

Classification and treatment of spontaneous carotid-cavernous sinus fistulas

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✓ An anatomical-angiographic classification for carotid-cavernous sinus fistulas is introduced and a series of 14 patients with spontaneous carotid-cavernous sinus fistulas is reviewed to illustrate the usefulness of such a classification for patient evaluation and treatment. Fistulas are divided into four types: Type A are direct high-flow shunts between the internal carotid artery and the cavernous sinus; Type B are dural shunts between meningeal branches of the internal carotid artery and the cavernous sinus; Type C are dural shunts between meningeal branches of the external carotid artery and the cavernous sinus; and Type D are dural shunts between meningeal branches of both the internal and external carotid arteries and the cavernous sinus. The anatomy, clinical manifestations, angiographic evaluation, indications for therapy, and therapeutic options for spontaneous carotid-cavernous sinus fistulas are discussed.

KEY WORDS • carotid-cavernous fistula • dural cavernous sinus fistula •
intracavernous carotid artery • cavernous sinus • angiography •
arterial embolization • detachable balloon catheter

CAROTID-CAVERNOUS sinus fistulas (CCSF's) are abnormal communications between the carotid artery and the cavernous sinus. They can be classified according to three criteria: 1) pathogenetically into spontaneous or traumatic fistulas; 2) hemodynamically into high-flow or low-flow fistulas; and 3) angiographically into direct or dural fistulas. A classification of CCSF's that is based on the presence or absence of antecedent trauma overlooks the anatomical and hemodynamic aspects of the fistulas, characteristics which have important prognostic and therapeutic implications. Unfortunately, there are no objective criteria for differentiating high-flow from low-flow lesions. This is a subjective distinction based on both angiographic findings and the severity of clinical symptomatology. An angiographic classification, however, provides an objective method for grouping CCSF's, determining prognosis, and planning therapy. We believe all CCSF's can be placed into one of four angiographic categories of abnormal communications (Fig. 1). Type A fistulas are direct shunts between the internal carotid artery (ICA) and cavernous sinus; Types B, C, and D are dural shunts. Type B are fistulas between meningeal branches of the ICA and the cavernous sinus; Type C are dural

shunts between meningeal branches of the external carotid artery (ECA) and the cavernous sinus; and Type D are those between meningeal branches of both the ICA and ECA and the cavernous sinus.

Traumatic CCSF's are almost always Type A direct fistulas formed by a tear in the cavernous portion of the ICA resulting in a high-pressure high-flow anomalous interconnection between the main arterial trunk and the cavernous sinus. This type of fistula rarely resolves spontaneously and requires treatment if there is progressive visual loss or an intolerable bruit or headache, or if a traumatic aneurysm enlarges beyond the cavernous sinus.

Spontaneous CCSF's may angiographically fall into any of these four categories. A spontaneous CCSF may develop following rupture of an intracavernous carotid aneurysm and result in a Type A direct shunt with high-flow characteristics indistinguishable from a traumatic fistula. The majority of spontaneous CCSF's are idiopathic, tend to appear in middle-aged women, and generally present with insidiously progressive glaucoma, proptosis, or a "red eye" — signs and symptoms that are usually less severe than those seen in direct fistulas.¹⁹ These idiopathic CCSF's are usually low-flow and can

Spontaneous carotid-cavernous fistulas

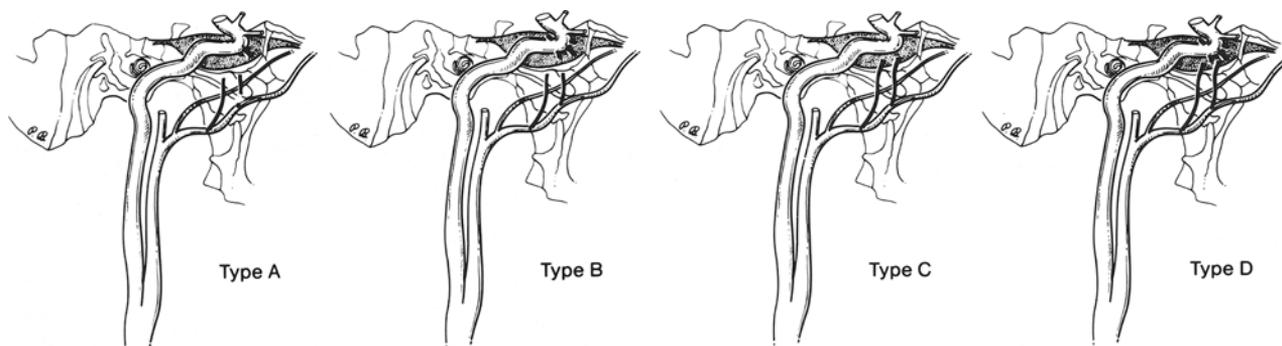


FIG. 1. Illustration of anatomical-angiographic categories of carotid cavernous sinus fistulas. Type A fistulas are direct shunts between the internal carotid artery and the cavernous sinus; Type B, C, and D fistulas are dural shunts. Type B are those between meningeal branches of the internal carotid artery and the cavernous sinus; Type C are those between meningeal branches of the external carotid artery and the cavernous sinus; Type D are those between meningeal branches of both the internal and external carotid arteries and the cavernous sinus.

