artery occlusion, progressive thrombosis, or development of abscess (1011,1012). Inadvertent thrombotic progression, if recognized earlier, can be treated with thrombolytics (1013). Distal unintended embolization of coils may produce focal neurologic dysfunction, including central retinal artery occlusion and other neuro-ophthalmic manifestations (1014, 1015). Perforation of the aneurysm during embolization is uncommon (between 0.5% in unruptured and 4% in previously ruptured aneurysms) (1016). In one rare case rupture of a paraclinoid aneurysm resulted in a carotid cavernous fistula (1017). In a prospective series 31% of patients had microemboli detected by transcranial Doppler (1018), and abnormal DWI sequences were found in 57% (8 of 14 patients) (1019) and 61% (45 of 74 patients) (1020). Late distal embolization occurs more commonly in patients with large aneurysms and in those with protruding coils (1021). Patients with antiphospholipid syndrome may also be at higher risk of thromboembolism following coil embolization (1022). Several techniques have been developed to retrieve coils that have embolized distally (1023).

FUSIFORM ANEURYSMS (ARTERIAL ECTASIA, DOLICHOECTASIA)

GENERAL CONSIDERATIONS

Large arteries of the carotid and vertebrobasilar systems may occasionally become enlarged and tortuous. This process is called by a variety of names, the most common of which are dolichoectasia (from the Greek words *dolichos,* meaning ''elongation,'' and *ectasia,* meaning ''distension''), ectasia, and fusiform aneurysm (1024). Some authors simply describe such arteries as ''tortuous'' (1025).

The intracranial arteries most commonly affected by fusiform aneurysms or dolichoectasia are the three largest: the internal carotid, the vertebral, and the basilar. Other large arteries that originate from these vessels, particularly the anterior cerebral, middle cerebral, posterior cerebral, and anterior communicating arteries, may also be affected in rare patients (714,1026–1028) (Fig. 41.54).

Many investigators believe that dolichoectasia results from atherosclerosis (1028,1029); however, in some cases, there is no evidence of atherosclerosis, even on histopathologic investigation (1030). In addition, arterial ectasia is occasionally found in children who have no evidence of atherosclerosis (1028,1031) and in patients with other disorders characterized by mesodermal dysgenesis, such as von Recklinghausen's neurofibromatosis (NF-1) (259,1032) and FMD (60). Malignant cells (1033), aspergillosis (168), lymphomatoid granulomatosis (175), and even HIV (1034) may weaken arterial walls and lead to fusiform dilatation. Surgical trauma may also play an etiologic role, especially following surgery for a craniopharyngioma (1035). De novo formation of fusiform aneurysms is unusual but has been reported (997). Histologic studies reveal fragmentation of the internal elastic lamina, neogenesis within the thickened intima, and intramural hemorrhage (1036).

Diagnosis of dolichoectatic arteries and fusiform aneurysms is traditionally accomplished via angiography. More

