## Dural Arteriovenous Fistula Presents Like an Ischemic Stroke

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**Objective:** We report the case of a patient with a dural arteriovenous fistula whose neurobehavioral syndrome was indistinguishable from that of an ischemic stroke.

**Background:** Case studies of dural arteriovenous fistulas primarily describe global cognitive changes like dementia, but detailed neuro-cognitive evaluations of dural arteriovenous fistula patients are rarely reported.

**Method:** We provide a dural arteriovenous fistula case of a patient who presented with aphasia and other symptoms of stroke. Background history, serial neuropsychological data, and angiographic images are presented.

**Results and Conclusions:** Serial neurocognitive data show the extent to which cognitive deficits are reversed with embolization. The case demonstrates that the mechanisms underlying neurocognitive deficits are specific to the fistula's unique hemodynamic features in addition to the location of the dural arteriovenous fistula.

**Key Words:** dural arteriovenous fistula, neurobehavioral manifestations, therapeutic embolization

(Cog Behav Neurol 2004;17:50–53)

Dural arteriovenous fistulas (DAVF) are arteriovenous shunts within the dural layer and account for approximately 1% of all strokes. DAVFs comprise 10 to 15% of cerebrovascular malformations, although some report that up to 20% of arteriovenous malformations (AVMs) are DAVFs.<sup>1</sup> Treatments for DAVFs include neurovascular embolization, radiosurgery, and surgical resection. Combined therapy and staged embolization treatments are also often employed.

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DAVFs can mimic other neurologic disorders and produce a broad spectrum of signs and symptoms, including headache, tinnitus, vertigo, Parkinsonism, and visual field and gait disturbances.<sup>1-4</sup> Studies of the neuropsychological sequelae of DAVFs typically report global changes in cognitive status, usually dementia.<sup>4-8</sup> Detailed neurocognitive evaluations of patients with DAVFs by standardized neuropsychological tests, however, are rarely reported. We present a patient with a DAVF followed with serial neuropsychological evaluations. Findings suggest a pathophysiologic mechanism that may account for similar deficits also seen in brain AVMs.

## CASE REPORT

An 80-year-old, right-handed woman with a 4-month history of rambling in conversations experienced an episode of severe confusion and was brought to an emergency room, where she had a generalized seizure. She was found to have a mild right hemiparesis that resolved within an hour. Brain imaging suggested a vascular malformation in the left temporooccipital region. She was placed on an anticonvulsant.

Two months after her initial presentation to the emergency room, she was referred for angiography and treatment planning and underwent a neuropsychological assessment.<sup>9</sup> Her educational history included a developmental reading problem and 2 years of post high school course work. The neurocognitive evaluation revealed mild to moderate Wernicke aphasia with greater impairment in auditory comprehension than reading, short-term memory loss, and ideational apraxia (Table 1, NE1). The patient was fully oriented on the Mini Mental State Examination (MMSE),<sup>10</sup> but could not spell world backward, name a pencil, follow a 3-step command, recall 2 of 3 words after a brief delay, or copy intersecting pentagons. Her conversational speech was fluent without dysarthria or dysphonia, but the aphasia examination revealed literal and verbal paraphasic errors, occasional perseveration, and a disorder of comprehension. There was mild to moderate impairment in repetition, Right-Left orientation was extremely slowed but accurate. Her writing mechanics suggested apraxia, but her oral reading was intact. With extended time and many self-corrections, she was able to draw the face of a clock with the numbers correctly placed inside a circle, but she was unable to place hands indicating a designated time. Ideomotor praxis was intact. Angiography demonstrated a left sigmoid

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TABLE 1. Neuropsychological Evaluation Test Scores (NE)				
Test <sup>9</sup>	NE 1	NE 2 (+2dy)	NE 3 (+10dy)	NE 4 (+6wk)
MMSE	*22/30	27/30	28/30	27/30
R-L	8/8		8/8	8/8
Commands	*11/15	14/15	15/15	14/15
CI	*7/12		*6/12	*6/12
Repeating	*11/16	*4/8	14/16	15/16
AN	*9		11	11
Oral	9/10		10/10	10/10
Reading	*7/10		9/10	10/10
BN	*35/60		42/60	42/60
Praxis	4/4		4/4	
Calculations	14/16		15/16	

\*Indicates impaired performance. MMSE, Mini-Mental Status Examination; R-L, Right-left discrimination; CI, Complex ideational material; Repeating, Repeating phrases; AN, Animal naming; Oral, Oral sentence reading; Reading, Reading sentences and paragraphs; BN, Boston Naming Test; P raxis, Following 4 commands to gesture object use.

sinus DAVF supplied by bilateral occipital arteries and the posterior division of the left middle meningeal artery. The left sigmoid sinus was completely occluded, and there was retrograde drainage into cortical veins (Fig. 1). Embolization of the left middle meningeal artery using Gelfoam and partial embolization of the right occipital feeder using NBCA glue were performed successfully. Two days post embolization, her MMSE improved to 27/30 (NE2). A test of comprehension employing commands demonstrated some improvement, but repetition remained impaired.

The patient underwent angiography 9 days after the initial treatment, targeting the distal left occipital artery for a second embolization. A superselective Wada test<sup>11</sup> was performed that disclosed no evidence of visual field or cranial nerve deficits, but the tortuosity of the targeted occipital artery and the presence of vasospasm precluded embolization. Another neuropsychological examination 1 day later (10 days after the initial embolization) revealed continued improvement in cognitive functioning (NE3). She showed a significant improvement in language and general cognitive functioning with only a mild Wernicke aphasia. Her MMSE score was improved, and she had fewer paraphasias in conversation as well as improvement in naming, comprehension, and repetition. Angiography, intraoperative arterial exposure, and proximal glue casting followed 6 weeks later. The distal branch feeders of the left occipital artery and the left transmastoid branch feeders were embolized using NBCA glue. Subsequent surgical resection sacrificed the remainder of the distal left occipital artery. Postoperative angiography showed that arterial feeders to the AVM were thrombosed and the DAVF was obliterated. The following day, neuropsychological evaluation revealed that the improvements in cognition seen previously were maintained (NE4).

