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Spontaneous Cervical Cephalic Arterial Dissection and Its Residuum: Angiographic Spectrum

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Cervical cephalic dissections are uncommon acute disruptions of the arterial wall occurring predominantly in middle-aged women. Clinically, most patients present with unilateral headache, oculosympathetic palsy, or ischemic neurologic symptoms. Usually, a single internal carotid artery, predominantly the right, is affected, but simultaneous multivessel dissections are evident in about one-third of patients. Angiographically, the appearance of the dissection varies, depending on its severity, extent, and the interval between onset and angiography. In the patients reported, the disruption was manifested initially by eccentric tapered stenosis in 47%, tapered stenosis and a dissecting aneurysm in 28%, occlusion in 18%, or a dissecting aneurysm alone in 7%. Subsequently, stenotic dissections resolved in 60%, improved in 20%, and progressed in 15%, while dissecting aneurysms diminished in half and resolved in one-fourth of patients. An angiographic residuum, temporally remote to its onset, was evident in 25% of dissections. Hence, carotid arterial dissections tend to resolve, sometimes progress, but seldom recur.

The more common disorders affecting the cervical cephalic arteries include atherosclerosis, fibromuscular dysplasia, and hemorrhagic dissection [1]. Dissection may be secondary to various conditions, including trauma and infection, or may occur spontaneously [2]. Recent articles have suggested that the incidence of spontaneous dissection is greater than suspected because resolution without any angiographic residuum is frequent [3, 4]. These observations prompted a review of our patients with clinical and angiographic evidence of dissections of the internal carotid artery (ICA).

Materials and Methods

Forty-two patients with spontaneous dissection of the cervical ICA have been evaluated at the Mayo Clinic during the last decade. The ages of the 27 women and 15 men were 21–65 years, with about 80% being less than 50 years old (table 1). Thirty-eight patients (91%) had headache, 28 (67%) had acute neurologic ischemia, 24 (57%) had incomplete Horner syndrome, 16 (38%) had subjective bruits, 15 (36%) had hypertension, and one (2%) had neck pain.

The initial angiographic study opacified the following arteries: bilateral ICAs and one vertebral artery in 30 patients; bilateral ICAs in seven; bilateral ICAs and bilateral vertebral arteries in one; a single ICA and a single vertebral artery in two; and a single ICA in two. Four renal arterial angiograms and one angiogram of the aortic arch, which also had been obtained, were pertinent to the evaluation, and data from these have been included in the study. Angiography was performed within 73 days in all cases, and 40 angiograms were obtained within 3 weeks after the onset of symptoms. In addition, follow-up angiograms of the dissected vessel, obtained 1–45 months (mean, 11 months) after the onset of symptoms, were available for review in 24 cases.

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TABLE 1: **Distribution of 42 Patients With Spontaneous Cervical Cephalic Arterial Dissection According to Gender and Age**

Gender	Age Decade					Total
	3d	4th	5th	6th	7th	
With fibromuscular dysplasia (six patients):						
Female	0	2	2	0	1	5
Male	0	1	0	0	0	1
With luminal irregularity (16 patients):						
Female	0	5	5	2	0	12
Male	0	0	1	1	2	4
With dissection only (20 patients):						
Female	1	4	5	0	0	10
Male	1	2	4	2	1	10
Total	2	14	17	5	4	42

Initial Angiographic Findings

Cervical and Cranial Location

In 57 arteries, angiography revealed occlusion or disruption of the arterial lumen. The latter was manifested by either an irregularly tapered stenosis or a dissecting aneurysm or both (fig. 1). Luminal irregularities remote from the site of dissection were also present in 22 patients and were absent in 20. Angiographically, the irregularity was not diagnostic in 16 patients but was consistent with fibromuscular dysplasia in the other six (figs. 2 and 3). These latter findings provided a basis for further classification of arterial dissections.

The 57 dissected vessels included 52 ICAs, four vertebral arteries, and one right subclavian artery (table 2). Of the 29 single-vessel dissections, 20 were in the right ICA and nine in the left. Thirteen patients had multiple simultaneous dissections: eight in both ICAs; two in both ICAs and a left vertebral artery; two in a right ICA and a left vertebral artery; and one in a right ICA and a subclavian artery. Tapered stenosis of the arterial lumen was observed in 27 vessels, tapered stenosis with concomitant aneurysm in 16, occlusion of an ICA in 10, and a dissecting aneurysm alone in four.