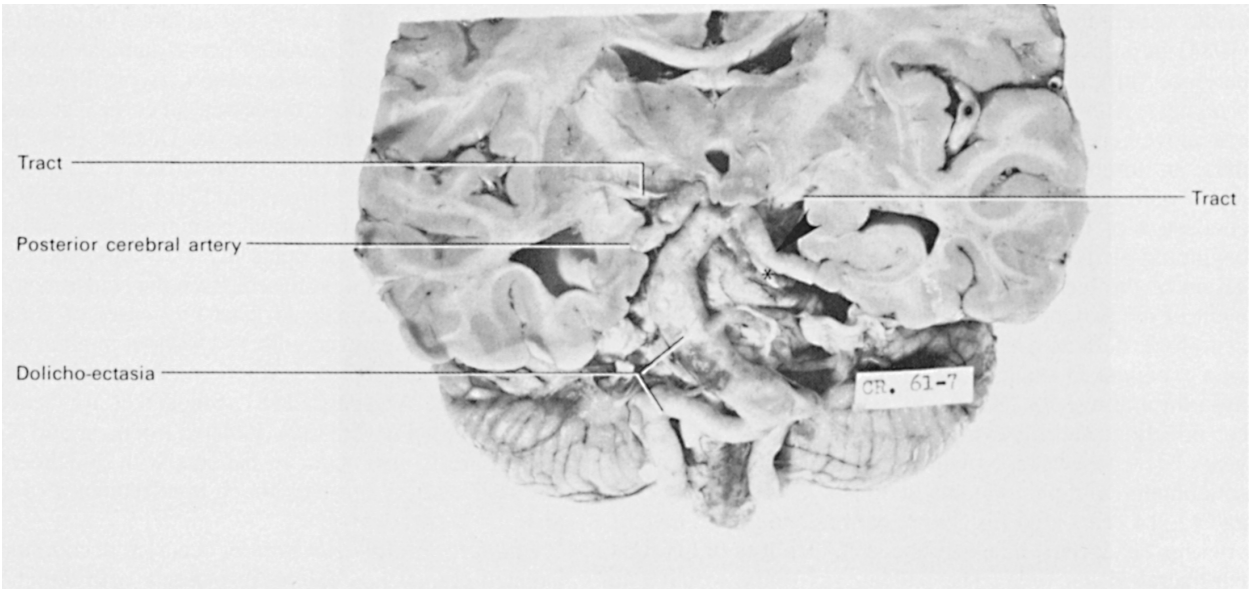

Figure 41.54. Diffuse dolichoectasia of the major arteries of the vertebrobasilar arterial system. The brain has been sectioned coronally, and the view is from in front and slightly below. Note dolichoectasia of the right vertebral artery, the basilar artery, and both posterior cerebral arteries. Also note that the right optic tract is compressed by the dolichoectatic right posterior cerebral artery which also erodes the adjacent internal capsule and mammillary body. The ectatic left posterior cerebral artery *(arrowhead)* compresses the left oculomotor nerve *(asterisk).* The patient had a left homonymous hemianopsia. (From Lindenberg R, Walsh FB, Sacks JG. Neuropathology of vision: an atlas. Philadelphia: Lea & Febiger, 1973.)

recently, MRI and MRA have offered alternatives to angiography. Computerized tomographic angiography (CTA) represents the latest technique available to detect and follow patients with dolichoectasia (1037). It offers the added advantage of demonstrating the relationship between the involved artery and the underlying bony structures of the skull base.

Ectatic and dolichoectatic arteries behave quite differently from saccular aneurysms. They do not arise from arterial bifurcations, almost always become symptomatic in middle-aged or elderly patients, are frequently calcified, and tend to be quite large. Most importantly, about 90% of saccular aneurysms first become symptomatic when they rupture, whereas fusiform aneurysms rarely cause SAH. Instead, they produce neurologic symptoms and signs by direct pressure on adjacent neural tissue (particularly cranial nerves), by impairment of circulation in adjacent neural tissue from direct compression of smaller feeding arteries and arterioles, by obliteration of nutrient vessels derived from the diseased artery, and by giving rise to emboli. In a series of 120 posterior fossa fusiform aneurysms reported by Drake and Peerless (1038), 50% presented with mass effect, 20% with hemorrhage, and 6% with TIAs.

Rupture of fusiform aneurysms may occur, however. This is particularly true of aneurysms involving the vertebral arteries (423).

The specific symptoms and signs produced by ectatic arteries are related to their location, size, and extent. Thrombus may form within the lumen of a fusiform aneurysm leading to distal embolization and associated transient or permanent neurologic sequelae (1039). When thrombosed, serpentine aneurysms may mimic intracranial tumors (1040).

The treatment of fusiform aneurysms is also different from that of saccular aneurysms (1028). Because fusiform aneurysms have no neck, they cannot be clipped. Surgical treatment, when appropriate, is directed toward isolating the aneurysm from the intracranial circulation. In some cases, the ectatic region may be excised without permanent neurologic deficit (1040). This is particularly true when the process affects a branch of the MCA. In many cases, however, excision or trapping of the aneurysm produces severe permanent neurologic deficits. In other cases, the affected artery is ligated proximal to the lesion, or the vessel is occluded using an inflatable balloon (1041). Detachable thrombogenic coils can also be utilized to occlude a dolichoectatic vessel. Their advantage over a detachable balloon includes occlusion of a shorter segment of normal artery, no traction on the parent vessel, and possibly safer and easier catheterization techniques. With feeding artery occlusion, a bypass procedure may be used to provide blood flow to the territory normally supplied by the occluded vessel but collateral flow provided by the revascularization procedure may allow the aneurysm to continue to enlarge (1042).

