Case Report

Multiple Cranial Neuropathy as a Feature of Internal Carotid Artery Dissection

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The traditional presentation of spontaneous internal carotid artery dissection includes ipsilateral hemicranial headache, oculosympathetic paresis, and contralateral focal cerebral ischemic deficits. However, we describe two cases with multiple cranial nerve involvement ipsilateral to the dissection as the principal feature. The first patient, a 36-year-old man, had involvement of the 9th, 10th, 11th, and 12th cranial nerves. The second case was a 53-year-old man with abnormalities of the 5th, 7th, 9th, 10th, and 12th cranial nerves. In both, magnetic resonance imaging revealed a ring-like area of abnormal signal intensity surrounding the carotid artery at the skull base. Carotid angiography was consistent with the suggestion of dissection on the magnetic resonance studies in both cases. The patients recovered without anticoagulation. Internal carotid artery dissection may thus present with multiple cranial nerve palsies, which could be mistaken for an infiltrating tumor of the skull base. Magnetic resonance imaging is useful in identifying the condition. (Stroke 1990;21:141-147)

Spontaneous dissection of the wall of the internal carotid artery (ICA) is relatively rare. The first reported case appeared in the literature in 1959, and a number of reports since then have further documented the clinical presentation of this disorder.

ICA dissection is generally divided into two syndromes. The first syndrome is characterized by ipsilateral hemicrania and oculosympathetic paresis. The second syndrome is more ominous and manifests with hemicrania and delayed contralateral focal cerebral ischemic deficits. When presenting with only mild signs, ICA dissection can be initially mistaken for other diseases such as Horton’s syndrome, cerebral cardiogenic emboli, or thrombotic infarctions.

Ojemann et al recognized the angiographic features of this condition (string sign), and angiography has remained the most reliable diagnostic test for ICA dissection.

We describe two cases of ICA dissection presenting primarily with the rapid evolution of multiple, lower cranial nerve palsies that gave rise to the initial diagnosis of an infiltrating skull base tumor.

Case Reports

Case 1

A 36-year-old right-handed white male lawyer initially developed a right occipital, nonthrobbing headache while jogging. The pain subsequently extended to the right supraorbital region and was intensified by movement of the jaw. Swallowing difficulties developed 2 days later, and slurring of his speech became apparent at the same time. He had also been aware of the earlier onset of a distortion of taste.

The patient's medical history was unremarkable; in particular, there was no history of significant trauma. However, he had played golf on the day before the onset of symptoms.

Examination revealed an intellectually intact individual with a mild dysarthria. Fasciculations were noted on the right half of his tongue, which deviated to the right on protrusion. The right sternocleidomastoid and trapezius muscles were weak, and the soft palate drooped to the right at rest and did not elevate on phonation. Palatal sensation was diminished on the right. There was no tenderness over the carotid arteries, and no asymmetries in pupil size or ptosis were noted. There was no long-tract sign, and no bruits were heard over the carotid arteries and mastoids.

An enhanced computed tomogram (CT scan) of the head was normal (Figure 1). A lumbar puncture was carried out, and the opening pressure was 170 mm water, increasing to 220 mm with right jugular compression and to 235 mm with left jugular compression. Cerebrospinal fluid analysis revealed two lymphocytes per high-power field; the protein concentration was 40 mg/dl and the glucose concentration was 49 mg/dl. The electrophoretic pattern was normal qualitatively and quantitatively.

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Magnetic resonance imaging (MRI) (Figure 2), using a resonance time of 500 msec and an echo time of 25 msec, revealed a crescent-like hyperintensity adjacent to the right jugular fossa on the anterior, median, and posterior aspects of the right ICA. This finding was consistent with extravasation of blood into the right ICA wall. The rest of the MRI study, with special attention to the posterior fossa and the vertebrobasilar system, was unremarkable. Carotid angiography demonstrated slight narrowing with minimal irregularity of the petrous portion of the right ICA (Figure 3). This finding correlated with the abnormality evident on MRI. The rest of the angiogram was normal.

The signs and symptoms resolved spontaneously without anticoagulation within 2 months. Follow-up MRI carried out 2 months after the onset of symptoms (Figure 4) showed no abnormalities, suggesting complete resolution of the dissection.

Case 2

This 53-year-old white man struck the back of his head while extricating himself from beneath his automobile, which he had been repairing at the time. A left occipital headache radiating down into the neck region developed soon after. The pain was dull in nature and did not vary with changes in head position. Numbness on the left side of his face and slurred speech developed 2 weeks later, followed by difficulty in swallowing. His medical history was unremarkable, and there was no other immediate significant history of trauma.

Examination revealed mild hypesthesia to pinprick over the left side of his face involving all three divisions of the 5th nerve. There was a left peripheral facial palsy, the left side of the palate failed to elevate, and fasciculations and atrophy were noted over the left side of his tongue, which deviated to the left on protrusion. Further examination of the nervous system was unremarkable; in particular, there were no features of a Horner's syndrome, and there were no long-tract signs.

CT of the skull base (Figure 5) revealed no significant abnormality. MRI of the same region (Figure 6) demonstrated an area of increased signal, which formed a ring-like zone around the left ICA just inferior to the left petrous bone. These features suggested a hematoma within the wall of the left ICA. The rest of the MRI study was unremarkable. Angiography confirmed the presence of a dissection of the left ICA (Figure 7). Anticoagulation was not instituted, and cranial nerve function gradually improved, with almost-complete resolution within 6 months. The only residual symptom was hypesthesia over the left side.
of the palate. MRI carried out at 6 months (Figure 8) showed complete resolution of the dissection.