Tapered Stenosis

Tapered stenosis of the ICA lumen affected the cervical segment in 12 vessels; the cervical and intracanalicular segments in 26; and the cervical, intracanalicular, and cavernous segments in 2 (table 3). A double-barrel lumen was present in two vessels with dissection, located proximally in one and distally in one (fig. 2). Three vertebral arteries were stenotic. Stenotic dissections of the vertebral arteries involved only the extradural segment of two vessels and the intradural and extradural segments of one vessel. These dissections were 2–4 cm long and were localized to the level of the first and second cervical vertebral bodies. Tapered proximal narrowing was characteristic of all stenotic lesions. Tapered reconstitution of the lumen occurred in 27 stenotic dissections of the ICA and in all three vertebral arterial dissections. In the other 13 ICAs with tapered stenoses, the lumen was reconstituted abruptly at the level of the entrance into or within the bony carotid canal. In four ICAs, an aneurysm lay immediately juxtaposed to an abruptly reconstituted lumen.

Aneurysmal Dilatation

Twenty-two aneurysms occurred in 20 dissected vessels. One ICA and the only subclavian artery affected had two aneurysms each. Thus, 18 aneurysms arose in 17 ICAs, two arose in two vertebral arteries, and two arose in the right subclavian artery. Twelve of the carotid aneurysms were located in the upper subcranial cervical region, and six were located in the midcervical region (table 4). Both vertebral aneurysms occurred in the intradural segment of the artery, and both aneurysms of the right subclavian artery were situated just proximal to the origin of the right vertebral artery. Both the small (<1 cm) and the large (>2 cm) aneurysms (seven cases) were saccular and wide-mouthed, while the intermediate (about 1 cm) aneurysms (11 cases) were ovoid and fingerlike (fig. 4). In 16 patients, an aneurysm lay within the stenotic dissected segment and was the only evidence of dissection in four (fig. 5). Dissecting aneurysms involving multiple different vessels, the left carotid and vertebral arteries, occurred in only one patient.

Occlusion

Of the 10 occlusions of the ICA, eight occurred distal to the carotid bulb while two occurred at its origin. In six vessels, the occlusion tapered to a point, resembling the configuration of a flame, whereas in two vessels the ends of the occlusions were rounded. The occlusions involved both ICAs in two patients, the right ICA in four, and the left ICA in two.

Intradural Location

Intracranial abnormalities included slowing of flow in the ICA–middle cerebral artery of 11 patients; collateral circulation through the ophthalmic artery or circle of Willis in nine; emboli within or occluding the supraclinoid ICA and distal cerebral branches in six; extension of a dissection from the intracanalicular segment into the cavernous segment of the ICA in two; extension of a cervical vertebral arterial dissection intradurally in one; and an intradural vertebral arterial aneurysm in two. In these 31 patients, primary dissections were not identified in the basilar or intradural ICA systems.

Follow-up Angiographic Findings

Follow-up angiograms of 29 dissected arteries with 16 aneurysms (24 patients) were available (table 5). The stenosis had resolved in 17 arteries, improved in six, and persisted unchanged in two. In four arteries, one of which had been explored surgically, progression or thrombosis occurred. Among the 16 aneurysms, six diminished, three remained unchanged, three were surgically removed, and one was not visualized owing to thrombosis of the parent vessel. Of the other three aneurysms, one resolved completely and two had

