Cranial nerve palsies in spontaneous carotid artery dissection

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Abstract

Two patients had isolated unilateral cranial nerve palsies due to spontaneous internal carotid artery (ICA) dissection without ischaemic cerebral involvement. One had acute glossopharyngeal and vagal, the other isolated hypoglossal nerve palsy. Reviewing all reported cases of angiographically confirmed ICA dissection in the literature, 36 additional cases with unequivocal ipsilateral cranial nerve palsies were analysed. While an isolated palsy of the IXth and Xth has not been reported previously, palsies of the XIIth nerve or the IXth to XIIth nerves were most frequently found. In these patients, lower cranial nerve palsies are probably the result of compression by an enlarging ICA due to mural haematoma. Symptoms and signs indicative of carotid dissection were concurrently present only in some reported cases. This raises the question of unrecognised carotid dissection as a cause of isolated cranial nerve palsies. When the dissection occurs in the subadventitial layer without relevant narrowing of the arterial lumen and when an aneurysm is thrombosed, angiography does not reliably yield the diagnosis. Therefore, carotid dissection might have been underestimated as a cause of isolated lower cranial nerve palsies before the advent of MRI. MRI demonstrates directly the extension of the wall haematoma in the axial and longitudinal planes. Some arteriopathies such as fibromuscular dysplasia and tortuosity make a vessel predisposed to dissection.

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The clinical presentation of spontaneous internal carotid artery (ICA) dissection is highly variable. Sudden, intense unilateral pain in the neck and face, ipsilateral oculosympathetic paresis, and transient monocular blindness (amaurosis fugax) associated with a contralateral transient ischaemic attack or stroke form the typical presenting triad suggesting extracranial carotid dissection.^{1 2 3} Lower cranial nerve palsies seem to be a rare finding, with the hypoglossal nerve being the most frequently affected (5%)⁴. However, when ICA dissection occurs without CNS involvement, it is not readily recognised as the cause of a cranial nerve palsy. Some of

the reported "idiopathic" jugular foramen (Vernet) and similar syndromes (Villaret, Collet Siccard) in the older literature might well have been caused by spontaneous carotid dissection.⁵ In those times ICA dissection as a cause of cranial nerve palsy was poorly known, angiography quite risky, and ultrasound and MRI not available. Out of 31 consecutive patients with ICA dissection diagnosed at our department within four years, two presented with cranial nerve palsies without cerebral involvement.

Case reports

PATIENT 1

A 42-year-old man had been in excellent health. He had no vascular risk factors. After strenuous activity (cutting wood) in military service he suddenly felt pain in the right upper jaw which later extended to the ear and the whole right side of his face. He suffered an episode of hazy vision on the right eye lasting about five minutes. The following morning pain had almost disappeared, but he had difficulty swallowing and could not drink properly. His voice had a nasal sound. Otherwise he felt well. In the hospital, three days after first symptoms had occurred, his voice was severely hoarse, almost aphonic, and nasal. On examination he had a drooping right palate arc. On phonation the palate as well as the dorsal wall of the pharynx deviated to the left. Sensation to touch of the right posterior pharyngeal wall was reduced and the gag reflex could not be elicited on the right side. No other signs were found. Chest x-ray, ECG, and routine blood laboratory tests were normal. Initially brainstem infarction was suspected and a posterior fossa CT scan was performed which was normal. Because of the right sided facial pain and the initial episode of presumed right amaurosis fugax, right ICA dissection was suspected.

Duplex sonographic examination of the carotid arteries performed the same day showed a patent carotid bifurcation, no atherosclerotic changes, and symmetrical blood flow velocities. Both common carotid arteries were also normal. Pulsed Doppler sonographic examination of the upper cervical segments of both ICAs disclosed a coiling of both with adjacent antegrade and retrograde flow in the high cervical portion. However, the blood flow velocities on the right side were twice as high as on the left side in both flow directions. This finding supported the diagnosis of an ICA dissection and anticoagu-

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lation with heparin sodium was started. The following day, a right common carotid arteriogram was performed which also revealed the ICA tortuosity. There were irregularities of the vessel wall about 3 cm distal of the bifurcation and extending cranially beyond the coiling. Equivocal slight stenosis on the anteroposterior projections was visible (fig 1). In addition, a pouch with trapped contrast media showed up on the late lateral views (fig 1 a2 and 1 a3) which is a quite distinct sign of dissection. These findings confirmed the suspected diagnosis of ICA dissection. They could, however, not explain the glossopharyngeal and vagal nerve palsy.