TABLE 1
Summary of 14 cases of spontaneous carotid-cavernous sinus fistula*

Case No.	Age (yrs), Sex	Other Illnesses	Type of Fistula	Treatment	Result	Follow-Up Period (yrs)
1	53, F	none	B	none	spontaneous resolution	16
2	63, F	none	C	none	spontaneous resolution	16
3	77, F	hypertension	D	none	spontaneous resolution	10
4	65, F	none	B	graded occlusion	good	9
5	64, F	hypertension	B	none	spontaneous resolution	6
6	68, F	hypertension	D on rt; C on lt	embolization of ECA × 3	good	6
7	65, F	hypertension	D	embolization of ECA	good	5
8	77, F	Parkinson's disease	B	none	spontaneous resolution	6
9	81, M	hypertension, diabetes	B	none	death from unrelated cause	1
10	66, F	hypertension	D	embolization of ECA	good	3
11	68, F	none	D	attempted embolization	stroke during angiography	2
12	67, F	none	D	none	poor: blindness	2
13	51, F	bilateral CS aneurysms	A	ICA & fistula occluded with balloon	good	1
14	62, F		A	ICA & fistula occluded with balloon	good	$\frac{3}{4}$

* CCA = common carotid artery; ECA = external carotid artery; CS = cavernous sinus; and ICA = internal carotid artery.

be angiographically divided into Types B, C, or D. Although these spontaneous dural CCSF's have a tendency to resolve without treatment, a sizeable number of patients will suffer progressive loss of vision, diplopia, or intractable glaucoma that warrant interventional therapy.

Table 1 summarizes 14 patients with nontraumatic CCSF's managed at Emory University Hospital. A review of these cases illustrates the usefulness of an angiographic classification of spontaneous CCSF's. Three cases are presented in detail to emphasize certain aspects of clinical presentation, angiographic diagnosis, and therapy.

Summary of Cases

Radiographic Features

There were 15 CCSF's in the 14 patients, because one patient (Case 6) had bilateral fistulas. Two patients

had Type A direct shunts that were angiographically indistinguishable from traumatic CCSF's with rapid transit of contrast material through the fistula and little or no filling of the intracranial circulation. In one patient, the fistula presumably developed after the rupture of a previously demonstrated intracavernous carotid aneurysm. Five patients had Type B dural fistulas fed from meningeal branches of the ICA. There were two Type C dural CCSF's between meningeal branches of the ECA and cavernous sinus. One of these was in a patient who had a Type D CCSF on the opposite side. A total of six Type D fistulas between meningeal branches of the ICA and ECA and the cavernous sinus were found.

The angiographic criteria for differentiating a fistula into high-flow or low-flow categories are quite subjective. High-flow fistulas fill the cavernous sinus and efferent veins within a fraction of a second and the

TABLE 2
Presenting symptoms and signs in 14 patients with
spontaneous CCSF*

Symptoms & Signs	No. of Cases
symptoms	
diplopia	14
eye redness	13
proptosis	10
headache	9
bruit	8
diminished vision	7
facial numbness	6
ocular pain	5
signs	
dilated episcleral veins	14
diminished vision	12
elevated intraocular pressure	10
sixth nerve paresis	10
chemosis	10
third nerve paresis	4
papilledema	4

* CCSF = carotid-cavernous sinus fistula.

intracranial branches of the ICA fill partially or cannot be visualized. An angiogram of a low-flow fistula will reveal slower drainage into the venous system and filling of the intracranial branches of the ICA.

Clinical Features

The presenting symptoms and signs in our 14 patients are outlined in Table 2. Symptoms were usually insidious in onset in patients with dural fistulas. Those with direct Type A fistulas, however, had more severe symptoms that presented more acutely. The length of time between the onset of the first symptoms and radiographic diagnosis of a CCSF ranged from 1 to 18 months with an average of about 5 months. The clinical features are similar to those reported in other series. The most common associated illness in our patients was hypertension, found in seven of the 14 patients. In keeping with other reports of spontaneous fistulas, 13 of our 14 patients were women and all were over 50 years of age, the average age being 66 years.

Treatment Modalities and Results

Of the 14 patients we evaluated, seven received no specific treatment for their CCSF. Five of these patients experienced spontaneous resolution of their signs and symptoms; one died within a year after diagnosis due to unrelated causes, without improvement in the ocular findings, and one patient went blind in the affected eye. The latter patient had extremely tortuous carotid arteries, precluding selective ECA catheterization and planned embolization of the ECA supply to the fistula. One patient suffered a dominant hemisphere stroke during angiography prior to a planned embolization of the CCSF, and no further treatment was attempted. The remaining six patients had interventional treatment because of progressive visual loss, corneal exposure, or