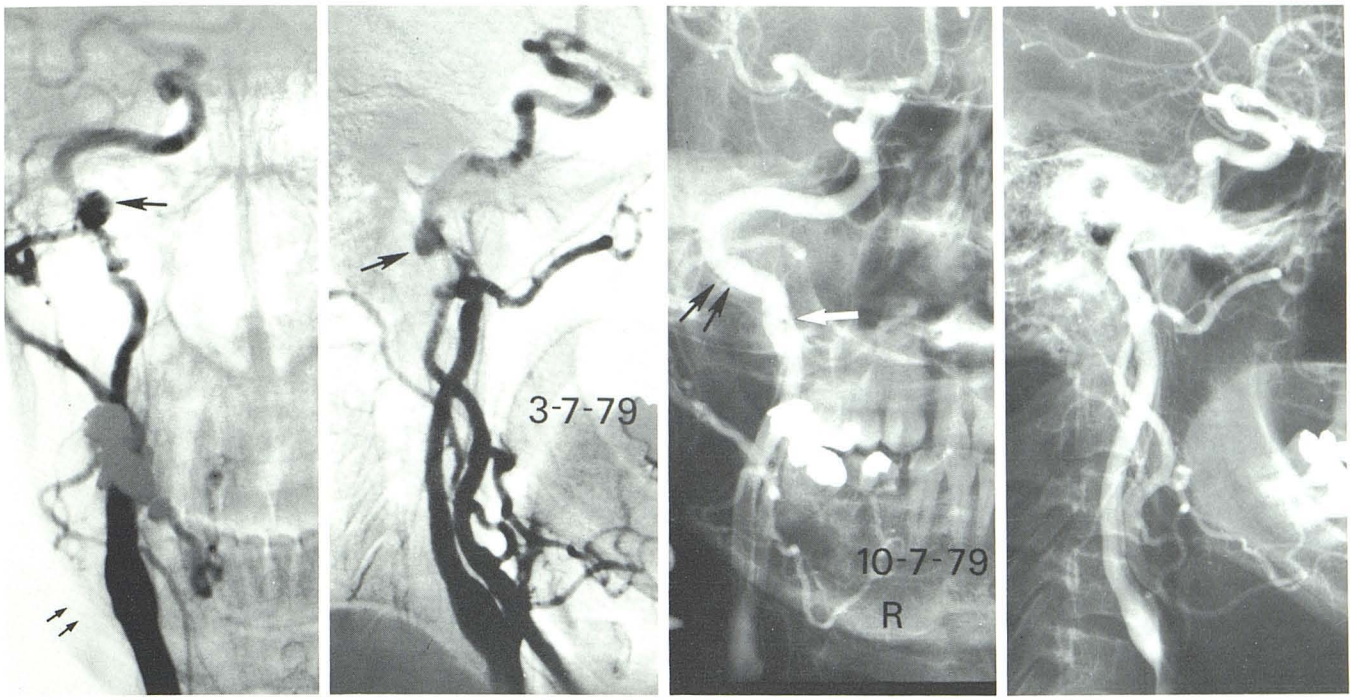


Fig. 1.—Dissecting aneurysm undergoing significant resolution. **A** and **B**, Markedly irregular, eccentric, tapered stenotic dissection with saccular subcranial dissecting aneurysm (*large arrows*) and minimal luminal irregularity of ICA proximally (*small arrows*). **C** and **D**, 7 months later. Resolution of tapered stenosis and resolution or thrombosis of aneurysm with only slight residual dilatation (*black arrows*) and residual intimal flap (*white arrow*). Note smooth proximal lumen.

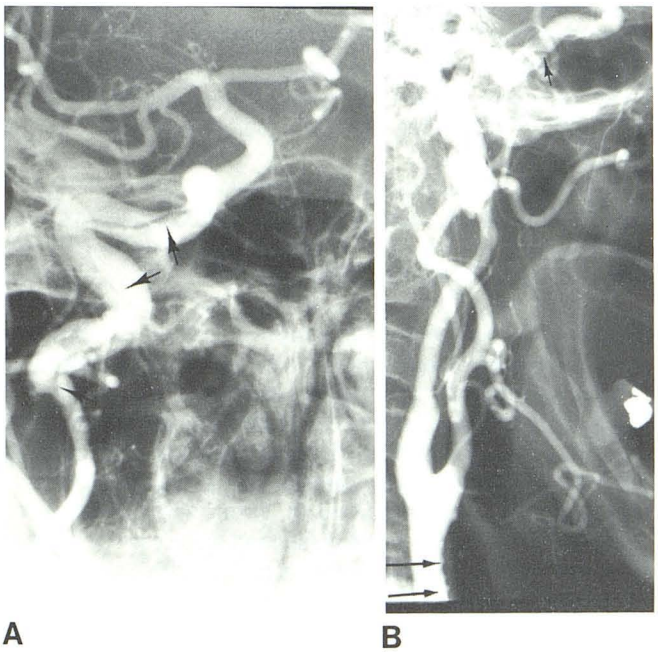


Fig. 2.—Dissection with demonstrable intimal flap. Tapered stenotic dissection of cervical ICA beginning distal to bulb and extending almost to carotid canal as well as intraluminal intimal flap (*short arrows*). Spiculated irregularity (*long arrows*) in distal common carotid artery. Also note irregular undulation of anterior wall in cervical dissection.

