Dural arteriovenous fistula–induced thalamic dementia: report of 4 cases

*Terrence F. Holekamp, MD, PhD,¹ Matthew E. Mollman, BS,¹ Rory K. J. Murphy, MD,¹ Grant R. Kolar, MD, PhD,² Neha M. Kramer, MD,⁵ Colin P. Derdeyn, MD,^{1,4,5} Christopher J. Moran, MD,^{1,4} Richard J. Perrin, MD, PhD,⁶ Keith M. Rich, MD,^{1,4} Giuseppe Lanzino, MD,³ and Gregory J. Zipfel, MD^{1,5}

Departments of ¹Neurological Surgery, ²Immunology, ⁵Neurology, and ⁶Pathology, Washington University School of Medicine in St. Louis; ⁴Department of Radiology, Mallinckrodt Institute of Radiology, Washington University School of Medicine in St. Louis, Missouri; and ³Department of Neurological Surgery, Mayo Clinic, Rochester, Minnesota

Nonhemorrhagic neurological deficits are underrecognized symptoms of intracranial dural arteriovenous fistulas (dAVFs) having cortical venous drainage. These symptoms are the consequence of cortical venous hypertension and portend a clinical course with increased risk of neurological morbidity and mortality. One rarely documented and easily misinterpreted type of nonhemorrhagic neurological deficit is progressive dementia, which can result from venous hypertension in the cortex or in bilateral thalami. The latter, which is due to dAVF drainage into the deep venous system, is the less common of these 2 dementia syndromes. Herein, the authors report 4 cases of dAVF with venous drainage into the vein of Galen causing bithalamic edema and rapidly progressive dementia. Two patients were treated successfully with endovascular embolization, and the other 2 patients were treated successfully with endovascular embolization followed by surgery. The radiographic abnormalities and presenting symptoms rapidly resolved after dAVF obliteration in all 4 cases. Detailed descriptions of these 4 cases are presented along with a critical review of 15 previously reported cases. In our analysis of these 19 published cases, the following were emphasized: 1) the clinical and radiographic differences between dAVF-induced thalamic versus cortical dementia syndromes; 2) the differential diagnosis and necessary radiographic workup for patients presenting with a rapidly progressive thalamic dementia syndrome; 3) the frequency at which delays in diagnosis occurred and potentially dangerous and avoidable diagnostic procedures were used; and 4) the rapidity and completeness of symptom resolution following dAVF treatment.

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INTRACRANIAL dural arteriovenous fistulas (dAVFs) are rare vascular malformations characterized by a direct shunt between dural arteries and a venous sinus and/or cortical vein. Most are idiopathic, though some can be related to prior surgery, trauma, or dural sinus thrombosis.^{8–10,17,30,44,47,55,64,71,80} Modes of presentation include 1) intracranial hemorrhage (ICH) due to cortical venous hypertension, 2) nonhemorrhagic neurological deficits (NHNDs) due to cortical venous hypertension, 3) symptoms of increased sinus drainage including pulsatile tinnitus and ophthalmological phenomenon, and 4) incidental.^{6,7,10} The natural history of dAVFs is strongly linked to the absence or presence of drainage into cortical veins,

which is termed cortical venous drainage (CVD). Borden-Shucart Type 1 dAVFs (those without CVD) rarely present with ICH or NHNDs and have a benign natural history.^{66,68,72} Borden-Shucart Type 2 and 3 dAVFs (those with CVD) commonly present with ICH or NHNDs and can have an aggressive natural history.^{3,9,10,19,24,71,80,81,84,91} In recent years, we⁷² and others^{24,71} have shown that mode of presentation also impacts natural history, as patients harboring dAVFs with CVD who present with ICH or NHNDs are at significantly higher risk for new neurological events as compared with those who present incidentally or with symptoms of increased sinus drainage.^{19,66,68} Based on these data, we proposed a modification to the

ABBREVIATIONS CVD = cortical venous drainage; dAVF = dural arteriovenous fistula; ICH = intracranial hemorrhage; NHND = nonhemorrhagic neurological deficit.

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* Dr. Holekamp and Mr. Mollman contributed equally to this work.

TABLE 1. Modified classification of intracranial dAVFs*

Zipfel	Borden-Shucart Type	Cognard Type	Venous Drainage	CVD	CVH	Presents w/ ICH or NHND	ICH Risk (%)	Death Risk (%)	Treatment
1	I	I, IIa	Dural sinus	No	No	No	<1	0	Elective for intractable symptoms
2A	II	IIb, IIa + b	Dural sinus	Yes	No	No	1.4–1.5	0	Elective to prevent ICH/NHND
2S	II	IIb, IIa + b	Dural sinus	Yes	Yes	Yes	7.4–7.6	3.8	Early to prevent ICH/NHND
3A	III	III, IV, V	Cerebral vein	Yes	No	No	1.4–1.5	0	Elective to prevent ICH/NHND
3S	III	III, IV, V	Cerebral vein	Yes	Yes	Yes	7.4–7.6	3.8	Early to prevent ICH/NHND

CVH = cortical venous hypertension.

