Spontaneous carotid-cavernous shunts
presenting diagnostic problems

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The diagnostic and therapeutic problems of 11 patients who presented the form fruste of a carotid cavernous fistula, that is, mild proptosis and conjunctival injection sometimes without a bruit, are discussed. Carotid angiography revealed a peculiar arteriovenous shunting in the region of the cavernous sinus, which may be overlooked since it does not resemble the classic picture of a carotid cavernous fistula. All 11 cases were of spontaneous onset. These fistulas appear to be more common in women who are postmenopausal, pregnant, or post-abortive. The etiology of these fistulas is uncertain but several theories are discussed. Conservative treatment is recommended unless there is an exacerbation of symptoms or the patient has a progressive loss of vision.

KEY WORDS: spontaneous carotid cavernous shunts, cavernous sinus, arteriovenous fistula, meningohypophyseal trunk, proptosis

We have been interested in the diagnostic and therapeutic problems presented by certain patients with dural arteriovenous shunts in the region of the cavernous sinus. Some have had the classic signs and symptoms of a carotid-cavernous fistula. Others have had only part of the symptom complex, such as a unilateral headache, proptosis, and conjunctival injection without bruit, or a bruit without other findings. Therefore, these cases have presented diagnostic problems from both the clinical and radiological standpoint; they often have been initially misdiagnosed as cases of Horton's headache, proptosis or sixth nerve palsy of undetermined etiology, unilateral Graves' disease, and orbital tumor. The principal cause of misdiagnosis has been the absence of a bruit. These fistulas have also been overlooked during cerebral angiography, or confused with orbital or nasopharyngeal angiomias and neoplasms.

All 11 of the cases reported here were of spontaneous onset. Three patients were men over 50 years of age. Six of the eight women were postmenopausal, ranging in age from 47 to 71 years. The other two women were in the reproductive period, being age 21 and 28 respectively. Four illustrative case histories are presented, and all of the cases are summarized in Table 1.

Related Anatomical Considerations
The first branch of the intracavernous internal carotid artery, the meningohypophyseal trunk or artery, arises from the dorsal posterior aspect of the initial curve of the internal carotid artery. Parkinson demonstrated it in 100% of his 200 cadaver dissections. Shortly after its origin, it divides into three subdivisions of equal caliber: tentorial, dorsal meningeal, and inferior hypophyseal. The tentorial branch is directed posteriorly and laterally to supply the tento-
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dium and falx, while the dorsal meningeal (or clival) branch goes inferiorly and posteromedially down the clivus to anastomose with its fellow on the opposite side. The inferior hypophyseal branch courses anteromedially or medially to supply the posterior lobe of the pituitary gland and dura over the floor of the sella.

The second branch of the intracavernous carotid artery, seen in 80% of Parkinson's dissections, is the artery to the inferior cavernous sinus. It arises from the inferior lateral aspect of the vessel about 4 to 5 mm in front of the origin of the meningohypophyseal trunk. It supplies the inferior cavernous sinus and the dura, traveling laterally to supply the Gasserian ganglion. Branches of this artery make a direct arterial anastomosis with the middle meningeal and accessory meningeal arteries near the foramen spinosum.

It is important to note that these branches run between the two leaves of the dura. Therefore, if one of them ruptures, there

<table>
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<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Proposis</th>
<th>Conjunctival Injection</th>
<th>Cranial Nerve Involvement</th>
<th>Bruit</th>
<th>Initial Clinical Diagnosis</th>
<th>Angiographic Site of AV Shunting</th>
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<tr>
<td>1</td>
<td>68</td>
<td>+</td>
<td>+</td>
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<td>+</td>
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<tr>
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<td>+</td>
<td>+</td>
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<tr>
<td>5</td>
<td>28</td>
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<td>+</td>
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<td>+</td>
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<td>+</td>
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<td>66</td>
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<td>0</td>
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<td>6th n.</td>
<td>0</td>
<td>internal carotid aneurysm</td>
<td>superior and inferior ophthalmic veins and inferior petrosal sinus</td>
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</tbody>
</table>

TABLE 1

Summary of findings in 11 cases of spontaneous carotid-cavernous fistula
may be a direct communication with an adjacent draining sinus or the cavernous sinus itself.

