

Spontaneous Dissection of Cervico-Cerebral Arteries

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SUMMARY: *Sixteen cases of spontaneous dissection of the cervical internal carotid artery (6 verified) are described. The mean age was 45 years. The clinical picture varied from simply headache and a bruit to hemiplegia and aphasia. Eleven patients had transient ischemic attacks. Headache, facial pain, a subjective bruit, oculo-sympathetic palsy and transient monocular blindness were present in various combinations in two-thirds of cases and their presence suggested the correct diagnosis. Examples of suspected dissection of the intracranial internal carotid, middle cerebral, posterior cerebral and extracranial vertebral arteries are also presented. Spontaneous dissection is more common than the literature indicates.*

RÉSUMÉ: *Nous décrivons 16 cas de dissection "spontanée" de l'artère carotide interne dans sa portion cervicale, dont 6 vérifiés anatomiquement. L'âge moyen des patients était de 45 ans. L'histoire clinique allait de la simple céphalée ou du bruit à l'hémiplégie avec aphasie. 11 patients eurent des épisodes vasculaires transitoires. Chez près de 2/3 des patients, mais en combinaisons variables, on retrouvait les symptômes et signes suivants qui peuvent orienter le diagnostic: céphalée, douleur faciale, bruit subjectif, paralysie oculo-sympathique et cécité monoculaire transitoire. Nous présentons également d'autres exemples de dissection possible de la carotide interne dans sa portion intracrânienne, des cérébrales moyennes et postérieures et finalement des artères vertébrales extra-crâniennes. La dissection spontanée est donc plus fréquente que ne l'indique la littérature.*

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In a previous paper on the surgical treatment of a single case of spontaneous carotid dissection (Ojemann et al., 1972) it was pointed out that the long stenotic segment of the internal carotid artery (ICA) visualized angiographically — the string sign — might be a reliable indicator of carotid dissection. In the following year, using the sign we were able to diagnose preoperatively two proven cases of non-traumatic carotid dissection. It became apparent that spontaneous dissection was not as rare as the nine clinical cases reported in the literature of the past twenty years would indicate. Because of the great length of the stenotic lesion in the carotid or the presence of total occlusion, surgery usually would not be undertaken, with the result that the nature of the disease generally remained unknown, and recognition of the frequency of the process was delayed. The string sign also was noted in a case of spontaneous dissection of the middle cerebral artery (Hochberg et al., 1975). We now have had 16 cases in which angiography has demonstrated a long, narrow column of contrast material which differs markedly from the typical short, stenotic lesion of atherosclerosis. The total number of cases in which the diagnosis of dissection has been made is 22 (See Table) and, although only 6 cases have been proven by surgical exploration or autopsy, 12 others appear to be certain candidates for the diagnosis of dissection while 4 cases are included as highly suspect. Some of the puzzling cases of the past are finding a diagnostic niche. Our experience may be a stimulus to further investigation of the diagnosis, incidence, treatment, and pathology of this disease. The term non-traumatic is used to indicate that no obvious precipitating

injury was recognized and is not intended to exclude minor or casual stresses and strains.

CLINICAL MATERIAL

Angiography

Because the diagnosis, in most of our cases, rested on the radiologic interpretation, it is necessary to establish the reliability of the clinico-radiologic correlation. Of the 11 cases of spontaneous dissection of the ICA (Ojemann et al., 1972), 5 that were surgically or pathologically verified showed a long, irregular filling defect which started 2 cm. above the bifurcation and extended throughout the extracranial course of the ICA. Four of the 6 verified cases in the present series also had the string sign. This distinctive appearance, the result of the dissecting hemorrhage compressing the natural lumen, rarely occurs with other types of cerebrovascular disease such as atheroma, fibromuscular dysplasia, arteritis, moyamoya and vasospasm. It has never been reported in a pathologically proven case of any of these other diseases and is therefore probably a reliable diagnostic finding. These long dissections usually extend only to the base of the skull without entering the petrous canal, but in our case 13 the intracranial precavernous segment was narrow and showed scalloped borders.

In some cases of dissection, both in the literature and this series, the ICA has been totally occluded. In all of these the occlusion has spared the carotid sinus and begun 2 cm. or more distal to the origin of the ICA. The patent segment gradually tapers to end in occlusion. At operation or autopsy the region of the sinus is soft to palpation and without evidence of atheroma (Figure 1). Intracranial occlusion of the distal ICA from any

cause may be associated with retrograde clotting extending into the neck which can mimic the tapered post-sinus occlusion of dissection but usually, in the acute stage, the tapering segment reaches much further distally than in dissection. In a tapered post-sinus occlusion there is transorbital collateral flow by way of the external carotid artery. If reflux occurs from the intracranial carotid downwards into the petrous portion or below, the evidence for dissection is strengthened.

Another distinctive type of carotid dissection takes the form of a localized aneurysmal sac or out-pouching of the cervical portion of the artery at the level between C1 and the base of the skull (Figure 1). The case of Hardin et al. (1964) was of this type and was proved surgically. Bostrom and Lilliequist (1967) described an asymptomatic aneurysm of the upper carotid which enlarged from 10 x 3 mm. to 12 x 7 mm., demonstrated by angiograms made one month apart. At autopsy four months later the lesion consisted of a blood-filled 1.5 cm. cavity which communicated with the arterial lumen by a smooth-walled neck 2 mm. wide. The wall of the carotid showed evidence of cystic medial necrosis. In traumatic carotid dissection similar pouches in the distal carotid have been described, although morphological proof of the diagnosis was not provided (Sullivan et al., 1973). From the available evidence one can be reasonably certain that an upper carotid outpouching is a tell-tale of dissection. Long carotid dissections may be combined with short dissection pouches.

Another angiographic appearance of dissection is illustrated in our case 7. When first studied because of recent transient ischemic attacks (TIAs), there was a typical carotid string sign 7 cm. long, but surgery was not undertaken. Eight days later another angiogram revealed that the residual lumen had widened from about 1 mm. to 2 mm. A third angiogram 7 months later showed full restoration of the lumen (Figure 1). At the proximal end of the former segment of narrowing, there was an oval-shaped intramural sac or pouch

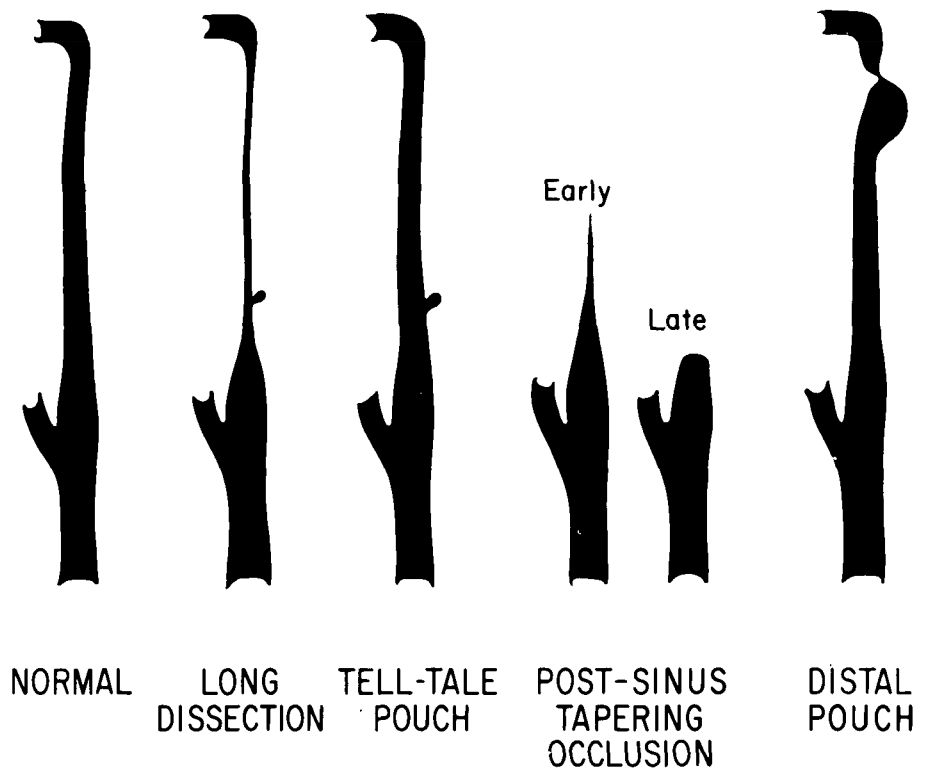


Figure 1—Diagrams of angiographic profiles in carotid dissection.