In patients who cannot tolerate loss of distal flow in the parent vessel of a fusiform aneurysm, the involved arterial segment may be reconstructed (1043).

Anson et al. (1043) reviewed the treatment of 40 patients with 41 fusiform or dolichoectatic aneurysms. Surgical procedures included direct clipping, trapping with bypass, proximal occlusion, resection with reanastomosis, transposition, aneurysmorrhaphy with thrombectomy, and wrapping. There was no surgical mortality, and the outcome at late follow-up was good (Glasgow Outcome Scale scores 1–2) in 78% of patients, although the patients with posterior circulation aneurysms did not do as well as those with anterior circulation aneurysms. Cardiopulmonary bypass, hypothermic circulatory arrest, and barbiturate cerebral protection are often utilized during resection of large fusiform aneurysms (311).

Medical therapy using antiplatelet drugs or anticoagulants may prevent permanent neurologic damage from emboli in selected patients with fusiform aneurysms (1044).

CLINICAL MANIFESTATIONS, TREATMENT, AND PROGNOSIS ACCORDING TO SITE

Fusiform Aneurysms of the Internal Carotid Artery and Its Branches

Dolichoectasia of the intracranial portions of the ICAs and their branches may be asymptomatic, or it may cause a variety of symptoms and signs that are related to the location and extent of the condition (1038). Thus, dolichoectasia of the petrous portion of the ICA may produce lower cranial neuropathies, whereas dolichoectasia of the intracavernous portion of the artery can enlarge the sella turcica and simulate a pituitary tumor, causing diplopia that is often painful (Fig. 41.53) (1045). Other authors have recognized abducens palsies related to fusiform aneurysms (1028). Epistaxis has been reported as a rare complication of a dolichoectatic aneurysm involving the cavernous sinus (1046).

As early as the 19th century, investigators observed that ectasia affecting the supraclinoid portion of the ICA had potentially severe effects on the adjacent optic nerves and chiasm (1050). The ipsilateral optic nerve was frequently flattened by the diffusely enlarged sclerotic artery that often compressed the nerve against the dural fold overlying the superior portion of the intracranial end of the optic canal. Histologic examination of affected nerves showed patchy loss of myelin and reduction of axons. Arcuate, altitudinal, and paracentral visual field defects coupled with increased cupping resembling ''low tension glaucoma'' have been found to be associated with dolichoectatic carotid changes, although this remains debated. In selected cases, surgical decompression could improve optic nerve dysfunction (1054).

The possible association between dolichoectatic carotid arteries and low-tension glaucoma is intriguing. Stroman et al. (1051) performed a prospective study in which they used high-resolution MR imaging to assess 20 patients with low-tension glaucoma and 20 age-matched control subjects and found no difference between the two groups in the diameter or length of the intracranial portion of the optic nerves, the cross-sectional area of the supraclinoid portion of the ICAs, or the distance from the intracranial optic nerves to the supraclinoid portion of the ipsilateral ICA. There was, however, a relative decrease in the cross section of the intracranial segment of the optic nerve and an increase in changes consistent with small-vessel ischemia. In a subsequent study, Gol-

nik et al. (1052) found that 20 patients with unexplained optic neuropathy had a statistically closer relationship of the intracranial segment of the optic nerve to the supraclinoid portion of the ipsilateral (undilated) carotid artery than did age-matched control subjects. Jacobson et al. (1053) reviewed MR imaging findings in 100 visually asymptomatic patients and determined that there was contact between the intracranial portions of one or both optic nerves and the supraclinoid portion of the ipsilateral ICA in 70% of cases. There appeared to be true compression of one optic nerve by the ipsilateral ICA in 5 patients and bilateral compression in 12 patients. The estimated odds of compression were significantly increased as the diameter of the supraclinoid portion of the ICA increased. These investigators concluded that supraclinoid ICA contact with the intracranial portion of the optic nerve occurs frequently compared with anatomic compression and that the risk of compression of the optic nerve is directly proportional to the diameter of the supraclinoid portion of the ICA.