**Representative Case Reports**

**Case 1**

A 68-year-old white man with diabetes was referred because of left frontal headaches and slight but progressive proptosis in the left eye of 8 months' duration. He had been followed initially with the diagnosis of a vascular headache. The patient had heard a bruit for several months, but it had stopped spontaneously just prior to admission. On examination he had a left proptosis with injection of the conjunctiva; the lids were not edematous. There was a slight left sixth nerve palsy. The fundi contained microaneurysms and exudates. Intraocular tension was 15 in the right eye and 15 in the left eye by applanation. There was no bruit heard on auscultation of the head, eyes, and neck. Carotid arteriography (Fig. 1) demonstrated shunting into the inferior petrosal sinus without opacification of the ophthalmic veins or other tributaries. On cursory examination one could easily mistake the early filling of the inferior petrosal sinus as "petroclinoid ligament calcification." There were tiny strands in the vicinity of the meningohypophyseal trunk. The patient was treated conservatively and is doing well 2 years later.

**Case 2**

A 57-year-old white man was in good health until 2 months before admission when he noted progressive development of a noise in his right ear, verified as a right temporal bruit by his physician. There was no history of trauma. Examination was completely normal except for a loud systolic bruit heard all over the head but loudest in the right temporal region. Specifically, the patient had no proptosis, conjunctival injection, or chemosis. Clinically an arteriovenous malformation over the right mastoid region was suspected. However, the angiogram showed a peculiar arteriovenous shunting in the region of the cavernous sinus. On the early films, the vessels of the meningohypophyseal trunk could be made out as well as opacification of the inferior petrosal sinus (Fig. 2 left). On a later film, contrast material was noted within the cavernous sinus (Fig. 2 right). No treatment has been instituted, and the patient's symptoms remain the same.

**Case 3**

A 54-year-old white woman had a subtotal thyroidectomy at age 18 and had been maintained on desiccated thyroid for a few years, but had then stopped taking her medication. Six months before admission she noted the onset of a sudden severe left-sided headache and thereafter developed proptosis, conjunctival injection, and slight chemosis with continual pain. On examination she had the above signs with no bruit over the orbits or neck. There was a left sixth nerve palsy and the left pupil was slightly larger than the right. The initial diagnosis was Graves' disease, but on ophthalmologic examination, it was found by applanation that the intraocular tension was 25 on the right and 35 on the left. Additional history was then obtained that at the onset of the headache, she had noted a roaring in the left ear which stopped spontaneously after a few weeks. Because of these findings, she had a...
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Fig. 2. Case 2. Left: Arteriogram showing early opacification of meningo-hypophyseal trunk vessels (arrow). Right: Later arterial film demonstrating contrast material within the cavernous sinus.

left carotid arteriogram which revealed early drainage via the inferior petrosal sinus during the arterial phase (Fig. 3 left). The carotid sinus and the superior ophthalmic veins were never visualized. In view of the progressive eye findings and the increased intraocular pressure, a clamp was placed on the left common carotid artery.

Over the next 6 months the proptosis receded and the diplopia and sixth nerve palsy

Fig. 3. Case 3. Left: Arteriogram showing drainage into inferior petrosal sinus only in the arterial phase (arrow). Right: Right brachial arteriogram showing opacification of the left ophthalmic vein during the arterial phase (arrow).

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disappeared. She had only a mild injection of the left eye with 20/20 vision. While in the hospital for evaluation of hypertension and neck pain 1 year after the initial arterial clamping, she suddenly developed acute proptosis of the left eye with marked chemosis and dilatation of the left pupil. A right retrograde brachial arteriogram showed cross filling via the anterior communicating artery into the internal carotid on the opposite side, with subsequent opacification of the left ophthalmic vein (Fig. 3 right). The left internal carotid artery proximal to the intracavernous portion failed to fill with contrast material and was presumed to be thrombosed. Because of the progression of symptoms, a craniotomy was performed with clipping of the internal carotid artery and ophthalmic artery intracranially. The patient has done well in the subsequent 2 years, except for a persistently dilated left pupil.

Case 4

A 49-year-old white woman (postmenopausal) experienced the sudden onset of severe left retroorbital pain 6 weeks before admission. She had associated left lacrimation, nasal congestion, and conjunctival reddening. The headaches had occurred frequently since initial onset, usually in the afternoon, but actually at any time of day or night. After several days she noted a drooping of the left lid. She was diagnosed as having Horton's headaches and treated with methysergide maleate (Sansert). Because of the persistent ptosis, a carotid arteriogram was done at her local hospital 3 weeks after the onset of the headaches; it was interpreted as inconclusive.