measuring 10 x 4 mm. Possibly, the occasional puzzling, small sac of this type represents a healed dissection. When a long dissection clot is undergoing absorption, the widening lumen may have scalloped or undulating margins.

In a case of pathologically proven middle cerebral dissection (Hochberg et al., 1975) the angiogram showed an elongated, thread-like residual lumen, indicating that the string sign applies also to this artery as well. In our case 20, a boy aged 13 with an acute stroke showed a typical filiform residual lumen but the diagnosis remained unverified.

Our experience with the angiographic profiles of dissection, particularly the string sign, has led us to suspect that cases of atypical stroke at other sites in which the involved artery shows a long, stenotic residual lumen also are the result of dissection. Examples are case 15, intracranial ICA; case 19, posterior cerebral artery; and case 20, the cervical vertebral artery.

CASE MATERIAL

Case 1. A hypertensive man, aged 59. one hour before admission, while eating, felt dizzy

and his left hand and left side of the face became weak and numb. Speech was slurred and there was a headache. Gradual improvement occurred but two hours later the deficit returned to its original severity. Examination showed a left facial weakness and almost total paralysis of extension, abduction and adduction of the left fingers with preservation of the hand grip. The left leg was not affected. Sensation was approximately normal. Improvement gradually occurred. An angiogram on the sixteenth hospital day showed a long segment of severe internal carotid stenosis extending from 4 cm. above the origin up to the base of the skull (Figure 2). The 1.5 cm. of artery proximal to the severe stenosis tapered gradually. On the eighteenth day the patient had another episode of numbness and weakness of the left face and left hand. Surgical exploration was undertaken and the ICA was enlarged and blue. A Fogarty catheter could not be inserted distal to the dissection. On aspirin therapy recovery was excellent and there was no recurrence in the following two years.

Case 2. A woman, aged 45, five days before admission experienced a 2-minute episode of scintillations, like bright snowflakes in her left eye. Three days before admission she developed prickling numbness of the right face, arm and leg and expressive dysphasia lasting about 40 minutes. One day before admission there was an episode of heaviness of the right arm and dysphasia and a recurrence of this on the day of admission. She was given heparin intravenously and examination 6 hours later showed normal neurological findings except for occasional hesitancy in speech. During the

examination she developed paraphasic errors, was unable to identify objects in the right hand, and pinprick was slightly decreased on the right side. There was slight weakness of the right side of the face and the right biceps. Ten minutes later the examination was again normal. The blood pressure was 120/70. A left carotid angiogram showed a long narrow column of contrast extending from 2 cm. distal to the origin of the internal carotid artery to the base of the skull. Surgery was undertaken immediately and showed the internal carotid artery enlarged to 2 or 3 times normal size and bluish in color. The artery was opened, revealing a dissection of the intima. Using a Fogarty catheter, the dissection clot and the elevated intima were completely stripped and removed and a good back-flow of blood was obtained. Two days postoperatively, she experienced brief tingling in the right hand and forearm. On the third day postoperatively there was tingling of the right face, arm and thigh. A carotid angiogram one week post-operative showed the artery widely patent and larger than normal.

Case 3. A man, aged 41, two days before admission while in the bathroom, suddenly felt weak and transiently a film obscured vision in his left eye. There may have been two previous spells of transient left monocular blindness. He was well until next day when, after breakfast, he developed a right hemiparesis which improved in 30 minutes. On the morning of the day of admission, he awakened with a right hemiplegia and aphasia. There was no history of trauma. On examination he was mute and had a complete right hemiplegia, right-sided sensory loss and right homonymous hemianopia. The blood pressure was 120/80. A left carotid angiogram showed a long irregular narrowing of the internal carotid artery starting 1 cm. distal to the carotid bifurcation and extending to the base of the skull. At the proximal end there was an irregular dilatation where the ICA was kinked acutely. At operation, the ICA was dilated and blue throughout its length. The proximal part of the dissection was resected and a Fogarty catheter was inserted, inflated and retracted, stripping the dissection clot and the elevated intima from the artery. Recovery was satisfactory. Pathological examination confirmed the presence of a dissection and showed irregularity of the media without cystic necrosis. This case was previously reported in detail by Ojemann et al. in 1972.

Case 4. A woman, aged 45, six days prior to admission on straightening up after bending, felt dizzy and strange. She ran downstairs, began to experience bright jagged lights in the left visual field, staggered and sat down. Within a minute she was aware of a low pitched buzz in the right ear, synchronous with the pulse. The flashing lights lasted 10 minutes and the buzzing about 6 hours. The pulsatile buzz returned the following day. Two days before admission, on awakening in the morning, she had the same strange feeling in the head. The day prior to admission, while seated at 10:00 a.m., the left fingers and hand tingled and the arm could not be lifted. This lasted 10 minutes. Four hours later there was

another similar spell with visual scintillations. On the day of admission she awakened at 2:00 a.m. with another spell and had another at 2:00 p.m. Examination on admission was normal. The central retinal artery pressure was 22 on the right, 45 on the left. There was a long, low pitched murmur over the right ICA. Three days after admission the patient had an episode of numbness and weakness of the left face, arm and leg and blurred vision in the left visual field. On the fourth hospital day the sound in the right ear disappeared and there was another episode of tingling of the left cheek and jagged lights in the left eye. On the eighth hospital day there were two further similar spells and recovery was no longer perfect. On the ninth hospital day the left face, arm and leg tingled and felt cold and the patient was blind in the left visual field. On the tenth hospital day two similar spells left her with a left hemiparesis and left visual field deficit. A right carotid angiogram showed complete occlusion of the right ICA 2 cm. from its origin. At operation the proximal 8 cm. of the ICA were dilated to about twice normal diameter and there was no palpable atheroma. Pulsation could be felt along the entire ICA. A blunt tipped wire corkscrew inserted in the ICA, encountered obstruction 1.5 cm from the origin. On rotation of the instrument old black blood issued, followed by bright red blood. The patient remained hemiplegic postoperatively and another angiogram showed a long narrow segment of the ICA extending from near the origin to the base of the skull. Pathologic examination showed the typical picture of dissection in the outer media of the ICA. There was no atheroma or cystic medial necrosis. It is possible that the dissection was the result of the surgical procedure, but the pathologic findings were typical of spontaneous dissection.