Patients with dolichoectasia of the ICAs may lose vision suddenly or in a slow, progressive fashion.

The optic neuropathy caused by dolichoectasia of the internal carotid and anterior cerebral arteries occurs equally in men and women. Most patients experience visual symptoms after 40 years of age. Visual loss is usually bilateral, but one eye may be affected weeks, months, or years before the other. The visual loss may be insidious in onset and slowly progressive, suggesting a compressive lesion, or it may be acute and nonprogressive, suggesting optic neuritis or ischemic optic neuropathy. Visual field defects are often altitudinal, but all types of field defects may occur, even central and cecocentral scotomas. Nonspecific visual field constriction is not uncommon (1054). Many patients have normal or pale optic discs when they are first examined, but some patients have swelling of one optic disc, usually in the eye with the more recent visual symptoms. The pathophysiology of the disc edema remains unclear.

Only rarely do bitemporal field defects occur in patients with ectasia of the supraclinoid portion of the ICA (1055) (Fig. 41.53). Such defects indicate damage to the nasal crossing fibers in the center of the chiasm and therefore cannot be ascribed to direct pressure by a laterally situated artery. Either the dilated vessels must act as a mass beneath the chiasm, or the process must specifically compromise the vascular supply to the center of the chiasm (1050,1056). In some patients, ectasia of the A-l or proximal A-2 segments of the ACAs cause them to compress one or both optic nerves from above, thus producing progressive visual loss amenable to surgical decompression (1057).

A homonymous visual field defect is an extremely rare finding in patients with a dolichoectasia of the ICA due to either compression of the ipsilateral optic tract by the ectatic vessel or from emboli that originate within this portion of the artery and obstruct vessels supplying the postchiasmal visual sensory pathway (1028).

Fusiform aneurysms of the distal anterior circulation (anterior and middle cerebral arteries) usually do not present with neuro-ophthalmologic symptoms. These patients may have a long history of headaches usually diagnosed as migraine. Rare rupture or thrombosis of the aneurysm may bring it to clinical attention, usually with a change in mental status and headache (1058).

A dolichoectatic PCA that arises from the ICA may produce an acute (occasionally painful and or recurrent) oculomotor nerve paresis, either from compression of the nerve or from interruption of its blood supply (1026).

Fusiform Aneurysms of the Vertebral and Basilar Arteries and Their Branches

Fusiform aneurysms (dolichoectasia) of the vertebral and basilar arteries and their branches are probably more common than similar lesions affecting the ICA and its branches (Fig. 41.54). Whereas visual loss is the only major symptom produced by dolichoectasia of the ICA and its branches, dolichoectasia that affects the vertebrobasilar arterial system may produce symptoms and signs related to: (*a*) ischemia; (*b*) compression of cranial nerves; (*c*) compression of the brainstem and cerebellum; and (*d*) hydrocephalus (1024, 1044,1059,1060).

Fusiform Aneurysms of the Vertebral Artery and Its Branches

Dolichoectasia of the vertebral artery and its branches occurs far less often than does dolichoectasia of the basilar artery and its branches and is also less common than saccular aneurysms (1044). Most fusiform aneurysm are asymptomatic or present with brainstem symptoms, but cases of hemorrhage have been reported. Dolichoectatic dilation of the vertebral artery may be related to dissection (1044,1061).

In most series, the complaints of patients with vertebral artery dolichoectasia are related to dysfunction of the cerebellum and caudal brainstem, including headache, dizziness, ataxia, dysarthria, dysphagia, and hemiparesis (1061). Ischemic involvement of the lateral medullary plate can produce symptoms of Wallenberg's syndrome with diplopia and symptoms of ocular tilt (1062). Intermittent diplopia may also be due to a skew deviation (1044).

Rare patients have dolichoectasia of one of the branches of the vertebral artery. Brainstem symptoms have included dysphagia, dizziness, nausea, and hemiparesis, along with signs of palatal weakness, ataxia, absent gag weakness, and gaze paretic nystagmus. Secondary embolic phenomena may produce distal ischemic findings, including homonymous hemianopsia. Ligation of fusiform aneurysms of the PICA may be carried out without inducing new neurologic symptoms if the origin is distal to the tonsillomedullary segment. Revascularization may be indicated (1063).