* Modified from Zipfel et al: *Neurosurg Focus* 26(5):E14, 2009. This modification to the Borden-Shucart angiographic classification system allows for risk stratification of patients with asymptomatic (2A and 3A) or symptomatic (2S and 3S) presentation.

Borden-Shucart classification system—based not only on angiographic appearance but also mode of presentation—to permit more accurate risk stratification and assist with clinical decision making for the type and timing of dAVF treatment (Table 1).^{68,91}

One relatively underappreciated type of NHND is progressive dementia resulting from dAVF-induced cortical venous hypertension, though correct diagnosis and treatment is increasing, thanks to improved imaging techniques.^{18,79} Dural AVF-induced progressive dementia can be differentiated as either cortical or thalamic in origin—2 categories that have relatively distinct patterns of clinical symptomatology^{18,54,62,80,85} and highly specific venous outflow patterns.^{18,80} Of the two, dAVF-induced thalamic dementia is less frequent, with only 15 published cases reported in the literature to date. All were individual case reports, and several lacked adequate clinical, radiographic, and/or treatment specifics to permit detailed assessment as to manner of presentation, underlying hemodynamic pathophysiology, and long-term outcome.^{18,21–23,32,49,53,62,65,73,76–78,86,88} None provided a comprehensive literature review.

Here, we report 4 cases of dAVF with CVD involving the vein of Galen that presented with a rapidly progressive thalamic dementia syndrome. The specific pattern of presentation, diagnostic imaging findings, method of treatment, and long-term patient outcome as well as a critical review of the literature are provided. Emphasis is placed on the differential diagnosis of this rare condition, the appropriate radiographic workup, the ease at which this condition can be misdiagnosed and/or potentially dangerous diagnostic procedures such as stereotactic biopsy used, and the rapidity and completeness by which the presenting symptoms can resolve.

Case Reports

Case 1

A 53-year-old right-handed man with a remote history of motor vehicle accident was admitted with a 10-day history of worsening confusion and memory problems. His examination revealed profound short- and long-term memory impairment, attention deficit, associative prosopagnosia, moderately severe verbal fluency and comprehension impairments, and emotional lability. He also exhibited constant moderately coherent provoked confabulation.

His Mini-Mental State Examination score was 24/30 (–4 orientation, –2 recall). Brain MRI demonstrated symmetric bilateral thalamic FLAIR signal hyperintensities and subtle, patchy gadolinium enhancement (Fig. 1A–C). Cerebral angiography demonstrated a modified Borden-Shucart Type 2S dAVF located near the confluence of the vein of Galen and straight sinus with arterial supply primarily from both middle meningeal arteries and the inferolateral trunks of the internal carotid arteries (Fig. 1D). Venous drainage occurred via anterograde flow directly into the patent straight sinus, as well as retrograde flow through a vermian vein and the vein of Galen. The dAVF was successfully embolized with ethylene vinyl alcohol (Onyx) using a transarterial approach via the posterior division of the left middle meningeal artery (Fig. 1F).

The patient's mental status began to improve during his hospitalization, and he was discharged home 6 days later. Brain MRI performed 2 years after embolization demonstrated complete resolution of bithalamic edema (Fig. 1E). MR angiography at 1 and 2 years after treatment revealed no evidence of recurrent dAVF. Neuropsychological assessment 1 year after embolization, including Wechsler Memory Scale III, revealed generally average function with only mild to moderate deficits in verbal fluency and memory recall (Working Memory Index 88, Visual Immediate Index 103, Visual Delayed Index 94, Confrontational Picture Naming 59/60, Verbal Memory Recall was 81%). His performance on all other tests was unimpaired. At the 6-year follow-up his Mini-Mental State Examination score was 30/30.

Case 2

A 59-year-old right-handed man was admitted with a 5-day history of altered mental status. His examination revealed severe short- and long-term memory deficits, mild verbal fluency impairment, and associative confabulation. CT demonstrated right thalamic hypoattenuation and subtle left posterior thalamic contrast enhancement (Fig. 2A and B). MRI revealed asymmetrical, bithalamic FLAIR hyperintensities (Fig. 2C). Cerebral angiography demonstrated a modified Borden-Shucart Type 3S tentorial dAVF deriving arterial supply from small branches of the left occipital artery as well as branches of both posterior meningeal arteries (Fig. 2D). Venous drainage was retrograde through an enlarged superior vermian vein, which refluxed into the vein of Galen and both internal cerebral

FIG. 1. Case 1. **A–D:** Images obtained at the time of diagnosis. Sagittal T1-weighted MR image without gadolinium showing no abnormality (A). Sagittal T1-weighted MR image with gadolinium showing bithalamic enhancement (B). FLAIR MR image showing medial bithalamic hyperintensities (C). Left lateral external carotid angiogram showing a modified Borden-Shucart Type 2S dAVF at the confluence of the vein of Galen and straight sinus (*arrow*) with arterial supply from the left middle meningeal artery (*arrowhead*) and venous drainage via anterograde flow into the straight sinus as well as retrograde flow into both internal cerebral veins (*block arrows*) (D). **E and F:** Images obtained 6 weeks after endovascular embolization of dAVF. FLAIR MRI showing resolution of bithalamic hyperintensities (E). Left lateral external carotid angiogram showing complete obliteration of the dAVF (F).

veins. A focally occlusive thrombus within the distal segment of the straight sinus impaired normal outflow. Transvenous endovascular coiling through the straight sinus and vermian vein successfully obliterated the dAVF (Fig. 2F).