The patient was then referred to this hospital for further evaluation. Upon specific questioning she stated that she had heard a swishing sound over her left eye from the onset of the severe headache and that the noise had persisted for about 4 weeks after which it spontaneously disappeared. Her eyes appeared injected during that time, but this was attributed to her cluster headaches. Neurological examination here 2 weeks after the bruit had disappeared was normal. There was no ptosis, pupillary asymmetry, proptosis, conjunctival injection, or bruit. Intraocular tensions were also normal. However, review of the arteriograms from the outside hospital revealed contrast filling of both superior ophthalmic veins (Fig. 4) and the left cavernous sinus during the arterial phase. It is worth noting that the above vessels had filled via a selective external carotid arteriogram.

Fig. 4. Case 4. Selective left external carotid angiogram showing contrast filling of both superior ophthalmic veins (arrows) in the late arterial phase.
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Discussion

Six of the 11 patients in our series did not have a bruit on examination, although nearly all had some degree of proptosis and conjunctival injection (Table 1). If chemosis was present, it was certainly very slight and not at all like the puffy lids which are associated with the classic pulsating exophthalmos. The absence of a bruit was the primary cause of misdiagnosis. It should be noted, however, that four of these six patients with no bruit had a history of a noise in the head which spontaneously disappeared before being seen by a physician.

Cases 5 and 6 were the only ones occurring in young women, the latter becoming symptomatic the day following an uncomplicated delivery and the former developing her symptoms suddenly 3 days after aborting a 3-month-old fetus. Dandy and Follis reported the association of pregnancy with carotid cavernous fistulas in 17 of 41 spontaneous cases, but Hamby had only two out of 27.

In Case 8, the superior ophthalmic vein filled on a single oblique projection only. On six subsequent injections, the angiograms were normal. In Case 9, the orbital bruit disappeared after the second injection of contrast material. Six subsequent injections were again normal.

The above examples led us to believe that at least some of these cases were partially thrombosed, or thrombosing, carotid-cavernous fistulas with one or two outlets remaining. In Cases 8 and 9 it is possible that the injection of contrast material precipitated the thrombosis of the last remaining outlet. Further suggestive evidence is the fact that Cases 3, 5, 6, 8, and 10 had increased intraocular tension in the involved eye. However, Case 1 had normal intraocular tension as did Case 4.

In contrast to Cases 8 and 9, Case 3 demonstrates that a previously nonvisualized (presumably thrombosed) vein may open up. This patient had improved for 6 months following a carotid ligation, then had an acute exacerbation at which time the superior ophthalmic vein was visualized for the first time.

In Case 2 there were no symptoms referable to the eyes, and in Case 4 none when the patient was seen at this hospital. Such examples naturally raise the question as to whether one can have a bona fide carotid-cavernous fistula demonstrable on x-ray without any clinical symptoms. Recently, a 15-year-old retarded boy with intractable seizures was admitted to this hospital in a stuporous state. A left carotid arteriogram done to rule out a subdural hematoma revealed a carotid-cavernous fistula with shunting into both superior ophthalmic veins. After the arteriogram, the eyes and head were re-examined but no proptosis, conjunctival injection, or bruit were detected. His mother stated that at age 4 or 5 he fell out of a car and had been hemiparetic since, so that the etiology here may well be traumatic. Nevertheless, the point to emphasize is that a carotid-cavernous fistula may be asymptomatic.

Case 4 is unique in our series because the dilated superior ophthalmic vein filled after selective external carotid angiography. The concept of anastomoses between the branches of the internal and external carotid arteries is not new. Elschnig in 1893 described these anastomoses after injecting colored fluid into the external carotid artery. Marx (1949) was probably the first to demonstrate the collaterals between the internal and external carotid arteries angiographically. Lie has reviewed the literature on the subject up to 1967. The second of his two case reports is that of a 53-year-old woman with a nonpulsating exophthalmos. A selective left external carotid angiogram demonstrated “a network of vessels” in the region of the cavernous sinus. She had a bruit over the left eye and later developed one on the right. Although no fistula was demonstrated, it was considered an abnormal form of a carotid-cavernous shunt. Lie’s thesis is that these spontaneous lesions are congenital.