Fusiform Aneurysms of the Basilar Artery and Its Branches

As is the case with ICA dolichoectasia, patients with dolichoectasia of the basilar artery and its branches may be asymptomatic, with the abnormality being found during a screening examination for unrelated symptoms or during a

routine autopsy (1029). Most patients who become symptomatic from this condition are 50 years of age or older and have atherosclerotic or hypertensive vascular disease. Men are affected more often than women (1043,1044).

Dolichoectasia of the basilar artery may cause a variety of neurologic difficulties, including isolated cranial neuropathy, multiple cranial neuropathies, mixed ischemic and compressive deficits, and evidence of hydrocephalus. Symptoms and signs of SAH are uncommon but TIAs or ischemic stroke have been reported (1024,1043).

Dysfunction of a single cranial nerve may be the only sign of a dolichoectatic basilar artery. The most common cranial nerves affected are the facial and the trigeminal nerves (1024). Damage to the facial nerve may cause hemifacial spasm, or isolated facial paresis (1024). Trigeminal nerve damage may be manifest as typical trigeminal neuralgia, atypical facial pain, or trigeminal neuropathy with decreased corneal sensation and facial hypesthesia and dysesthesia (1064) (Fig. 41.55). Other individual cranial nerves, including the vestibulocochlear, glossopharyngeal, vagus, and hypoglossal, may become compromised (1065). Even the ocular motor nerves may become damaged. Cases of isolated oculomotor nerve paresis, with and without pupillary involvement, are documented in the literature (680, 1024,1066,1067) (Fig. 41.56), including one bilateral (1068). Isolated trochlear and abducens nerve paresis also occur in patients with dolichoectasia of the basilar artery but with much less frequency (1024).

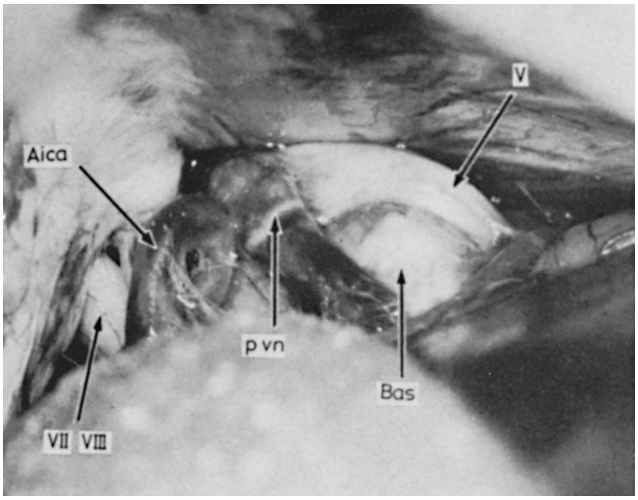

Figure 41.55. Basilar artery ectasia causing trigeminal neuralgia. The patient was a 71-year-old man with an 8-year history of left trigeminal neuralgia affecting the first and second divisions of the trigeminal nerve. The patient initially responded to carbamazepine, but he was unable to tolerate the drug on a long-term basis. An angiogram showed evidence of ectasia of the basilar artery, and it was decided to perform a posterior fossa microvascular decompression of the trigeminal nerve. Surgical view shows that the left trigeminal nerve *(V)* is compressed and elevated by the dilated, tortuous basilar artery *(Bas). Aica,* anterior inferior cerebellar artery; *VII VIII,* facial and vestibuloacoustic nerves; *p vn,* petrosal vein. (From Lye RH. Basilar artery ectasia: An unusual cause of trigeminal neuralgia. J Neurol Neurosurg Psychiatry 1986;49:22–28.)