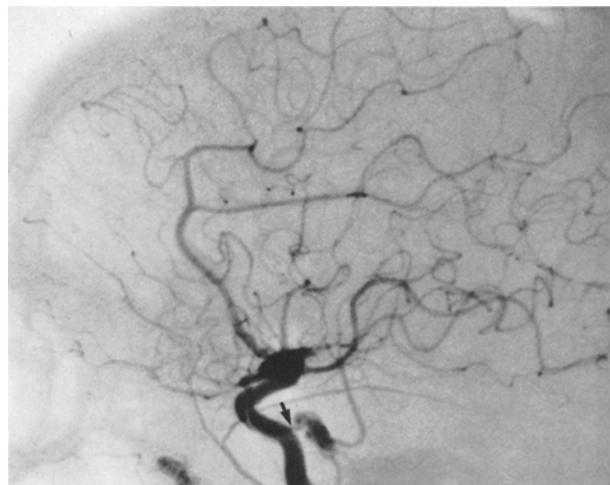


FIG. 2. Case 5. Lateral left internal carotid arteriogram reveals a Type B fistula (arrow) at the posterior portion of the intracavernous carotid artery. The left external carotid arteriogram was normal.

cosmetically offensive proptosis. Three of the six treated patients underwent some form of particulate embolization of the ECA supply to the fistula, all with dramatic resolution of their ocular findings. A graded common carotid artery occlusion using a Crutchfield clamp was performed on one patient, with subsequent resolution of signs and symptoms. The remaining two patients had the fistula and ICA occluded with a Debrun detachable balloon and experienced significant improvement. In both cases, attempts to occlude the fistula with the balloon while sparing the carotid artery failed.

Illustrative Case Reports

Case 5

This 64-year-old woman with controlled hypertension began having left-sided headaches and blurred vision in January, 1976. The symptoms subsided spontaneously for a few months, but abruptly increased one evening when she was awakened from sleep with a severe left hemicranial headache, diplopia, decreased vision in the left eye, a subjective bruit, and redness in the left eye. The patient did not seek medical attention until she developed obvious proptosis, at which time she was diagnosed as having orbital pseudotumor. Steroids had no appreciable effect and she was referred for consultation approximately 1 year after the onset of symptoms.

Physical examination disclosed the following findings in the left eye: 5 mm of axial nonpulsatile proptosis, engorgement and tortuosity of the episcleral veins, and slight loss of visual acuity (20/30). Extraocular movements were full and visual field analysis and funduscopy gave normal results. Intraocular pressures were 19 mm Hg in the right eye and 27 mm Hg in the left. A bruit was present over the left globe.

Spontaneous carotid-cavernous fistulas

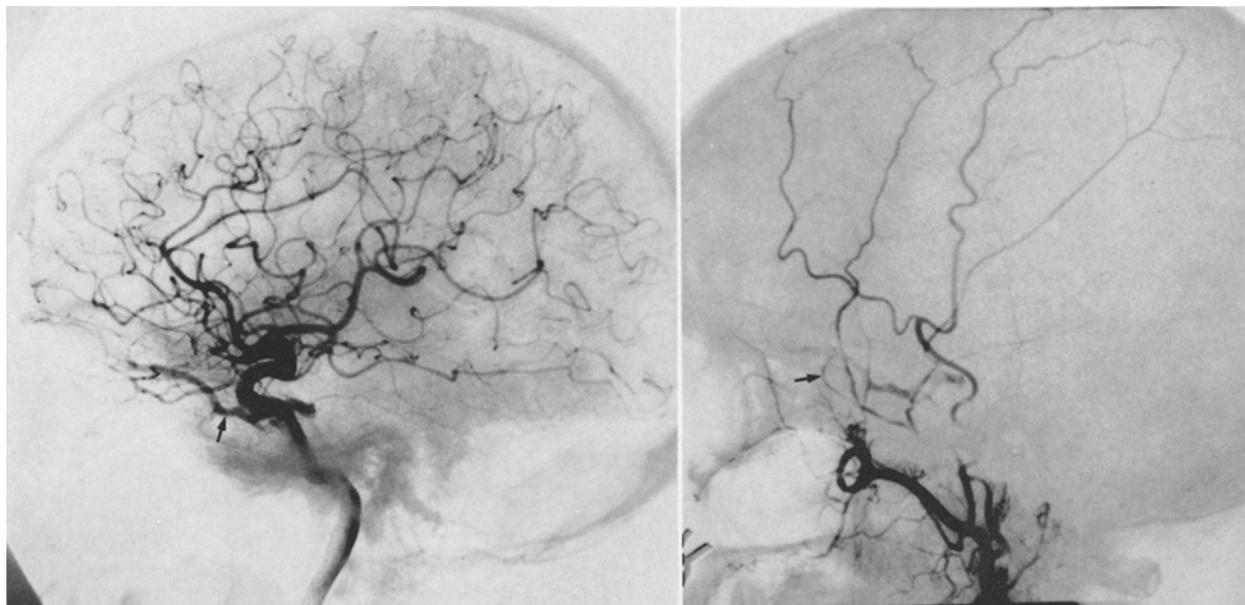


FIG. 3. Case 10. *Left:* Lateral left internal carotid arteriogram showing a Type D fistula with drainage into the superior ophthalmic vein (*arrow*). *Right:* External carotid arteriogram also filling the fistula through a branch of the middle meningeal artery (*arrow*).