## DISCUSSION

Our case demonstrates that the neurologic presentation of dural arteriovenous fistulas can mimic focal neurologic disorders of the brain. Although the main lesion was outside of the



FIGURE 1. Angiogram showing venous congestion caused by sinus occlusions (occluded unopacified transverse sinus, A, dotted line indicates sinus) and concomitant dural fistula. The congestion is centered over the posterior temporal lobe with retrograde venous drainage from isolated sinus segment (isolated segment of transverse sinus, B), back through the vein of Labbe C), into the middle cerebral vein (D), and into vein of Trolard (E). Additional drainage of congested temporal lobe retrograde to the occipital temporal vein (F). Arterialized blood supplied to the fistula (fistula rete \*) via shunts fed by the middle meningeal artery (G) and the posterior auricular artery (H).

brain parenchyma, this patient's Wernicke aphasia, transient right hemiparesis, confusion, memory loss, and ideational apraxia were indistinguishable from symptoms found in patients with focal brain lesions from stroke, brain tumors, or infection. On presentation to the emergency room, the rapid resolution of her symptoms raised the possibility that the deficits were postictal. The patient's family reported that prior to her emergency room presentation, she was rambling in conversation. This is suggestive of aphasia, but the etiology is unclear. However, 2 months after her hospitalization, the neurocognitive examination demonstrated a persistent aphasia, making it unlikely that her syndrome was due to a postictal state.

The spectrum of signs and symptoms associated with DAVFs has been attributed to any one or combination of 6 pathophysiologic factors: the arteriovenous shunt, cerebral hypoxia and ischemia, increased venous pressure, retrograde drainage and secondary engorgement of basal sinuses, sinus obstruction, and subarachnoid hemorrhage secondary to involvement of the pial venous system.<sup>12</sup> Passive venous hypertension and/or congestion is often cited as an important etiological factor in the development of deficits in DAVFs.<sup>5,6,13–16</sup> In one series, 20% of the DAVF patients with persistent cortical venous reflux suffered a progressive dementia syndrome due to generalized venous hypertension.<sup>13</sup> Venous hypertension, due to increased inflow, reduction of outflow, or both, can lead to retrograde transmission of pressure into the cortical veins, which in turn impairs parenchymal venous drainage, thereby causing ischemia. Reduced regional cerebral blood flow and increased regional oxygen extraction fraction have been associated with cognitive symptoms, with the impaired hemodynamics presumed to arise from the venous congestion at the site corresponding to the symptoms.<sup>17</sup> Parenchymal circulation delay was also seen in areas with reduced regional cerebral blood flow.

In a review of 100 cases, Awad et al found that DAVF location determined the type of symptoms, but drainage characteristics were responsible for the focal neurologic complications.<sup>18</sup> Lasjaunias et al also related focal central nervous system symptoms to the venous territory and not to the arterial supply characteristics.<sup>14</sup> Focal neurologic deficits in cases of nonhemorrhagic brain AVMs have also been postulated to be the result of venous hypertension rather than "cerebral steal," as has been argued in the past.<sup>19</sup> In the context of DAVFs, meningeal arterial steal has been associated with cranial nerve deficits due to the dural arteries' supply to the cranial nerves.<sup>14</sup> However, since the intracranial internal carotid and vertebrobasilar systems supply the brain, but contribute little to the dura, arterial steal is no longer considered a tenable explanation for symptoms associated with DAVFs.<sup>2,7,14,20</sup>

This case illustrates the effects of embolization on neurocognitive functioning with longitudinal psychometric data.

The DAVF characteristics responsible for the observed neurologic deficits in this patient, we believe, were the bilateral arterial supply from 3 arterial feeders combined with the complete occlusion of the left sigmoid sinus causing significant venous backpressure into the cerebral cortex. This mechanism of venous congestion and consequent hypoperfusion is a recognized mechanism for neurologic deficits found in brain AVMs.<sup>21</sup> Global venous hypertension may present with nonspecific symptoms such as headache and reduced level of consciousness. When the venous pressure is transmitted to a more restricted region, focal symptoms can occur. Ischemic lesions due to arterial occlusions may be indistinguishable clinically from those due to venous occlusion. Arterial ischemic lesions, however, appear on imaging in a specific arterial territory, whereas venous ischemic lesions often cross arterial vascular territories and have associated edema. Arteriography is often needed, as in this case, to diagnose the underlying cause of the ischemia.

In this patient, the presumed reduction of pressure from the arterial side with the 2 arterial embolizations lowered the arterial inflow into the DAVF, resulting in an alleviation of neurologic symptoms. The cognitive deficits and resulting improvements in cognition, however, were associated with areas not adjacent to the embolized arteries but rather in regions distant from the treatment site. A possible mechanism for the neurologic recovery was the normalization of local hemodynamics by embolization in the occipital region and the subsequent reduction of parenchymal ischemia due to reduced venous backpressure in the left temporal lobe.

Various interrelated mechanisms may contribute to the presenting syndromes in any particular DAVF. The mechanisms associated with a patient's symptom profile are specific to the fistula's hemodynamic features and the location of that DAVF, with a behavioral syndrome that may be indistinguishable from that arising from an ischemic stroke.

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