

Carotid Cavernous Fistulae: Indications for Urgent Treatment

Van V. Halbach¹
Grant B. Hieshima¹
Randall T. Higashida¹
Murray Reicher²

Angiographic and clinical data from 155 patients with carotid cavernous fistulae were retrospectively reviewed to determine angiographic features associated with increased risk of morbidity and mortality. These features included presence of a pseudoaneurysm, large varix of the cavernous sinus, venous drainage to cortical veins, and thrombosis of venous outflow pathways distant from the fistula. Clinical signs and symptoms that characterized a hazardous carotid cavernous fistula included increased intracranial pressure, rapidly progressive proptosis, diminished visual acuity, hemorrhage, and transient ischemic attacks. Cortical venous drainage from the carotid cavernous fistula is secondary to occlusion or absence of the normal venous outflow pathways and is associated with signs and symptoms of increased intracranial pressure and an increased risk of intraparenchymal hemorrhage. Angiographic demonstration of a cavernous sinus varix, with extension of the sinus into the subarachnoid space, is associated with an increased risk of fatal subarachnoid hemorrhage. Identification of these high-risk features provides a basis for making decisions about treatment.

Carotid cavernous fistulae (CCFs) are spontaneous or acquired connections between the carotid artery and the cavernous sinus, and can be classified as direct or indirect. Direct connections between the internal carotid artery and cavernous sinus may occur as a consequence of trauma, ruptured intracavernous carotid aneurysms, collagen deficiency syndromes, arterial dissection, fibromuscular dysplasia, and direct surgical trauma [1–10]. Indirect fistulae are usually supplied from dural branches of the external carotid artery but can be supplied from dural branches of the internal carotid artery. Although the cause is often unknown, factors associated with the development of indirect fistulae include pregnancy, sinusitis, trauma, surgical procedures, and cavernous sinus thrombosis. Symptoms caused by CCFs are related to their size, duration, location, adequacy and route of venous drainage, and presence of arterial and venous collaterals [11].

Surgical and angiographic techniques that have been described for the closure of CCFs include carotid occlusion; trapping procedures; direct surgical exposure and closure; and embolization with muscle, glue, thrombus, wires, and, more recently, detachable balloons [12–19].

Unfortunately, the natural history of CCFs is incompletely understood. Spontaneous closure, which is more common in indirect than direct CCFs, as well as closure following diagnostic angiography, has been documented by Seeger et al. [20]. Carotid compression therapy has been successful in closure of 17% of direct and 30% of indirect CCFs [21]. While aggressive forms of therapy may be successful in closure of CCFs, no technique is without risk. Ideally, the decision to institute a potentially hazardous treatment should be based on full understanding of the disease's natural history.

To identify those patients whose poor natural history mandated the need for emergent or aggressive therapy, we evaluated the angiographic and clinical data from 155 patients with CCFs. The delineation of high-risk features enables rational choices to be made regarding the timing and method of treatment.

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¹ Department of Diagnostic and Interventional Neuroradiology, UCSF Medical Center, San Francisco, CA 94143. Address reprint requests to V. V. Halbach.

² Department of Radiology, UCLA Medical Center, Los Angeles, CA 90024.

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Materials and Methods

The radiographic findings and clinical records of 150 patients treated for CCF over the past 10 years were reviewed retrospectively. There were 122 direct fistulae and 28 indirect fistulae. Radiographic and clinical findings from five additional patients who died before treatment could be instituted were also reviewed. Before treatment, specific attention was given to the signs, symptoms, and angiographic features associated with a poor clinical outcome, and these were recorded.

Results

Several angiographic and clinical signs and symptoms were found to be associated with a poor clinical outcome, all of which occurred prior to treatment. These are summarized in Table 1.

