Carotid-cavernous sinus fistulas and venous thrombosis.

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AJNR Am J Neuroradiol 1980, 1 (2) 141-148
http://www.ajnr.org/content/1/2/141

This information is current as of September 3, 2023.
Carotid-Cavernous Sinus Fistulas and Venous Thrombosis

Radiographic signs of cavernous sinus thrombosis were found in eight consecutive patients with an angiographic diagnosis of carotid-cavernous sinus fistula; six were of the dural type and the ninth case was of a shunt from a cerebral hemisphere vascular malformation. Diagnostic features consisted of filling defects within the cavernous sinus and its tributaries, an abnormal shape of the cavernous sinus, an atypical pattern of venous drainage, and venous stasis. Progression of thrombosis was demonstrated in five patients who underwent follow-up angiography. Because of a high incidence of spontaneous resolution, patients with dural-cavernous sinus fistulas who show signs of venous thrombosis at angiography should be followed conservatively.

Spontaneous closure of dural arteriovenous fistulas involving branches of the internal and/or external carotid arteries and the cavernous sinus has been reported by several investigators [1-4]. The cause of such closure has been speculative, although venous thrombosis recently has been suggested as a possible mechanism [3]. This report demonstrates the high incidence of progressive thrombosis of the cavernous sinus associated with dural carotid-cavernous shunts, proposes a possible mechanism of the thrombosis, and emphasizes certain characteristic angiographic features which are clues to thrombosis in evolution, with an associated high incidence of spontaneous "cure."

Materials and Methods

We reviewed the radiographic and medical records of eight consecutive patients studied at our hospital in 1977 who had an angiographic diagnosis of carotid-cavernous sinus fistula. An additional case of a cerebral hemisphere arteriovenous malformation which drained in part into the cavernous sinus was included. Eight of the nine patients were reevaluated clinically at least 1 month after their initial angiography, and five patients had follow-up angiography one or more times. One patient also had orbital venography after initial angiography. The angiography in most cases included bilateral selective internal and external carotid angiographies and also vertebral angiography in some patients.

Observations

Angiographic Features

The cases were divided into three groups according to the initial angiographic appearance of the fistula: (1) dural-cavernous sinus fistula, six cases; (2) direct internal carotid-cavernous sinus fistula, two cases; (3) distant cerebral arteriovenous malformation draining into the cavernous sinus, one case. Significant
angiographic features are summarized in table 1.

Although direct carotid–cavernous sinus fistulas generally are regarded as more common, most fistulas in this series were dural, probably reflecting the referral pattern of our hospital.

Five of the six dural–cavernous sinus shunts were of the low flow, low pressure type [1] and opacified only the ipsilateral cavernous sinus and its venous connections. Four of these shunts involved the posterior part of the cavernous sinus and one, the anterior part. Feeding vessels were dural branches of one or both external and/or internal carotid arteries. The sixth dural-cavernous fistula was posttraumatic, and on the initial examination showed rapid flow from dural branches of both internal and both external carotid arteries into both cavernous sinuses.

The two direct fistulas both drained into the midportion of the cavernous sinus. One was due to a lateral tear of the internal carotid artery and was of the high flow, high pressure type. The other was secondary to a ruptured carotid aneurysm and showed only an intermediate flow rate.

Seven of the nine cases exhibited features suggesting at least partial sinus or venous thrombosis on the initial angiograms, and the other two showed evidence of thrombosis on subsequent angiographic examinations. Features of coexistent venous thrombosis included one or more of the following: (1) apparent deformity of abnormal shape of the opacified part of the cavernous sinus, not related to position of the patient or location of the shunt; (2) intraluminal filling defects within the cavernous sinus and/or tributaries; (3) an atypical venous outflow pattern; (4) unusually slow clearing of the contrast medium from the cavernous sinus, including puddling or layering; and (5) a change in the pattern of venous drainage between angiographic examinations, including the development of collateral pathways. Three patients developed angiographic features of venous thrombosis on the side opposite the fistula.