Case 5. A man, aged 45, 12 hours before admission had a brief episode of blurred vision. On the morning of admission he fell while shaving and was found lying on the floor speechless and with a right hemiplegia. On admission he showed a right hemiparesis and was mute except for yes and no, but could comprehend almost fully. Ophthalmodynamometry was 75 on the right and 35 on the left. Angiography showed a long irregular segment of stenosis of the left cervical ICA, beginning 4 cm. distal to the origin. At the upper end of the narrowing near the base of the skull there was a dissection sac approximately twice the width of the artery. Contrast passed slowly along the stenotic segment. At operation (Dr. E. H. Tarlov) a Fogarty catheter was used to remove a large clot obstructing the ICA but intima was not identified. The patient, who had begun to improve prior to operation, continued to recover and restoration was finally almost complete. The patient later recalled that on the day before the hemiplegia there had been several brief spells of visual disturbance whose nature he could not remember. For 10 days before the paralysis he had a viral illness which was epidemic at the time and was subject to an extremely persistent, almost incessant non-productive cough with gagging and near-vomiting.

Case 6. A man, aged 67, one day before admission experienced weakness and buckling of the left leg for 10 minutes. On the morning of admission the left hand was weak and clumsy for an unstated period. Three hours later the left hand became weak and numb. Examination showed slight weakness of the left hand, arm and leg, extinction of the left visual field, slight dystereognosis and constructional dyspraxia. The blood pressure was 130/70. The patient was subject to a severe chronic cough. A carotid angiogram showed the right ICA tapering to complete occlusion 3.5 cm. distal to the origin (Figure 3). Collateral flow via the ophthalmic artery filled the ICA retrogradely to the base of the skull. At emergency surgical exploration there was no disease of the carotid sinus region. A satisfactory back-flow could not be obtained using a Fogarty catheter. A second carotid angiogram 5 days later again showed collateral flow via the ophthalmic artery and retrograde filling into the distal petrous carotid artery, evidence of primary occlusion of the cervical artery.

Case 7. A man, aged 45, 6 years before had a subarachnoid hemorrhage which arose from an aneurysm at the right carotid bifurcation intracranially. Angiography showed three aneurysms on the right side and two on the left and the right common carotid artery (CCA) was ligated. The patient was well until one week prior to admission when during an influenza epidemic he developed a left-sided headache, chiefly above the eye but also on the side of the head and side of the nose. For one week he also had faint intermittent shimmering scintillations in the left visual field, less definite than the migraine scintillations he was subject to. From the onset of the headache the left upper lid drooped slightly and the left pupil was smaller. Everything tasted unusually sweet. For three days at the onset the patient had a pulsation deep in the left ear. This disappeared for a few days, but then returned. On the day of admission there was a 15 minute spell of numbness of the right fingers, slurred speech and sagging of the right side of the face. On admission neurological examination was normal except for a left oculo-sympathetic palsy. On the day after admission he had a further similar spell lasting 25 minutes and on day three, three more spells. The central retinal artery pressure was 60 on the right, 30 on the left. A left carotid angiogram showed a string sign extending from 3 cm. distal to the bifurcation up to the base of the skull (Figure 4A). The patient was dysphasic for 24 hours after the angiogram. Heparin was started and there were no further spells. Eight days later, repeat angiograms showed that the lumen had widened from about 1 mm. to 2 mm. and an oval-shaped intramural sac 10 mm. x 4 mm. had formed at the proximal end of the dissection (Figure 4B). He remained well and the central retinal artery pressure returned to normal. A third angiogram 7 months later showed full restoration of the lumen with a tell-tale pouch at the origin.

Case 8. A woman, aged 46, while in the throes of an acute upper respiratory infection

associated with an unusually severe persistent cough, developed a superficial, non-throbbing pain and soreness in the left posterior frontal region high on the side of the head. Superimposed on the persistent pain were jabs of severe pain which occurred every 10 minutes and caused the patient to wince. The hair in the area was sensitive to comb. At the same time she noticed a pulsatile sound in the left ear, like an air-pump. Pain, which seemed related to the parietal pain, developed deep in the left upper neck posteriorly just behind the insertion of the sternocleidomastoid muscle. It was not related to head movements. She noted that the left eye was half-closed due to drooping of the upper lid and on examination left miosis was noted. A left carotid angiogram showed a sac 1.5 cm. vertically and 2.5 mm. wide which protruded posteriorly from the ICA between C1 and the base of the skull and also bulged forward to narrow the lumen to about 2.5 mm. The sac was rounded and shaped like a pharmaceutical capsule or bullet (Figure 5). Angiography one year later showed the sac about the same size but the arterial lumen was no longer narrowed. Another study after 15 months showed no change. The head pain persisted on and off at about the same severity during the next 14 months. The bruit disappeared in the 5th month. The pain in the upper neck lasted 2 months, disappeared for 8 months, then returned and was still present on and off 16 months later. It was then pulsatile and could be evoked by exertion. Ptosis and miosis lessened but were evident after 28 months.

Case 9. A woman, aged 43, for one year often felt dizzy, nauseated and faint on arising in the morning. Associated with this, there was frequently a left-sided pounding headache that extended to involve the teeth and back of the neck. Resting for one to two days often gave relief. At about this time the patient began to have an intermittent pulsating, sh-sh-sh, sound in the right ear. Two months before admission she experienced white punctate, bright silver sparkles in both visual fields for 15 minutes. The left-sided headache became more persistent and dizziness and faintness more frequent. The bruit lasted a few hours at a time. A bruit heard in the right side of the neck was attributed to a venous hum. Three weeks prior to admission she developed a central scotoma in the right eye only, associated with silver sparks, all in 15 to 30 minutes. Because of the continuation of headache and wooziness, four-vessel angiography was undertaken. The right ICA 3 cm. distal to its origin was narrowed to 2 mm. by a lateral filling defect; the left ICA showed a long segment of severe narrowing reaching to the base of the skull, the right vertebral artery (VA) was severely stenosed 2 cm. from its junction with the basilar artery and the left VA was entirely occluded just proximal to the origin of the atlantic branch. The nature of the four-vessel vascular disease remained obscure, but the angiographic picture in both ICAs was consistent with dissection.

Case 10. A man, aged 31, suddenly had difficulty in getting his words out, uttering

only hesitant monosyllables, and the right hand and fingers became weak and numb. A mild left-sided headache had been present for 1½ hours. The deficit had almost cleared completely in the next 3 hours when the patient fell asleep. On awakening the next morning the speech deficit and weakness were as bad as at the onset or worse. The right leg was numb. Examination showed non-fluent expressive aphasia, difficulty in comprehending complex speech and a right central facial weakness. The blood pressure was 120/62. A left carotid angiogram resulted in a subintimal injection. In the next 18 days the patient gradually improved but then for 10 minutes he had scintillations in the left inferior quadrant of both eyes as if looking through broken glass. Letters in the same quadrant appeared upside down. He had migraine headaches since the age of 10. An arch aortogram 14 days from the onset showed slight stenosis of the left ICA, thought to be insignificant hemodynamically. A selective catheter angiogram of the left carotid 3 days later showed 3 regions of narrowing in the ICA, the first at the entrance to the carotid canal, the second immediately within the skull and the third in the precavernous siphon. The abnormal regions appeared to be dissections. Another left carotid angiogram 1 week later showed two small 3 mm. collections of contrast material medial and anterior to the left ICA, just proximal to the entrance into the carotid canal of the petrous bone. These lesions were consistent with a local dissection. No generalized disease was discovered. The sedimentation rate was 8 mm. and no source of embolism was found. The serum lipids and the CSF were normal. The patient continued to recover and one year later speech was almost normal.

