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Spontaneous internal carotid artery dissection: early diagnosis and management in 44 patients

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Abstract First symptoms and initial clinical, ultrasonographic and neuroradiological findings ascertained a mean of 5.6 days (SD = 5.6 days), 7.7 days (7.0), and 11.2 days (8.0) after symptom onset were analysed in 44 patients who suffered a spontaneous internal carotid artery dissection (ICD) verified by magnetic resonance imaging, angiography, or both. Common symptoms signalling dissection were unilateral headache in 68%, transient ischaemic attack in 20%, and cerebral infarction in 9%. Severe pain preceded cerebral ischaemia by more than 3 days in 60% of those patients who eventually suffered a stroke. However, only 2 were admitted because of pain alone and 33 for evolving neurological deficits. During the first month, ipsilateral severe headache occurred in 89%, neck pain in 36%, ipsilateral cerebral ischaemia in 82%, ocular ischaemia in 16%, oculosympathetic palsy in 48%, and cranial nerve palsy in 5%. Recent "trivial" head or neck trauma was elicited in 41%. Doppler and duplex sonography confirmed the clinical suspicion of ICD in 91.5% and in 96% of those with a significant stenosis or occlusion. MRI demonstrated a thickened vessel wall in all 33 imaged carotid dissections and a mural haematoma in 30. None of the 32 patients who received anticoagulant treatment subsequently deteriorated. Monitoring anticoagulant treatment with ultrasonographic follow-up studies demonstrated recanalization in 70% and persistent occlusion in 30%. The results demonstrate that familiarity with the initial symptoms, especially headache, and performance of an ultrasonographic study without delay are the cornerstones of an early diagnosis. Immediate anticoagulation to prevent fatal cerebral embolism seems the appropriate treatment when intracranial dissection is excluded, although its efficacy has not yet been proven by a controlled study.

Key words Carotid artery disease · Dissection · Magnetic resonance imaging · Ultrasonics · Anticoagulants

Introduction

Only about 4% of cerebral infarctions (CI) occur in patients under the age of 40 years [28, 29, 34] and about 10% in those under 50 years [53]. Atherosclerosis is the cause in less than 25% and a wide array of aetiologies has to be considered with increasing probabilities in young stroke patients [1, 6, 9, 12, 24, 28, 32, 33]. Cerebral emboli from a cardiac source is the most frequent cause, followed by non-atherosclerotic cerebral vasculopathy. Dissection is the most important non-atherosclerotic vasculopathy of the arteries supplying the brain and is recognised with increasing frequency to be the cause of stroke in younger adults (1–22%; 76% in a group without predisposing conditions) [1, 6, 9, 12–14, 18, 24, 28, 32, 33]. The increasing number of reported cases is not likely to be due to a growing incidence but rather to increasing famil-

iarity with this clinical entity and to progress in investigational methods, making diagnosis easier [45]: (1) decreasing risk and higher resolution of angiography [25, 47]; (2) recognition of distinct findings with ultrasonography (US) [19, 20, 30, 37, 48];(3) almost pathognomic findings with magnetic resonance imaging (MRI) [31, 40, 42, 43, 49, 56]. Whereas US and angiography yield characteristic but only rarely specific findings, confirmation of suspected dissection with MRI showing a mural haematoma is now easy. The real incidence of carotid and vertebral artery dissection is unknown. An average annual incidence of angiographically verified spontaneous dissections of 2.6 per 100000 has been calculated [45]. From the cited studies it can be estimated that about one of five CI in younger adults is caused by a dissection. Internal carotid artery (ICA) dissections (ICD) are predominant [3, 8, 14, 26, 27, 44]. Data on the patients reported show that ICD is still frequently not considered in the differential diagnosis of headache and cerebral ischaemia and is therefore still underdiagnosed or its diagnosis delayed. Early diagnosis seems mandatory, since younger patients are mainly affected and because there is much evidence that CI may be prevented by instant anticoagulation. The objective of this study was (1) to establish the first clinical features that should raise suspicion of an ICD and (2) to analyse the value of US and neuroradiological studies in the management of these patients.