A cerebral arteriogram revealed a left Type B CCSF filling from the left ICA with drainage into the ipsilateral superior ophthalmic vein (Fig. 2). The left external carotid and vertebral angiograms were normal. Conservative noninterventional treatment was recommended and all of the patient's symptoms gradually resolved.

Comment. Because we angiographically documented a low-flow low-pressure Type B dural CCSF and the patient had minimal findings, no treatment was recommended. This case illustrates the insidious evolution of symptoms and signs that are characteristic of dural CCSF's, and the frequent occurrence of spontaneous resolution.

Case 10

This 66-year-old woman was seen in consultation 1 week after she developed diplopia and redness of her left eye. A subjective bruit was transiently noticed. There was a past medical history of hypertension.

On examination, the patient had 2 mm of proptosis of the left eye with large tortuous episcleral veins. Visual acuity was 20/25 in the right eye and 20/30 in the left, with perimetric evidence of an inferior arcuate scotoma in the left eye. The left optic disc was swollen and retinal veins were engorged. There was bilateral abduction paresis. No bruit was heard. Cerebral angiography revealed a left Type D CCSF filled by both the left ICA and ECA (Fig. 3).

Two months after discharge, the patient developed an acute change in symptoms. In the left eye, proptosis increased to 7 mm, visual acuity dropped to 20/200 with a left Marcus Gunn pupil, and there was greater

conjunctival and eyelid edema and chemosis. The fundoscopic examination showed greater swelling of the left optic disc with multiple small blot and splinter hemorrhages in both retinas. Intraocular pressures were 15 mm Hg in the right eye and 35 mm Hg in the left. The bilateral abduction paresis persisted unchanged. Again there was no bruit. Repeat cerebral angiography showed no change in the size or drainage pattern of the CCSF compared to the prior examination.

The left middle meningeal and internal maxillary arteries were embolized with Ivalon fragments (Fig. 4). Within 1 week there was dramatic resolution of her previous findings. Proptosis was reduced 3 mm, conjunctival edema resolved, abduction was 90% normal on the right, 60% normal on the left, and there was no edema of the lids or conjunctiva. Visual acuity and intraocular pressures gradually improved; after 1 month, acuity was 20/25 in both eyes, and intraocular pressures were normal bilaterally. At the 2½-year follow-up examination, vision was 20/20 in both eyes and there was no proptosis. The patient complained of mild intermittent diplopia.

Comment. This case illustrates that: 1) Symptoms of a CCSF may be present in the contralateral eye, presumably by drainage of the fistula through the intercavernous sinus to the opposite side. 2) Bilateral selective ICA and ECA angiography is necessary to best outline all of the arterial contributions to these fistulas. 3) At a time when the patient deteriorated clinically, there was no significant change in angiographic appearance of the CCSF. As the symptoms and signs are primarily related to venous hypertension, the exacerbation resulted from a retinal vein occlusion in the left