Clinical Features

The significant clinical features in our patients are summarized in table 2. The classic signs of a carotid–cavernous fistula, including sudden onset of pulsating exophthalmos, a prominent bruit behind the affected eye, marked venous congestion and chemosis, impaired eye movement, and reduced visual acuity were present only in the patient with a traumatic direct internal carotid–cavernous sinus fistula (case 8). The patient with a direct fistula due to a ruptured aneurysm (case 7) had only diplopia and bilateral sixth nerve palsies.

The five spontaneous dural–cavernous shunts all occurred in patients 65 years old or older; four were women. They usually were insidious in onset and had features similar to those observed by Newton and Hoyt [1]. The most common in order of frequency were ipsilateral headache, proptosis, conjunctival injection, elevated intraocular pressure, sixth nerve palsy, and a subjective or objective bruit.

Classic findings of aseptic cavernous sinus thrombosis include headache; sixth and occasionally third, fourth, and fifth nerve palsy; proptosis; decreased visual acuity; chemosis; and venous congestion [5]. Because our patients exhibited most of these findings in various combinations (table 2), it was difficult to determine clinically if and when a dural–cavernous sinus fistula had progressed to partial or even complete sinus thrombosis. Except for bruit, which was present in only some of our patients, the clinical findings of the two entities show considerable overlap. At some time during their evaluation many of our patients demonstrated an acute, usually transient increase in symptoms, particularly headache and chemosis, which probably heralded the onset of venous thrombosis. The patient with posttraumatic direct fistula (case 8) developed signs of cavernous sinus thrombosis on the side opposite the fistula, and the patient with distant cerebral vascular malformation (case 9) exhibited bilateral orbital abnormalities which suggested bilateral cavernous sinus thrombosis.

The five patients with spontaneous dural–cavernous fistulas showed variable degrees of clinical improvement on follow-up examinations. Most symptoms, including the bruit, regressed with time. However, elevated intracranial pressure and diplopia usually persisted much longer, even after the dural fistula was no longer demonstrable at angiography. The patient with posttraumatic dural–cavernous fistula (case 6) showed only slight clinical improvement despite interval angiographic changes consistent with thrombosis and partial obliteration of the fistula. The patient with posttraumatic direct carotid–cavernous shunt (case 8) eventually underwent surgical entrapment of the fistula, and the patient with a distant arteriovenous malformation draining into the cavernous sinus (case 9) had the malformation resected. The patient with a direct fistula secondary to a ruptured cavernous aneurysm (case 7) refused clinical and angiographic follow-up.

Representative Case Reports

Case 1

A 65-year-old woman developed right periorbital pain and headache after a “sinus infection” 1 year prior to admission, followed by diplopia 3 months later. Two months before admission she noted proptosis and a “swishing noise” on the right. Physical examination at admission showed right exophthalmos, chemosis, and sixth nerve palsy.

CT scanning demonstrated nontumoral right eye proptosis and prominent enhancement of the right cavernous sinus. Bilateral carotid angiographies revealed a right-sided dural–cavernous fistula draining exclusively into the right inferior petrosal sinus (fig. 1A). Slow flow, puddling, and layering of the contrast agent in the right cavernous sinus (fig. 1B) were interpreted as reflecting partial right cavernous sinus thrombosis. A few hours after angiography, increased swelling and chemosis developed about the right eye. This had improved by the following morning, and the subjective bruit had disappeared. One month later, despite mild residual right proptosis, chemosis, and sixth nerve palsy, repeat angiography showed disappearance of the fistula (figs. 1C and 1D). Clinical examination 4 months later revealed complete resolution of the proptosis and chemosis, and only diplopia on far right lateral gaze persisted.

Case 2

A 74-year-old woman with Parkinson’s disease had gradual onset of “red eye” and painless proptosis on the left, beginning 8 months
TABLE 1: Summary of Angiographic Features in Cavernous Sinus Shunts

