New Concept in Cavernous Sinus Dural Arteriovenous Fistula
Correlation With Presenting Symptom and Venous Drainage Patterns

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Background and Purpose—An extradurally located cavernous sinus dural arteriovenous fistula (CSDAVF) exhibits different clinical behavior from other dural arteriovenous fistulas (DAVFs) located between 2 dural leaves. The aim of this study is to define angiographic types of CSDAVF associated with presenting symptom (Sx) and venous drainage patterns.

Methods—CSDAVFs during a mean of 23-month follow-up period of 58 patients (17 to 73 years, male:female ratio = 8:50) were retrospectively analyzed. The 3 types of CSDAF, ie, proliferative (PT), restrictive (RT), and late restrictive (LRT) types, were categorized by the degrees and patterns of prominent arteriovenous shunt as well as venous flow. The status of the venous connection with CS and the presenting Sx patterns classified as orbital (OrbSxP), ocular (OcuSxP), cavernous (CavSxP), and cerebral (CerSxP) were associated with angiographic types as well as symptom onset, age, and gender. Correlations of discrete and categorical variables were statistically analyzed using the χ² or Fisher exact test.

Results—PT (n = 23) and RT (n = 23) of CSDAVF were more common than LRT (n = 12) (P = 0.016) in patients with younger than 65 years and were related to OrbSxP (P = 0.015) and CavSxP (P = 0.038) in contrast to LRT to OcuSxP (P = 0.004). Early onset of Sxs was related to the OrbSxP (P = 0.08) and CavSxP (P < 0.001). CerSxP (5%) was noted in RT or LRT. OrbSxP was related to the superior ophthalmic venous drainage (P = 0.026) and CavSxP to the inferior petrosal sinus (P = 0.046) and posterior fossa venous drainages (P = 0.014). Seven patients revealed chronological progression of CSDAVF from PT to LRT and even to complete healing.

Conclusions—CSDAVF presents as 3 distinctive angiographic types and is associated with presenting Sxs and venous drainage patterns. (Stroke. 2005;36:1134-1139.)

Key Words: angiography arteriovenous fistula
between CS and cerebral veins takes place.21 Such a variation of venous drainage in CS and a variety of symptomatology make the optimal therapeutic modality decision and timing difficult in CSDAVF.18,22

We retrospectively analyzed 58 patients with CSDAVF and propose a concept for the typing and progression of CSDAVF related to the presenting Sxs and venous drainage patterns.

Materials and Methods

Patients

We have retrospectively analyzed 58 patients with CSDAVF and reviewed their angiographic findings and medical records during the mean 23-month follow-up. Ages ranged from 17 to 73 years (mean 57; male-to-female ratio, 8:50). Selective angiography of the internal carotid artery (ICA) and external carotid artery and vertebral arteries was obtained by high-resolution biplane digital subtraction angiography. Each patient and/or the patient’s family gave informed consent at the time of the examination.

Angiographic Typing

Based on the cerebral angiographic findings, we defined 3 types of the disease. The proliferative type (PT) revealed numerous arterial feeders to CS. Arterial feeders, especially from the middle meningeal artery and ophthalmic artery, converge around CS in addition to the dural branches of ICA. The number of feeders was not quantified because of many feeders with small size and large number. There was a large amount of arteriovenous shunt (avs), and the shunt flow was relatively rapid while filling CS as well as the antegrade and retrograde venous routes (Figure 1A). The venous phase showed well-preserved antegrade flow into IPS. Both CSs usually filled completely with shunted flow regardless of the involved side of CS. There was a bulging in the sinus wall because of the shunted venous flow (Figure 1B).