Hemorrhage

Hemorrhage occurred in 13 (8.4%) of 155 patients with direct CCFs, and was associated with a poor outcome. In eight patients, hemorrhage occurred at least 3 weeks after the event, usually trauma, that was responsible for the development of the fistula. The remaining five patients, all with pseudoaneurysms, hemorrhaged within a week of onset of the fistula. Four patients experienced massive subarachnoid hemorrhage, which was rapidly fatal in all cases. None were associated with a sentinel bleed, as is common with aneurysms. Three of the patients with subarachnoid hemorrhage had a large varix of the cavernous sinus that extended beyond the normal sinus wall confines. Although three of the patients who suffered fatal subarachnoid hemorrhage demonstrated a large varix of the cavernous sinus, 18 other patients who did not bleed also demonstrated this finding. We define a cavernous sinus varix as a dilated cavernous sinus with extension into the subarachnoid space.

Figure 1 shows a varix of the cavernous sinus involved in a CCF. There is occlusion of the inferior petrosal drainage

and narrowing of the junction of the cavernous sinus with the superior ophthalmic vein. Angiographic differentiation between a cavernous sinus varix and a pseudoaneurysm may be difficult or impossible. In a pseudoaneurysm, the walls of both the carotid artery and cavernous sinus have been torn and only thrombus or brain parenchyma remain. Figure 2 shows a pseudoaneurysm in the fourth patient who suffered a massive subarachnoid hemorrhage and died before treatment could be instituted. The onset of the pseudoaneurysm is usually coincidental with the trauma, as opposed to the delayed onset of a varix, which develops after the occlusion of venous outflow pathways.

Four patients suffered intracerebral hemorrhage; one had a fatal pontine bleed, another had a temporal lobe hemorrhage, one had temporal and frontal lobe hemorrhages, and the last had multifocal hemorrhages. Each of these patients exhibited cortical venous drainage from the fistula, which correlated with the site of the hemorrhage. All four patients had occlusion of the normal outflow pathways of the cavernous sinus and diversion of flow into cortical veins. Figure 3 shows cortical venous drainage from a direct CCF in a patient who subsequently developed a frontal and temporal hematoma. Although all four patients who suffered from an intracerebral hemorrhage demonstrated cortical venous drainage secondary to occlusion of outflow pathways, this finding was also observed in seven other patients who did not bleed. All these patients had fluctuating neurologic signs presumed to be from the cortical venous hypertension. After closure of the CCF with detachable balloons there was prompt resolution of symptoms in all patients. The four patients who hemorrhaged suffered no further hemorrhages after their fistulae were closed with detachable balloons.

Five patients (3.2%) had external hemorrhage with one case of otorrhagia and four cases of massive epistaxis. Of the four cases of epistaxis, two were fatal. All four of the cases had a pseudoaneurysm or venous pouch that entered the sphenoid sinus via a communication through a basal skull fracture. Figure 4 is a lateral common carotid angiogram showing a traumatic CCF and a pseudoaneurysm posteriorly with communication into the sphenoid sinus through a clival fracture. The patient suffered a massive fatal epistaxis before treatment could be instituted. The patient with otorrhagia (Fig. 5) had minor external bleeding from dilated draining veins involving the ear canal. The two surviving patients with epistaxis and one patient with otorrhagia were treated with detachable balloon embolization and had no further episodes of hemorrhage.

Twelve patients (7.7%) have subconjunctival hemorrhages from rupture of dilated arterialized veins. None of these patients had a poor outcome. After closing the CCFs in these patients, no further episodes of hemorrhage occurred.

Increased Intracranial Pressure and Cortical Venous Hypertension

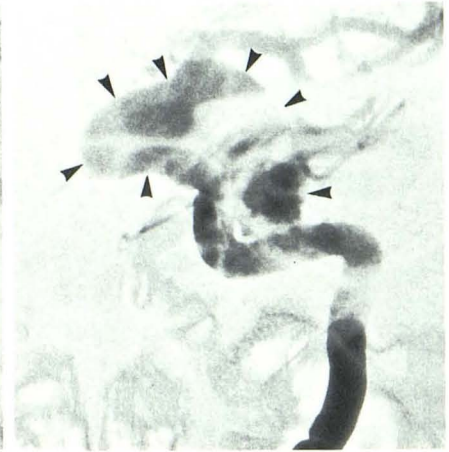
Twelve patients (7.7%) presented with signs and symptoms of increased intracranial pressure, including nausea, vomiting, papilledema, and waxing and waning neurologic signs. Eleven