MR images of the upper cervical region performed eight days after the first symptoms

yielded the explanation. A large ICA mural haematoma was demonstrated as a hyperintense signal on the sagittal and axial sections of the T1- and T2-weighted and proton density spin echo sequences (fig 2). It extended from beneath the coiling up to the petrosal carotid canal. In the axial plane, a large sub-adventitial extension of the haematoma was demonstrated with expansion of the vessel wall in the carotid space. This led to displacement of the parapharyngeal space (fig 2 b1 and 2 b2) where the glossopharyngeal nerve crosses the ICA medially leading from the jugular foramen to the root of the tongue. In this region the Xth cranial nerve is also in close contact with the ICA (fig 3). Cranial MRI was normal.

After one week of heparin treatment, oral anticoagulation with a coumarin was started. No visual or TIA-like symptoms occurred. Six weeks after admission, hoarseness and nasal voice were still present, but swallowing was possible again. Pharyngeal sensation was normal on both sides, no palate droop was found any more and pharyngeal wall shift on phonation was only minimal. Doppler sonography still showed a higher blood flow velocity on the right side. A second MRI scan eight weeks after admission showed complete resolution of the mural haematoma. Anticoagulants were continued and after a total of 11 weeks all symptoms and signs had cleared. A persistent slight asymmetry of blood flow velocities at Doppler examination was considered to be due to possible slight residual stenosis. Coumarins were stopped and aspirin started.

PATIENT 2

A 45-year-old man had untreated mild arterial hypertension for several years. During the past two years he suffered from intermittent slight nuchal pain. Two days before admission nuchal pain intensified but did not change its well known quality. There was no special event and no trauma. On the day of admission he noticed difficulty with articulation, a heavy tongue, and problems to eat. Slightly slurred speech was noted by his wife. Brainstem infarction was suspected and a CT scan performed which was normal. On admission he clearly stated that he could not properly move the food with his tongue, while oral sensibility, chewing, and swallowing were not disturbed. Examination showed moderate left hypoglossal palsy with tongue deviation to the left when thrusting it out and deviation to the right when phonating. No other signs were found. Chest and cervical spine x ray, electrocardiogram, and routine blood laboratory tests were normal.

Doppler and Duplex sonographic examination of the cervical carotid and vertebral arteries and transcranial Doppler sonography on the day of admission were normal. Repeat ultrasound study the following day revealed occlusion of left ICA with typical signs indicating dissection.⁶ Transcranial Doppler examination showed good collateral supply from the right carotid and from the verte-

Figure 1 (Patient 1) Right common carotid arteriogram (a1 to a3 sequential lateral views, b1 and b2 sequential anteroposterior projections). Lateral views demonstrate only minimal wall irregularities (arrows) without recognisable stenosis. On late images (a2 and a3) a pouch can be seen wherein contrast material is trapped (double arrow) situated in the ascending loop of a carotid artery loop (open arrow). Wall irregularities are much more marked on anteroposterior projections (small arrows in b1 and b2), starting about 2 cm distal to carotid bifurcation and extending beyond the artery loop (open arrow). There seems to be only slight stenosis in this segment of ICA.

Figure 2 (Patient 1) MR images at 1.5 tesla. The sagittal right paramedian T1 weighted (TR = 400 ms, TE = 18 ms) section (a1) demonstrates mural haematoma "looping" along the ICA coiling as hyperintense signal (arrows). There is only minimal stenosis of the vessel lumen represented by the adjacent low signal ("flow void"). The T1 weighted (TR = 500 ms, TE = 16 ms) adjacent axial sections at the atlas level (b1 and b2) demonstrate large expansion of ICA vessel wall by mural haematoma. The hyperintense signal surrounds in a semilunar shaped way all three coils of the loop (arrows in b1). Also the contralateral left ICA shows a coiling (three sectioned lumina (flow voids)) without mural haematoma (see b1). Note the large posterior expansion of the mural haematoma (arrows) into the region where the glossopharyngeal nerve passes on image b2. The same, trefoil leaf-shaped, hyperintense signal is demonstrated on T2 weighted (TR = 3,000 ms, TE = 90 ms) axial sections (c) at the atlas level (arrows) identifying the underlying pathology as blood.