<table>
<thead>
<tr>
<th>Type of Shunt/Case No.</th>
<th>Location of Shunt</th>
<th>Feeding Vessels</th>
<th>Pattern of Venous Drainage</th>
<th>Signs of Thrombosis</th>
<th>Angiographic Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dural-spontaneous:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Posterior right CS</td>
<td>Right ICA, right ECA</td>
<td>Right SOV only</td>
<td>Puddling, layering, flow pattern, change between exams</td>
<td>Complete obliteration</td>
</tr>
<tr>
<td>2</td>
<td>Posterior left CS</td>
<td>Left ICA &gt; left ECA &gt; right ICA</td>
<td>Left IOV &gt; Left SOV</td>
<td>Puddling, filling defects—left SOV, flow pattern</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Anterior left CS</td>
<td>Left ECA only</td>
<td>Left SOV, left IOV</td>
<td>Filling defects, flow pattern, change between exams</td>
<td>Decreased flow through shunt, filling defect—left SOV</td>
</tr>
<tr>
<td>4</td>
<td>Posterior left CS</td>
<td>Left ECA &gt; left ECA &gt; right ICA</td>
<td>Cerebral veins &gt; left SPS &gt; left SOV</td>
<td>Flow pattern</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Left basilar plexus</td>
<td>Left ICA &gt; left ECA &gt; right ICA</td>
<td>Left SOV only</td>
<td>Puddling, layering, flow pattern</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Right CS, left CS</td>
<td>Right ICA, right ECA, left ICA, left ECA</td>
<td>Bilateral SOV, IOV, IPS</td>
<td>Change between exams</td>
<td>Progressive incomplete obliteration of both CSs and veins</td>
</tr>
<tr>
<td>Direct-ruptured</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>aneurysm, 7</td>
<td>Mid left CS</td>
<td>Left ICA only</td>
<td>Bilateral IPS only</td>
<td>Flow pattern</td>
<td></td>
</tr>
<tr>
<td>Direct-trauma, 8</td>
<td>Mid right CS</td>
<td>Right ICA only</td>
<td>Bilateral SOV, IOV, IPS, right cerebral veins</td>
<td>Flow pattern change between exams</td>
<td>Progressive obliteration of left CS and veins</td>
</tr>
<tr>
<td>Distant AVM, 9</td>
<td>Mid right CS</td>
<td>Right cerebral veins</td>
<td>Right SOV, right IOV</td>
<td>Flow pattern, change between exams</td>
<td>Obliteration right CS, new collaterals</td>
</tr>
</tbody>
</table>

Note.—Data on six women and three men aged 29–83 years. CS = cavernous sinus; ECA = external carotid artery; ICA = internal carotid artery; IOV = inferior ophthalmic vein; IPS = internal petrosal sinus; SOV = superior ophthalmic vein; SPS = superior petrosal sinus; AVM = arteriovenous malformation.

TABLE 2: Summary of Clinical Findings in Nine Cases of Cavernous Sinus Shunts

<table>
<thead>
<tr>
<th>Onset/Case</th>
<th>Orbital Pain</th>
<th>Noise</th>
<th>Diplopia</th>
<th>Cranial Nerves</th>
<th>Bruit</th>
<th>Retinal Vein Dilation</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Ocular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Right</td>
<td>Left</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gradual:</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>VI</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<td>2</td>
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<td>0</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>VI</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>VI</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>VI</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Sudden:</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>V1</td>
<td>0</td>
</tr>
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<td>+</td>
<td>0</td>
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<tr>
<td>7</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>VI</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>8</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>III, VI</td>
<td>0–VI</td>
<td>+</td>
</tr>
<tr>
<td>9</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Note.—All cases except case 6 had proptosis, chemosis, and conjunctival injection. Only cases 8 and 9 had disc blurring.

* Findings on both sides.

prior to admission. Physical examination 4 months before admission revealed left exophthalmos, conjunctival injection, and increased intraocular pressure. A sudden, marked increase in proptosis and chemosis developed 1 day before admission.

CT scanning showed only left proptosis. Bilateral internal and external carotid angiographies 4 days after admission revealed a shunt from left ascending pharyngeal branches as well as cavernous branches of the right and left internal carotid arteries to the posterior part of the left cavernous sinus (figs. 2A and 2B). Despite its posterior location, the fistula drained exclusively in an anterior direction, into the left inferior and superior ophthalmic veins (fig. 2C). Very slow clearing of contrast medium from the cavernous sinus as well as pudding, layering, and filling defects in the right superior ophthalmic vein suggested evolving venous thrombosis (fig. 2C). Despite an apparent uncomplicated angiographic procedure, the patient developed sudden left hemiparesis 6 hr later, from which she recovered only partially. When she was transferred to a nursing home 1 month later, the proptosis and chemosis had improved but had not resolved completely.