Case 3
A 29-year-old woman developed transient numbness of her left arm and face as an isolated symptom. Examination of the nervous system was entirely normal. MRI 5 days later (Figure 9) showed an area of increased signal intensity surrounding the upper cervical segment of the right ICA, with minimal impingement on the lumen. These features were consistent with extravasation of blood into the wall of the ICA. The carotid angiogram (Figure 10), however, was normal. This patient was not anticoagulated and remains in good health 1 year after the event.

Discussion
The first two cases are important in that they indicate that ICA dissection may present solely with multiple cranial nerve palsies. This condition should therefore be considered in the differential diagnosis of multiple cranial neuropathy.

The pathogenesis of 9th, 10th, 11th, and 12th cranial nerve involvement may be explained on the basis of compression. All these nerves lie close to the ICA, in the vicinity of the jugular bulb, and could be compromised by lateral extension of the expanded ICA wall. However, involvement of the 5th and 7th cranial nerves as illustrated in case 2 is more difficult to explain. Cohen et al8 reported a case of fibromuscular dysplasia with dissection of the left ICA and hypesthesia in the first division of the left 5th nerve. They explain this finding on the basis of poststenotic dilatation of the intracranial portion of the ICA, compressing the first branch of the 5th cranial nerve. In our case 2, poststenotic dilatation of the intracranial ICA was not evident on the angiogram. On the basis of anatomic studies of the cavernous portion of the ICA, another possibility is that dissection of
FIGURE 5. Case 2. Computed tomogram of head after injection of contrast agent. Nonspecific irregular enhancing lesion inferior to left jugular canal.

the ICA may have resulted in occlusion of the inferior cavernous sinus artery (lateral trunk), which supplies the trigeminal ganglion, resulting in ischemic neuropathy. A similar hypothesis has also been suggested to explain a case of 6th nerve palsy with ICA dissection. Our experience with these cases concurs with the reports of Goldberg et al and Lieschke et al, which indicate that MRI is a sensitive method for detecting dissection of the ICA. A further example of the usefulness of MRI in the diagnosis of ICA dissection is evidenced by case 3.

The findings of this and other studies also emphasize the fact that CT is less sensitive than MRI in the diagnosis of ICA dissection. More importantly, while many reports indicate that angiography is the diagnostic procedure of choice in the diagnosis of ICA dissection, it is apparent that the angiogram may be negative or may show only minimal abnormalities in the presence of significant dissection. MRI can define dissection with greater precision and has the further advantage of being noninvasive. We propose that MRI is the diagnostic procedure of choice for screening patients with suspected ICA dissection. However, angiography is still necessary to demonstrate patency of the vessel and status of the distal circulation, which are important in planning therapeutic strategies, with particular respect to the institution of anticoagulation.

Of the three cases described, one showed normal ICA anatomy on angiography while the other two demonstrated only minimal luminal involvement. The decision not to anticoagulate was based on the stable clinical picture and the absence of significant luminal compromise. The excellent clinical outcome in all three patients and the complete radiologic resolution of the defects tend to indicate that ICA dissection may resolve spontaneously without anticoagulation in select clinical situations.

The clinician should be aware of the possibility of ICA dissection when confronted with a patient presenting with multiple lower cranial nerve palsies, even in the absence of the other typical signs. On clinical grounds, this condition can be differentiated from posterior fossa disorders, and particularly from intradural and extradural vertebral artery dissections; intradural and extradural vertebral artery dissections have been shown to have two types of presentation, either subarachnoid hemorrhage or brainstem infarction, and usually the lateral medullary syndrome of Wallenberg. Of 51 patients with cranial nerve palsies, all had long-tract or cerebellar signs. ICA dissection should also be considered in patients presenting with subtle deficits suggestive of hemispheric pathology (as illustrated by case 3).

When ICA dissection is suspected, MRI of the neck and skull base should be the investigation of choice since it is apparent that extravasated blood is
FIGURE 6. Case 2. Magnetic resonance image of head. Resonance time 600 msec and echo time 20 msec at level inferior to petrous bones. Signal of increased intensity surrounding left internal carotid artery (arrow).

FIGURE 7. Case 2. A. Left carotid artery angiogram early after injection of contrast agent shows no stenosis at bifurcation. B. Same angiogram, but subtracted and late after injection of contrast agent. Some dye remains at cervical level of internal carotid artery (ICA) in nondependent fashion (arrow). (The patient was lying on his back.) This has been called “twisted ribbon sign” and is due to entrapment of contrast in false lumen caused by dissection. Note that defect disappeared before ribbon siphon portion of ICA.
readily detected in the vessel wall by this imaging technique. MRI also has the advantage of clearly imaging the posterior fossa anatomy and in this way helps rule out disorders that can present similarly. Angiography is still indicated in the presence of a positive MRI because it provides information about the extent of the dissection of the vessel wall and about the overall vascular stream.

ICA dissection presenting with multiple lower cranial neuropathy in the absence of Horner’s syndrome and contralateral neurologic deficit appears to carry a good prognosis. Such lesions are readily demonstrable on MRI, and in patients in whom angiography shows little or no involvement of the lumen, anticoagulation does not appear to be indicated.

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References

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