Patients and methods

Forty-four consecutive patients [15 women (34%) and 29 men; mean age 46 years (range, 25–69 years)] with spontaneous ICD proven by angiography, MRI or CT were analysed. They were admitted from 1988 to 1992. The patients and their relatives were specifically interviewed regarding: (1) vascular risk factors; (2) head or neck trauma, unusual neck movements, head position, sports activities, and strenuous exercises during the month preceding ICD, (3) warning symptoms (head or anterior neck pain, amaurosis fugax, transient ischaemic cerebral attacks, tinnitus, oculosympathetic paresis) preceding stroke; (4) current (presenting) symptoms (pain, also regarding location, intensity, quality and duration; symptoms of local cranial nerve or sympathetic compression; cerebral or ocular ischaemia); and (5) reason for referral (Table 1). During follow-up, the course and cumulative occurrence of symptoms were recorded. All patients underwent repetitive neurological examinations, extensive blood, blood chemistry and coagulation studies, electrocardiogram, chest and cervical spine radiology. All patients were interviewed and examined personally by the author according to a standardized protocol including a specially designed questionnaire with particular emphasis on headache. Forty-three underwent extra- and transcranial pulsed-wave Doppler sonography; in addition 40, had extracranial duplex sonography of the arteries to the brain. In 28 patients cerebral catheter angiography was performed; in 30 MRI of the neck and head, in 16 both MRI and angiography, in 14 MRI only, in 12 angiography only and in 2 patients CT of the neck vessels only (high resolution, thin slice, contrast enhanced with reformations). In 40 patients one or more cranial CT scans were obtained (Table 2). After hospital discharge, regular clinical and ultrasonographic follow-up at 6- to 8-week intervals was possible in 40 patients. All 30 patients who

Table 1 Internal carotid artery dissection: clinical findings [44 patients (%)]

A)	First Symtoms [a]				
	Pain: headache	30 (68)			
	anterior neck pain	2			
	Transient ischaemic attack	9 (20)			
	Cerebral infarction	4 (9)			
	Horner's syndrome	1			
	Amaurosis fugax	1			
B)	Cumulative symptoms				
	Pain:	40 (91)			
	Ipsilateral		39		
	Headache		39		
	Anterior neck/neck only		16/1		
	Cerebral ischaemia	38 (86)			
	Infarction:		27		
	severe			13	
	moderate			8	
	slight			5	
	Transient ischaemic attack		9 (20)		
	Amaurosis fugax		7 (16)		
	Horner's syndrome	21 (48)			
	Tinnitus/Objective carotid bruit	3/4			
	Ipsilateral carnial nerve palsy	2			
C)	Warning symptoms [a]	23 (85)			
	[before infarction established (n = 27)]				
	Headache	20 (74)			
	< 1 day		4		
	3–7 days		7		
	> 7 days		9		
	Transient ischaemic attack	9 (33)			
	Horner's syndrome	5 (19)			
	Amaurosis fugax	2			
D)	Presentation				
	Pain and infarction	26 (59)			
	Painful Horner's syndrome	6 (14)			
	Pain and transient ischaemic attacks	5 (11)			
	Cerebral ischaemia only	4			
	Painful cranial nerve palsy	2			
	Pain only	2			
E)	Risk factors	38			
	Vascular:		29		
	Migraine/hypertension/			17 (39) / 10	
	smoking/hypercholesterolaemia/			8 / 7	
	contraceptive pills			4	
	Preceding trivial trauma		18 (41)		
	Vascular and trauma		10		
F)	Course				
	Complete recovery	26 (59)			
	Persistent symptoms	18 (41)			
	Handicap: slight		5		
	moderate		7		
	severe		4		
	Death	2			
G)	Treatment				
	Heparin immediately	32			
	Coumarin	34			
	Antiplatelet agents	8			
	Complications	0			
H)	Delay (in diagnosis and treatment)	38 (86)			
	Physician's delay	13			
	Patient's delay	11			
	Patient's and physician's delay	6			
	Without consequences (CI)	6			