TABLE 2: **Initial Angiographic Findings in 42 Cases of Cervical Cephalic Arterial Dissection**

	No. of Patients	Carotid		Left Vertebral	Right Subclavian
		Right	Left		
Dissection:					
Single vessel	24	20	9	0	0
Multiple vessels	13	8	8	0	0
...		2	2	2	0
...		2	0	2	0
...		1	0	0	1
Total	42	33	19	4	1
Angiographic features:					
Tapered stenosis alone	15	10	2	0	
Tapered stenosis with dissecting aneurysm	12	3*	1	0	
Dissecting aneurysm alone	0	2	1	1*	
Occlusion	6	4	0	0	
Total	33	19	4	1	

* Multiple (two aneurysms in vessel).

only a tiny focal residual dilatation. Although none of the aneurysms appeared to enlarge, one aneurysm in the cervical ICA and one in the intracavernous ICA evolved during the 4 and 45 month interval, respectively, between examinations. A small stenotic lesion, possibly a recurrent dissection, evolved at the origin of the ICA in the same patient in whom

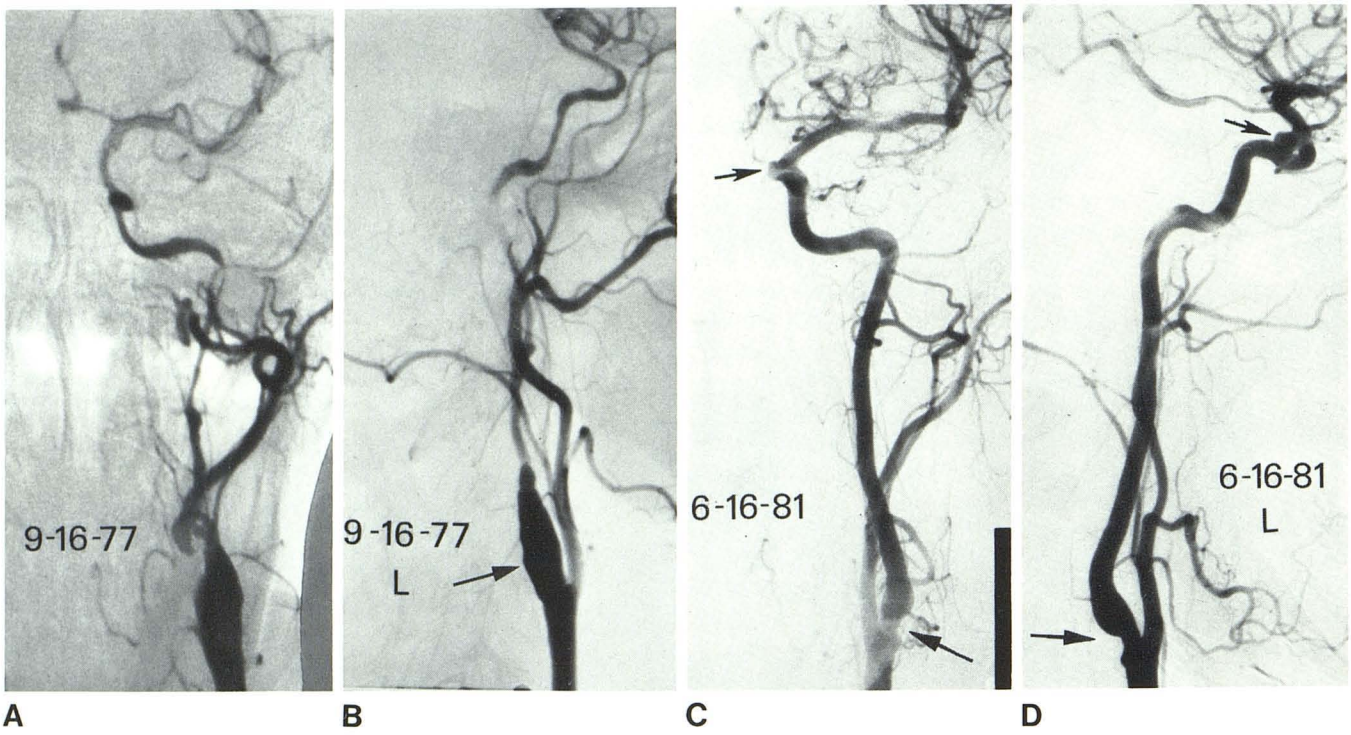


Fig. 3.—Arterial dissection with subsequent development of distal aneurysm. **A** and **B**, Tapered stenotic dissection of cervical and intracanalicular segments of ICA accompanied by irregularity of carotid bulb (*long arrows*). **C** and **D**, Almost complete resolution of stenotic dissection and intimal irregularity, but an aneurysm of cavernous ICA (*short arrows*) and a stenotic area (?recurrent dissection) (*long arrows*) have developed.

