlusion, from our study. Of the 19 patients identified, seven were women and 12 were men; their ages ranged from 13 to 59 (mean 44) years.

Two patients had arterial hypertension and another had a pterional meningioma contralateral to the cerebrovascular symptoms. Five patients presented with transient ischemic attacks, seven with a minor stroke, six with a major stroke, and one with epileptic seizures (Table 1). Head and neck pain occurred in seven patients and an incomplete Horner’s syndrome in two. Angiography demonstrated a string sign in 17 patients, a double lumen with occlusion in one, and multiple carotid scalloped defects with distal occlusion in the other; fibromuscular dysplasia (FMD) was present in one patient. Anticoagulation was never used, and medical therapy consisted of antiplatelet drugs in most cases. Five patients (26%) were treated surgically.

Results

Three patients (16%) died during the acute phase of their illness. Of the remaining 16 patients, three require total care, five have permanent deficits
TABLE 1. Summary of Course of 19 Patients With Occluding Cervical Carotid Dissection

<table>
<thead>
<tr>
<th>Pt/age/sex</th>
<th>Symptoms</th>
<th>At presentation</th>
<th>Control</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Follow-up (yr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/59/M</td>
<td>Major stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Supportive</td>
<td>Hemiplegic</td>
<td>9</td>
</tr>
<tr>
<td>2/38/M</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Recanalization, wedge-shaped defect</td>
<td>Endarterectomy, postoperative occlusion</td>
<td>Permanent deficit</td>
<td>8</td>
</tr>
<tr>
<td>3/44/F</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Recanalization, FMD</td>
<td>Medical</td>
<td>Permanent deficit</td>
<td>9</td>
</tr>
<tr>
<td>4/59/M</td>
<td>Major stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Supportive</td>
<td>Death</td>
<td>–</td>
</tr>
<tr>
<td>5/55/M</td>
<td>Major stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Supportive</td>
<td>Hemiplegic</td>
<td>9</td>
</tr>
<tr>
<td>6/16/M</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Persistent occlusion</td>
<td>STA-MCA bypass</td>
<td>Permanent deficit</td>
<td>13 (recurr...</td>
</tr>
<tr>
<td>7/42/M</td>
<td>Major stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Supportive</td>
<td>Hemiplegic</td>
<td>8</td>
</tr>
<tr>
<td>8/52/M</td>
<td>Major stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Supportive</td>
<td>Death</td>
<td>–</td>
</tr>
<tr>
<td>9/13/M</td>
<td>Seizures</td>
<td>String sign</td>
<td>Recanalization</td>
<td>Medical</td>
<td>Normal</td>
<td>12</td>
</tr>
<tr>
<td>10/44/M</td>
<td>TIA</td>
<td>String sign</td>
<td>Recanalization</td>
<td>STA-MCA bypass</td>
<td>Normal</td>
<td>8</td>
</tr>
<tr>
<td>11/49/F</td>
<td>TIA</td>
<td>String sign</td>
<td>Recanalization, kinking</td>
<td>Medical</td>
<td>Normal</td>
<td>8</td>
</tr>
<tr>
<td>12/48/F</td>
<td>Major stroke</td>
<td>String sign, FMD</td>
<td>Not done</td>
<td>Supportive</td>
<td>Death</td>
<td>–</td>
</tr>
<tr>
<td>13/46/F</td>
<td>TIA</td>
<td>String sign</td>
<td>Recanalization</td>
<td>Medical</td>
<td>Normal</td>
<td>5</td>
</tr>
<tr>
<td>14/48/M</td>
<td>TIA</td>
<td>Scalloped defects, distal occlusion</td>
<td>Recanalization, dissecting aneurysm, kinking</td>
<td>Medical</td>
<td>Normal</td>
<td>5</td>
</tr>
<tr>
<td>15/22/F</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Not done</td>
<td>Open at carotid exploration</td>
<td>Normal</td>
<td>6</td>
</tr>
<tr>
<td>16/57/M</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Persistent occlusion</td>
<td>Medical</td>
<td>Permanent deficit</td>
<td>10 (recurr...</td>
</tr>
<tr>
<td>17/57/F</td>
<td>Minor stroke</td>
<td>Double lumen with occlusion</td>
<td>Not done</td>
<td>Carotid exploration</td>
<td>Permanent deficit</td>
<td>7</td>
</tr>
<tr>
<td>18/58/F</td>
<td>TIA</td>
<td>String sign</td>
<td>Not done</td>
<td>Medical</td>
<td>Lost to follow-up</td>
<td>–</td>
</tr>
<tr>
<td>19/23/M</td>
<td>Minor stroke</td>
<td>String sign</td>
<td>Recanalization, occlusion at ophthalmic artery</td>
<td>Medical</td>
<td>Normal</td>
<td>6</td>
</tr>
</tbody>
</table>

Pt, patient; M, male; F, female; TIA, transient ischemic attack; FMD, fibromuscular dysplasia; STA-MCA, superficial temporal artery—middle cerebral artery.