Case 11. A hypertensive man, aged 52, four weeks before admission noted profuse tearing of the right eye followed by machinery-like noises in both ears, greater on the right. The following evening the scalp on the right side was tender and throbbing and there was a mild, deeper headache. The right eyelid drooped. The tearing, discomfort and bruit fluctuated or disappeared intermittently for the next 10 days and then ceased. The scalp on the right side was hyperesthetic and combing the hair was painful. Eight days prior to admission there was a 5 minute episode of tingling and clumsiness of the left forearm and hand. Five days before admission a similar spell involved the entire left arm. Two days before admission the prickling involved the entire left arm and left leg for several minutes.

On examination the only neurological abnormalities were smallness of the right pupil by 1 mm., right-sided ptosis and a slight drift of the left arm. The blood pressure was 160/100. A right carotid angiogram showed a rounded 8 mm. aneurysmal sac in the segment between C1 and the base of the skull. Immediately distal to the sac, the carotid artery was narrowed acutely to less than 1 mm. in diameter. The choroidal blush appeared at 6 seconds (normal 3.5). The patient was placed on antiplatelet aggregants. Six months later there was a 7 minute episode of transient

blindness in the right eye. Angiography showed the dissection unchanged.

Case 12. A woman, aged 29, one evening while reading, developed a complete right hemiplegia and speechlessness. After 5 minutes aphasic utterances began to emerge and within 45 minutes full recovery had occurred. She had been receiving oral contraceptives for about 15 months. For 4 days before the episode she had a frontal headache slightly to the left of the midline. A left carotid angiogram showed a long segment of severe narrowing in the left ICA extending to the base of the skull. The residual lumen was about 1.5 mm. The opposite carotid system was normal.

Case 13. A man, aged 74, had a 15 minute episode of numbness and weakness of the left face and arm and slurred speech. Four months later a right carotid angiogram showed a long segment of severe stenosis extending from the origin of the right ICA to the base of the skull. The residual lumen was about 2 mm. At the proximal end of the narrowed column there was a 5 mm. tell-tale outpouching or dissection sac. The precavernous ICA showed the irregular scalloped outline described in some cases of post-traumatic dissection. A left carotid angiogram opacified the right anterior cerebral artery and contrast passed through the border zone anastomoses to the right middle cerebral territory.

Case 14. A diabetic obese woman of 45 within one day experienced a sudden change in personality. She became apathetic, slowed, vague, forgetful, late and neglectful. She had no insight. She was thought to be depressed. On driving her auto she veered across the center line to the left and went off the left side of the road. She reported 3 or 4 episodes of numbness of the fingers and hand on the left side. In the previous year she had noted a pulsating bruit in the head. She was subject to migraine headaches. Angiography showed a long segment of stenosis in the cervical portion of the right ICA extending to the base of the skull with restoration of the normal lumen in the petrous bone. The supraclinoid ICA was totally occluded. The appearance was identical with cases of proven dissection. The nature of the distal occlusion was obscure. After remaining in the above condition for 6 weeks the patient returned to normal.

Case 15. A man, aged 53, noted that light coming from the left side was too bright. A few hours later on bending to tie his shoe he heard a sh-sh-sh- sound in the left ear. The next day he noted the left palpebral fissure was narrowed and the left pupil smaller. There was a mild ache in the left side of the face involving the eyeball, forehead, cheek, and upper teeth. There was a bitter taste on the left side of the tongue affecting all kinds of food. Smell was not affected. Gradual improvement occurred and at the time of examination 6 weeks later only a slight pain persisted deep in the orbital region and above the eye. The face was almost normal. The head noise had become more prominent. For one

week there had been fluttering in the lower visual fields "like looking at a fan". The patient had had hypertension for 5 years. There was a history of a single migrainous spell with visual aura 6 years before.

A striking feature of the history was that 4 years before, he had had a similar syndrome involving the right side of the head — ptosis, miosis, dysgeusia and intense headache. All cleared in about one week. There was no bruit at that time.

Neurological examination was normal except for the presence of a slightly smaller pupil on the left and a high-pitched bruit on the left side over the mastoid process, cheek, forehead and eye. The bruit ceased with carotid compression. The blood pressure was 170/95. The central retinal artery pressure was 70 on the right, 60 on the left. A left carotid angiogram showed the ICA smoothly narrowed to a width of 1.5 to 2 mm. in the 2 to 3 cm. before entering the petrous canal.

Case 16. A man, aged 45, awakened one morning with a persistent left frontal and retro-orbital headache. Ten days later left-sided ptosis, miosis and conjunctival redness were noted. Angiography on day 14 showed a string sign in the upper half of the left ICA in the neck. The patient noted a bruit in the left ear at night or when he was quiet. The eye signs and headache disappeared at the end of 4 weeks. There were no TIAs.

Case 17. A man, aged 68, 24 hours before admission had several 10-minute episodes of weakness of the right hand and arm and slurred speech. Neurological examination was normal. The blood pressure was 170/90. A left carotid angiogram showed the ICA narrowed to a thread for 1 cm. proximal to the anterior clinoid process. Heparin was given intravenously but in the next 24 hours there were several further episodes of loss of speech, weakness of the right hand and arm and right homonymous hemianopia. After 24 hours there were no further spells. The column of contrast tapered gently toward the distal end.

Case 18. A man, aged 45, while recovering from an acute upper respiratory infection, developed paralysis of the left side of the tongue and a loud pulsating gushing sound in both ears that was audible to people at a distance of 2 feet. The bruit varied from a swish to a squeak. There was also pain in the paraspinal region of the upper cervical region. When first examined 9 weeks later the left side of the tongue was weak and shrivelled. The noise in the head had disappeared except at times of excitement. It was not audible on examination. The blood pressure was 190/120. An angiogram was not made, and no follow-up information was obtained. It is postulated that carotid dissection involved the hypoglossal nerve which is applied to the posterior aspect of the carotid artery.

Case 19. A girl, aged 15, 3 hours before admission complained of nausea, right-sided headache, blurred vision and difficulty seeing to the left. One half hour before admission she

made an ordinary dive into a swimming pool. Five minutes later she complained of nausea, right-sided headache, staggered and fell. On examination there was a severe left hemiparesis with anosognosia. On the second hospital day a right carotid angiogram showed the right middle cerebral artery narrowed to a hair-line over a distance of 2 cm. with slowed circulation distally. Anticoagulation with heparin and warfarin was begun. On the thirteenth hospital day the patient became comatose and died 12 hours later. Pathologic examination showed a 3 x 3 cm. hematoma in the right frontal lobe in the region of a 2-week-old infarction. Serial sections of the right middle cerebral, anterior cerebral and terminal ICA showed a typical dissection between the elastica and the media. This case has been reported previously by Hochberg et al (1975).

Case 20. A boy, aged 13, the night before admission developed a bad headache after playing ice hockey. One hour before admission, while walking on the street, he said he felt weak but continued to walk for another 10 minutes when he developed right facial weakness and dysphasia. Examination showed right facial weakness and fluent dysphasia. The CSF was normal. In the following 12 hours the right facial weakness disappeared and the dysphasia improved. At 1:30 p.m. the day after admission the patient developed a complete right hemiplegia and became mute. Angiography disclosed narrowing of the left middle cerebral artery for a distance of 8 mm. from its origin (Figure 6). The lumen was narrowed to a hair-line but further distally the middle cerebral artery widened to approximately normal diameter. A right internal carotid angiogram filled the left anterior cerebral artery and generous collateral flow passed through the border zone anastomoses to fill the superior division of the left middle cerebral artery.