TABLE 3: **Extent of Internal Carotid Arterial (ICA) Dissection**

Extent	ICA Segment			Total
	Cervical	Cervical and Intra-canalicular	Cervical, Intra-canalicular, and Cavernous	
Fibromuscular dysplasia	2	2	0	4
Dissection with irregularity	4	11	2	17
Dissection alone	6	13	0	19
Total	12	26	2	40

TABLE 4: **Site and Configuration of 18 Dissecting Aneurysms of Internal Carotid Artery**

Extent	Configuration		Site	
	Saccular	Elongated	Subcranial	Midcervical
Fibromuscular dysplasia	1	2*	2*	1
Luminal irregularity	3	5	6	2
Dissection only	3	4	4	3
Total	7	11	12	6

* Multiple.

an aneurysm developed in the cavernous ICA. Wedge-shaped intimal flaps were also a residual change that was noted in five patients.

Angiographic Classification

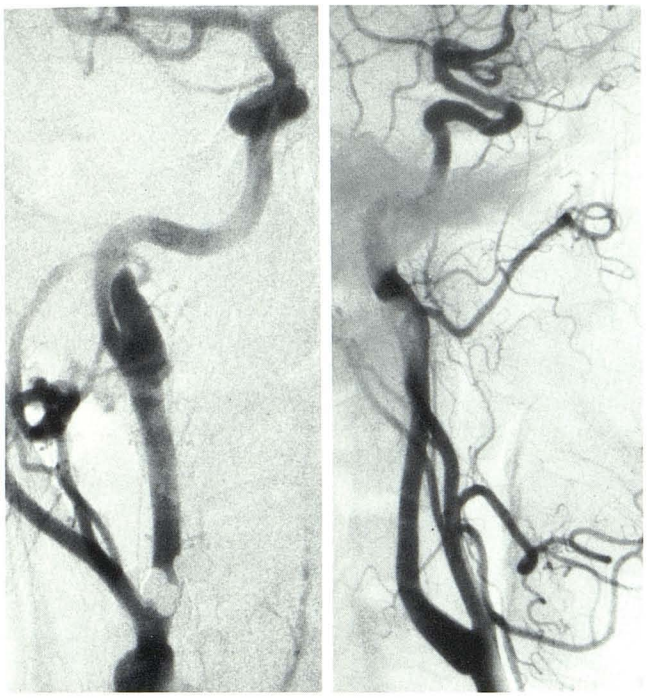
Dissection Only

When the angiographic abnormality was dissection only, men and women were affected with equal frequency. Of the 20 patients with dissections only, 15 had unilateral and five had bilateral involvement. A tapered stenosis involved the cervical and intracanalicular segments of 13 ICAs and the cervical segment of six ICAs. In six vessels with occlusion (two bilateral and two unilateral), the carotid bulb was spared. Angiography of the five flame-shaped, somewhat pointed occlusions was done within 20 days after the onset of symptoms. Concomitant aneurysms were found in five. All 14 of the vertebral arteries investigated appeared to be normal.

Follow-up angiograms on 15 dissected arteries and five dissecting aneurysms (12 patients) revealed that the tapered stenosis resolved in nine, became less pronounced in two, progressed or were occluded in three, and persisted unchanged in one and that the dissecting aneurysm diminished in two, persisted as a minimal focal residuum in one, resolved completely in one, and remained unchanged in one. Somewhat paradoxically, a cervical carotid aneurysm in one patient evolved during the 4 month interval between examinations.

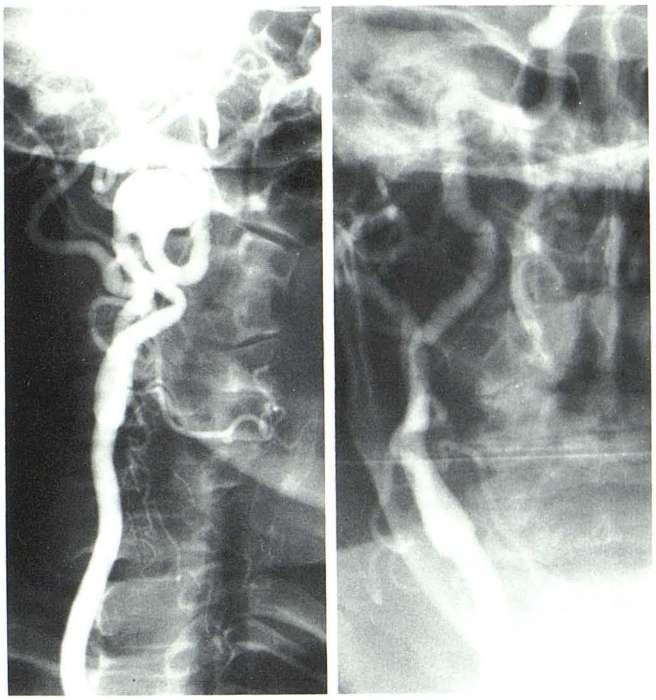
Dissection with Remote Luminal Irregularity

Of the 16 dissections associated with luminal irregularity, 12 occurred in women. The irregularity was noted in a verte-



A B

Fig. 4.—Fingerlike, ovoid dissecting aneurysm projects parallel to arterial lumen.