TABLE 1: Complications in Direct and Indirect Carotid Cavernous Fistulae

Complication	Direct (n = 127)	Indirect (n = 28)	Total (n = 155)
Hemorrhage			
Intracerebral	4 (3.1%)	0	4 (2.6%)
Epistaxis	4 (3.1%)	0	4 (2.6%)
Otorrhagia	1 (0.8%)	0	1 (0.6%)
Subarachnoid	4 (3.1%)	0	4 (2.6%)
Total	13 (10.2%)	0	13 (8.4%)
Increased intracranial pressure	11 (8.7%)	1 (3.6%)	12 (7.7%)
Decreased visual acuity	41 (32.3%)	7 (25.0%)	48 (31.0%)
Blindness	4 (3.1%)	3 (10.7%)	7 (4.5%)
Rapidly progressive proptosis	2 (1.6%)	0	2 (1.3%)
Cerebral ischemia	1 (0.8%)	0	1 (0.6%)
Fatalities	5 (3.9%)	0	5 (3.2%)

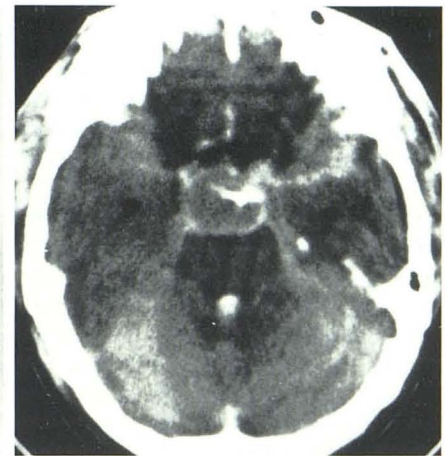
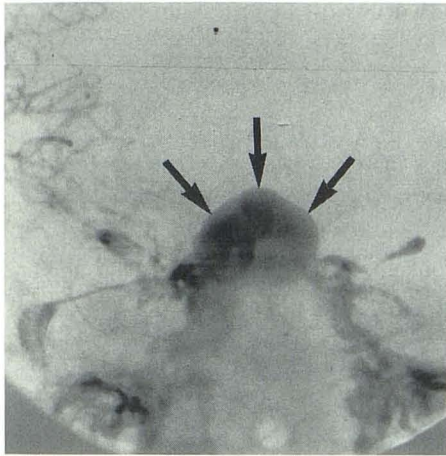
Fig. 1.—A, Internal carotid artery injection, lateral view, showing a varix (arrowheads) of cavernous sinus involved in direct carotid cavernous fistula.

B, Anteroposterior view of same patient. Note extension of varix (arrowheads) outside normal cavernous sinus confines.



A

B



A

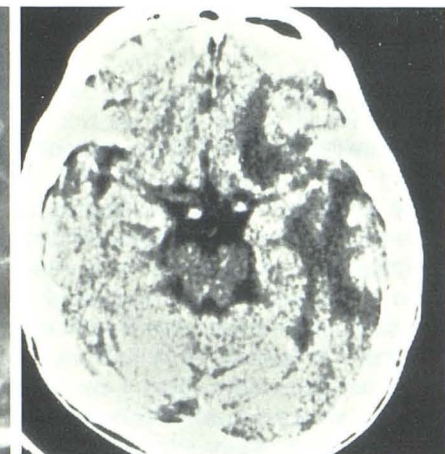
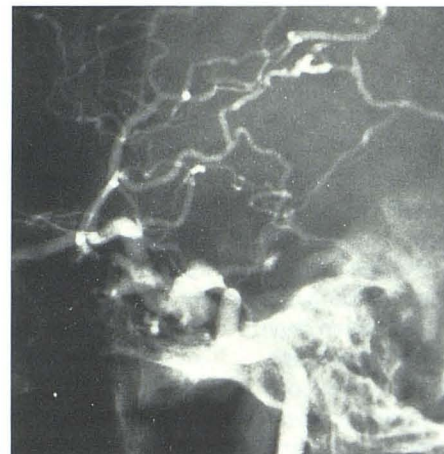
B

C

Fig. 2.—Anteroposterior (A) and lateral (B) internal carotid injection showing pseudoaneurysm (arrows) involving carotid cavernous fistula. C, Unenhanced CT scan at level of pseudoaneurysm shows massive subarachnoid hemorrhage.