[a] Combinations possible

Table 2 Internal carotid artery dissection: neuroradiology findings (*MCA* middle cerebral artery, *CT* computed tomography, *MRI* magnetic resonance imaging, *FMD* fibromuscular dysplasia

Summary		
(44 patients; 48 dissected carotid arteries)		
Stenosis:	29 (60%)	
Slight		3
Moderate		10
High grade		19
Occlusion	19 (40%)	
Pseudoaneurysm	3	
Fibromuscular dysplasia (FMD)	6	
Tortuosity	12	
Angiography		
(28 patients; 29 dissected carotid arteries)		
Stenosis:	16	
Slight		2
Moderate		3
High grade		11
Long/short distance		10/6
Smoth/irregular		13/3
Occlusion	11	
Flame shaped		9
Pseudoaneurysm	3	
FMD/tortuosity	6/12	
Atherosclerotic plaques	2	
"Normal"	2	
MRI of the neck vessels		
(30 patients; 33 dissected carotid arteries)		
Stenosis	19	
Occlusion	14	
Thickened vessel wall	33	
Mural haematoma	30	

	Brain MRI (30 patients)	Brain CT (14 patients)
Infarction	17	8
Territorial (peripheral)	9	4
Basal ganglia	1	0
Large MCA infarction	7	4
Normal	13	6

underwent MRI had at least one repeat MRI examination 3–6 weeks after the initial (diagnostic) scan. No patient had repeat of angiography.

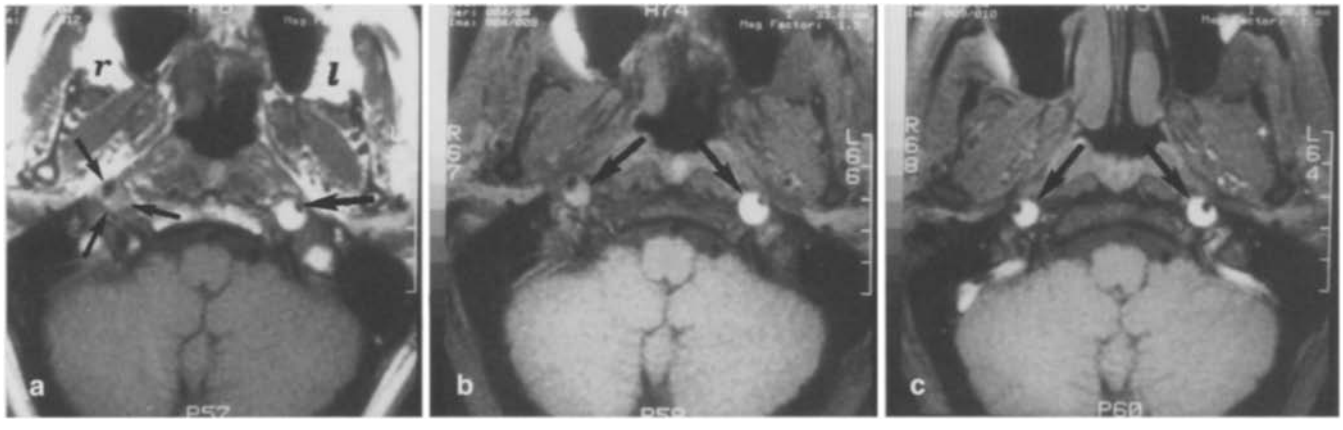
Cerebral angiography

Transfemoral selective common carotid arteriograms were obtained in 28 patients with 29 carotid dissections. The indication was cerebral ischaemic symptoms. Maximal stenosis was graded as slight, moderate (more than 50% diameter reduction in any plane) and high grade (more than 80% diameter reduction) or complete occlusion. Stenosis was further classified into short or long distance and regular or irregular with respect to the bordering vessel wall. Special attention was paid to vessel tortuosities (coiling or kinking), aneurysms, signs of fibromuscular dysplasia and atheromatous plaques (Table 2).