Multiple unilateral and bilateral cranial neuropathies occur more commonly than isolated cranial neuropathy. Any of the cranial nerves (except for the olfactory and optic nerves) may be affected. Oculomotor nerve paresis may occur in combination with other cranial neuropathies (680,1060,1066,1067,1069–1072). Trochlear nerve paresis may also be seen in combination with other cranial nerve pathology (1044). The often affected nerves are those that course through the cerebellopontine angle (i.e., the trigeminal, abducens, facial, and vestibulocochlear nerves) (1024, 1025,1044). The symptoms and signs in such patients thus mimic those produced by CPA neoplasms, such as vestibular schwannomas and meningiomas. Some patients develop both hemifacial spasm and ipsilateral trigeminal neuralgia, whereas others develop progressive trigeminal, abducens, and facial paresis (1044). Progressive paresis of the lower cranial nerves may develop, mimicking the symptoms and signs produced by tumors in the region of the clivus and foramen magnum (e.g., chordoma, nasopharyngeal carcinoma, meningioma). In some cases, both upper and lower cranial nerves are affected, a consequence of the extensive nature and erratic course of the ectatic artery (1060).

A variety of acute brainstem and cerebellar symptoms and signs may develop in patients with dolichoectasia of the basilar artery and its branches. In some cases, the symptoms and signs develop acutely and resolve within about 24 hours. Patients in whom this occurs may be thought to have suffered a TIA from atherosclerotic vertebrobasilar insufficiency (1044,1073) (Fig. 41.57). In other cases, neurologic symptoms and signs develop suddenly, do not resolve, and may even progress (1074). Hydrocephalus develops in some patients with basilar artery dolichoectasia (1075). In most cases, increased ICP is caused by obstruction of the 3rd or 4th ventricle, the floor of which is compressed and elevated by the ectatic portion of the artery. In other patients, an ectatic basilar artery extends into the floor of the 3rd ventricle and exerts a water-hammer pulse that is transmitted toward the foramina of Monro, thus impairing outflow from the lateral ventricles. Afferent system dysfunction may occur if the dolichoectatic basilar artery compresses the optic tract (1076), chiasm, optic nerve, or even visual radiations.

Coppeto (1077) described three women with long-standing, recurrent, migraine-like phenomena with expanding hemianopic scintillating scotomas and characteristic ''build-up.'' Each patient was eventually found to have severe basilar or vertebrobasilar dolichoectasia. One patient underwent partial occlusion of the dolichoectatic basilar artery segment, after which she had no further visual disturbances during an 8-year follow-up period. It is possible that the ectasia and the migraine in these patients may have been independent phenomena.

Dolichoectasia of the PCA and other branches of the basilar artery may cause neurologic dysfunction (including partial and complete oculomotor palsy) (502,1074). Distal involvement of the posterior cerebral artery may rarely occur (1078). Also uncommon is dolichoectasia of the anterior inferior cerebellar artery producing facial sensory loss (718). MRI may document progressive enlargement of dolichoectatic aneurysms of the vertebrobasilar system (1079).