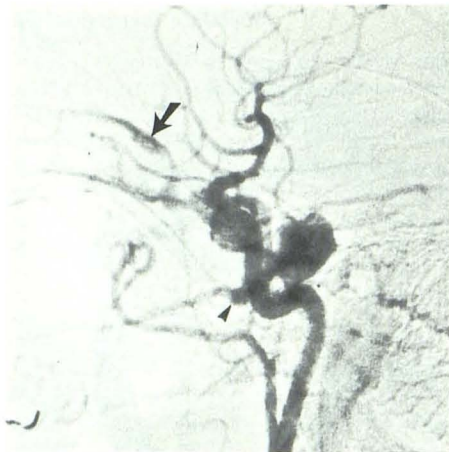
Fig. 3.—A, Lateral internal carotid injection showing cortical venous drainage from a direct carotid cavernous fistula.

B, Noncontrast CT scan with frontal and temporal hematomas.



A

B



4



5

Fig. 4.—Common carotid injection, lateral view. Note arrowhead at site of clival fracture creating a communication of precavernous carotid artery with sphenoid sinus. Arrow indicates drainage to superior ophthalmic vein.

Fig. 5.—Left internal carotid artery injection, lateral view, showing venous drainage from cavernous sinus to superior (arrows) and inferior petrosal sinus. Superior petrosal drainage resulted in engorgement of ear veins, causing otorrhagia.

patients demonstrated cortical venous drainage resulting from occlusion or hypoplasia of the normal cavernous sinus drainage pathways. After closure of the fistulae in these patients there was complete resolution of their symptoms of increased intracranial pressure. Figure 6 shows cortical venous drainage from an indirect CCF that resulted in severe headaches, retroorbital pain, papilledema, and fluctuating level of consciousness.

Elevation of intracranial pressure also occurred in one patient who demonstrated drainage to the sigmoid and transverse sinuses. This patient suffered a traumatic direct CCF and presented with increased intracranial pressure (40 mm Hg) and coma. Angiography demonstrated venous drainage through the inferior petrosal sinuses to the sigmoid sinuses. Figure 7 is a subarachnoid pressure tracing made during balloon occlusions of the fistula. During occlusion of the fistula there was an immediate drop in intracranial pressure and concomitant improvement in the patient's clinical status. However, during detachment, the balloon shifted, and the fistula reopened. This was associated with a prompt increase in intracranial pressure and clinical deterioration. After reclosure of the CCF with a second balloon, the intracranial pressure again fell to normal (7 mm Hg) with immediate improvement in the patient's clinical status.

Decreasing Visual Function

Twenty-five percent (7/28) of indirect CCFs and 32.3% (41/127) of direct CCFs were associated with diminishing visual acuity. Seven (4.5%) of the 155 patients had total visual loss in the involved eye, with bilateral blindness occurring in three patients (1.9%). Minor decreases in visual acuity were reversible with closure of the fistula. Visual decline can progress rapidly, and if it is severe (to the point of light perception only) it is often irreversible, even with prompt closure of the fistula. This event occurred in five of six patients. However, one exception was a patient who progressed to total blindness and was treated 48 hr later with balloon embolization and

closure of the fistula. He gradually recovered vision over the next 4 days to a 20/40 acuity.

Progressive Proptosis

Proptosis occurred in 75% (21/28) of indirect CCFs and in 77% (98/127) of direct CCFs. Rapidly progressive proptosis associated with severe pain and visual deterioration occurred in two patients, and in each case angiography revealed thrombosis of the superior ophthalmic vein distal to the site of the fistula. One patient developed a direct CCF after a motor vehicle accident with few symptoms for a period of 2 years. After a bout of retching and vomiting, he developed severe retroorbital pain, visual loss, and progressive proptosis. Follow-up angiography demonstrated interval thrombosis of the distal superior ophthalmic vein and diversion of venous drainage to cerebral veins (Fig. 8). After the fistula was closed, the patient's symptoms and proptosis abated.