Magnetic resonance imaging

Five-millimetre imaging contiguous axial and sagittal sections of the head and neck using T1- and double-echo T2-weighted spin-echo sequences were acquired using a General Electric Signa 1.5 T imaging system. Most patients were imaged using the fat-suppression technique [11](Fig. 1b, c). In those patients in whom MRI was performed but not angiography, stenosis was classified by measuring the maximal degree of luminal (flow void) narrowing at the dissection site (Fig. 1). An absent flow void was considered to be an occlusion. The thickness of the vessel wall and mural haematoma (hyperintense signal on T1- and T2-weighted images) were assessed (Fig. 1, Table 2).

Fig. 1 a–c Magnetic resonance images at 1.5.T. Axial T1-weighted sections (repetition time, 420 ms; echo time. 16 ms); **a, b** 3 days after initial symtpoms; **c** 4 days later; **b, c** with fat-suppression technique. Initial pictures clearly delineated the semilunar-shaped hyperintense signal of mural haematoma eccentrically surrounding the severely narrowed lumen (flow void) on the left side (*large arrow* in **a**). Right-sided dissection might be missed unless asymmetrical thickening of vessel wall is recognized (*three arrows* in **a**), since typical hyperintense signal is not (yet) present. Fat-suppression technique allows delineation of mural haematoma and vessel wall thickening more clearly on both sides (*arrows* in **b**). Severe stenosis due to large eccentric mural haematomas in both internal carotid arteries is clearly shown on a repeat MRI scan 4 days later (*arrows* in **c**)



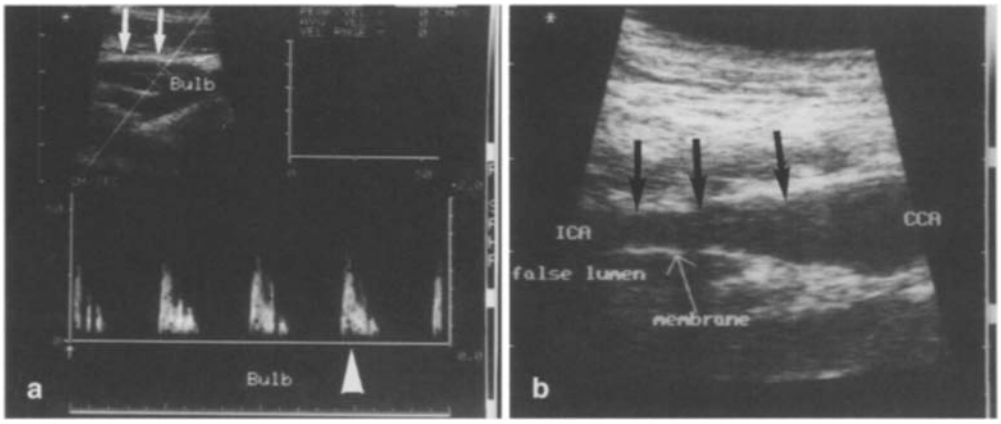


Fig. 2 Duplex sonography (**a**) demonstrates patent bulb without atherosclerotic wall changes *(arrows)*. Doppler sample in the bulb demonstrates only short systolic flow signal without diastolic flow (stump flow) *(arrowhead)*. B-mode imaging of bulb and proximal internal carotid artery (**b**) illustrates tapering luminal narrowing *(black arrows)* and a membrane *(white arrow)* separating true from false lumen

Table 3 Internal carotid artery dissection: ultrasonographic findings (43 patients; 47 dissected carotid arteries)

Occlusion of internal carotid artery	33	
Neuroradiologically confirmed		19
Neuroradiologically high-grade stenosis		14
High cervical stenosis of ICA	10	
Neuroradiologically confirmed		10
(High grade/moderate/low grade)		(1/7/2)
Normal findings	4	
Neuroradiologically low-grade stenosis		3
Neuroradiologically moderate stenosis		1
Sensitivity		
General	91.5%	(43/47)
Occlusions	100%	(19/19)
Moderate or high-grade stenosis	95.7%	(22/23)[a]
Low-grade stenosis	40%	(2/5)
Follow-up (40 patients, 44 dissected carotid arteries)[b]:		
Complete recanalization of occlusion	18	
Persistent occlusion	11	
Resolution of stenosis	9	
Persistent stenosis	1	
Recanalization of occlusion with residual stenosis	1	
Persistent normal finding	4	