Case 3

An 83-year-old man had sudden onset of left photophobia and eye pain, headache, diplopia, and left ptosis 5 months before hospital admission. All symptoms remitted, except for a slight ptosis. However, 4 weeks prior to admission, the left eye pain recurred, associated with blurred vision, diplopia, tense left periorbital swelling, and left proptosis. He had noted a pulse-synchronous murmur in the region of his left eye for several months. Physical examination showed marked left exophthalmos, mild left ptosis, chemosis, conjunctival injection, lid edema, increased intraocular pressure, and marked retinal venous congestion.

Cerebral angiography revealed a very small dural fistula to the anterior part of the left cavernous sinus (fig. 3A), which drained slowly into the left superior and inferior ophthalmic veins (fig. 3B). The posterior part of the left cavernous sinus was opacified during the venous phase by an uncal vein and did not have any visible communication with that part filled by the fistula (fig. 3C). Because sinus thrombosis-in-evolution was suspected, the patient was treated conservatively. Three months later, the patient no longer had headache, eye pain, diplopia, or bruit but still had mild chemosis, proptosis, and conjunctival injection. Repeat carotid angiography showed only minimal, slow filling of the previously seen fistula, and thrombosis of the intraorbital part of the right superior ophthalmic vein had developed (fig. 3D). Three months later, all eye findings had resolved, except for minimal proptosis.
Case 9

A 30-year-old man with a 7-year history of seizures was otherwise well, until he developed sudden redness and swelling of the right eye 10 days prior to admission. This was followed by gradual right proptosis, occasional blurring of vision on the right, and generalized headaches. Examination revealed marked right conjunctival injection and proptosis and increased right ocular pressure. Funduscopic examination showed a pale optic disc on the right and an indistinct disc margin on the left. There were no spontaneous venous pulsations in either eye.

Right carotid angiography demonstrated a large right posterior cavernous sinus angiography.
Fig. 3.—Case 3. A. Left external carotid angiogram. Fistula from ascending pharyngeal and internal maxillary branches (arrows) to anterior cavernous sinus, demonstrating abrupt posterior cut-off (arrowhead). Internal carotid angiogram was normal. B. Left common carotid angiogram. Same shunt and posterior cut-off (white arrow). Drainage solely into superior and inferior ophthalmic veins (arrows). C, Venous phase. Posterior cavernous sinus opacified by uncal vein (arrow). D. Venous phase of left common carotid angiogram 3 months later. Even slower opacification of fistula and decreased caliber of ophthalmic veins. Intraluminal filling defect in superior ophthalmic vein (arrow).

frontal, high convexity arteriovenous malformation with significant venous drainage into the right cavernous sinus (fig. 4A), which emptied exclusively via enlarged superior and inferior ophthalmic veins (fig. 4B). Left carotid angiography showed findings of partial left cavernous sinus thrombosis (fig. 4C).

The patient continued to have severe right frontal headaches. Seven months later the right proptosis worsened suddenly. Funduscopic examination showed a pale optic disc on the right with distended right retinal veins, and papilledema on the left. There was increased intracocular pressure on the right. Repeat right carotid angiography for preoperative evaluation of the cerebral arteriovenous malformation revealed interval development of right cavernous sinus thrombosis (fig. 4D).

After successful removal of the arteriovenous malformation, the patient continued to have increased intracocular pressure bilaterally, but the eye findings improved gradually and were normal on follow-up examination 3 years later.

Discussion

While some investigators have concluded that the cavernous sinus is largely an unbroken venous channel [6, 7], others believe the sinus is not cavernous, but rather a plexus of venous channels which divide and coalesce [8]. In either case, the main venous connections that normally drain toward the cavernous sinus are the superior and inferior ophthalmic veins, the sphenoparietal sinus, and other direct cerebral veins, and probably the superior petrosal sinus [9]. Usual routes of egress are the inferior petrosal sinus and the plexus of the foramen ovale. Anterior and posterior intercavernous sinuses along the wall of the sella turcica as well as a large basilar plexus behind the dorsum sellae connect the two cavernous sinuses [7].