Case 21. A youth, aged 18, suddenly developed pain in the right side of the neck posteriorly at the C3 level as if the neck was stiff. In the following 5 minutes numbness appeared over the left side of his body. At the onset there were shimmering lights before his eyes for a few seconds. On arrival in the emergency department 40 minutes later there was tingling in the entire left side from the top of the head to the sole of the foot. The patient knew that he did not see to the left and examination showed a left homonymous hemianopia. The left limbs were of approximately normal power and there was no headache, speech difficulty or confusion. An angiogram after 14 hours showed a severely narrowed segment in the right posterior cerebral artery beginning at about the origin of the posterior communicating artery and extending 2.5 cm. distally. At the distal end the lumen was narrowed to less than 0.5 mm. in diameter (Figure 7). After one month the left homonymous hemianopia persisted but sensation on the left side had returned and only a slight residual tingling remained. Another angiogram after 3 months showed no change in the configuration of the stenotic lesion. The

long segment of narrowing led to the tentative conclusion that dissection had occurred. The patient had engaged in a strenuous basketball game 24 hours before the onset of the illness.

Case 22. A man, aged 35, while vigorously scolding his son, felt something snap on the right side of the upper neck posteriorly. He immediately felt weak, lost his equilibrium, crawled to an adjoining room and lay on a bed. He immediately noted diplopia, burning of the eyes, extension of the headache forward to the back of the eye, numbness of the left arm, right arm and right leg, dysphagia and incoordination of the right arm and leg. He was unable to sit up because of imbalance. Examination showed a right lateral medullary syndrome. An angiogram on day four showed a 3-4 cm. elongated narrowing of the right vertebral artery from C1 to C3 (Figure 8). The signs and symptoms cleared in about 2 weeks and another angiogram after 23 days showed the vessel lumen restored to normal. The angiographic appearance was unusual for embolism. Vasospasm was another possibility, but in the absence of evidence for such an entity, dissection was suspected.

Discussion

The Clinical Manifestations —Carotid Cases

When the cases in which the diagnosis of dissection was definite or highly likely (cases 1 to 16) are analysed, the clinical picture is different from the literature where 6 of 9 patients presented with a hemiplegia. This difference might be anticipated since, except through surgical exploration, criteria for the diagnosis during life have not been available previously. At the time of initially consulting a physician, only 3 of the 16 present patients were hemiplegic or aphasic. Four had a slight hemiparesis or monoparesis, 6 had a normal neurological examination except for an oculo-sympathetic palsy (OSP) and in 3 the examination was entirely normal. The ages were 29, 31, 41, 43, 45 (six patients), 46, 52, 53, 59, 57, and 74, on the average much younger than in atherosclerotic stroke cases. Ten were male, six were female.

The initial symptoms in the 16 cases were: TIAs 5; transient monocular blindness, TIAs and hemiparesis 3; headache and bruit 3; headache, bruit and TIAs 1; headache and aphasia 1; photophobia, bruit and dysgeusia 1; tearing, bruit, headache and TIAs 1; and scintillation, bruit and TIAs 1.

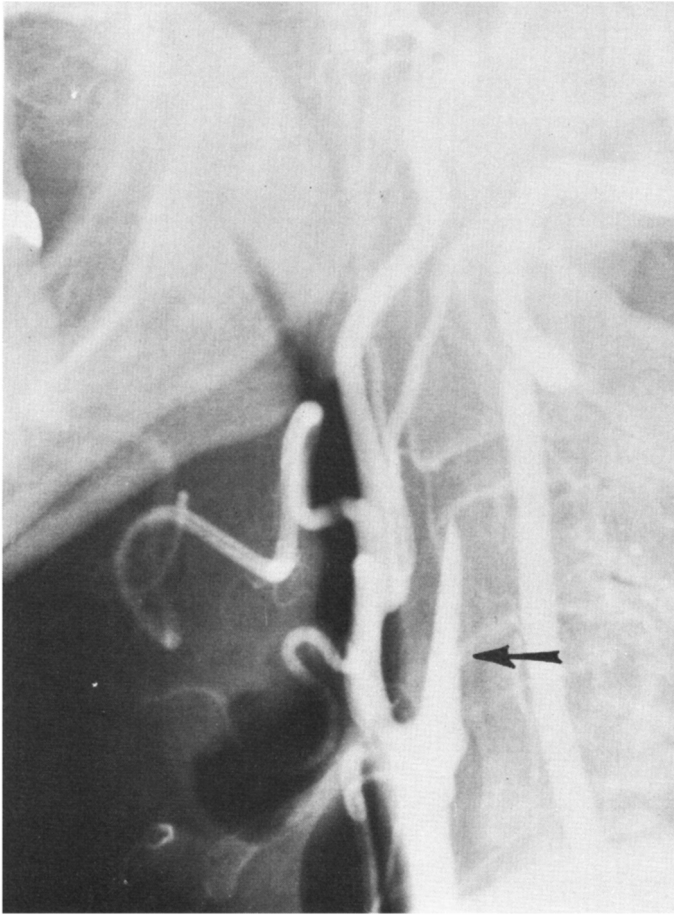


Figure 2—Angiogram in case 1.

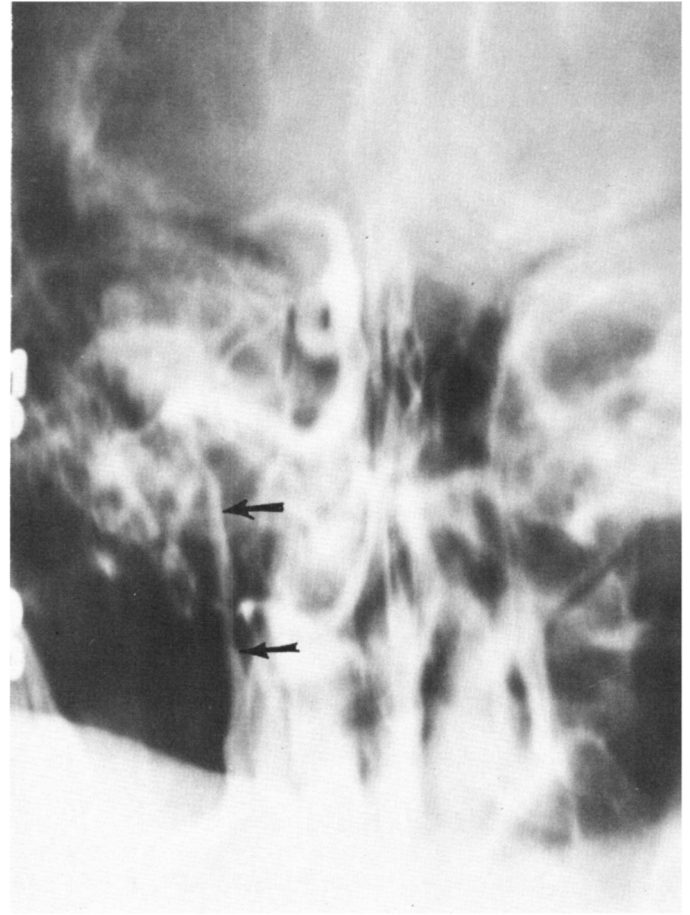


Figure 3—Angiogram in case 6.