Figure 3 Drawing of an anatomical axial section through the neck at the level of the first cervical vertebral body (atlas). The close relationship between the ICA and the IXth, Xth XIth and XIIth cranial nerves is highlighted in the magnified cutting. (1) nasal cavity; (2) maxillary sinus; (3) facial muscles; (4) jaw muscles; (5) neck muscles; (6) atlas body; (7) parotid gland; (8) parapharyngeal space. (Compare with axial MR image: fig 2 b1).

brobasilar system across the circle of Willis. MR images of the upper cervical region were performed two days after the first symptoms. They showed a mural haematoma in the prepetrosal left ICA segment with severely narrowed lumen (flow void). Caudal to it there was an isodense mass adjacent to the artery, probably representing an aneurysm (fig 4). Three sectioned lumina (flow void) on both sides indicated bilateral coiling. There were no signal changes in the brain, brainstem, or cerebellum. Bilateral carotid angiography was performed three days after the first symptoms. It showed bilateral coiling before the ICAs entered the petrosal canal. On the left side contrast filling was markedly delayed and a saccular aneurysm attached to the coiling filled. The segment immediately distal to the coiling was severely stenosed (fig 5). On the day of admission heparin treatment was begun and seven days later changed to oral anticoagulation. Control ultrasound examination seven days later showed partial resolution of the occlusion with a now present, though diminished flow signal in the left ICA. Hypoglossal palsy was still slightly present but subjective discomfort was minimal.

Review of the literature

All reported series and cases of spontaneous ICA dissection were reviewed looking for unequivocally documented symptoms and signs of cranial nerve involvement. Swallowing difficulty alone was not considered to indicate IXth or Xth nerve dysfunction because local expansion of the aneurysm towards the pharynx might be the cause.⁷ Unilateral "scalp tenderness", a frequently encountered symptom in some series,⁸ was not considered to indicate trigeminal nerve involvement unless there was a clear sensory deficit reported. "Dysgeusia" (unpleasant, strange taste) does, in our opinion, indicate facial or glossopharyngeal nerve dysfunction. As Maitland did,⁹ we suspect a compression of the IXth rather than of the VIIth nerve or of the chorda tympani to be the cause.^{8,10} Detailed testing of taste, which should allow a differentiation, was not reported in these cases. If a nerve compression by the expanding wall haematoma (pseudoaneurysm) is the pathogenetic mechanism, the IXth nerve is more at risk. If, however, interruption of the nerve's blood supply is the working mechanism, the IXth and VIIth may be affected equally. Finally, isolated tinnitus was not considered a sign of VIIIth nerve dysfunction.

Only cases in whom the diagnosis had been supported by angiography showing a typical picture were considered.^{1,3,8,11,12}

Results

Thirty-six patients reported under the heading of carotid dissection with signs of ipsilateral cranial nerve palsies were found in the literature and analysed together with our two patients (table 1).^{2,8,9,10,12-31} In 35, the age and sex were given: the average age was 44 years,

Figure 4 (Patient 2) MR images at 1.5 tesla. The proton density weighted ($TR = 2740$ ms, $TE = 30$ ms) contiguous axial sections at the level of the skull base (a1 and a2) demonstrate narrow lumen (flow void) of left ICA (arrowhead in a1) and a surrounding mass with mixed iso and hyperintense signal (arrows in a1). A little more cranially a hyperintense signal adjacent to the flow void signal indicates mural haematoma (arrow in a2). These signal changes are even better visualised on T1-weighted ($TR = 800$ ms, $TE = 11$ ms) axial sections with fat suppression technique (b1 and b2): The mass asymmetrically surrounding the narrowed vessel lumen (arrows in b1) represents the aneurysm. The slit-like residual lumen (arrowhead in b2) is compressed by the mural haematoma shown as hyperintense signal (arrow in b2).