A B

Fig. 5.—Large dissecting aneurysm of distal cervical ICA with intimal flaps proximally. **A**, Preoperative frontal view. **B**, Postoperative lateral view demonstrates fibromuscular dysplasia more clearly.

bral artery of nine patients, the proximal part of a dissected ICA of five, the opposite ICA of one, and the common carotid artery of one. The irregularities were minimal, easily overlooked, and varied from serrations on one or more margins of the arterial lumen to divots in the lumen or even spiculation of the lumen in the distal common and proximal ICAs (fig. 1A).

In 11 ICAs, the dissection was unilateral. Five patients had multiple vessels dissected (both ICAs in two, both ICAs and the left vertebral artery in two, and one carotid and one vertebral artery in one). The lesion affected the cervical segment of seven ICAs; the cervical and intracanalicular segments of 11; and the cervical, intracanalicular, and cavernous segments of two. Eleven dissecting aneurysms occurred concurrently in eight ICAs and two vertebral arteries. Nine aneurysms were stituated within the dissected part of eight ICAs and one vertebral artery, while the aneurysm was the only evidence of dissection in one vertebral artery.

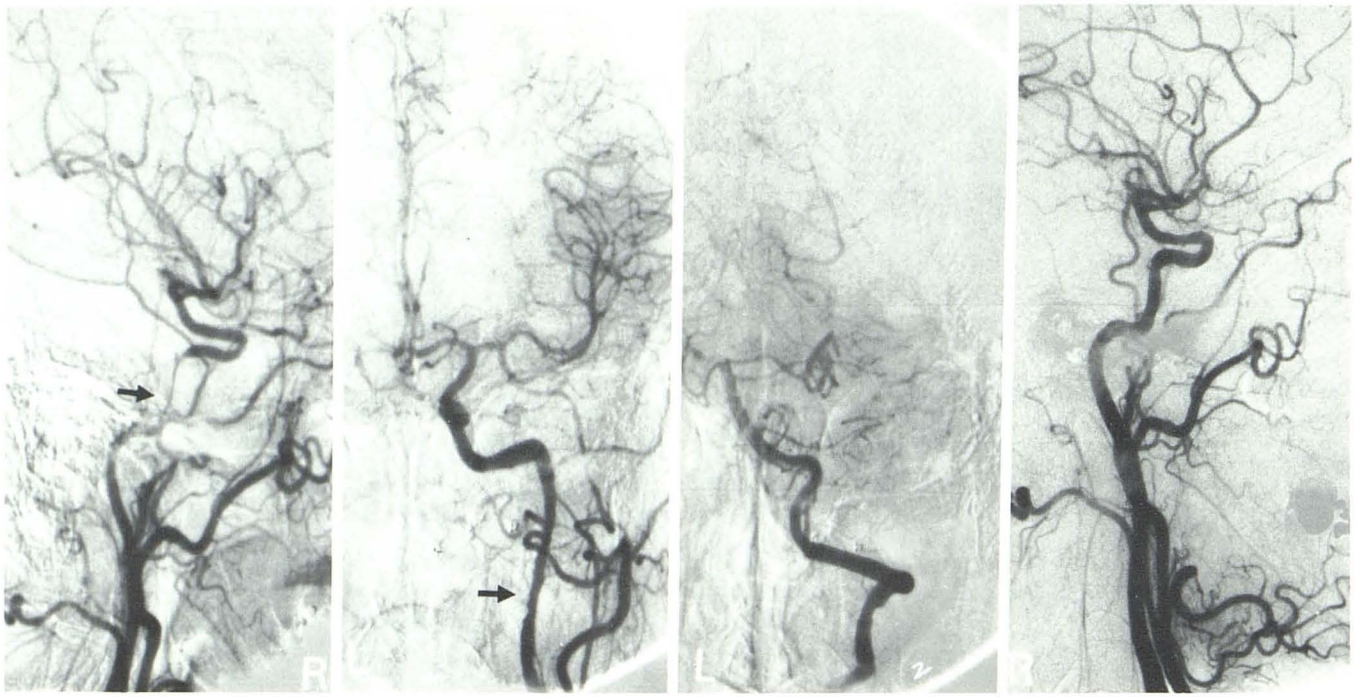
TABLE 5: **Follow-up Angiographic Findings in 29 Cases of Spontaneous Dissection and 16 Cases of Spontaneous Aneurysm of the Cervical Cephalic Arteries (24 Patients)**