[a] Eight stenoses were correctly assessed with US, 14 (neuroradiologically high grade) stenoses were overdiagnosed as occlusions
[b] Three patients were not followed-up: two died, one was lost

Ultrasonography

In 43 patients extra- and transcranial pulsed-wave Doppler sonography was carried out. In addition, in 40 extracranial duplex sonography of the brain-supplying arteries was performed (Fig. 2a, b). The method used and the diagnostic criteria applied have been published previously [48]. US examination was performed before neuroradiological confirmation in all patients except one; the sonographer was thus unaware of the final diagnosis (Table 3).

Results

Clinical findings

Among those 27 patients in whom CI was eventually established, 23 (85%) reported *premonitory symptoms* or signs preceding neurological cerebral deficit: headache in 20 (74%), transient ischaemic attacks (TIA) in 9 (33%), oculosympathetic paresis in 5 (19%), and amaurosis fugax in 2 (7%). Severe unilateral pain preceded CI by more than 3 days in 16 patients (Table 1). Considering the ultimate diagnosis and the overall clinical course, there was a delay in diagnosis and subsequent treatment in 38 patients (86%). In 6 cases this had no consequences (no CI). In 13 patients the first physician consulted did not recognize the danger of an impending stroke, whereas 11 patients did not seek medical advice until stroke had developed. In 6 patients both the patient and the doctor were to blame for the delay. The mean delay from first symptoms to hospital admission was 5.6 days (SD = 5.6, range 0–22).

In most patients symptoms developed over time. With respect to the *cumulative symptoms* assessed in each patient, *pain* was again the most frequent symptom (40 patients, 91%) with certain particularities: (1) location – unilateral (on the dissection side): 39 (89%); head: 39 (89%); anterior neck: 16 (36%); neck only: 1 patient; temple 32: (73%); forehead: 31 (70%); orbit 25 (57%); ear: 8 (18%); occiput: 6 (14%); occiput only: 3 patients; jaw: 5 (11%); (2) intensity – severe: 29 (66%); moderate: 8 (18%); slight: 3 (7%); (3) quality – never experienced before: 100%. The second most frequent symptom was *oculocerebral ischaemia:* 36 patients (82%) had amaurosis fugax, TIA or CI or a combination of these; 27 (61%) had established CI; 9 (20%) TIA only; and 7 (16%) amaurosis fugax (isolated in 2, associated with TIA in 3 and with CI in 2 patients). Six patients (14%) had never had ischaemic symptoms. The third most frequent symptom was *oculosympathetic paresis:* 21 (48%), in most cases developing

in the first week, following pain and ischaemia. In 5 patients it preceded established stroke and in one it was the first-ever symptom. A carotid *bruit* was audible in 4 patients. Three patients had transient pulsatile tinnitus. Interestingly, the tinnitus preceded cerebral ischaemia and had disappeared by the time they presented with CI and ICA occlusion. Transient tinnitus reappeared 10–15 days later when US demonstrated beginning recanalization and high-grade stenosis. Two patients had isolated *cranial nerve palsy* on the dissection side, involving the twelfth nerve in the first and the ninth and tenth in the second. Both had accompanying ipsilateral pain, but no cerebral ischaemia.