ranging from 28 to 60 years. Thirty were men (86%, mean age 47 years) and five women (mean age 39 years). All but one patient²⁸ showed an ICA narrowing of variable degree: 11 patients had a high grade stenosis with a string sign, seven a long irregular slight narrowing, 15 a segmental narrowing, and one a tapering occlusion. Seventeen (49%) had aneurysmal dilatation: nine had a fusiform pseudoaneurysm, two of them bilaterally; three had aneurysmal pouches (fig 1); three had a saccular aneurysm (fig 5); one had a double lumen; and in one patient no details on the "aneurysm" were available. In three patients detailed angiographic results were not given. In 35 patients the site of the dissection could be located: the prepetrosal segment was involved in all, 11 showed extension of the dissection to the (lower) cervical segment, 11 to the intrapetrosal, and eight to the cavernous segment. In three patients this information was not provided. In 26 patients data about tortuosity of the ICA were given: 10 patients (38%) had some degree of tortuosity, four had kinking, three coiling, and three moderate tortuosity. Four patients had documented findings suggestive of fibromuscular dysplasia (FMD). Tortuosity as well as the signs of FMD may be masked by the dissection itself. Only a

minority of the reported cases had documented cerebral four vessel angiography and comments concerning the presence of FMD and tortuosity in other cranial arteries were frequently lacking. Therefore, the relative frequency of these vascular alterations is certainly underestimated.

In 16 patients MRI scans were performed, 14 of which had a mural haematoma demonstrated. In two patients the MRI showed no hyperintense signal. In one of these the delay was 8 weeks and the haematoma might have resolved.²⁸ In the other patient no details on MR technique or examination time were given in the presence of a typical angiographic result.¹⁰ In our two patients MRI demonstrated tortuosity (fig 2 b1) and also an aneurysm (fig 4 b1) in addition to the mural haematoma.

In 27 patients an ipsilateral XIIth nerve palsy was reported, 10 times isolated, six times in conjunction with IXth, Xth, and XIth nerve palsies, four times with Xth nerve palsy, five times with IXth and Xth nerve palsies, once with XIth nerve palsy, and once with IXth, Xth, VIIth, and Vth nerve palsies. Five patients had dysgeusia, three isolated, one in conjunction with Vth, and one with VIth nerve palsy. Among seven patients with a Vth nerve palsy, it was isolated in four. All patients with Vth, VIth, or IIIrd nerve palsy had an extension of the dissection up to the cavernous segment. The compiled frequency of the affected cranial nerves in the 38 cases was as follows (table 1): XIIth: 27 times; IXth (including the five patients with dysgeusia): 18 times; Xth: 17 times; XIth: 7 times; Vth: 7 times; VIIth/VIth/IIIrd: once each. In 20 patients (53%) an ipsilateral oculosympathetic paresis (Horner's syndrome) was observed.

In 13 patients information about outcome of cranial nerve palsies was given: eight patients had recovered after three weeks to two years, usually within two to four months; five patients had persistent palsies after three to 10 months and, hence, some might well have recovered later on.

Ten patients (26%) had ischaemic symptoms. Six had a transient and two a persistent hemiparesis contralateral to the dissection, and two had an ipsilateral amaurosis fugax.

Discussion

Peripheral cranial nerve palsy caused by ICA dissection is only rarely observed. In the large reported series totalling 356 patients with ICA dissection, no cranial nerve palsies were described.^{2,3,32-43} Most of these patients with angiographically confirmed ICA dissection were presenting with cerebral ischaemic signs and symptoms. On the other hand, only 8 (21 %) of 38 patients presenting with cranial nerve palsies due to ICA dissection collected from the literature (table 1) showed cerebral ischaemic involvement.