In 1963 Hayes described the external carotid-internal carotid artery anastomosis following the Dandy trap procedure for carotid-cavernous fistulas. In three patients he showed that collateral channels opened up between the external carotid and cavernous sinus following surgery, thereby explaining the recurrence of the symptoms and signs.

Newton and Hoyt described 11 cases similar to some of ours. By means of selective external and internal angiography and
subtraction films, they demonstrated the meningohypophyseal trunk in five cases and the artery to the inferior cavernous sinus in three instances. They also demonstrated cases with the arterial supply from the external carotid, namely, the internal maxillary artery in seven cases and the ascending pharyngeal artery in two. In their series, five patients did not have a bruit, and two had the onset of symptoms following abortions.

The etiology of carotid-cavernous fistulas has never been certain. There is evidence that intracavernous aneurysms may rupture to produce a carotid-cavernous fistula. In 1951 Sugar proposed that the residual remnant of the primitive trigeminal artery might be the basis of an intracavernous aneurysm. In essence, he elaborated on Padget's theory of aneurysm formation, that of resolution of the trigeminal artery, leaving a nubbin on the carotid artery from which a presumed leak occurred. Dandy and Follis suggested that spontaneous fistulas may be caused by the rupture of an arteriosclerotic carotid artery into the cavernous sinus. Although this mechanism may play a part in traumatic fistulas, it seems an unlikely explanation of spontaneous ones.

On the basis of our arteriograms and those demonstrated by Newton and Hoyt, we are in accord with their suggestion that a mechanism for the formation of carotid-cavernous fistulas is the rupture of the intradural terminations of the intracavernous branches where many of them inosculate with branches from the other side or with small branches of the external carotid. Probably the greatest contribution made by these detailed serial angiograms has been the serial visualization of the smaller individual branches of the meningohypophyseal trunk and other intracavernous branches which heretofore were not visible in the larger carotid-cavernous fistulas with large, abrupt venous drainages. Case 2, for example, reveals visualization of the meningohypophyseal branches first, then the inferior petrosal sinus (or at least the intradural portion surrounding these branches), and later opacification of the cavernous sinus. This implies perhaps a slow leak from these vessels. In other cases there may be a leak at the site of an anastomotic rete formed between the external and internal carotid artery intradurally in the region of the cavernous sinus.

Whether some of these dural arteriovenous shunts are congenital, as proposed by Lie, remains conjectural. It is certainly interesting that this condition usually occurs in postmenopausal women, older men, or in younger women just after a delivery. Even Lie's Case 2 was in a 53-year-old woman. In the case of the single diabetic patient in our series, it could be suggested that the small vessels were perhaps more arteriosclerotic and brittle and therefore susceptible to rupture. Only one patient (Case 3) was hypertensive. One patient (Case 8), a 67-year-old woman, was in the act of straining at the onset of symptoms. Perhaps straining may also have played a part in the females who noted the bruits after their deliveries. Certainly on the basis of this small series, we cannot say whether diabetes, hypertension, or straining play an etiologic role in these spontaneous cases.

Although the symptoms of a carotid-cavernous fistula may be easy to diagnose by clinical history, the radiological findings may be confusing or so subtle that they are overlooked. The usual rapid venous drainage through enlarged draining veins is not seen. Instead, there is opacification of one or two of the intracavernous branches with or without later visualization of the cavernous sinus. A single inconspicuous drainage route through the inferior petrosal sinus, ophthalmic veins, or superficial Sylvian veins may be the only abnormality noted.

The etiology of these lesions is uncertain. However, it is possible that they represent partially thrombosed carotid-cavernous fistulas. The role of the intracavernous branches of the internal carotid artery, particularly the meningohypophyseal trunk, is important in the pathogenesis of this condition. Although it may be that these lesions are congenital as suggested by Lie, the possibility remains that the meningeal network in the cavernous sinus is a collateral response to the thrombosing fistula between the internal carotid artery and the cavernous sinus. In some cases, at least, there may be a break of these small intracavernous vessels between the leaves of the dura, or a leak from the intradural rete formed between the anastomosis of these
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tiny vessels and the branches of the external carotid artery.

Treatment of these cases should be conservative unless there is an exacerbation or unless the patient has progressive loss of vision.

Summary

A peculiar arteriovenous shunting in the region of the cavernous sinus has been illustrated in 11 cases of spontaneous onset. The absence of bruit and mild nature of characteristic symptoms have been emphasized as diagnostic hazards. The arteriographic findings have been described in detail.

References


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