PT revealed 2 subtypes according to the location and lesion extent. The diffuse proliferative type (DPT) involved the whole CS. Numerous feeders from ophthalmic artery and middle meningeal artery converging to CS were the characteristic feature of DPT in addition to the dural feeders of the cavernous segment of ICA (Figure 1). The posterior proliferative type (PPT) was confined to the posterior portion of CS (Figure 2). This type usually recruited branches of external carotid artery, as well as the ipsilateral and contralateral ICA. However, some rare cases, as one of the patients in our study, were only supplied by ICA corresponding to the Barrow type B.7

The restrictive type (RT) showed multiple arterial feeders, but not as many as the number of arterial feeders in PTs. Differentiation between PT and RT was that each arterial feeder converging to the wall of the dilated CS could be delineated in RT but not in PT. The venous phase showed obliteration of antegrade flow most commonly of IPS and opening of retrograde flow into SOV and/or cortical veins. AVS still existed, although the flow was not as rapid as in PT. CS wall margin was not delineated completely because of deformity (loss of normal sinus contour) and lobulation of CS wall. Such lobulated change of CS margin seemed to be caused by a deformed outline of the sinus wall caused by fibrosis or change caused by organizing thrombosis.1,2,5

The late restrictive type (LRT) showed only a few arterial feeders with sluggish retrograde venous flow. Constrictive change (pruning) of the draining veins in the retrograde venous outflow was the outstanding feature of this type. There was flow stasis in CS or draining vein even in the late venous phase. Such sluggish venous outflow seemed to be related to the constrictive venous change caused by the long-standing elevated venous pressure.

According to the angiographic types defined, each patient’s angiograms were reviewed by 2 neuroradiologists (S.J.K., J.H.L.) blinded to the clinical information. Each neuroradiologist independently decided on the type of angiography.

Presenting Symptom Patterns

Because the Sxs and signs related to CSDAVF are diverse,23 we divided them into 4 patterns. The orbital Sx pattern (OrbSxP) was caused by the retrograde venous flow and/or pressure from CS including chemois, exophthalmos, periorbital pain, and eyelid swelling. The cavernous Sx pattern (CavSxP) included ptosis, diplopia, anisocoria, and ophthalmoplegia caused by cranial nerve deficits that seemed to be related to bulging or elevated pressure of CS and/or perhaps to the steal phenomenon of blood supply to the cranial nerves.23 The ocular Sx pattern (OcuSxP) related to increased venous pressure draining the eyeball was decreased vision (when
there is a patient symptom or interocular difference of >0.2, ie, 2 lines difference in the Snellen visual acuity chart or its equivalent), increased intraocular pressure (>20 mm Hg or a interocular difference of >5 mm Hg), severe ocular (eyeball) pain, glaucoma, and retinal hemorrhage. The cerebral Sx pattern (CerSxP) was related to reflux of AVS into SMCV or of the petrosal vein into the perimesencephalic vein and cerebellar vein, thus leading to infarction or venous congestion in the basal ganglia, brain stem, or cerebellum leading to seizure or to hemorrhage. Patient age and gender were also correlated with Sx pattern and angiographic typing.

**Statistical Analysis**

Agreement in typing CSDAVF by 2 neuroradiologists was analyzed using \( \kappa \) statistics. The relationship between the angiographic types, Sx and venous drainage patterns, age (older than 65 years), gender, and onset (<1 month, from 1 to 3 months, >4 months) were analyzed using the \( \chi^2 \) or Fisher exact test, as appropriate. Continuous variables (age and onset) were converted to categorical variables. \( P<0.05 \) was considered to indicate a statistically significant difference. The SPSS statistical software package (version 10.1 for Windows) was used for all statistical analysis.

**Results**

**Relation of Angiographic Types and Presenting Symptom Pattern**

Fifty eight patients presented with PT (n=23), RT (n=23), or LRT (n=12) of CSDAVF. Blind review by 2 neuroradiologists for the 3 types revealed excellent agreement (\( \kappa=0.865 \)). The Sx patterns were CavSxP (71%), OcuSxP (64%), OrbSxP (53%), and CerSxP (5%) in order of frequency (Table 1). The angiographic types were correlated to the presenting Sx patterns. PT and RTs were related to OrbSxP (\( P=0.015 \)) and CavSxP (\( P=0.038 \)) in contrast to LRT to OcuSxP (\( P=0.004 \)). PT was related to the absence of OrbSxP in contrast to RT, which was related to the presence of CavSxP (Figure 1). CerSxP (n=3; 5%) were noted in RT or LRT. Two patients revealed venous infarction (Figure 3), and a patient with dilated cortical veins presented with seizure without any brain parenchymal changes. PT revealed DPT in 15 patients and PPT in 8 patients (Table 2).

