Spontaneous Angiographic Conversion of Intracranial Dural Arteriovenous Shunt
Long-Term Follow-Up in Nontreated Patients

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Background and Purpose—Dural arteriovenous shunt (DAVS) is a disease in which abnormal arteriovenous communications develop within the dura. Some case series have suggested DAVS may evolve over time, but the natural history is poorly understood. In this study, we aimed to define the incidence and clinical characteristics of patients with DAVS showing spontaneous angiographic pattern conversion.

Methods—We assessed clinical and angiographic features of patients with angiographic conversion without any treatment from a single center database consisting of 335 DAVS cases. Spontaneous angiographic conversion was defined as complete occlusion of a pre-existing DAVS or conversion of a benign into an aggressive lesion on follow-up diagnostic subtraction angiography.

Results—One hundred twelve patients were followed without treatment after the initial diagnosis of DAVS. Overall, we saw pattern conversion on angiography in 18 of the 112 cases (16.1%). Fourteen patients showed spontaneous occlusion of the shunt (12.5%); the most common locations of spontaneous obliteration were the transverse and cavernous sinuses. Four patients showed conversion to an aggressive lesion from benign DAVS (4.0%); all of these cases were associated with occlusion of the ipsilateral draining vein.

Conclusions—DAVS is a dynamic disorder, which will show chronological progression. Spontaneous angiographic obliteration or conversion into an aggressive type may occur on follow-up of untreated DAVSs. (Stroke. 2010; 41:1489-1494.)

Key Words: cerebral angiography • dural arteriovenous fistula • intracranial vascular disorders

Dural arteriovenous shunt (DAVS) is a disease in which abnormal arteriovenous communications develop within the dura. A wide variety of factors have been demonstrated to be associated with DAVS formation such as sinus thrombosis, infections, surgery, postpartum status, and various coagulopathies all may provide a conducive environment for disease development. Angiogenic growth factors also seem to be associated in the genesis of dural arteriovenous fistula. Basic fibroblast growth factor and vascular endothelial growth factors have been shown to be present in the dural walls of these patients. The presence of cortical venous reflux is an important angiographic feature that is implicated with increased intracranial hemorrhagic rates and a grave outcome with annual mortality reaching 10%. Thus, aggressive curative management is recommended for such lesions. On the other hand, DAVSs without cortical reflux are considered benign and conservative management consisting of palliative or observational treatment is advocated. Despite the recent growth in the technical management of this disease, its natural history remains poorly understood. Some isolated case series have hinted that some of these patients may undergo an angiographic pattern conversion on long-term follow-up. However, many of these patients had been treated palliatively and there is still controversy on whether or how often these lesions may actually show an evolutive pattern conversion without any treatment alterations.

The purpose of this study is to define the incidence, clinical, and angioarchitectural characteristics of patients with cranial DAVS showing natural temporal pattern conversions on diagnostic subtraction angiography (DSA) based on the long-term follow-up data from a large single-center database.

Materials and Methods

The patients were selected from a single institution database consisting of 335 intracranial DAVSs (Borden Type I, n=164; II/III, 161)}
Table 1. Summary of Patients Showing Spontaneous Obliteration of DAVS