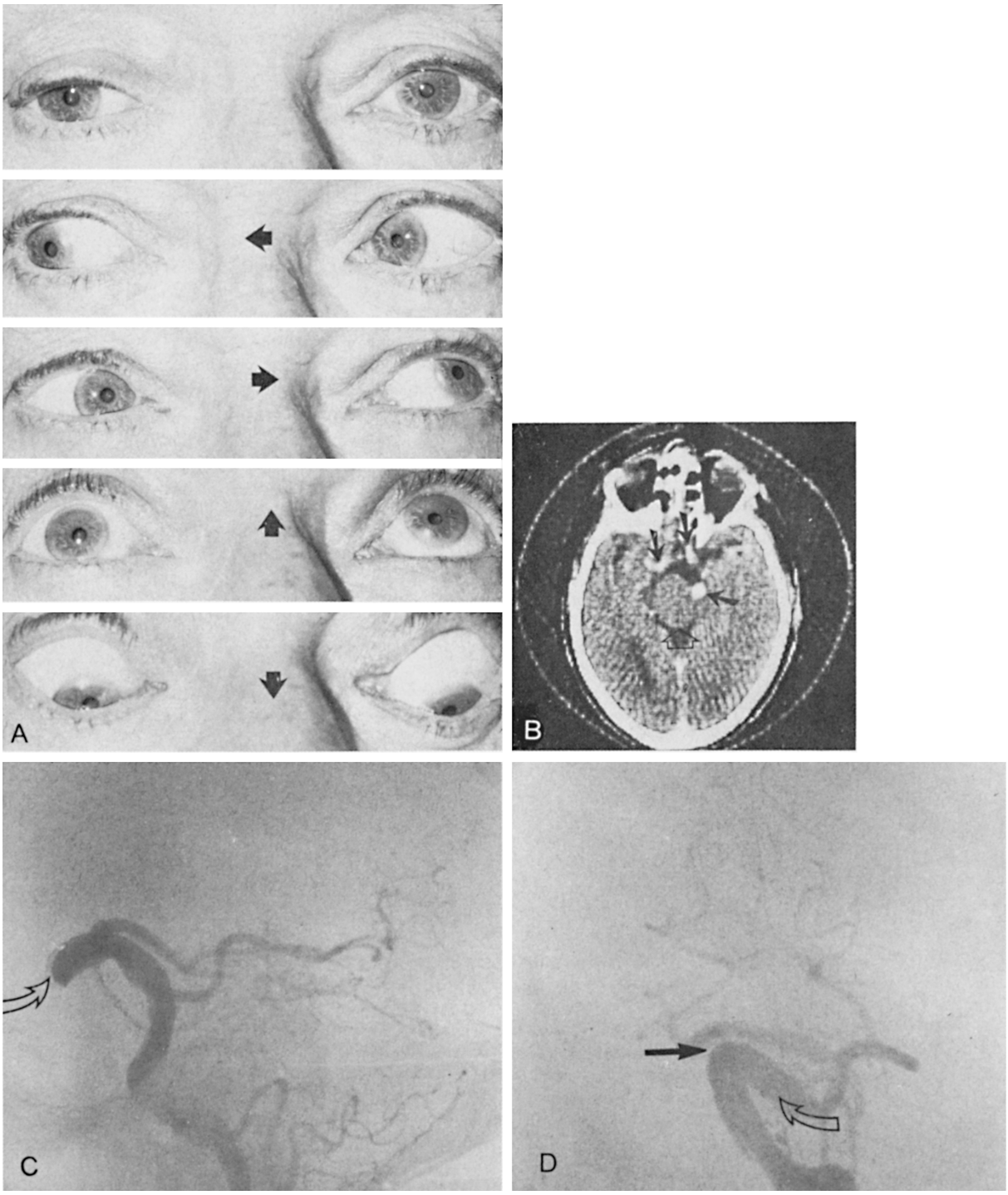

Figure 41.56. Isolated oculomotor nerve paresis caused by a fusiform aneurysm (*dolichoectasia*) of the basilar artery. The patient was a 61-year-old hypertensive man with a 3-year history of diplopia. *A*, Appearance of the patient shows a moderate right ptosis associated with marked limitation of elevation and mild limitation of depression and adduction of the right eye. The right pupil is the same size as the left. It reacted normally to light and near stimulation. *B*, A CT scan shows small areas of opacification consistent with enlargement of the basilar artery in the perimesencephalic, parasellar, and suprasellar cisterns *(solid arrows).* The brainstem is displaced to the left *(open arrow). C* and *D*, Vertebral angiogram, lateral and anteroposterior views, respectively, shows marked dolichoectasia of the basilar artery. At its most distal portion *(solid arrow),* it is causing a mass effect on the right posterior cerebral artery which also is somewhat ectatic and dilated. The anterior portion of the basilar artery extends into the left suprasellar cistern *(open arrows).* (From Trobe JD, Glaser JS, Quencer RC. Isolated oculomotor paralysis: the product of saccular and fusiform aneurysms of the basilar artery. Arch Ophthalmol 1978;96:1236–1240.)