Thirty-four patients were treated with heparin or a coumarin, 8 with acetylsalicylic acid only. Two patients who presented on the second day with temporal lobe herniation due to large middle cerebral artery (MCA) infarction with oedema had only been treated for raised intracranial pressure. Thirty-two patients were started on heparin the day of admission, when ICD suspected on clinical grounds had been corroborated by the US findings. None showed any subsequent clinical deterioration. Six (young) patients not treated with heparin on the day of admisson because of TIA – despite the recommendation to do so after US examination – developed severe MCA infarction during the following 2 days. Two were then treated with heparin without subsequent further deterioration. Among the 8 treated with acetylsalicylic acid only, 6 had large MCA infarctions and the responsible physicians considered heparin too risky, 2 had only moderate stenosis and no signs or symptoms of ischaemia.

Ultrasonography

The mean delay from first symptoms to US examination was 7.7 days (SD = 7.0, range, 6 hours to 22 days). Of the patients, 85% were studied within 2 days of hospital admission. The delay between admission and US is explained by the fact that in several cases the correct diagnosis was not suspected initially and thus US not performed immediately.

Extracranial Doppler revealed absent flow in the ICA and a high resistance flow profile in the ipsilateral common carotid artery consistent with ICA occlusion in 33 patients (Table 3). *Duplex-sonography* confirmed absent ICA flow in all 33 and showed high resistance (stump) flow in the proximal bulb segment (Fig. 2a). In addition, B-scan confirmed a patent bulb and the absence of atheromatous lesions, thus rendering atherosclerotic thrombosis unlikely [48] (Fig. 2b). *Transcranial Doppler* demonstrated reduced velocities in the ipsilateral MCA and collateral flow across the anterior communicating artery. Four patients with an absent MCA flow signal (embolic MCA occlusion, verified by a "dense cerebri media sign" on initial CT scan in 2 patients) developed severe MCA infarction, with a lethal outcome in 2. Ten patients had a high cervical (retromandibular) stenosis. Duplex, sonography again confirmed the absence of atherosclerotic lesions, but the stenotic signal could be detected in only 2 patients. Thus, if only duplex sonography had been performed, 8 stenosing dissections would have been missed but all occlusions would have been detected. Findings of combined US examination were normal in 4 patients: one of them had a moderate and 3 a minimal stenosis in the neuroradiological studies. Sensitivity for detecting occlusions was thus 100% and for moderate or high-grade stenosis 95.7%.

Neuroradiological findings

The mean delay from first symptoms to the first neuroradiolgical study (angiography in 21, MRI in 21 and CT in 2 patients) was 11.2 days (SD = 8.0; range 1–36). The mean delay from first symptoms to angiography (28 patients) was 13.8 days (SD = 8.8; range 1–36) and to MRI (30 patients) 13.5 days (SD = 10.4; range 2–48). Six patients showed multiple vessel dissections: bilateral ICD in 4 and vertebral artery dissection in addition to ICD in 2. In the 4 patients with bilateral ICD only the side with the more severe stenosis was symptomatic. A summary of the combined neuroradiological findings and details of angiography and MRI is listed in Table 2. In 2 patients with large MCA infarctions and subsequent fatal temporal lobe herniation CT (thin slice, high dose contrast with coronal and sagittal reformations) of their carotids was performed showing tapering occlusions in both. MRI did not demonstrate a mural haematoma in 3 of the 33 imaged dissected ICAs although the vessel wall was thickened. In one patient, imaging was performed the very first day of symptoms; repeat MRI 4 days later showed the typical hyperintense signal. In the other 2 patients, the delay between symptoms and MRI was more than 6 weeks; thus the haematoma had probably changed to an isointense signal. MRI was unable to identify aneurysm formation proven by angiography in 3 patients. Tortuosity (kinking or coiling) was identified in 12 patients and 18 vessels. The degree of stenosis as classified with both methods was not compared in these patients, since stenosis might well have changed owing to a time lag between the two examinations. However, it should be mentioned that 2 patients presenting with painful Horner's syndrome had normal angiograms 20 and 29 days after symptom onset. MRI performed the following day showed a (small) mural haematoma. US, performed several days before angiography, was normal in the first and showed a moderate stenosis in the second.