This discrepancy may be due to not noticing ipsilateral cranial nerve palsies in the presence of signs of contralateral stroke. It might

Figure 5 (Patient 2) Bilateral common carotid arteriogram (a1 to a3: sequential lateral views, b1 and b2: sequential anteroposterior projections of the left side; c and d lateral and anteroposterior views respectively of the right side). Left side: Lateral views demonstrate delayed ICA filling (a1) simulating "string sign" in the early images (arrows in a1), saccular aneurysm attached to coiling (arrowhead in a2 and a3,) and poor contrast of the segment distal to the aneurysm (open arrow in a2 and a3) because of stenosis (see b); contrast material is trapped in the aneurysm (a3). Anteroposterior views demonstrate aneurysm attached to the coiling (arrowhead in b1 and b2) and high grade stenosis immediately distal to the coiling (open arrow in b1 and b2). Right side: Lateral (c) and anteroposterior (d) view show prepetrosal ICA coiling (arrowhead). Collateral blood supply from right ICA to left MCA across circle of Willis (arrows in d).

also indicate that ICA dissection as a cause of lower cranial nerve palsies is not readily recognised. In a recent review of 32 patients with XIIth nerve palsy,⁴⁴ carotid dissection was considered to be the cause in only one patient, and this was a traumatic dissection. Unfortunately, no details on the diagnostic examination of these patients are given. Therefore, the question remains, whether some of the reported patients with so called traumatic XIIth nerve palsies or some of those attributed to brainstem ischaemia²⁵ in reality had a carotid dissection causing the nerve palsy. In our own series of 31 patients with ICA dissection, the two patients reported here (6%) presented with isolated cranial nerve palsies. In retrospect, we recall another two patients who presented with isolated XIIth nerve palsy and ipsilateral neck pain. CT scan did not support the suspected diagnosis of a cervical tumour or infection. This was several years ago when ICA dissection was not considered as a possible cause and hence no further investigations were performed.

However, the relatively frequent observation of isolated cranial nerve palsies without central involvement (79%) as opposed to the rare observation of cranial nerve palsies in ICA dissection patients with cerebral ischaemic symptoms could indicate that these two groups have a different pathogenesis. It is, for instance, conceivable that the former had subadventitial rather than subintimal dissections, since the subadventitial dissection is less likely to obstruct the vessel lumen and would rather extend outwardly. Several facts from our analysis support such a hypothesis: (1) the higher prevalence of men in the ICA dissection patients with cranial nerve palsies (86%) as opposed to the general ICA dissection population (51%),⁴ (2) like in traumatic ICA dissection,³² there is a high prevalence of aneurysms in spontaneous ICA dissection patients with cranial nerve palsies (49%), while aneurysms in ICA dissection patients with hemispheric signs are less frequent (10–33%)^{31,33}; (3) the higher prevalence of extreme vessel tortuosity in the ICA dissection patients with cranial nerve palsies (27%) compared with the general population (3–12%).⁴⁵ Tortuosity may indicate a structure wall anomaly making the vessel predisposed to external mechanical injury. Tortuosity also does alter flow dynamics which may cause sheer injury to the vascular wall again making the affected vessel predisposed to dissection. Could more frequently occurring trivial and unreported traumas of a physically more active male population be favouring subadventitial dissection in the presence of a predisposing arteriopathy?

In the case of subadventitial dissection, muscular medial layer may impede narrowing of the lumen and the mural haematoma may considerably expand the circumference of the vessels (figs 2b and 4b). The close topographical relationship of the most frequently involved XIIth, IXth, and Xth nerves with the carotid artery in the upper cervical parapha-