Although four of the five could resume their previous activities, and seven are asymptomatic; one patient was lost to follow-up 3 years ago (Table 1). The duration of follow-up ranged from 5 to 13 (mean 8.2) years. Symptoms recurred after 6 months and 3 years in patients aged 16 and 57 years, respectively; both patients had permanent occlusion. One remains severely disabled, and the other could not resume his previous activities.

Ten of the 16 living patients had control angiography and subsequent yearly Doppler ultrasonography. Control angiograms were obtained 2 weeks to 6 months and, in one case, 4 years after the initial arteriogram. Recanalization was demonstrated in eight patients, and the occlusion was unchanged in the other two (Table 1). In three patients the healed artery was completely normal. Kinking was evident in the recanalized artery in two patients, and in one a dissecting aneurysm developed, which remained unchanged on a second control angiogram performed 3 months later. Intracranial occlusion of the internal carotid artery (ICA) beyond the ophthalmic artery was seen in one patient.

After spontaneous reopening that was documented angiographically, patient 2 had carotid endarterectomy for a wedge-shaped stenotic area believed to represent a residual atheroma; the artery occluded postoperatively without symptoms. Two patients (15 and 17) had no control angiography and underwent carotid exploration; the vessel failed to reopen in one case, and in the other the artery was found to be open at surgery 3 months after the onset of symptoms. Two other patients (6 and 10) had an extracranial—intracranial bypass; in the latter recanalization was seen on the control angiogram. There was no operative mortality or morbidity in the five patients treated surgically.

The eight patients with recanalized ICAs remain clinically stable 5 (two cases), 6 (one case), 8 (three cases), 9 (one case), and 12 (one case) years after recanalization. Six are asymptomatic and two have permanent neurologic deficits.
Discussion

The most common angiographic appearance of a carotid dissection is a long tapered narrowing or occlusion distal to the bulb (string sign). While stenotic lesions tend to improve or resolve angiographically in approximately 85% of vessels within a few months, the natural history of complete occlusion is poorly understood. Some aspects, including recurrence of the dissection, occurrence of embolic phenomena, healing time, the so-called “angiographic residuum,” and long-term prognosis, remain unclear. The clinical manifestation of occlusive dissection was severe and the prognosis poor in one third of our cases. Most such patients were >50 years old, suggesting that concomitant atherosclerotic disease may play some role in older patients; conversely, only three of the 13 patients with less severe clinical manifestations were >50 years of age. Overall, long-term outcome was favorable and the overall carotid reopening rate was good (47%).

It is generally believed that no occluded ICA will reopen after 6 months, with most recanalizations occurring within 1 month. An even faster healing (1 week) has been reported, and we document healing after 15 days in one of our patients. Moreover, in one patient recently operated on for traumatic carotid dissection, we found that the artery had spontaneously opened only 3 hours after angiography had demonstrated complete occlusion. These findings emphasize that recanalization may occur early, possibly as a result of the simple vis a tergo of blood and before the time-consuming spontaneous resorption by fibrinolytic processes. Reports of spontaneous reopening of occlusive carotid dissections are few. In the Mayo Clinic series, only eight (17%) carotid arteries were completely occluded and only one vessel was found to be reopened on follow-up angiography. Conversely, in an extensive study of this subject, Bogousslavsky et al found a reopening rate of 43%, which is very similar to our results.

Even with recanalization, residual vascular abnormalities are notable. Houser et al found an angiographic “residuum” in 25% of healed dissections. In our patients with reopened ICAs, angiographic abnormalities could have been preexistent (FMD, atheromas, tortuositities) or neoformed (luminal defects, dissecting aneurysms, embolic intracranial occlusions). Residual wedge-shaped defects, probably representing a coincidental atheroma or scar and not a recurrent dissection, may be left undisturbed.

The role of mechanical stress on arterial loops in the pathogenesis of carotid dissection has been rarely stressed, but this factor may be involved in a significant number of cases. In occluding dissections, intramedial hemorrhage dissects into a subintimal rather than a subadventitial plane, and late pseudoaneurysm formation is rare; the “angiographic” course is often resolution or improvement. Conversely, when the dissection occurs in an area of looping, the incidence of subsequent aneurysm formation is high. In one series, aneurysms were identified in five of nine ICA lesions and four occurred in tortuous arterial segments.

Intracranial embolization was evident on the control angiogram in only one of our 19 cases, while in another series it occurred in 14% of patients. It is likely that complete occlusion partly forestalls early embolization.

Recurrent dissection is considered rare, with only one instance in a series of 30 cases followed for a mean of 3.2 years and none in 36 patients reported by Mokri et al. Our long-term follow-up confirms the duration of recanalization. However, in spite of the rarity of local recurrence, these patients have remained at some vascular risk. One patient developed a dissection in a renal artery, while another developed a dissection in the opposite carotid artery. These findings suggest that some unknown disorder of the blood vessel wall predisposes to dissection. Some forme fruste of FMD was suspected in patients without evident predisposing factors.

In conclusion, the reopening rate of occluding carotid dissection is satisfactory (47%) and long-term prognosis for patients with reopened arteries is fairly good, but residual vascular abnormalities are notable and must be followed over time.

References


KEY WORDS • angiography • carotid artery diseases • dissection
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