Follow-up

Patients were followed-up for a mean of 29 months (SD = 22; range 6–78; geometric mean 22). Twenty-six (59%) recovered completely; 16 (36%) had persistent symptoms, impairing daily life in 11 (25%) and 2 (5%) died as a consequence of dissection-related CI. All 30 patients with an initial MRI examination underwent a second study 6–8 weeks later. In every case the mural haematoma had decreased in size and luminal narrowing was stationary or reduced. Repeat US examination (40 patients, 44 dissected carotid arteries) demonstrated a recanalization in 28 of the 40 vessels with previously abnormal US findings (70%; 8 occlusions and 20 stenoses according to the neuroradiological studies) and a persistent occlusion in 11 arteries (30%; all were already occluded at the initial neuroradiological study). Eleven of 19 (58%) occlusions thus did not recanalize, whereas no patient demonstrated a progression from a stenosis to an occlusion. Two patients (5%) suffered recurrent dissection of the contralateral carotid artery 15 and 56 months later. These were detected because of clinical symptoms and confiremd with MRI. Patients without new symptoms during follow-up did not undergo regular MRI but only US studies. Thus, clinically asymptomatic recurrent dissections not causing changes in the US studies would have escaped detection.

Discussion

Clinical aspects

Early diagnosis seems essential for the successful management of patients with ICD since stroke, mostly due to embolism from local thrombosis at the site of dissection, may be prevented by early anticoagulation. The first suspicion of ICD is based on clinical peculiarities. The clinical presentation of ICD, however, may be quite variable, ranging from the patient with unspecific headache or even no corresponding clinical symptoms and signs to the patients with early severe stroke and rapidly ensuing death. These clinical extremes may not give rise to further investigations, the underlying cause (ICD) remaining unrecognized. In the presence of the characteristic triad with unilateral headache,ipsilateral oculosympathetic paresis, and hemispheric ischaemic symptoms, the diagnosis is straightforward. This combination of findings was, however, presented initially by only 8 patients (18%) in this series.

Pain would have been a crucial symptom for earlier diagnosis since it was a warning symptom in 74% of those eventually suffering a stroke and one of the first symptoms in 72% of the whole group. In 16 (59%) of the stroke patients pain preceded cerebral ischaemia by more than 3 days. Six (14%) presented with "painful Horner's syndrome" [17, 22, 35, 54]. However, only 2 patients were admitted because of headache alone [7] and 33 because of subsequently evolving neurological deficits. Headache associated with dissection had distinct features: unilateral on the dissection side; located in the temple forehead and orbit; severe intensity; and a quality never before experienced [21]. The extracranial cerebral arteries are known to be pain sensitive [38] and the pain in ICD signals the onset of mural haemorrhage. This pain seems to be a key symptom as regards starting anticoagulation before CI has occurred, once US findings have corroborated the suspicion of ICD. Only 4 patients (9%) in this series had never experienced pain and presented with isolated cerebral ischaemia as the first symptom.

Further characteristic features of these patients which could arouse suspicion of ICD are: young age and low frequency of atherosclerotic risk factors or signs of general atherosclerosis. However, migraine (39% of the presented patients) seems overproportionally frequent as noted in a previous study [16]. Without discussing the distinction between traumatic and nontraumatic dissections, I found that asking whether a minor trauma had preceded the symptoms of ICD (in 41% of this series) may yield another clue to the correct diagnosis [3, 51].

Diagnostic work-up

Until recently, diagnosis had to be confirmed with angiography, which is still an invasive method that can involve risks [25, 47]. Today, nonivasive methods such as MRI and US allow early definite diagnosis in most cases [2, 4, 5]. US is a quick and noninvasive way of substantiating clinical suspicion reliably at least in those carotid dissections which should be treated with anticoagulants immediately. General experience shows that the only chance of preventing infarction is to start heparin treatment as early as possible [3, 5, 36, 37, 39, 44, 52]. To await confirmation by angiography or MRI may be harmful (6 patients in this series). US is readily available and most equipment can be taken to the emergency room especially pulsed wave systems for extra – and transcranial Doppler studies. For an experienced operator (technician or physician) confirmation of an ICA occlusion or a significant stenosis takes about 10 min. The combined Doppler and duplex examination in this series proved sufficiently sensitive to detect occlusion (100%) and moderate or high-grade stenosis (95.7%). Low-grade stenoses (less than 50% diameter reduction) can escape diagnosis (3 of 5 missed) but are rare and probably harbour a low risk of embolic complications. I stress that duplex examination alone, though providing important additional morphological information (B-scan) [48], is not sufficiently reliable, since retromandibular (high cervical) stenosis, unless high grade, may be missed. Eight dissections in this series would have been missed without additional (extracranial) pulsed wave Doppler examination. Further important in-