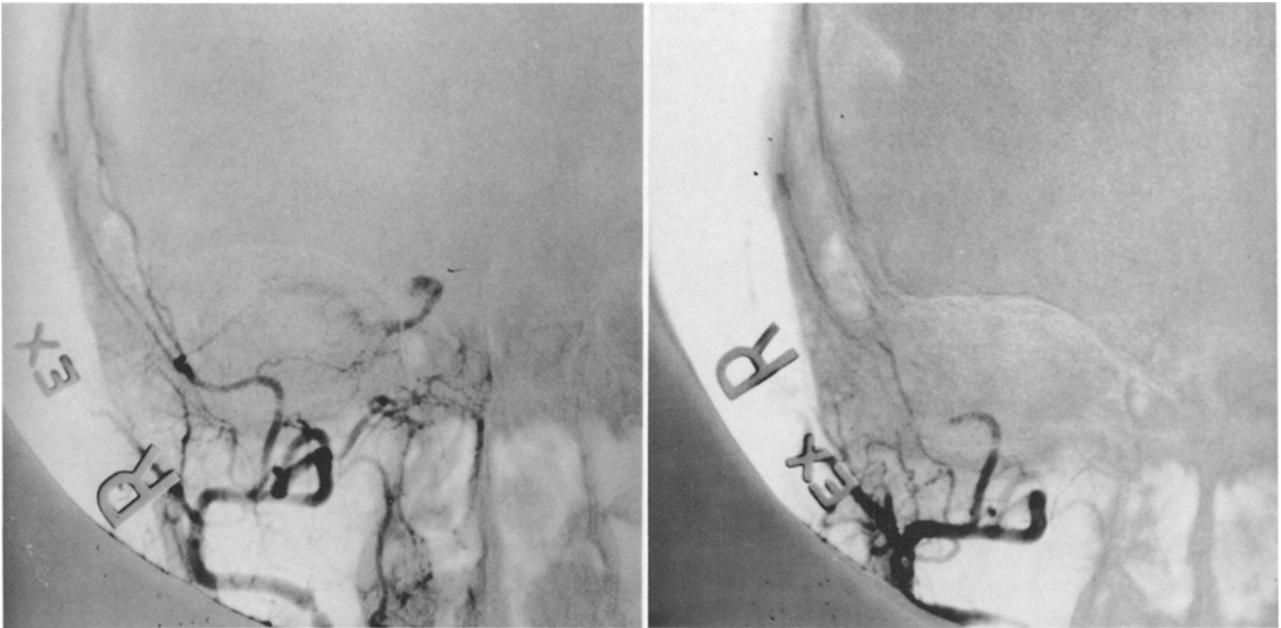


FIG. 4. Case 10. External carotid arteriograms before (*left*) and after (*right*) therapeutic embolization of the middle meningeal and internal maxillary arteries.

eye. 4) At no time during this patient's course was a bruit auscultated; thus, despite serious oculopathy from a low-flow CCSF, a bruit may not be heard which can be a cause of misdiagnosis.³⁴ 5) If the decision not to treat a CCSF is made, the patient must be carefully followed for progressive visual deterioration. 6) Incapacitating headaches or bruit, uncontrollable glaucoma, or evidence of anterior segment hypoxia warrant careful consideration of interventional therapy.

Case 13

This 51-year-old woman began having left-sided headaches which were followed by blurring of vision and intermittent diplopia. Cerebral angiography revealed large bilateral intracavernous ICA aneurysms (Fig. 5 *left*). Gradual occlusion of the left ICA was performed over 7 days using a Crutchfield clamp. The patient tolerated the procedure well, but shortly after the clamp was closed she developed a subjective bruit that increased in intensity over the next few days. Her right eye became proptotic and injected, and she developed blurred vision and diplopia.

Examination showed right-sided proptosis, episcleral vascular congestion, and a right sixth nerve palsy. Visual acuity was normal. A loud bruit, maximal over the right orbit, was also heard over the right side of the neck and head. Cerebral angiography showed a right Type A CCSF (Fig. 5 *right*). The fistula drained through the superior ophthalmic vein, bregmatic vein, and intercavernous sinus to the left cavernous sinus and both jugular veins. The left middle cerebral artery (MCA) and anterior cerebral artery filled from the right carotid

artery. A left common carotid arteriogram documented occlusion of the ICA and identified a superficial temporal artery (STA).

The patient was discharged and monitored as an outpatient. However, progressive proptosis, chemosis, and decreased visual acuity prompted readmission for a therapeutic procedure. With the intent to occlude the right CCSF with a Debrun detachable balloon, and because the patient only had a patent right carotid artery, she underwent bilateral STA-MCA bypasses to augment her blood flow in the event that the right carotid artery had to be sacrificed during balloon occlusion of the fistula. A xenon-133 cerebral blood flow (CBF) study prior to the bypass procedure showed that mean blood flow decreased by only 20% of baseline during manual compression of the right ICA. According to the criteria outlined by Miller, *et al.*,¹⁷ this reduction in flow indicated that the right cerebral hemisphere would likely tolerate acute carotid artery occlusion without prior bypass. Despite the findings, the prospect of possible sacrifice of the only patent carotid artery seemed ominous. It was elected to provide what would theoretically be maximal collateral protection. A left STA-MCA bypass was performed first and the patient was allowed 10 days to recover from this procedure and give the bypass time to mature. The right-sided anastomosis was then done immediately prior to the balloon occlusion. The Debrun balloon was easily placed within the fistula but, upon inflation and detachment, the ICA became nearly occluded. Subsequent injection of the right carotid artery disclosed that the right cerebral hemisphere was adequately supplied by