**Relation of Presenting Symptoms and Venous Drainage Patterns**

The differences in the presenting Sx patterns were related to the venous drainage patterns. OrbSxP was related to the absence of IPS (\( P<0.001 \)) and SOV drainage (\( P=0.026 \)). CavSP was related to the presence of IPS (\( P=0.046 \)) and posterior fossa venous (\( P=0.014 \)) drainage. CerSxP was related to the presence of a CS connection with cerebral or posterior fossa veins (\( P=0.015 \)).
Relation of Angiographic Types and Venous Drainage Pattern
The difference in the venous drainage was related to the presenting type differences because venous drainage pattern was partly included in defining each type (Figure 4).

Onset
Early onset of Sxs <1 month was related to OrbSxP (P=0.08) and CavSxP (P<0.001). Although acute onset of presenting Sxs within 1 month tended to be more common in PT, and subacute onset within 3 months tended to be more common in RT in our study, there was no statistical significance possibly suggesting that the presenting Sxs were more related to other factors such as the shunt flow amount or extent of the lesion than to Sx duration itself.

Gender
There was a tendency toward a female preference (86%) in CSDAVF (Table 2). It is interesting that 15 patients with DPT were only found in females in contrast to PPT in 3 male and 5 female patients (P=0.032). A female preponderance was also noted in LRT.

Progression and Follow-up
We could compare the type change on 16 angiograms from 11 patients (from 1 month to a 6-year follow-up period). Among them, 7 patients revealed change of the type during follow-up. Changes from PT to RT were noted in 3 (4, 5, and 7 months later in each patient). One of these patients received no treatment, one received particle embolization, and one received coil embolization.

Changes from PT into LRT were noted in another 3 patients. One patient did not receive any treatment because of having multiple sclerosis; this patient revealed LRT 13 months later. Another patient with 4 angiographic follow-ups and treated by 2 sessions of arterial particle embolizations, revealed RT 5 months later, and progressed into LRT 2 years later, and was cured 6 years after initial presentation (Figure 5). The other patient revealed LRT on the 19-month follow-up after coil embolization, which had left a small residual flow into the SOV.

Spontaneous cure of LRT was noted in a patient with complete disappearance of the shunt at the time of embolization attempt 4 days after her first angiogram.

There was no change of type in 4 of our patients with RTs (mean 2 months; range from 1 to 4 months). Three patients were followed-up before treatment and 1 patient after partial particle embolization.

Discussion
CSDAVF revealed 3 distinctive angiographic types at the time of presentation: PT (40%), RT (40%), and LRTs (20%). Our type classification was associated with 4 presenting Sxs

<table>
<thead>
<tr>
<th>Type</th>
<th>Male</th>
<th>Female</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proliferative-diffuse</td>
<td>0</td>
<td>15*</td>
<td>15</td>
</tr>
<tr>
<td>Proliferative-posterior</td>
<td>3</td>
<td>5</td>
<td>8</td>
</tr>
<tr>
<td>Restrictive</td>
<td>4</td>
<td>19</td>
<td>23</td>
</tr>
<tr>
<td>Late restrictive</td>
<td>0</td>
<td>12*</td>
<td>12</td>
</tr>
<tr>
<td>Total (%)</td>
<td>7 (14)</td>
<td>51 (86)</td>
<td>58 (100)</td>
</tr>
</tbody>
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*Female preponderance in the diffuse proliferative and the late restrictive types.

Table 2. Gender Difference According to the Angiographic Type of CSDAVF

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*S* Female preponderance in the diffuse proliferative and the late restrictive types.