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex/Age, Years</th>
<th>Location/Borden Type</th>
<th>Initial Symptom(s)</th>
<th>Initial Venous Features</th>
<th>DSA Interval, Months</th>
<th>Symptom Changes</th>
<th>DSA Changes</th>
<th>Associated Clinical Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/47</td>
<td>TS, L/II</td>
<td>Bruit</td>
<td>Intact</td>
<td>76</td>
<td>Resolved</td>
<td>Sinus intact</td>
<td>Clival chordoma operation</td>
</tr>
<tr>
<td>2</td>
<td>M/62</td>
<td>TS, R/II</td>
<td>Bruit</td>
<td>Intact</td>
<td>140</td>
<td>Resolved*</td>
<td>Sinus intact</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>M/66</td>
<td>CS, R/II</td>
<td>Eye swelling, diplopia, brisk</td>
<td>Slow flow, RI PS/SPS occlusion</td>
<td>23</td>
<td>Improved</td>
<td>Venous phase not available</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F/63</td>
<td>CS, bilateral/I</td>
<td>Bruit, injection</td>
<td>Slow flow, intact distal veins</td>
<td>112</td>
<td>Fluctuation</td>
<td>Venous phase not available</td>
<td>Anticoagulation due to DVT</td>
</tr>
<tr>
<td>5</td>
<td>M/62</td>
<td>CS, bilateral/I</td>
<td>Headache, brisk, visual loss</td>
<td>Slow flow, L PS/SPS occlusion</td>
<td>74</td>
<td>Improved</td>
<td>Bilateral CS oblation</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>F/41</td>
<td>TS, L/II</td>
<td>Pontine hemorrhage; 4 months ago</td>
<td>Slow flow, L TS stenosis</td>
<td>55</td>
<td>Improved</td>
<td>L TS stenosis</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>M/51</td>
<td>HG, R/II</td>
<td>Bruit, CN 12 palsy</td>
<td>Intact</td>
<td>12</td>
<td>Fluctuation</td>
<td>Sinus intact</td>
<td>Chemotherapy due to CLL</td>
</tr>
<tr>
<td>8</td>
<td>F/37</td>
<td>TS, L/II</td>
<td>Bruit, occipital pain</td>
<td>Intact</td>
<td>36</td>
<td>Resolved</td>
<td>Sinus intact</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>F/52</td>
<td>JB, L/II</td>
<td>Bruit</td>
<td>Intact</td>
<td>17</td>
<td>Resolved</td>
<td>Sinus intact</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>F/38</td>
<td>HG, R/II</td>
<td>Bruit</td>
<td>Intact</td>
<td>8</td>
<td>Resolved</td>
<td>Sinus intact</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>M/42</td>
<td>CS, R/II</td>
<td>Diplopia, brisk, exophthalmos</td>
<td>Slow flow, RI PS/SPS occlusion</td>
<td>22</td>
<td>Resolved</td>
<td>Venous phase not available</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>M/43</td>
<td>Parietal convexity, L/III</td>
<td>Intact</td>
<td>Intact</td>
<td>114</td>
<td>Resolved</td>
<td>Sinus intact</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>M/45</td>
<td>SSS/III</td>
<td>Parietal hemorrhage</td>
<td>Intact</td>
<td>1</td>
<td>Improved</td>
<td>Venous phase not available</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>F/60</td>
<td>TS, L/II</td>
<td>Intact</td>
<td>Intact</td>
<td>119</td>
<td>Resolved</td>
<td>Venous phase not available</td>
<td></td>
</tr>
</tbody>
</table>

*Development of ipsilateral symptomatic carotid stenosis.

M indicates male; F, female; TS, transverse sinus; L, left; R, right; CS, cavernous sinus; HG, hypoglossal canal; JB, jugular bulb; SSS, superior sagittal sinus; CN, cranial nerve; IPS, inferior petrosal sinus; SPS, superior petrosal sinus; SOV, superior ophthalmic vein; DVT, deep vein thrombosis; CLL, chronic lymphocytic leukemia.

The database has been collected prospectively since 1989 by a team of both neuroradiologists and neurosurgeons in a multidisciplinary clinic. The evaluations included a detailed medical history, full neurological examination, and imaging findings. The diagnosis of DAVS was based on DSA findings.

The standard management protocol for patients with intracranial DAVS at the institution throughout the period was observational or palliative therapy (flow reduction with particles or partial embolization) for patients with intolerable symptoms for benign type of lesions without cortical reflux (Borden Type I); and for aggressive type lesions with cortical reflux (Borden Type II/III): curative endovascular or surgical therapy for disconnection of cortical venous reflux. Manual compression therapy was not implemented on any of the patients. In general, the patients were followed on an annual basis in the multidisciplinary clinic. The patients were instructed to return for an investigation when they noticed any changes of symptoms on follow-up. The mean duration of follow-up for these patients was 40.4 months (range, 1 to 187 months).

Results

The 112 patients with intracranial DAVS (male:female = 41:71; age, mean = 56.5 years; range: 3 to 89 years) who were followed without any therapy were located at the cavernous (n = 41), transverse (n = 37), sigmoid (n = 8), jugular bulb (n = 7), foramen magnum (n = 7, including the hypoglossal canal and marginal sinus), and others (n = 9). Three cases showed synchronous multiple lesions (superior sagittal and transverse sinus, tentorium and transverse sinus, and sigmoid sinus and torcula). Eleven cases were lost to follow-up. The mean duration of follow-up for these patients was 40.4 months (range, 1 to 187 months).