Finding	Fibromuscular Dysplasia	Luminal Irregularity	Dissection Only
Spontaneous dissection:			
Resolved	2	6	9
Improved	1	3	2
Unchanged	1	0	1
Progressed to occlusion	0	1*	3
Total	4	10	15
Spontaneous aneurysm:			
Resolved	0	1	0
Focal residuum	0	1	1
Decreased	2	2	2
Unchanged	0	1	2
Surgically removed	3	0	0
Vessel occluded	0	0	1
Total	5	5	6

* Postoperative.

Fibromuscular Dysplasia

Five of the six patients with fibromuscular dysplasia were women. Fibromuscular dysplasia was noted angiographically in the renal arteries of four patients and in the ICAs of three patients (fig. 6); moreover, the diagnosis was confirmed pathologically in the resected aneurysms of two patients. The dissection affected a single vessel (ICA) in four patients and multiple vessels in two (both the ICA and the left vertebral artery in one and the right carotid and subclavian arteries in the other). The stenosis was limited to the cervical segment in two patients but extended from the cervical into the intracanalicular segment in two. Multiple, intermediate-sized aneurysms occurred in one patient, while a single large aneurysm occurred in the ICA of the other patient. Multiple aneurysms arose in a right subclavian artery. Two occlusions of the ICA were present, one at the origin and the other distal to the bulb. Two patients showed luminal irregularity of the ICA



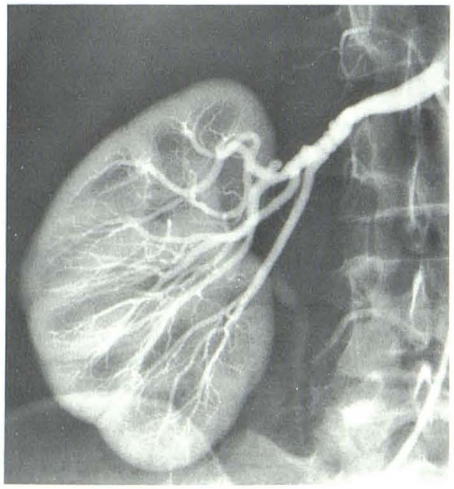


Fig. 6.—Arterial dissection in fibromuscular dysplasia. Dissection in bilateral ICAs (**A** and **B**, *arrows*) and left vertebral artery (**C**) with renal arterial fibromuscular dysplasia (**E**). **D** shows resolution of dissection in right ICA after 8 months.

resembling that noted in the preceding category of patients (fig. 3B). A vertebral (probable arterial) dissection resembling atheromatous disease occurred in a 40-year-old woman with classic dissections of both ICAs.

Discussion

Atherosclerosis [5] and fibromuscular dysplasia [6–8] are well known entities, but the radiologic, clinical, and pathologic features of hemorrhagic dissection [1–4, 9–31], including its residuum [1, 2, 24], have received less attention in the literature. Patients with spontaneous dissection differ from those with atherosclerosis or fibromuscular dysplasia without concurrent dissection because both of these disorders predominate in patients who are usually more than 50 years old, while patients with spontaneous dissection are often less than 50 years old [3, 4]. A definite gender predilection has yet to be confirmed, although O'Dwyer et al. [2] and Ehrenfeld and Wylie [3] have reported that the genders are affected equally. However, males predominated in the series of Fisher et al. [4] while two-thirds of our patients were women. Recently, Mokri (Mokri B, personal communication) described two clinical syndromes: oculosympathetic palsy (incomplete Horner) and hemicrania with or without cerebral ischemic symptoms in 62% of patients and hemicrania with delayed neurologic deficit in 58%. Both syndromes are suggestive of dissection.

Clinically and angiographically, spontaneous cervical cephalic arterial dissections appear to be acute vascular epi-

sodes that often resolve, sometimes progress, and seldom recur. The arterial site of predilection is the ICA in most patients, although dissections have been noted in vertebral [4, 12, 16] and common carotid arteries. These dissections, affecting the right ICA in 48% and the left ICA in 21% of our patients, usually involve a single vessel; however, multiple simultaneous dissections occurred in one-third of our patients, including both ICAs in 19%, both ICAs and a vertebral artery in 5%, an ICA and vertebral artery in 5%, and a single carotid and subclavian artery in 2%. Such simultaneous involvement is suggestive of a systemic disorder; furthermore, reports of coexistent intracranial dissections tend to support this concept [4, 9, 19]. Nevertheless, concurrent angiographic evidence of disease in other peripheral arteries has not been recorded nor was it noted in the present series, except for fibromuscular dysplasia.