Table 1 Patients with spontaneous ICA dissection and cranial nerve palsies

Reference	Age/Sex	Side	Site of ICA dissection	Angiography	Tortuosity	FMD	MRI (delay)	Cranial nerve palsies
Kramer ¹⁴	41/F	left	prepetrosal	segmental narrowing, aneurysm prepetrosal	no	yes	no	IX, X(?), XII left
Gros ¹⁵	38/M	left	prepetrosal	fusiform aneurysm	no	no	no	XII left
Labauge ¹³	38/M	left	prepetrosal	slight stenosis, pseudoaneurysm	?	?	no	XII left
Cohen ¹⁶	31/F	left	pre and intrapetrosal, cavernous (?)	segmental narrowing pseudoaneurysm	?	yes	no	V (1 + 2) left
	38/M	left	pre and intrapetrosal cavernous (?)	segmental narrowing pseudoaneurysm	?	yes	no	V left
Fisher ⁸	45/M	left	cervical + prepetrosal	string sign up to petrosal canal, pouch	?	?	no	dysgeusia chorda tympani left ?
	53/M	left	prepetrosal	segmental narrowing	?	?	no	dysgeusia chorda tympani left ?
Barbizet ¹⁷	42/M	right	cervical, prepetrosal, intrapetrosal, cavernous	string sign emboli in MCA branches	no	no	no	III and V right
Bradac ¹⁸	58/M	left	prepetrosal	string sign, saccular aneurysm	no	no	no	IX, X, XI, XII left
Goodman ¹⁹	41/M	left	prepetrosal	string sign	no	no	no	XII left
	59/M	right	prepetrosal	string sign	kinking	no	no	XII right
Maitland ⁹	37/F	left	pre- and intrapetrosal, cavernous	string sign	no	no	no	VI left, dysgeusia (chorda tympani?)
Hommel ²⁰	55/M	left	prepetrosal	segmental narrowing saccular aneurysm	no	no	no	XII left
	54/M	right	prepetrosal	segmental narrowing fusiform aneurysm	yes	no	no	X, XII right
	44/M	left	cervical, prepetrosal	string sign	no	no	no	IX, X, XII left
Gauthier ¹²	?	left	?	stenosis; no further details	?	?	no	IX, X, XII left
	?	left	?	?	?	?	no	IX, X, XII left
Mokri ²	?	?	?	stenosis, no further details provided	?	?	no	dysgeusia
Mokri ²¹	47/M	bilateral	prepatrosal left	segmental stenosis pseudoaneurysm	kinking	no	no	IX,X,XI,XII left
Benrabah ²²	40/M	left	prepetrosal right cervical, prepetrosal, cavernous	string sign	kinking ?	no	no	none right V left (hyperesthesia)
	50/M	left	cervical, prepetrosal, cavernous	string sign	?	no	no	V1 left (hyperesthesia)
Vighetto ²³	48/M	left	prepetrosal	string sign	yes	?	no	XII left
Goldberg ²⁴	49/M	left	pre- and intrapetrosal	long irregular narrowing	no	no	yes, delay: 16 days	IX,X,XI,XII left
Francis ¹⁰	46/F	left	high cervical, intrapetrosal, cavernous	tapered narrowing	no	yes	yes, delay: ?	V left, dysgeusia
Hess ²⁵	41/M	right	cervical, prepetrosal	tapering occlusion	no	no	yes, delay: 13 days	X,XII right
Lieschke ²⁶	42/M	right	prepetrosal	string sign	kinking	no	yes, delay: about 10 days	X,XII right
Waespe ²⁷	41/M	bilateral	prepetrosal bilateral	pseudoaneurysm bilateral, irregular slight narrowing	no	no	yes; delay: about 10 days	IX,X,XI,XII left
Bradac ²⁸	28/M	right	cervical, prepetrosal	long irregular narrowing	no	no	yes, delay: 10 days	none right
	41/F	left	cervical, prepetrosal	long irregular slight narrowing, pseudoaneurysm	coiling	no	yes, delay: 15 days	X,XII right
	53/M	left	prepetrosal	pseudoaneurysm, no stenosis	kinking ?	no	yes, delay: 8 weeks no hyperintense signal	XII left
Dal Pozzo ²⁹	58/M	left	prepetrosal	slight segmental narrowing	no	no	yes, delay: 19 days	IX,X,XI,XII left
	45/M	right	prepetrosal	slight segmental narrowing	?	?	yes, delay: 40 days	XI,XII right
	54/M	left	cervical, pre- and intra petrosal	slight irregular segmental narrowing, small pouch	?	?	yes, delay: 27 days	IX,X,XII left
Vernay ³⁰	60/M	left	prepetrosal	segmental stenosis	no	no	yes delay: 8 weeks	XII left
Panisset ³¹	36/M	right	pre- and intrapetrosal	slight stenosis	?	?	yes, delay: ?	IX,X,XI,XII right
	53/M	left	pre- and intrapetrosal, cavernous?	stenosis, double lumen	yes	?	yes, delay: ?	V,VII,IX,X,XII left
Sturzenegger	42/M	right	cervical, prepetrosal	long distance irregular wall, slight stenosis, small pouch	coiling bilateral	no	yes, delay: 8 days	IX,X right
	45/M	left	pre- and intrapetrosal	segmental severe stenosis saccular aneurysm within coiling	coiling bilateral	no	yes, delay: 2 days	XII left