Figure 4. Diagram showing incidence of venous drainage pattern according to the presenting types. 1 indicates superior ophthalmic vein; 2, superficial middle cerebral vein; 3, perimesencephalic vein; 4, cerebellar vein; 5, inferior petrosal sinus; 6, loss of normal (lobulated) contour of the cavernous sinus; 7, sluggish flow or decreased size and number of the draining veins.
patterns. The early onset of Sxs (<1 month) was related to OrbSxP and CavSxP in contrast to OcuSxP in LRT. There was a female preponderance (86%) in CSDAVF, and DPT appeared only in females. LRT tended to involve relatively older (older than 65 years) female patients.

Compared with the previous classification based on the venous drainage or arterial supply pattern, our typing system shows the overall status of extradurally located CSDAVF, ie, the proliferative change in CS wall, CS configuration, ie, the presence of lobulated scarring or venous occlusion, the fistula flow amount, and venous flow restriction.6,10 The reflux of venous flow into SOV, one of the most common drainage patterns in CSDAVF, is not properly classified in the Cognard classification.10

Four patterns regarding the complex symptomatology of CSDAVF have an advantage in the assessment of CSDAVF and, to the best of our knowledge, have not been previously described. PT and RT tended to be related with OrbSxP and CavSxP in contrast LRT with OcuSxP. Correlation of the Sx patterns with the type of lesion is required in defining the status of CSDAVF. In addition to the emphasis on the venous drainage pattern as described in previous classifications, relationship with OrbSxP, CavSxP, OcuSxP, and CerSxP contributes to the evaluation of the disease status and even of disease progression. More malignant Sx patterns such as OcuSxP and CerSxP require close follow-up or radical management to avoid loss of vision or neurological deficits.

LRT is related to ocular complications when the venous drainage is only to SOV. Whether SOV is the only or just one of the draining veins, close follow-up with ocular pressure measurement and management is needed. Cerebral or cerebellar venous infarction can be followed when there is reflux into the cerebral and/or cerebellar veins. Our study showed that venous infarction is noted in patients (5%) with LRT. It is difficult to explain why some patients in LRT have serious complications, whereas others are able to be healed. Centrifugal venous run-off from CS to the opposite direction of the remaining venous routes seems, in our opinion, to partially affect the patient’s final status. LRT seems to be the most serious type of the disease or the final stage of the progression if it is left untreated.

Analysis of CS confluence is a key factor in analyzing CSDAVF and in predicting the prognosis.10 The physiological process, capture of laterally approaching SMCV draining into tentorial sinus by CS, affects postnatal venous drainage pattern because CS drains into SOV and IPS prenatally.19–21,25,26 Serious neurological complications can be expected if DAVF is not treated in case there is communication between CS and the subarachnoid veins via the sphenoparietal sinus or directly into SMCV or via superior petrosal sinus into the petrosal vein and then into the perimesencephalic and cerebellar veins.17,18 A study performed with 3-dimensional CT angiography in 250 patients revealed that ≈27% of the adult study population did not reveal any communication between CS and the SMCV.25

Subsequent progression from one type of CSDAVF to another was demonstrated in 7 patients. In some patients, CSDAVF seems to progress or to be healed from PT to RT and then finally to LRT, as shown in the diagram (Figure 5). Since Piton et al27 described the progression of DAVF in the lateral sinus, Cognard et al have denied the progression of DAVF, because there was no progression in 84 of their study patients with type I DAVF.10 Davies et al agreed that there is no evidence of progression unless any kind of treatment is engaged.16 However, Cognard et al have reported that 7 patients, 6 of whom were partially treated, experienced an alteration in venous drainage during the follow-up period.28 It is also uncertain why DAVF reveals progression in cases in which partial embolization was performed. Although Satomi et al reported that a benign DAVF carries only a 2% (2 out of 112 cases) risk for cortical venous drainage,18 close follow-up is recommended even with the benign nature of venous drainage when the Sx pattern of CSDAVF changes.

**Conclusions**

CSDAVF presented with 3 distinctive types related to 4 presenting Sx patterns and also the status of CS confluence. This concept associated with presenting Sxs and CS venous drainage patterns provided a practical basis for assessing the disease status and for planning a therapeutic decision.

**Acknowledgments**

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