Overall, angiographically demonstrated pattern conversion was seen in 18 of the 112 cases (16.1%, mean age, 50.1 years; male:female = 10:8). In terms of the patients with DSA follow-up, 52.9% (18 of 34 patients) showed angiographic pattern conversion.
Fourteen patients showed spontaneous occlusion of the DAVS (12.5%, 14 of 112 patients; mean age, 50.6 years; male:female = 8:6; Table 1; Figure 1). The mean interval between diagnosis and spontaneous obliteration on DSA was 56.4 months (range, 1 to 140 months). Two shunts located at the superior sagittal sinus and the parietal convexity showed conversion from an aggressive lesion to complete occlusion (Cases 12 and 13). The rest of the spontaneous occlusions were initially benign lesions. The most common location of spontaneous occlusions were the transverse sinus (TS) (n=5) and cavernous sinus (n=4). Other locations included the hypoglossal canal (n=1), jugular bulb (n=1), clivus (n=1), and the superior sagittal sinus (n=1). In all cases of cavernous sinus DAVSs, which showed spontaneous occlusion, the main angioarchitecture of the shunt consisted of small arterial feeders with slow stagnant flow. A case of TS DAVS was associated with progressive occlusion of the shunt and the ipsilateral TS (Case 1). Six cases maintained sinus patency with spontaneous occlusion localized to the fistulous compartment. The initial symptoms had resolved or showed improvement in 12 patients. Two patients had been re-examined due to recent symptom fluctuations. No case of reopening of a spontaneously occluded shunt was observed.

Four patients showed conversion of a benign DAVS to an aggressive lesion (4.0%, 4 of 99 patients; mean age, 48.3 years; male:female = 2:2; Table 2; Figure 2). Changes in symptoms such as loss/change of bruit or newly developed orbital symptoms had prompted the follow-up imaging study in all 4 patients. The mean interval from diagnosis and conversion into an aggressive type was 12.3 months (range, 5 to 23 months). These lesions were located at the TS (n=1), cavernous sinus (n=1), sigmoid sinus (n=1), and hypoglossal canal (n=1). All 4 cases were associated with an underlying or subsequent development of an occlusion of the ipsilateral draining sinus or vein. In a case of TS DAVS, increased flow with dilated feeders in addition to progressive occlusion of the draining suboccipital vein contributed to aggressive conversion (Case 15). Transvenous embolization (Cases 15, 17, 18) or surgical disconnection of the cortical reflux (Case 16) was performed for these patients.

**Discussion**

The analysis of our results has shown that intracranial DAVSs are a dynamic disease that may undergo spontaneous angiographic pattern conversion. According to a long-term follow-up of a large single-center database, approximately 16% of the patients showed angiographic pattern conversion without any treatment.

Various theories have been proposed to explain the angiographic conversions. Awad et al suggested the presence of several stages in the natural history of DAVS with subsequent progression from 1 stage to the next. Sinus thrombosis, which is often implicated in the pathogenesis of the disease, is also frequently associated with its natural evolutive history. Thrombosis of the recipient sinus or the ultimate fibrosis of the intraluminal thrombus and the inflamed sinus wall may cause occlusion of the fistulous communication. Concomitant hemorrhage may promote thrombosis of the shunt by mass effect or vasospasm.

However, not all spontaneous occlusions have been associated with angiographically visible sinus thrombosis. Luciani et al hypothesized that spontaneous closures may occur by changes in the structure of the sinus wall. They proposed that intrinsic compression of the shunts within the sinus wall caused by focal increase in the sinus size may be a key mechanism. Saito et al suggested that changes in flow dynamics such as reduction of shunt flow from the external carotid artery in addition to the venous changes may be an important factor. Despite the angiographic absence of thrombosis, histological study of DAVS lesions suggests that...
microscopic thrombosis is always present and plays a key role in the release of angiogenic growth factors and the pathogenesis of DAVSs.\textsuperscript{5} Our cases show that spontaneous occlusions may occur with or without angiographically visible sinus thrombosis. Thrombosis localized to a compartment or accessory mural channel of the drainage sinus may have caused shunt occlusions with an angiographically intact main sinus.\textsuperscript{28}

Sinus thrombosis, which is often associated with shunt occlusion, could also be the cause of aggressive conversion.\textsuperscript{18} In our patients with aggressive conversion, all cases showed occlusion of the venous outflow as the main pathogenic mechanism of dangerous rerouting. Increase in flow with extension of the shunt seemed to be an additional aggravating factor in 1 of our cases (Case 15).