Cerebral Ischemia

Delayed cerebral ischemia occurred in one patient who had a direct CCF treated by internal carotid ligation 30 years before developing repeated transient ischemic attacks. Repeat angiography revealed the fistula supplied by massively enlarged anterior and posterior communicating arteries as well as retrograde flow through an enlarged ophthalmic artery (Fig. 9). Tortuosity of the communicating arteries precluded endovascular treatment; therefore, a surgical clipping of the supraclinoid carotid artery to complete a surgical trapping procedure was attempted. Immediately after applying the clip there was massive cerebral edema and the patient died.

Discussion

Retrospective review of the radiographic and clinical findings of 155 patients with CCF revealed several features associated with a poor clinical outcome.

Fig. 6.—Common carotid injection, lateral view, early (A) and later (B) arterial phase. Arrowheads (B) outline cavernous sinus involved in indirect carotid cavernous fistula with venous drainage to cortical veins.

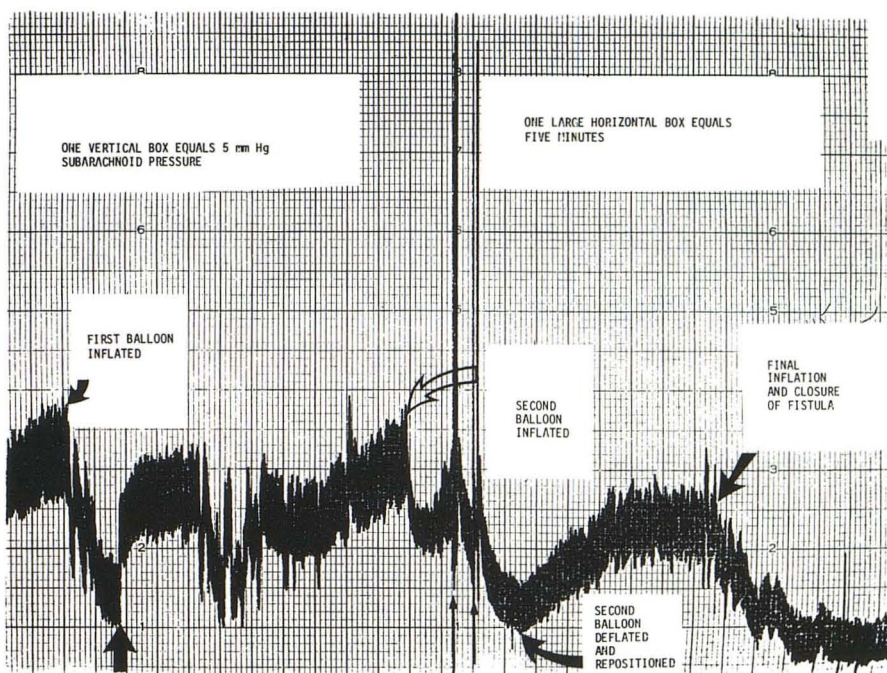
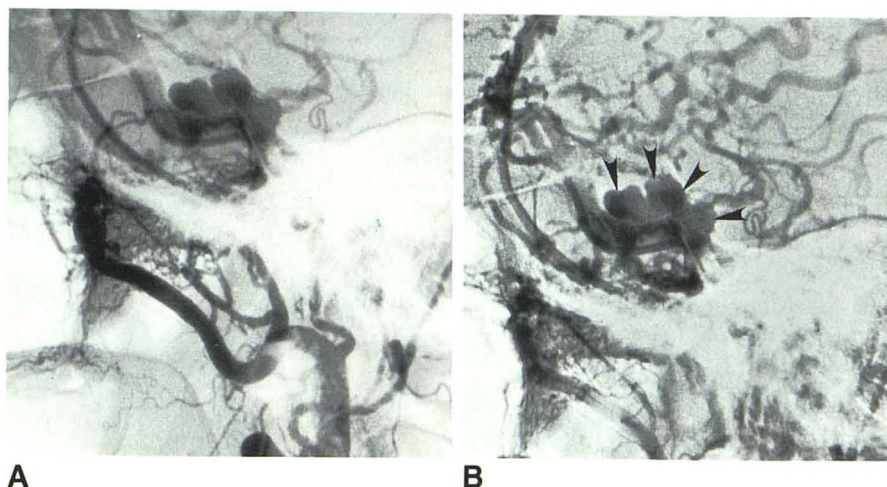