Year indicates year of publication; ICA, internal carotid artery; MCA, middle cerebral artery; FMD, fibromuscular dysplasia; TIA, transient ischaemic attack; ?, means that the respective criterion could not be analysed because of lack of data.

ryngeal space (fig 3) are a strong argument for a nerve compression by the expanded carotid wall as the underlying mechanism. In most cases the dissection begins 2 to 4 cm distal to the carotid bulb extending cranially and showing a maximal distension at the C1 or C2 vertebral level. At this point the IXth cranial nerve crosses the ICA laterally, the Xth lies posteriorly and medially, and the

XIIth crosses the ICA a little more caudally, in general at the C3 level. The fact that in patients with involvement of upper cranial nerves, dissection usually extended into the petrosal and cavernous segment (table 1) is another argument for a compression to be the causative mechanism. Nerve ischaemia due to compromised blood supply is a second possible mechanism.^{46 47} The IXth to XIIth nerves

Horner's syndrome	Pain	TIA/stroke	Special remarks
yes left	ear, face, neck left	no	dissection very likely
no	temple left	right arm + face transient paresis right arm transient	dissection probable
no	face left	no	persistent XII nerve palsy
yes left	eye, cheek left	no	"Raeder's syndrome"
yes left	temple left	no	"Raeder's syndrome"
yes	face, eye left	right arm + face transient	
yes	face, eye left	no	
yes	neck, temple, head right	hemiplegia left persistent	
yes	neck left	no	
yes	temple left	no	
yes	neck right	hemiparesis left transient	
no	face, neck, eye left	no	recovery of VIth nerve palsy within 2 months
no	eye, temple left	no	recovery of XIIth nerve palsy within 2 months
			control angiography after 2 months: persistent aneurysm, no stenosis
no	temple, occipital right	no	recovery of Xth nerve palsy within 3 days and of XIIth within 6 months
			control angiography after 8 months: persistent aneurysm, no stenosis
no	yaw, ear, face left	hemiplegia right aphasia, persist	XIIth nerve palsy persistent
			control angiography after 3 months: normal ICA left, MCA branch occlusion
yes	face left	no	no details on angiography
no	face, neck left	right arm, transient	no details on angiography
?	?	?	no further details provided
yes left	occipital left	no	asymptomatic after 2 1/2 y
no right	no		
yes	face, neck, eye left	no	
yes	face, eye left	no	
yes left	neck, temple left	no	
yes	face left	no	
yes	face, neck left	no	MRI = normal (?) delay not given
no	orbital right	hemiparesis left transient	
no	temple, eye right	no	
no	neck, face left	no	bilateral dissection, right side asymptomatic
no	neck right	no	XII paresis persistent after 4 months
yes	head	amaurosis fugax	XII paresis persistent after 6 months
no	head, neck left	no	XII paresis persistent after 1 year, dissection not proved
no	neck	no	CT also positive. Follow up MRI after 13 months; haematoma resolved.
yes	neck right	no	CT also positive. Follow up MRI after 14 months: haematoma resolved
yes	eye, front left	no	CT negative. Follow up MRI after 6 weeks: smaller still persistent haematoma.
yes	occipital left	no	
no	occipital, eye right	no	recovery within 2 months
no	occipital, neck left	no	recovery within 6 months
no	yaw, ear, face right	amaurosis fugax	bilateral coiling at MRI. Follow up MRI after 7 weeks: haematoma resolved.
no	nuchal and arteriography)	no	clinical recovery within 10 weeks. bilateral coiling (at MRI)
			Partial resolution of stenosis after 7 days (Ultrasound). Clinical recovery within 3 weeks.

are supplied by the ascending pharyngeal artery and the Vth and VIIth by the middle meningeal artery.^{46 47} Both arteries arise from the external carotid artery which may be directly affected by dissection or compressed by the expanding dissected ICA. There are few data supporting this pathogenesis. The distinction of these two mechanisms is, however, irrelevant for the diagnostic and thera-

peutic management of the patients.