In general, the angiographic findings in the present series, consisting of a tapered stenosis in 46%, tapered stenosis with a concomitant dissecting aneurysm in 27%, occlusion in 20%, and only a dissecting aneurysm in 7% of affected vessels, agree with those noted by others [1–4]. The angiographic hallmark of dissection occurring in the cervical segment of the ICA in 30%, the cervical and intracanalicular segments in 65%, and the cervical, intracanalicular, and cavernous segments in 5% is a long, tapered, usually eccentric stenosis that begins distal to the carotid bulb and is associated with irregularity of the lumen. This irregularity is more prominent along one surface of the vessel. The reconstitution of the lumen may be abrupt if the dissection ends at the level of or within the carotid canal. The configuration of dissecting aneurysms varies, depending on size. Round, saccular aneurysms either may be small (less than 1 cm) with a wide neck or less often large (more than 2–3 cm). Most aneurysms are intermediate in size (about 1 cm), ovoid, extend like a finger parallel to the vessel, and appear to be flattened along the surface that faces away from the artery. If the original intimal tear lies at the site of the aneurysm, the typical location of the aneurysm in the midcervical and subcranial parts supports the conclusion of Fisher et al. [4] that dissection originates in these regions. Nevertheless, we cannot exclude the possibility that the buttressing effect of the bony carotid canal could act like a dam, which might force the blood through the media to form an aneurysm before its rupture through the intima, where it reenters the lumen of the ICA. A tapered occlusion that spares the carotid bulb (which occurred in 80% of our patients) is typical of dissection, but it may be mimicked by other conditions.

The angiographic appearance of arterial dissection depends on its severity and extent and the temporal interval between its onset and angiography. Stenotic dissections tend to resolve, although resolution in our series did not match the 80% described by Quisling et al. [1]. In our series, the stenosis resolved in 59% of patients, improved in 20%, progressed to occlusion in 14%, and remained unchanged in 7%. Dissecting aneurysms behave somewhat similarly, but the improvement is not as striking. In our series, the aneurysm resolved completely or with minimal focal dilatation in 25% of patients, diminished in 50%, and remained unchanged in 25%. The evolution of an aneurysm, like that noted in two of our patients, must be unusual, although O'Dwyer et al. [2] documented this phenomenon angiographically in a dissected cervical ICA. The development of a similar aneurysm in the cavernous ICA at the termination of an extensive dissection is unexpected, but probably coincidental; however, an intracranial vertebral arterial aneurysm was visualized on the initial angiogram of another patient in our series.

As the clinical syndromes and angiographic findings of the dissection are better recognized, the role of angiography in the diagnosis and treatment of cervical cephalic arterial dissection is likely to expand, since angiographic depiction of intracranial emboli (15% of our patients), the routes of collateral circulation, and ICA–middle cerebral artery slow flow may influence treatment. Currently, it seems reasonable to recommend angiography for patients whose clinical syndromes are typically associated with dissection, since residual intimal flaps or cervical arterial aneurysms were found in about one-fourth of our patients when angiography was not done soon after clinical onset. Similarly, follow-up angiography of acute dissection should be considered, because an occlusion or an aneurysm may develop within the dissected segment (almost 15% of our patients).

Finally, angiographic categorization of patients with spontaneous dissection into those with and without fibromuscular dysplasia can be substantiated pathologically; however, pathologic confirmation of other remote intimal irregularities that we previously termed "idiopathic regressing arteriopathy" [31] was not possible. Wirth et al. [8] described a single similar irregularity that proved to be a form of fibromuscular dysplasia. Nevertheless, patients whose angiograms reveal intimal irregularities that are not typical of fibromuscular dysplasia are more likely to be female, less likely to have occlusion of the ICA, and more likely to have involvement of the vertebral artery when compared with those whose only angiographic finding is that of dissection. Until the pathologic findings are available, we can only postulate, despite having many reservations, that remote intimal irregularities represent a forme fruste of fibromuscular dysplasia. Regardless of the cause, we agree that the incidence of spontaneous dissection is significant in patients who present with unilateral headaches, oculosympathetic palsy, or ischemic neurologic symptoms and are less than 50 years of age.

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