TIA's occurred in 11 patients and consisted of various combinations of transient monocular blindness (TMB), scintillations, weakness, paresthesias or dysphasia. The estimated number of TIA's in the 11 cases was 1 (3 cases), 2 (3 cases), and 3, 4, 5, 6 and 13 respectively. Two cases had TIA's superimposed on a stroke that temporarily improved and 3 cases had no TIA's. In no case did a persistent deficit occur without a TIA or prodrome of some kind.

Headache or facial pain was present in 10 cases, a subjective bruit in 8 cases (in 3 it was objective also), oculo-sympathetic palsy (OSP) in 5 cases, TMB in 3 cases and scintillations or photophobia in 5 cases. In 14 of the 16 cases, one or more of these manifestations was present in different combinations: 2 cases had 4 of the 5, 4 cases 3, 3 cases 2 and 5 cases 1. In conventional athero-thrombotic carotid disease, these 5

manifestations may occur but, save for TMB, they are not common and usually occur only singly. An analysis of 25 personally verified atherosclerotic cases showed 8 (32%) with one of the above symptoms, 7 with TMB and one with a headache which was intermittent. Thus, approximately $\frac{2}{3}$ of cases had symptoms and signs suggesting dissection of the carotid artery rather than atherosclerosis. Many of the same features were present in the 10 cases reported by Greiner (1976).

It was our impression that TIA's associated with dissection tend to represent a broader territory of cerebral ischemia than those with arteriosclerotic stenosis. For example, the combination of weakness and numbness of the hand and face with or without dysarthria or dysphasia occurred in 6 of 13 patients with TIA's. In arteriosclerotic TIA's the area of involvement is more restricted, for example, tingling or

numbness involving the hand or face rather than both and weakness or numbness rather than both.

The period of evolution of symptoms leading to the diagnosis was shorter than in arteriosclerotic cases and was as follows: One hour, 3 hours, 12 hours, 16 hours, 1 day, 1½ days, 3 days, 5 days, and 10 days (3). When the signs and symptoms are derived from the carotid system and evolve at a brisker pace than in arteriosclerotic cases, one might epitomize the process as "carotid allegro".

Only 3 cases were hypertensive. The diagnosis could be made on the angiographic appearance in 15 cases. The string sign was present in 10 cases, a distal carotid pouch in 3, the two combined in 1 case and a tapering post-sinus occlusion in one. One case with a string sign was later verified pathologically. In only one case was the angiographic appearance of fibromuscular dysplasia

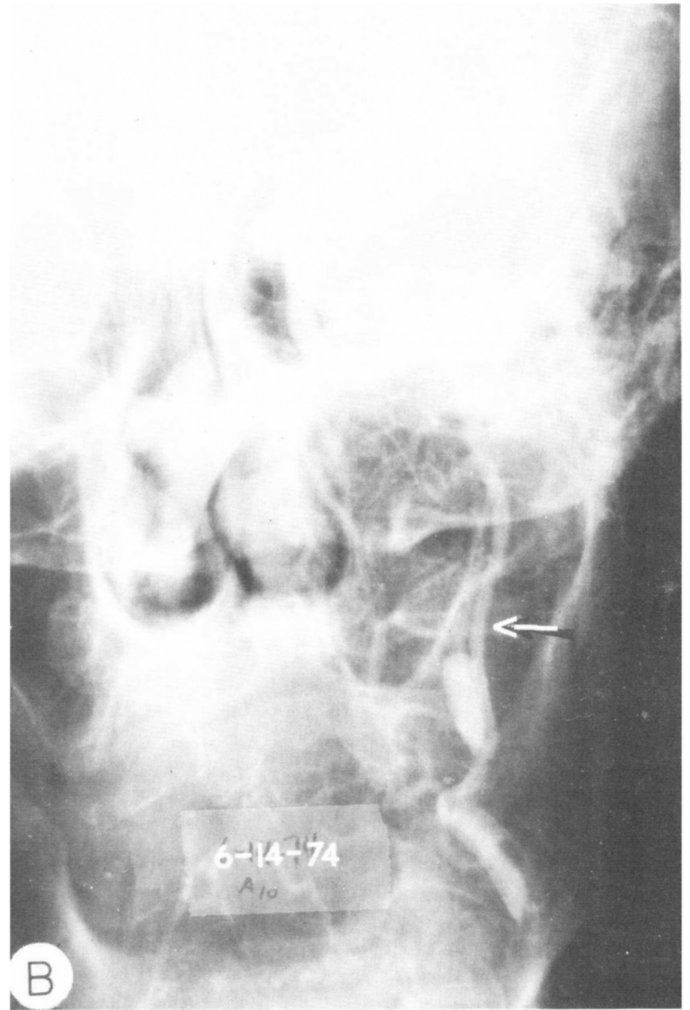
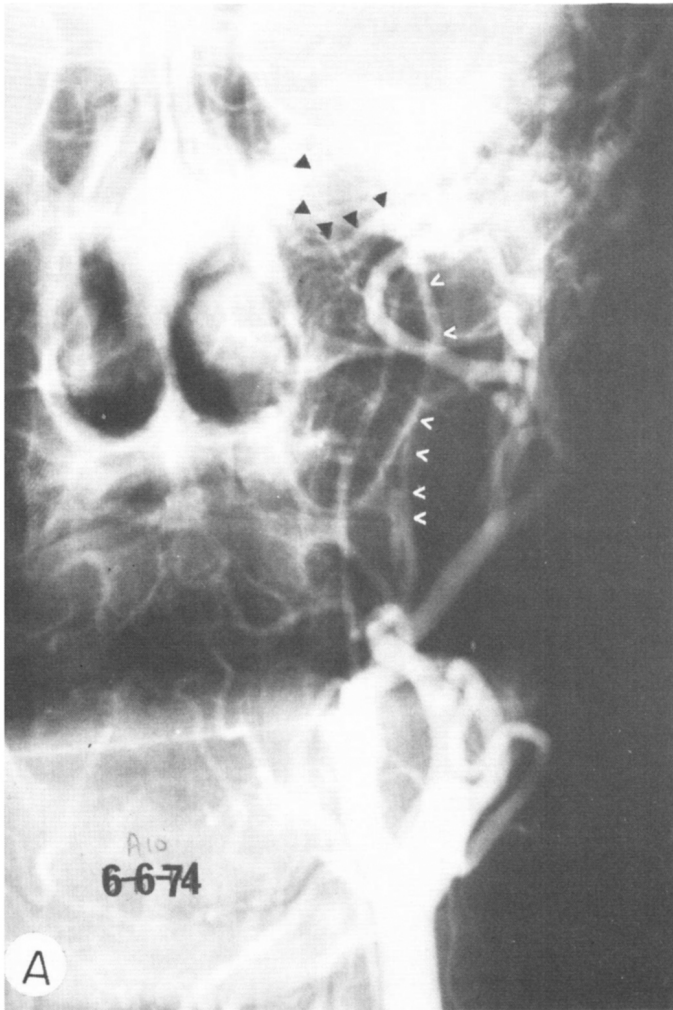


Figure 4—Angiograms in case 7. A. Day 8, B. Day 16.