The proof of carotid artery dissection as the cause of cranial nerve palsies is a problem not yet solved. Angiography, considered as the standard by most authors, can detect a compromise of the vessel lumen only above a certain degree. The recognition of a low grade narrowing of the lumen is difficult, with considerable interobserver disagreement.⁴⁸ In the case of a subintimally located dissection the mural haematoma will in the first place protrude the intima into the vessel lumen and produce a variable degree of stenosis up to occlusion. These cases are easily depicted by angiography.^{1 3 8 11 12} As the majority of ICA dissections seem to occur in the subintimal layer¹ angiography is the diagnostic method of choice especially if there are clinical or ultrasound signs of cerebrovascular circulation compromise.⁶ Yet, even in these cases, angiography is by no means always pathognomonic.^{28 29 31 49} The advent of magnetic resonance angiography allows evaluation of luminal caliper non-invasively as well.

However, if the dissection occurs in the subadventitial layer, considerable wall expansion into the carotid space and displacement of the adjacent parapharyngeal space may occur (figs 2 b and 4 b) The resulting haematoma can compress adjacent structures such as the lower cranial nerves (fig 3) without causing relevant vessel narrowing. These cases may not be detected by angiography. Not only can no vessel anomaly be seen, but also the clinical picture with isolated cranial nerve palsies is not even considered an indication for angiography. The question therefore arises whether the reported prevalence of subintimal dissection^{1 3 4} is merely a consequence of patient selection and the diagnostic method used, since angiography seems an inadequate method to detect subadventitial dissection.

The reported cases raise questions about the significance of aneurysms and tortuosity found on angiography both for establishing the diagnosis and for looking for an underlying arteriopathy.

Aneurysms are reported on angiography in up to 33% of patients with ICA dissection.^{3 11 33} We have observed three patients with an aneurysm among 27 patients with ICA dissection (11%): two had small pouches (one of them patient 1) and one (patient 2) had a saccular aneurysm. It is often impossible to determine whether the (saccular) aneurysms were present as congenital malformations before dissection occurred or whether they are the result of a persistent communication between the dissection cavity and the regular lumen of the artery.^{15 27 28}

Aneurysms of the extracranial ICA segment are sometimes associated with FMD.⁵⁰ FMD in ICA dissection has been reported in as many as 22%.³ However, FMD cannot be considered the main aetiology of ICA dissection. The sex ratios observed in FMD and ICA dissection strongly argue against that: FMD affects up to 80% women,⁵⁰ and in the present review series 86% of patients with

ICA dissection were men. In addition, intracranial aneurysms occur frequently with FMD,⁵⁰ but only exceptionally with ICA dissection.

Tortuosity, next to FMD, is the second most common arterial wall anomaly cited to make a vessel predisposed for dissection.^{3 42 51} In our review (table 1) 10 (38%) of 26 patients whose angiograms could be judged had some degree of tortuosity. Seven (27%) had a kinking or coiling. According to large series of consecutive, selected carotid arteriograms slight tortuosity is present in 11% to 31%, but extreme tortuosity (kinking and coiling) in only 3% to 12%.⁴⁵ Tortuosity seems to be more frequent in those patients with dissection who have aneurysm formation. Among the 10 patients with tortuosity we found seven with aneurysms (70%), whereas the 16 patients without tortuosity had only five aneurysms (31%). Whether extreme tortuosity alone, not associated with atherosclerosis or dissection, can cause cerebral ischaemia or lower cranial nerve compression is a matter of debate.⁴⁵

Application of MRI and a large index of suspicion may clarify several of the unsettled issues. MRI is the only non-invasive technique to demonstrate mural haematoma and to show its longitudinal extension (fig 2 a) and the degree of wall expansion (fig 2 b and 4 b).^{24 26-31 49} Newer techniques such as chemical shift imaging (fat suppression) (fig 4 b1, b2) make MRI more sensitive to even small arterial dissections.⁴⁹ MRI additionally gives information about surrounding tissues. The information provided by MRI at present is complementary to that of angiography.

In conclusion, carotid dissection should be considered in isolated or combined lower cranial nerve palsies, especially if associated with ipsilateral neck pain, headache, and Horner's syndrome. Descriptive terms like "carotidynia", Raeder's syndrome, "migraine with miosis", and all the cranial nerve syndromes with famous eponyms⁵ should be avoided in favour of a causal diagnosis. Today, MRI provides a sensitive non-invasive method for the diagnosis of ICA dissection if performed within the first weeks.

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