Spontaneous carotid-cavernous fistulas

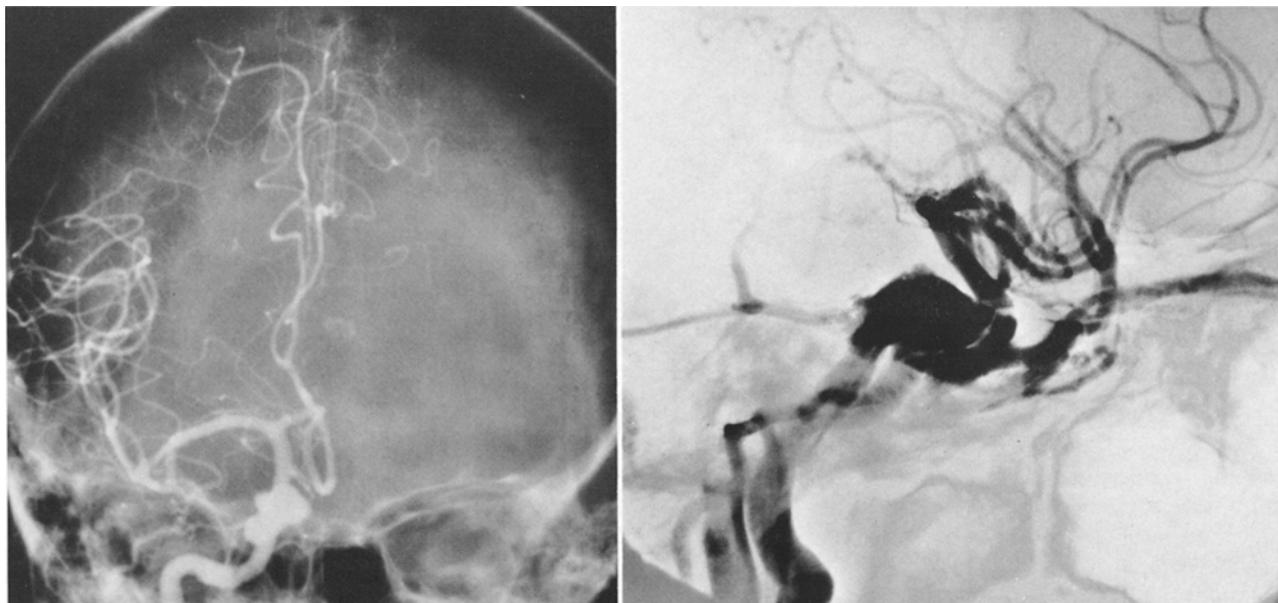


FIG. 5. Case 13. *Left:* Preoperative right internal carotid arteriogram, anteroposterior view, revealing an intracavernous aneurysm. The patient also had a giant left intracavernous aneurysm. *Right:* Postoperative right internal carotid arteriogram, lateral view, showing a Type A fistula at the site where the intracavernous aneurysm had previously been demonstrated.

the right STA-MCA bypass (Fig. 6). The patient experienced immediate resolution of proptosis, chemosis, and bruit, and did not suffer an ischemic deficit.

Comment. The right-sided CCSF in this case presumably developed as a result of rupture of the known right intracavernous carotid artery aneurysm; this is one mechanism for the occurrence of spontaneous CCSF's.²⁰ Prior occlusion of the contralateral ICA may have altered vascular pressure or flow dynamics so as to contribute to rupture of the aneurysm. In this case, the right CCSF was a Type A direct and spontaneous fistula with high-flow characteristics.

This case was quite a challenge in that one carotid artery had already been sacrificed. In treating 54 traumatic CCSF's with detachable balloons, Debrun, *et al.*,¹⁰ had to sacrifice the carotid artery to close the fistula in 20 (37%) of their cases. Although not entirely dependable, there are maneuvers to help determine a patient's ability to tolerate acute ICA occlusion. Miller, *et al.*,¹⁷ have used noninvasive CBF measurements to predict the safety of acute occlusion and found the following: 1) ligation is safe if CBF is more than 40 ml/min/100 gm during carotid occlusion, regardless of change from control flow; 2) ligation is safe if CBF during occlusion ranges between 20 and 40 ml/min/100 gm, provided that the reduction from control flow is no more than 25%; 3) ligation is always unsafe if CBF during clamping is less than 20 ml/min/100 gm, regardless of change from control flow.

Another maneuver is to occlude the ICA with the inflated but undetached balloon for several minutes



FIG. 6. Case 13. Left common carotid arteriogram, lateral view, demonstrating occlusion of the internal carotid artery by the detachable balloon (*arrow*). The middle cerebral artery (MCA) is filling through the superficial temporal artery-MCA bypass.

while examining the patient for evidence of an ischemic neurological deficit. While the inflated balloon is occluding the carotid artery and fistula, the contralateral common carotid artery and the vertebrobasilar circu-

lations can be visualized via the opposite femoral artery not only to determine the collateral circulation but also to be certain that there is no contribution to the fistula from these arteries. It should again be emphasized that these maneuvers do not guarantee that a given patient will tolerate carotid occlusion. An ischemic event has been known to occur hours to days following occlusion of the carotid artery.^{5,22,23,29,38,39} These delayed ischemic events may be secondary to an embolus from the occluded carotid artery or to a low-flow state. Treatment with anticoagulant or antiplatelet medication following carotid artery occlusion may reduce the incidence of embolic complications.

Discussion

Knowledge of the anatomy of the cavernous sinus and the intracavernous ICA is essential to an understanding of the etiology and treatment of CCSF. This anatomy has been studied and reported in detail by Parkinson.²⁶

Traditionally, the cavernous sinus has been regarded as a contiguous network of anatomically separated sinusoids rather than actual veins. A rent in the wall of the intracavernous carotid artery or rupture of one of its branches that traverse and are surrounded on all sides by the sinus cavity, produces an arteriovenous fistula without concomitant venous injury in contradistinction to fistulas elsewhere in the body.⁷

The anatomy of spontaneous low-flow dural CCSF's concerns itself primarily with the small vessels emanating from the intracavernous carotid artery, supplying the regional dura and cranial nerves, and their collateral connections to various branches of the ECA's (Fig. 7). The most proximal trunk of the intracavernous carotid artery which can be implicated in low-flow CCSF's is the meningo-hypophyseal trunk. This fairly constant trunk arises from the apex of the initial proximal curve of the intracavernous carotid artery within the cavernous sinus. It usually gives off three branches: the tentorial artery (artery of Bernasconi-Cassonari), the dorsal meningeal artery to the clivus, and the inferior hypophyseal artery. Of these branches, the dorsal meningeal artery to the clivus is perhaps most often implicated in low-flow CCSF's. This vessel may anastomose within the dura of the clivus with collateral branches from the neuromeningeal trunk of the ascending pharyngeal artery, a branch of the external carotid system. Therefore, this allows a possible route for therapeutic intervention of meningo-hypophyseal feeders via the ECA system.