Cavernous DAVSs deserve special mention because these lesions often manifest the dynamic chronological sequence of this disease.\textsuperscript{17,24} Satomi et al has shown that in some cases of cavernous DAVSs, closure of the posterior drainage routes, inferior petrosal sinus and pterygoid plexus followed by the anterior drainage routes, ophthalmic veins, occur in a chronological progression with some progressing to complete obliteration.\textsuperscript{17} Suh et al proposed a new classification of the cavernous DAVS suggesting a chronological progression from the proliferative type to the restrictive, often with inferior petrosal sinus obliteration, and a late restrictive type, pruning of draining veins, with only few arterial feeders and sluggish retrograde venous flow.\textsuperscript{24} Progressive venous drainage restriction by longstanding elevated venous pressure is a plausible mechanism for such a disease course.\textsuperscript{17,24} It is noteworthy that all the patients with cavernous DAVS in our series who progressed to spontaneous obliteration showed similar features as the late restrictive type. Three of these patients had occlusions of the posterior drainage routes on the initial DSA and later progressed to complete occlusion. However, 1 patient who showed progressive occlusion of the drainage routes transformed into an aggressive type with rerouting to the cortical veins rather than spontaneous occlusion (Figure 2).

Our experience demonstrates the discrepant natural course of the DAVS lesion in regard to the location of the lesion, venous thrombosis, and changes in flow. No specific site was immune from angiographic conversion. Sinus occlusion was associated with both shunt obliteration and aggressive conversion. The underlying venous topography in terms of location of the shunt and underlying cortical venous connection may predict the symptoms and the drainage patterns.\textsuperscript{29} The subsequent development of associated factors such as venous thrombosis, whether it extends into the shunt nidus resulting in occlusion or occludes the venous drainage route and causes dangerous rerouting, or extension/regression of high flow shunts within the underlying individual venous topography may ultimately influence the clinical manifestation and the natural history of the patient.

Although our results show that the incidence of angiographic conversion is not as rare as previously considered, we do not believe that observational management should be advocated for patients with aggressive drainage pattern. The annual morbidity rate is too high and outweighs the probability of long-term spontaneous occlusion. On the other hand, benign lesions with slow flow shunts, especially in the cavernous sinus, were prone to spontaneous occlusion. Thus, aggressive treatment may be avoided for such patients.

A change in symptoms should be considered as an indication for further investigation because it may be a manifestation of an underlying aggressive conversion. The symptoms of DAVS are related to the venous drainage pattern of the shunt.\textsuperscript{24,30} Changes in symptoms may reflect the angiographic flow changes of the DAVS. In fact, more than half of the patients (18 of 34 cases [52.9%]) who were followed with DSA due mostly to symptomatic changes showed angiographic pattern conversion. Although symptomatic resolution was often observed with patients showing obliteration of the shunt, this was not indicative of cure. In 2 cases, the patients who showed resolution of a previous symptom were associated with a rerouted aggressive conversion.

Considering the frequency of spontaneous angiographic conversions, the results of our study call on more vigilance for clinicians in terms of follow-up of these patients, especially those showing symptom changes. It also underscores the importance of a valid noninvasive imaging method for
follow-up of the large proportion of these patients who are followed conservatively.\textsuperscript{31}

The limitation of this study is that a DSA was performed only in patients who developed symptom changes. Because of the timespan of our study, we chose DSA as the gold standard for depiction of angiographic conversions. More recently, it has become our practice to use time-resolved contrast MRI and CT angiography in the follow-up of these patients and any suspicious findings are then investigated with DSA. Once these noninvasive vascular imaging techniques have proven to be equal or superior to DSA, then we will obviously rely on them primarily for guidance of management.

In conclusion, DAVS is a dynamic disease, which will show chronological progression. Spontaneous angiographic obliteration or conversion into an aggressive type may occur on follow-up without any treatment. Multiple factors such as thrombosis, shunt flow, and underlying venous topography were key factors influencing the angiographic outcome.

Disclosures

None.

References


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