formation which can be provided by transcranial Doppler only is the adequacy of the collaterals in the case of high-grade stenosis or occlusion. Low ipsilateral MCA flow with dampened profile is a frequent finding but usually recovers in the first few days (unpublished personal data). Transcrainal Doppler offers the possibility of detecting those cases with persistent low flow that are at risk for infarction and might benefit from revascularization in the case of recurrent TIA despite anticoagulation. Finally, US is the method of choice for monitoring anticoagulation [5]. Since recanalization may occur within a few hours [5, 55] or take several months, the duration of anticoagulant treatment can be tailored individually.

Once anticoagulation has been started neuroradiological diagnostic confirmation is no longer urgent. Though recent reviews still consider angiography the current gold standard test [3, 39, 44] experience form this series and others [5, 31, 37, 56] suggests that noninvasive MRI is more sensitive and more specific. Angiographic features of dissection are very variable, often characteristic, but, except for the finding of a double lumen or an intimal flap, nonspecific [23, 36, 41, 44, 46]. Angiography missed two dissections in this series whereas MRI was always reliable when not only the haematoma signal but also evidence of a thickened vessel wall were sought [4, 22]. The typical hyperintense semilunar-shaped signal of mural haemorrhage in T1- and T2-weighted sequences may be absent in the very early and also the late stage of dissection when deoxyhaemoglobin is not yet or no longer present [10, 15]. Even small lesions can be depicted with the use of the fat-suppresion technique (Fig. 2 b, c) [10, 11, 15, 25, 40, 56]. MRI does have certain shortcomings which are, however, ususally irrelevant with respect to early patient management: e.g. MRI is less sensitive when searching for an underlying arteriopathy such as fibromuscular dysplasia or tortuosity; aneurysms (especially small pouches or intimal flaps) are usually not recognized; intracranial extension of the dissection may be difficult to determine due to the interference of artefacts related to low flow.

Therapeutic aspects

Two main mechanisms may lead to cerebral ischaemia in patients with ICD: arterio-arterial embolism from local thrombus formation and insufficient distal flow due to severe stenosis or occlusion [52]. It has been shown that low flow favours embolism [50]. Serial transcranial Doppler examination has demonstrated that low flow ipsilateral to the dissection in many patients may improve within a few days. In both situations anticoagulation might be beneficial. Although there is no prospective randomized study available that proves the benefit of anticoagulants, most authors recommend early treatment with heparin and later with a coumarin [3, 5, 19, 20, 36, 39, 44, 46, 52]. Clinical evidence from this series also favours this regimen since none of the 32 patients who received early heparin treatment showed any subsequent deterioration; yet 6 young patients not immediately heparinized suffered severe stroke due to embolic MCA occlusion. I am not aware of any report in the literature showing clinical deterioration due to anticoagulant treatment. Anticoagulation should be withheld in cases of intracranial dissection, which is rare in the carotid artery and its branches.

In conclusion, a young or middle-aged patient without vascular risk factors presenting with acute severe unilateral headache should raise the suspicion of carotid dissection; all the more if there is associated ocular or cerebral, transient or permanent ischaemia, Horner's syndrome or cranial nerve palsy or a preceding minor trauma. Combined Doppler and duplex examination should be the first diagnostic step. If there is stenosis or occlusion, anticoagulation with heparin should be started without delay. Currently, diagnostic confirmation is obtained noninvasively with MRI. Treatment is monitored with serial US studies.

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