Fig. 7.—Subarachnoid pressure tracing during embolization of direct carotid cavernous fistula. Small curved arrow (top left) indicates the time first balloon was inflated with fistula closure. Straight arrow (bottom left) is the point at which first balloon shifted during detachment with partial reopening of fistula. Curved open arrow (top middle) indicates the time of inflation of second balloon with closure of fistula. Large curved arrow (bottom middle) indicates the time when balloon was deflated and repositioned in cavernous sinus. Slanted arrow (top right) indicates final balloon inflation with complete closure of fistula. Thin straight arrows (bottom middle) indicate artifacts from patient coughing.

Hemorrhage is a devastating complication of a carotid cavernous fistula. The first reported case, in 1908, was by deSchweinitz and Holloway [22]. In 1930, Sattler [23] reviewed 322 cases of CCF reporting the incidence of 1.5% fatal epistaxis and 0.9% intracerebral hematomas. In our series, 13 (8.4%) of 155 patients had hemorrhage, with four (2.6%) having fatal subarachnoid hemorrhage. Although three patients with subarachnoid hemorrhage had a varix of the cavernous sinus, this angiographic sign was not specific, being found in 18 other patients who did not bleed. However, considering the outcome of subarachnoid hemorrhage in our series (100% fatal), we feel that any patient with angiographic findings of a cavernous sinus varix should have emergent

treatment to eliminate the risk of subarachnoid hemorrhage. Patients with angiographic evidence of a pseudoaneurysm have a grave prognosis and should also receive immediate treatment.

All 11 patients with cortical venous drainage were symptomatic with signs and symptoms suggestive of increased intracranial pressure. All had hypoplasia or occlusion of normal cavernous sinus outflow pathways with diversion of venous outflow into the cortical veins. Following closure of the fistula these symptoms abated. Kosnik et al. [24] in discussing symptoms of dural arterial venous malformations stated: "Flow problems in these patients are created more by the increased venous pressure and consequent passive conges-

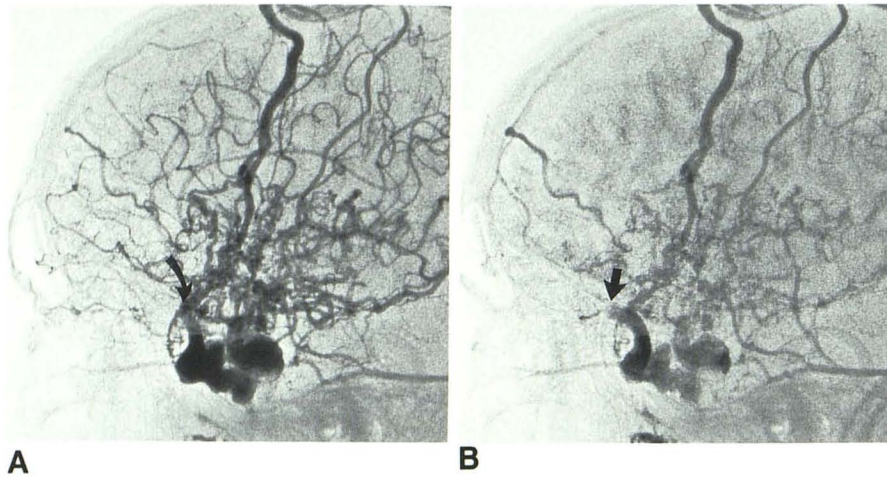


Fig. 8.—A, Mid-arterial internal carotid injection, lateral view, showing thrombosis of mid portion of superior ophthalmic vein (arrow) with diversion of venous drainage from fistula to cortical veins.

B, Late venous phase, same projection. There is stasis in superior ophthalmic vein (arrow).