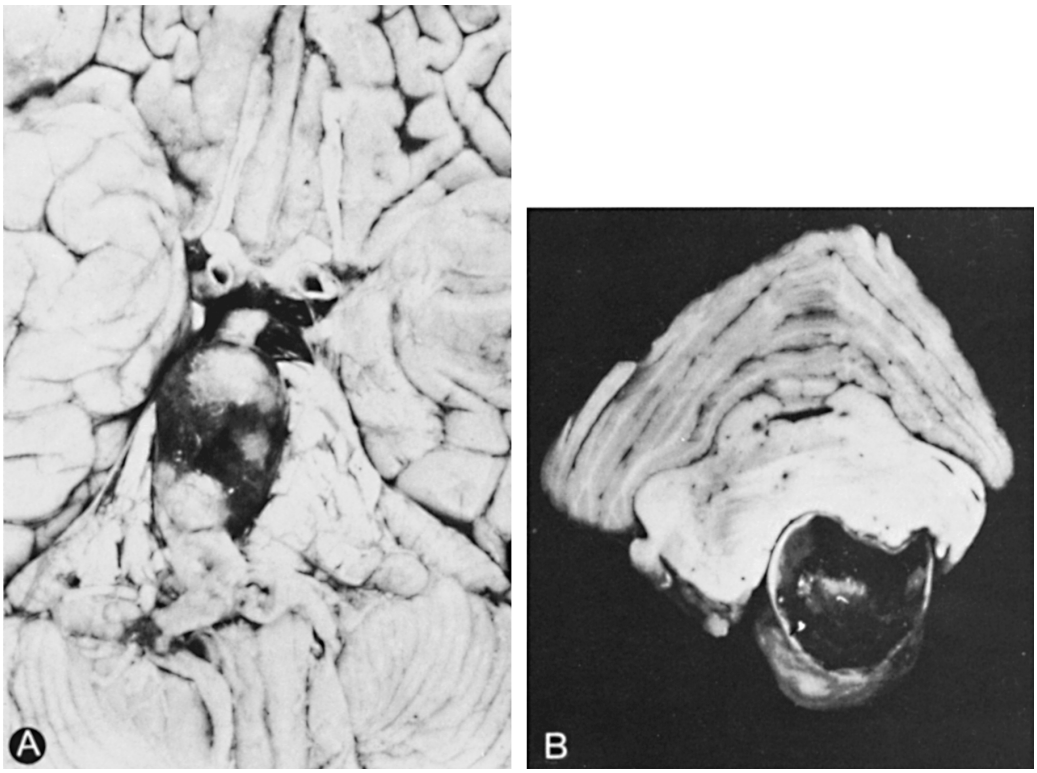

Figure 41.57. Fusiform aneurysm of the basilar artery (dolichoectasia) causing transient ischemic attacks. The patient was a 50-year-old man with hyperlipidemia and angina pectoris who experienced episodes of binocular diplopia that lasted 1–3 minutes. During one of these episodes he also experienced transient left hemiparesis, paresthesias, nausea, and vomiting. The patient was thought to have vertebrobasilar atherosclerotic disease, and he was treated with anticoagulation. He died 4 months later from rupture of the thoracic aorta. *A*, View of the base of the brain in this patient shows a large fusiform aneurysm (*dolichoectasia*) of the midportion of the basilar artery. *B*, Cross-section of the pons shows marked compression by the aneurysm. Note that the ''aneurysm'' is filled with premortem thrombus and atheroma. (From Nijensohn DE, Saez RJ, Reagan TJ. Clinical significance of basilar artery aneurysms. Neurology 1974;24:301–305.)

CONCLUSIONS

Intracranial aneurysms occur with high frequency and have a significant impact on public health. Most aneurysms present with subarachnoid hemorrhage, but 10% cause symptoms by mass effect, distal embolization, or thrombosis. Early clinical recognition and the use of improved and improving diagnostic techniques permit accelerated intervention, which itself continues to improve, with resultant decrease in overall mortality and morbidity and improvement in long-term prognosis. Neuro-ophthalmic signs and symptoms are commonly seen with aneurysms presenting either with mass effect or subarachnoid hemorrhage. Aggressive treatment to prevent or reverse delayed secondary ischemia and endovascular techniques for both ruptured and unruptured aneurysms promise to have a continuing impact on the outlook for patients with intracranial aneurysms. Better prospective studies will refine those indications for intervention in patients with cerebral aneurysms. The greater morbidity percentages seen in the newer surgical series are likely associated with the attempt to quantitate function, such as the Rankin scale (950) and other psychosocial parameters (981), although low functional morbidity continues to be reported (6%) (1080).

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