The trunk of the intracavernous carotid artery most commonly contributing to low-flow dural CCSF's is the artery of the inferior cavernous sinus, also called the "inferolateral trunk" or the "lateral main-stem artery." The artery of the inferior cavernous sinus corresponds to the proximal or carotid remnant of the embryonic dorsal ophthalmic artery.¹⁴ It is identified in 80% to 85% of anatomical specimens.²⁴

The main trunk of the artery of the inferior cavernous sinus arises from the lateral aspect of the intracav-

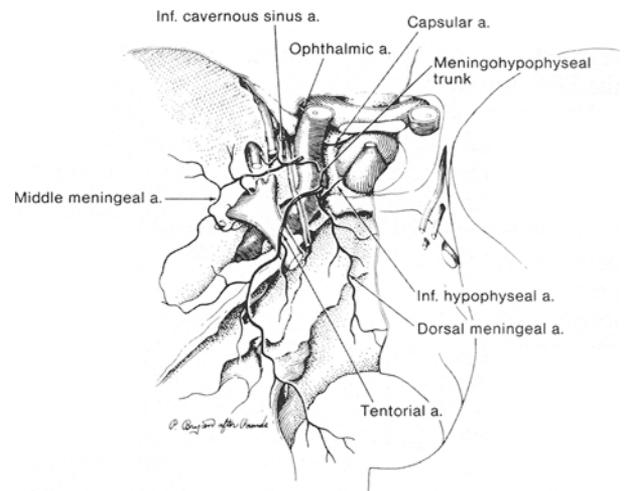


FIG. 7. Diagram showing the normal vascular anatomy as seen from above and the relationships in the region of the cavernous sinus. Adapted with revision from Slusher, *et al.*³³

ernous carotid artery and curves over the sixth cranial nerve, after which it usually gives rise to three branches¹⁴ supplying the dura and cranial nerves in the cavernous sinus region. The first is a superior or tentorial ramus which supplies the roof of the cavernous sinus and the third and fourth cranial nerves as they enter the sinus. This branch may at times also give rise to the tentorial artery instead of the meningo-hypophyseal trunk. The second is an anterior ramus which usually divides into two branches: a medial and a lateral branch. The medial branch courses to the superior orbital fissure region supplying dura and the third, fourth, and sixth cranial nerves as they enter the orbit. This branch may ultimately anastomose with the ophthalmic artery. The lateral branch is more important, and courses to the foramen rotundum which supplies dura in the region of the temporal fossa and terminates at the artery of the foramen rotundum. This branch may anastomose with the distal internal maxillary artery via the artery of the foramen rotundum and with the middle meningeal artery via small temporal rami. The third branch of the inferior cavernous sinus artery is a posterior branch, which is usually evident and which also divides into two rami: a medial branch to the sixth nerve, Gasserian ganglion, and motor root of the fifth cranial nerve, and a lateral branch which also supplies the Gasserian ganglion and adjacent dura. The medial branch courses to the region of the foramen ovale and can anastomose with the accessory meningeal artery of the proximal internal maxillary artery. The lateral branch can anastomose with the cavernous branch of the middle meningeal artery as it emerges from the foramen spinosum.

It is clearly evident that the blood supply to the cavernous region functions as a "balanced" circuit, interconnecting branches of the cavernous carotid artery and external carotid system. It is precisely for this reason that these low-flow dural CCSF's may be

Spontaneous carotid-cavernous fistulas

reached, for therapeutic purposes, through branches of the ECA system.

Four types of CCSF's are distinguishable on anatomical and radiographic grounds: 1) direct high-flow shunts between the ICA and the cavernous sinus (Type A); 2) dural shunts between meningeal branches of the ICA and the cavernous sinus (Type B); 3) dural shunts between meningeal branches of the ECA and the cavernous sinus (Type C); and 4) dural shunts between meningeal branches of both the ICA and ECA and the cavernous sinus (Type D). Peeters and Kröger²⁷ have discussed the first three types of fistula but do not include our Type D. Traumatic CCSF's are direct Type A fistulas with high-flow characteristics. Such a fistula may also occur spontaneously from rupture of an intracavernous aneurysm. The CCSF's of the last three types are dural fistulas, usually occur spontaneously, and generally are low-flow.