Although much of our evidence is indirect, we believe that the frequent occurrence of spontaneous "cure" of dural-cavernous fistulas is, in most instances, due to partial or complete thrombosis of the cavernous sinus and/or its tributaries. Venous thrombosis associated with carotid-cavernous shunts is not rare, as emphasized by the angiographic findings of thrombosis in all eight of our cases studied in 1977. Taniguchi et al. [2] reported unusual angiographic findings in 11 cases of spontaneous carotid-cavernous fistulas, some of which were thought to reflect thrombosed, or thrombosing fistulas, with only one or two outlets remaining. Brismar and Brismar [3] presented six consecutive patients with spontaneous carotid-cavernous fistulas draining into the orbital veins, all of whom showed extensive thrombosis of the cavernous sinus and its draining veins at orbital venography. These authors have emphasized the
importance of venography in providing information concerning thrombosis of the orbital veins and sinuses at the skull base [3, 10]. We believe that not only the fistula, but also the venous thrombosis may be recognized at carotid angiography and that venography is not necessary for further evaluation in most instances.

Regarding the etiology of thrombosis, spontaneous obliteration seems to occur most often with the low flow, low pressure dural-cavernous shunt, which is considered secondary to a ruptured cavernous arterial branch of the internal or external carotid artery [1]. The shunt may elevate venous pressure within the involved sinus to a critical point at which flow in one or more of the tributaries which normally drain toward the sinus is decreased, resulting in venous stasis. The venous hypertension also may lead to edema and ischemic and inflammatory change in the surrounding tissues, particularly within the orbit [11]. In some cases, there may be associated damage to the venous wall itself. The hypothesis that a combination of stasis and adjacent tissue damage may lead to venous and/or sinus thrombosis is supported by experimental observations that even brief venous stasis following surgical trauma to tissues adjacent to a vein results in venous endothelial damage [12]. It is mediated by a chemotactic effect, causing leukocytes to adhere to and migrate into the venous wall, resulting in endothelial desquamation and subsequent thrombosis.

Thrombosis associated with elevated intracavernous venous pressure apparently can occur even without a primary adjacent vascular disorder, since cavernous thrombosis occurred in our patient with a distant vascular malformation which drained into the cavernous sinus (case 9). It also is noteworthy that thrombosis seems to have been precipitated in the cavernous sinus opposite a high flow, high pressure, direct carotid-cavernous shunt (fig. 5), probably reflecting elevation of pressure in the opposite cavernous sinus to the point of stasis of one or more tributaries. A similar mechanism may be responsible for the unusually high incidence of thrombosis of intracranial venous sinuses associated with dural arteriovenous shunts involving the transverse sinus [13].

It has been shown experimentally that angiographic contrast media may exaggerate the process of leukocytic accumulation, when surrounding tissues are damaged and venous stasis is induced [14]. Direct effects of the contrast medium on the vascular endothelium as well as the effect on red blood cells, causing clumping and aggregation, may
CAROTID-Cavernous Sinus Fistulas

Several of our patients noted a sudden increase in symptoms on the side of the fistula at some time during their clinical course. In two patients, this abrupt change prompted hospitalization and diagnostic cerebral angiography. Another patient developed increased severity of symptoms immediately after angiography, and a fourth patient developed symptoms in the eye opposite the fistula while angiography showed progressive decrease in venous opacification on that side (fig. 5). Aggravation of findings in these patients included increased ocular pain, decreased visual acuity, diplopia, and lid swelling. The amount of shunting demonstrated at angiography was small compared to the severity of the clinical eye findings. This situation, together with angiographic evidence of some venous thrombosis in all the patients, suggests that the abrupt change in symptoms probably was due to development of venous thrombosis.

Our experiences with cavernous sinus fistulas have led us to several conclusions: (1) the association of venous thrombosis with dural-cavernous shunts is the rule rather than the exception; (2) venous stasis and adjacent tissue injury probably precipitate the thrombosis; (3) angiography (including venography) may hasten the thrombosis; (4) clinical eye findings that are disproportionately marked when compared to the size of the shunt suggest venous thrombosis, a process other than the fistula itself, as a major cause of the symptoms; and (5) a diagnosis of thrombosis in evolution is associated with a high incidence of spontaneous "cure" and warrants a trial of conservative management.

REFERENCES