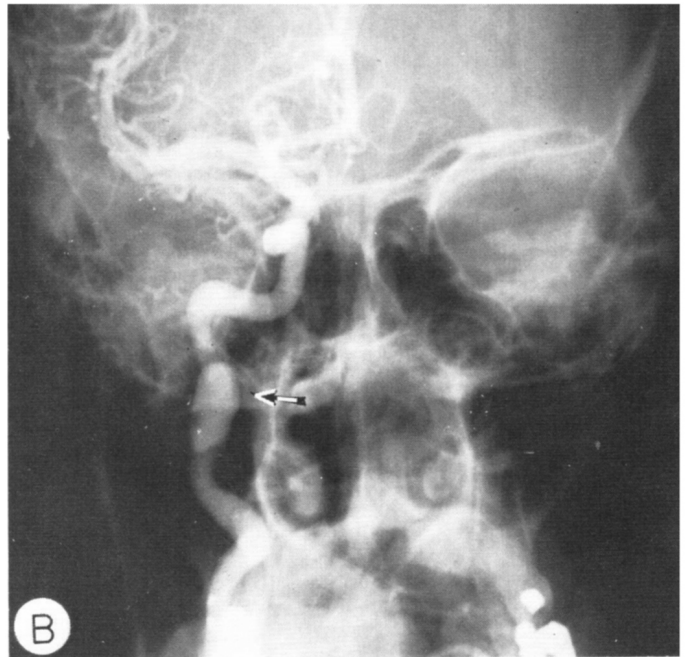
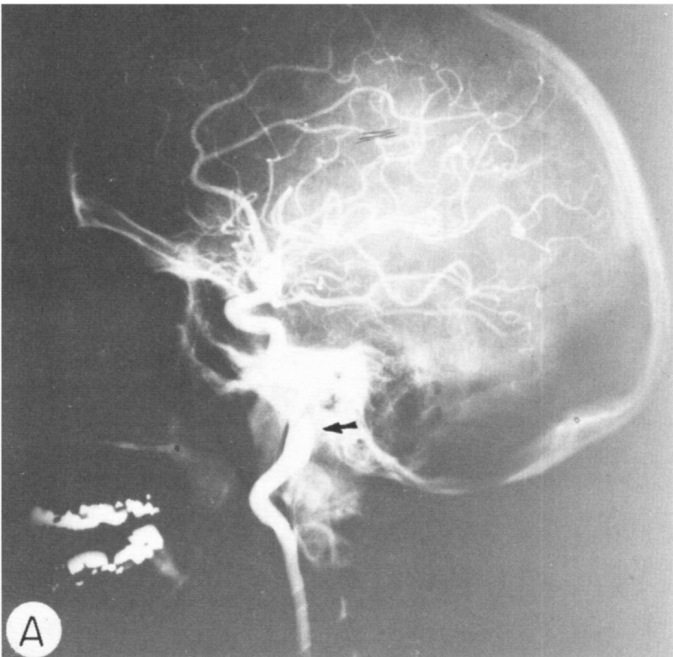


Figure 5—Angiogram in case 8. A. Lateral view, B. A-P view.



Figure 6—Angiogram in case 20.

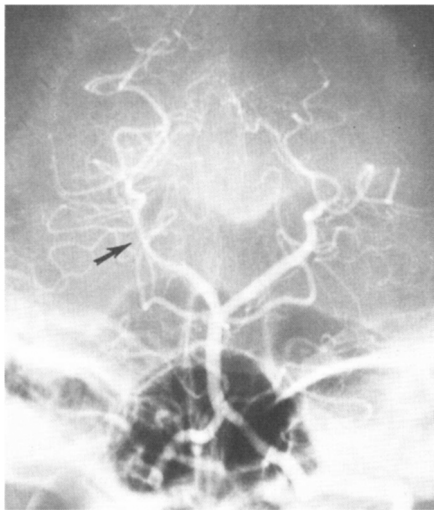


Figure 7—Angiogram in case 21.

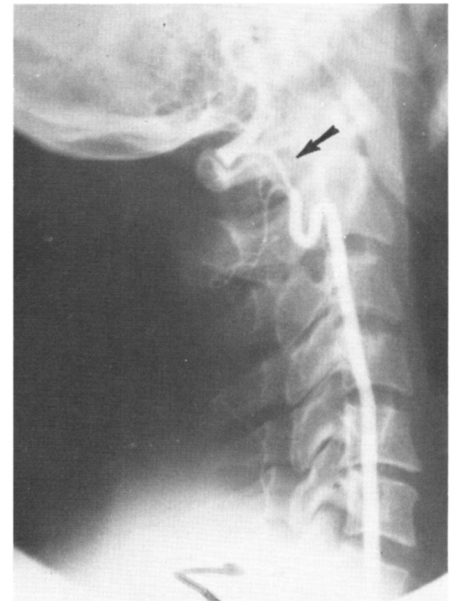


Figure 8—Angiogram in case 22.

present, a finding in the three cases of Lederman and Salanga (1976).

The development of dysgeusia due to involvement of the chorda tympani is exemplified perhaps in cases 7 and 15. Involvement of the hypoglossal nerve as it lies applied to the carotid sheath is postulated in case 18. Recognition of oculosympathetic palsy (OSP) may be hampered when there is a contralateral facial weakness and widening of the palpebral fissure, in which case pupillary size is a more reliable guide. In one case, perhaps two, the dissection was bilateral.

Our experience with these cases has shed light on some heretofore puzzling problems, but lack of confirmation of the diagnosis in several of our cases prejudices any inclination to extrapolate our findings further. Some typical cases of Raeder's trigeminal syndrome and of migraine associated with OSP should be suspected of harboring a dissection as in our cases 7, 8 and 15 and the cases of Lederman and Salanga (1976). The previously obscure cases in which a subjective bruit suddenly appeared only to vanish a few days or weeks later can probably be ascribed to dissection as can cases in which an initially narrowed carotid lumen has been found completely restored a few months later.

Angiographically, the string sign may be produced by early life occlusion of the supraclinoid carotid artery, e.g. moyamoya, with resultant "atrophy" and smallness of the artery in the neck. In an adult with such an occlusion the carotid canal will be smaller on the involved side. Fibromuscular dysplasia has been reported in conjunction with the string sign but pathological studies have not been made. In one case of multiple dissections, fibromuscular dysplasia was present but trauma was probably a factor (Ringel et al., 1977).

Traumatic dissection can produce the same picture as in the spontaneous group. There may be other as yet unrecognized causes of dissection and of a long string sign. The time between the onset of the dissection and the development of symptoms is probably variable, judging from the findings in post-traumatic cases. In Yamada et al.'s (1967) series of ICA cases only 10% had a deficit in the first hour after trauma, 50% were still asymptomatic after 10 hours and 17% after a day or more. For middle cerebral cases, Hollin et al (1966) found that the interval varied from several hours to 2 weeks (2 cases 12 hours, 3 cases 1 day, and 1 case each 2 days, 3 days, 4 days, 5 days, 1 week and 2 weeks respectively). A precipitating event in the

non-traumatic cases thus need not coincide with the appearance of symptoms. The appearance of a bruit, however, may accurately signal the time of onset of dissection.

Prognosis and Management

In the cases of carotid dissection reported in the literature the diagnosis was most often based on pathologic confirmation at autopsy, a method of selection in which the group becomes heavily weighted for patients with severe deficits. In the present series the only patient who died had undergone carotid surgery with resultant progression of the hemiplegia. One patient with a severe hemiplegia and aphasia recovered satisfactorily following surgical exploration and use of a Fogarty catheter. Of the other 5 patients who presented with a hemiplegia or hemiparesis, 4 made a virtually complete recovery, whereas one remained moderately impaired. Five patients progressed no further in their illness than the stage of TIAs and 3 patients had no hemispherical manifestations at all. The picture in over half the cases was that of a benign illness and in the remainder recovery was surprisingly satisfactory.