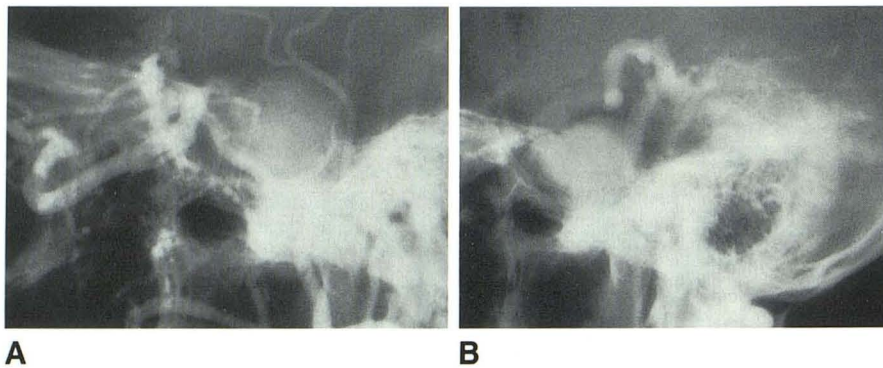


Fig. 9.—A, Common carotid injection, lateral view, shows retrograde flow in ophthalmic artery filling a carotid cavernous fistula.

B, Vertebral artery injection, lateral view, shows enlarged posterior communicating artery supplying fistula.

tion of the brain than by arterial steal." Lasjaunias et al. [11] stated in a review of neurologic manifestations of dural arterial venous malformations that "these symptoms appear to be related to chronic passive congestion due to retrograde increased venous pressure towards the venous drainage routes of the normal brain." Of the 11 patients with cortical venous drainage, all with signs and symptoms of increased intracranial pressure, four suffered intracerebral hemorrhage. Turner et al. [25] reported three intracerebral hematomas associated with CCF, each of which demonstrated cortical venous drainage in the region of the hemorrhage. Therefore, patients who demonstrate cortical venous drainage are at risk for intracerebral hemorrhage and should receive emergent therapy.

Increased intracranial pressure can also be present in patients with fistula drainage to the sigmoid sinuses. Proposed mechanisms of elevated intracranial pressure in dural fistulae involving the sigmoid and transverse sinuses include an increase in cerebral blood volume and dural sinus pressure with concomitant diminution of cerebral fluid absorption resulting in an increase in intracranial pressure [26–28].

Decreased visual acuity is an ominous finding, and, if untreated, may result in blindness. Many factors contribute to the ischemic changes in the orbit associated with CCF. These include venous hypertension and diminished arterial pressure

resulting from reversal of flow in the supraclinoid carotid artery and steal away from the ophthalmic artery into the fistula [29]. In our series, visual decline was observed in 31% (48/155) of patients, with blindness occurring in 4.5% (7/155). Because severe visual loss is often irreversible, and mild degrees of visual loss are completely reversible, we feel that all patients with CCFs should have frequent ophthalmologic examinations as well as daily self-evaluation of visual function. Any visual decline is an indication that urgent treatment is needed.

Rapidly progressive proptosis may signify spontaneous thrombosis of venous outflow pathways to the orbit. Because of the severe symptoms associated with this occurrence, such a finding is an indication for immediate treatment.

Fortunately, cerebral ischemia occurred in only one patient with CCF, but this patient died after surgical closure of the fistula. This symptom may signify the cerebral vasculature's impaired autoregulation secondary to chronic long-standing steal and may indicate the need for staged closure to prevent normal perfusion pressure breakthrough and cerebral edema during treatment.

In conclusion, patients presenting with major external hemorrhage such as epistaxis have a high mortality rate and require emergent treatment. Patients with cortical venous

drainage have symptoms of increased intracranial pressure and are at risk to develop intraparenchymal hemorrhage. These patients should also be given urgent treatment. The angiographic demonstration of a cavernous sinus varix is associated with an increased risk of development of fatal subarachnoid hemorrhage and should be treated immediately. Visual loss associated with CCFs is common, and if treated promptly can be entirely reversible. Severe visual loss, however, is often irreversible; therefore, treatment is warranted for any rapid or severe visual decline. With the recognition of these signs, symptoms, and angiographic features that indicate an increased morbidity and mortality from CCFs, immediate definitive treatment can be instituted to improve the outcome.

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