The etiology of most spontaneous CCSF's remains speculative. They have been attributed to rupture of a pre-existing intracavernous aneurysm,^{6,20,21,28,34,36} a theory borne out by our Case 13. Although a ruptured intracavernous aneurysm may result in a CCSF, it is an unusual cause and cannot account for the dural type of fistulas. Lie¹⁵ proposed that some of these fistulas are congenital. Taniguchi, *et al.*,³⁴ believed that spontaneous CCSF's may represent a partially thrombosed CCSF in which "the meningeal network in the cavernous sinus is a collateral response to the thrombosing fistula between the internal carotid artery and the cavernous sinus." Newton and Hoyt¹⁹ speculated that spontaneous low-flow CCSF's form after the rupture of one of the thin-walled dural arteries that normally traverse the cavernous sinus, a theory we favor. Extensive preformed dural arterial anastomoses not directly involved in the fistula may contribute collateral blood supply and dilate, resulting in an angiographically demonstrable fistula which can be indistinguishable from a congenital vascular malformation.¹

Certain factors may predispose to the rupture of these small arteries, including pregnancy,^{12,19,26,31,34} arterial hypertension associated with pregnancy,^{35,39} minor trauma, straining,^{19,34} atherosclerotic vascular disease,^{19,33,34} and collagen vascular disease.

It is clear that low-flow dural CCSF's have a relatively high incidence of spontaneous resolution. This incidence ranges from 10% to 60% in various series in the literature.^{11,19,27,33,36} In several cases patients improved shortly after angiography.^{19,33} Five (36%) of our 14 patients experienced spontaneous resolution. Because spontaneous resolution of nontraumatic dural CCSF's may occur, we believe that an initial conservative approach is indicated. Because of the characteristic symptoms and signs of a low-flow CCSF, De Keizer¹¹ proposed that angiography may even be deferred. We think differently, that complete and selective cerebral arteriography provides a definitive diagnosis, illustrates the anatomy of the fistula, and may induce an enigmatic change that leads to spontaneous resolution.^{3,19,37}

If the decision not to treat a CCSF is made, the patient must be carefully followed for progressive visual deterioration. This monitoring should include periodic determination of vision, pupillomotor activity, intraocular pressures, visual fields, proptosis measurements, gonioscopy, and direct and indirect funduscopy.

Based on our experience, we propose the following indications for treatment of a spontaneous CCSF: 1) Visual deterioration. This may result from a combination of reduced arterial perfusion and venous hypertension with accompanying glaucoma. Intraocular pressures may rise as a result of venous hypertension or secondary to rubeosis iridis, a neovascularity of the iris induced by prolonged ischemia which contributes to overall ocular necrosis. 2) Obtrusive diplopia related to vascular engorgement and enlargement of the extraocular muscles or to neural compression within the cavernous sinus. 3) Intolerable bruit or headache. 4) "Malignant" proptosis with untreatable corneal exposure. These indications are not absolute and depend on the general physical condition of the patient, the severity of the symptoms, and the anatomy of the fistula which, in turn, determines the treatment modality. The fact that spontaneous low-flow dural CCSF's are rarely life-threatening emphasizes the importance of minimizing the morbidity and mortality of any therapeutic procedure.

If treatment of a dural CCSF is indicated, an intense angiographic search should be carried out for an ECA supply to the fistula. The angiographic evaluation of dural CCSF's should, therefore, include subselective catheterization and angiography of: 1) the distal internal maxillary artery for visualization of the artery of the foramen rotundum; 2) the proximal internal maxillary artery for visualization of the middle meningeal and accessory meningeal arteries; and 3) the ascending pharyngeal artery. These vessels should be studied using lateral magnification subtraction angiography to map the collateral pathways to the cavernous carotid region for possible therapeutic intervention via transarterial embolization.

Mullan¹⁸ has developed and modified a technique of CCSF occlusion with carotid artery preservation by inserting various thrombogenic materials into the venous side of the fistula. This is a complex procedure with which we have no personal experience. Parkinson²⁵ has advocated a direct surgical approach to the cavernous sinus.

In a CCSF that is supplied entirely by the ECA or that has an ECA contribution, we elect to embolize that component if treatment is indicated. This can be carried out safely and will often induce spontaneous thrombosis of the fistula with a dramatic resolution of symptoms.^{4,13,16,30,36} Various substances can be used as embolic material, including Gelfoam, Silastic beads, isobutyl-2-cyanoacrylate, and Ivalon. Because of the risk of recanalization, we no longer use Gelfoam and prefer Ivalon.

Treatment of a spontaneous dural CCSF fed entirely

by the ICA may be quite challenging and may require sacrifice of the carotid artery. One of our early patients was treated by graded common carotid artery occlusion using a Crutchfield clamp. Although the patient did well, we do not recommend this procedure for treating CCSF's. The fistula itself may remain patent and recruit blood from other sources. Such circumstances increase the risk of cerebral ischemia and preclude the later use of balloon embolization techniques.

The use of detachable balloon catheters has revolutionized the treatment of Type A direct CCSF's.^{2,8-10,32,36} The small-diameter vessels that often constitute the dural fistulas usually do not allow the introduction of a balloon, and the balloon must occlude both the carotid artery and fistula in order to cure the latter. This may be necessary even with the Type A direct CCSF, but is to be expected if one chooses to treat a dural Type B fistula with a detachable balloon.

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Manuscript received May 17, 1984.

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