The natural history of the dissection itself is variable. In some cases the residual lumen becomes nar-

SYNOPSIS OF TWENTY-TWO CASES OF PROBABLE OR PROVEN SPONTANEOUS
DISSECTION OF CERVICO-CEREBRAL ARTERIES

Case	Age	Sex	TIAs	ICA Verified			Examination	OSP	Period of Evolution	Angio-gram
				No. TIAs	HA	Bruit				
1	45	F	TMB, numbness of R. face, arm & leg, dysphasia	5+	0	0	Very slight residuum	0	5 days	string sign
2	41	M	TMB, R. hemiparesis	2	0	0	Right hemiplegia, aphasia	0	2 days	string sign
3	45	F	Scintillation, L.-sided weakness & numbness	13	0	+	L. hemiparesis & hemianopia	0	10 days	occlusion
4	45	M	TMB	1+	0	0	R. hemiparesis, aphasia	0	12 hours	string sign
5	59	M	Temporary improvement & relapse. One TIA of numbness & weakness of the L. face & hand		+	0	L. hemiparesis	0	1 hour	string sign
6	67	M	Weakness of L. leg. Weakness of L. hand.	12	0	0	L. hemiparesis	0	1½ days	occlusion
ICA Unverified										
7	45	M	Scintillation, numbness of R. fingers, R. facial weakness, dysarthria	6+	+	+	WNL, except for →	+	10 days	string sign
8	46	F	None		+	+	WNL, except for →	+	—	distal pouch
9	43	F	Scintillations		+	+	WNL, except for →	0	—	string sign & pouch
10	31	M	Deficit with temporary improvement. One episode of scintillations		+	0	Aphasia, R. facial weakness	0	16 hours	distal pouch
11	52	M	Tingling of L. arm & leg. Tearing R. eye	3	+	+	Slight drift L. arm	+	4 weeks	distal pouch
12	29	F	45 min. TIA of R. hemiplegia & speechlessness		+	0	WNL	0	—	string sign
13	74	M	15 min. weakness & numbness of L. face & arm and slurred speech		0	0	WNL	0	—	string sign
14	45	M	Numbness of the R. hand	4	+	+	Abulia	0	3 days	string sign
15	53	M	None		+	+	WNL, except for →	+	1 day	string sign
16	45	M	None		+	+	WNL, except for →	+	10 days	string sign
17	68	M	Weakness of R. hand and slurred speech, loss of speech, R. hemianopia	10+	0	0	WNL	0	48 hours	string sign
18	45	M	None	0	+	+	Paralysis of L. side of tongue	0	Immed.	—
Middle Cerebral Artery										
19	15	F	Nausea, blurred vision to the left	1	+	0	L. hemiparesis, anosognosia	0	3 hours	string sign
20	13	M	"Weakness"	1	+	0	R. facial weakness, fluent aphasia	0	16 hours	string sign
Posterior Cerebral Artery										
21	18	M	None	0	+	0	L. hemianopia, L. sensory loss	0	Immed.	string sign
Vertebral Artery										
22	35	M	None	0	+	.0	Lateral medullary syndrome	+	Immed.	string sign

TIA = Transient Ischemic Attack. TMB = Transient Monocular Blindness. HA = Headache. OSP = Oculo-sympathetic Palsy.
ICA = Internal Carotid Artery. WNL = Within normal limits.

rowed to the point of occlusion. Or a vessel only narrowed initially is later found to be occluded. In the case of Bostrom and Liliequest (1967) a distal dissection sac enlarged by several millimeters in one month. As illustrated in our case 7, a patient treated with heparin intravenously, the residual lumen which was severely narrowed initially soon widened and at angiography 7 months later the lumen was fully restored. At the proximal extremity of the dissection there remained a tell-tale mural pouch. According to Sullivan et al. (1973), in traumatic dissection, short dissection sacs in the distal carotid may undergo progressive thrombosis and become obliterated. It has been suggested that one of the mechanisms of symptoms in a dissection is the dislodgement of thrombus formed at the site of occlusion with resultant embolism, but in the present cases there is little or no evidence for this hypothesis. We have witnessed this, however, in 2 cases of post-traumatic dissection.

The proper management of dissection lesions is largely unknown. Whether prompt surgical exploration and removal of the dissection clot and intima using a Fogarty catheter is advantageous must await further experience. If the vessel is entirely occluded and the neurologic deficit has originated within the previous 12 hours, surgery is probably indicated. When only a string sign is present, should one operate or treat with anticoagulants or antiplatelet aggregants? The question cannot be answered reliably. Distal pouches are probably beyond surgical approach and there are no guidelines regarding the advisability of anticoagulants. There is the possibility that in some cases anticoagulant therapy may hinder thrombotic occlusion of a sac or even promote further dissection, but this we have not experienced. The natural history of further cases is needed for guidance.

Pathogenesis

In 7 of the 11 previously described cases the involved internal carotid artery as well as the opposite carotid

and the aorta showed cystic medial necrosis. In our only case studied pathologically this change was not noted. Dissections begin at two sites of predilection — 2 to 3 cm. distal to the origin and in the distal carotid 2 to 4 cm. from the base of the skull, but a dissection occasionally originates between these two regions.

Whether these cases are truly of non-traumatic origin is a question that must be raised. Obvious external trauma was not a factor in our cases. In 3 cases symptoms began during a period of heavy coughing associated with influenza or an acute upper respiratory infection and we tentatively suspect that coughing may in some way produce tearing of the carotid intima. It is conceivable that extension of the neck may stretch and disrupt the carotid intima.

Non-carotid Cases

Middle cerebral artery. Only two examples are included here, both demonstrating the string sign. The development of the clinical picture was more abrupt than in carotid cases. TIAs were fewer and the neurologic deficit was more severe.

Chang et al. (1975) described spontaneous dissection of the terminal internal carotid artery and adjacent middle cerebral artery bilaterally 5 weeks apart in an 8-year-old boy. The arteries of the intracranial internal carotid system showed an abnormal amount of splitting, fraying, disintegration and irregularity. In the angiogram an unusual arterial fold was present in the middle cerebral artery suggesting that dissection may assume still another appearance. The re-entry of the dissection into the middle cerebral artery distally may have provided a by-pass. Gagne et al. (1977) reported the case of a 6-year-old child with a fatal dissection of the anterior and middle cerebral arteries. It is possible that with early recognition of the syndrome before the deficit is maximal, effective measures, surgical or other could be devised.

The other arteries. Insofar as surgical or pathologic confirmation of the

diagnosis is lacking in these two cases, further remarks are presented with reservation. In the first, a youth, aged 18, showed an unexplained 2.5 cm. long segment of severe stenosis of the right posterior cerebral artery which persisted unchanged after 3 months. There had been a sudden stroke. The second patient, a man, aged 35, suddenly suffered a lateral medullary syndrome which was associated with a 3.5 cm. long segment of severe stenosis of the right vertebral artery from C1 to C3. These cases are mentioned only to suggest that the principle of spontaneous arterial dissection may extend more widely than suspected.

Ringel et al. (1977) described the remarkable case of a 49-year-old man who, while skiing, developed acute dissections of both internal carotid arteries and both vertebral arteries. The carotid arteries and the right vertebral artery were occluded at the level of the second cervical vertebra and the left vertebral artery at the sixth cervical vertebra. The involved arteries showed fibromuscular dysplasia. Minor falls during skiing must be suspected as a cause of trauma in this case as in other cases in our experience.

Dissection in the basilar artery system has been reported by Perier et al. (1966), Escourolle et al. (1972) and Pesendorfer and Platthy (1973).

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