The patient's mental status began to improve during

FIG. 2. Case 2. **A–D:** Images obtained at the time of diagnosis. Axial noncontrast and contrast CT scans showing subtle left posterior thalamic enhancement and right thalamic hypoattenuation (A and B); FLAIR MRI showing left thalamic hyperintensities on the right more than on the left (C). Oblique right vertebral artery angiogram demonstrating a Modified Borden-Shucart Type 3S dAVF (*thin arrow*) deriving arterial supply from small branches of the right posterior meningeal artery (*arrowhead*). Venous drainage occurred via an enlarged vermian vein, refluxing through the vein of Galen into both internal cerebral veins (*block arrows*) (D). **E and F:** Images obtained 7 weeks after endovascular embolization of dAVF. Axial noncontrast CT showing resolution of right thalamic hypoattenuation (E). Oblique right vertebral artery angiogram demonstrates complete disconnection of the dAVF (F). A pretreatment postgadolinium MR image and a posttreatment MR image were unavailable.

his hospitalization, and he was discharged to a rehabilitation facility 7 days later. A head CT obtained 7 weeks after treatment demonstrated complete resolution of the thalamic edema (Fig. 2E). He was eventually discharged

home, where his mental status continued to improve, according to his family. Unfortunately, the patient died of a myocardial infarction approximately 3 months after treatment, making further clinical and radiographic assessments impossible.

Case 3

A 60-year-old right-handed man was admitted after 4 days of rapidly worsening confusion and memory deficits. Electroencephalography showed frontal slowing, and brain MRI demonstrated asymmetrical bilateral thalamic FLAIR hyperintensities with moderate heterogeneous gadolinium enhancement (Fig. 3A–C). Due to this enhancement, as well as PET scan results consistent with hypermetabolism, neoplasms such as high-grade glioma and lymphoma were included in the differential diagnosis. This prompted a stereotactic biopsy, which was aborted after obtaining only 2 cores when bleeding was noted from the biopsy needle. CT scanning performed postprocedure showed a small, right thalamic hemorrhage. Pathology revealed moderate gliosis, hyalinized dilated microvascular changes, petechial hemorrhages, and patchy microinfarctions (Fig. 4). Given the patient's clinical history, these histology results were considered potentially consistent with Wernicke encephalopathy. Despite 6 weeks of vitamin supplementation for alcohol abuse, the patient's mental status continued to worsen. Cerebral angiography was then performed, which demonstrated a modified Borden-Shucart Type 3S torcular dAVF supplied by branches of the right occipital artery. Venous drainage was retrograde through a dilated vermian vein into the vein of Galen and internal cerebral veins (Fig. 3D). No filling of the straight sinus was noted, consistent with thrombosis. Transarterial Onyx embolization through the right occipital artery branches was performed, but vermian CVD persisted. Two days later, a midline suboccipital craniotomy was performed for clip ligation of the CVD. Intraoperative angiography demonstrated complete dAVF obliteration (Fig. 3F).

The patient's mental status began to improve during the hospitalization, and he was discharged to a rehabilitation facility 2 days later. Brain MRI performed 6 months after treatment showed complete resolution of thalamic FLAIR hyperintensities (Fig. 3E) and the associated gadolinium enhancement. His cognitive function returned to normal, and he resumed work as a truck driver. At 3-year follow-up his Mini-Mental State Examination score was 30/30 and the patient's wife felt that his cognitive function was even better than his prediagnosis baseline.

Case 4

A 71-year-old right-handed man was admitted after 6 months of progressively worsening forgetfulness, attention deficit, confusion, and daytime sleepiness. Four weeks prior to presentation, he had developed mild left-sided weakness and left-hand numbness that remained unevaluated until his rapid cognitive decline became more apparent. His examination revealed hypersomnolence and Kokmen Short Test of Mental Status score of 30/38 (–2 orientation, –1 attention, –1 calculation, –1 information, –3 recall). He also exhibited mild left-sided weakness in

FIG. 3. Case 3. **A–D:** Images obtained at the time of diagnosis. T1-weighted MR image without gadolinium showing no abnormality (**A**). T1-weighted MR image with gadolinium showing enhancement in bilateral thalami (**B**). FLAIR MR image showing left greater than right bilateral thalamic T2 signal hyperintensities (**C**). Right lateral common carotid angiogram demonstrating a Modified Borden-Shucart Type 3S (arrow) deriving arterial supply from the right occipital artery (arrowhead). Venous drainage occurred via an enlarged right tentorial vein and then refluxed into the vein of Galen and both internal cerebral veins (block arrows, **D**). **E and F:** Images obtained 6 weeks after surgical ligation of the dAVF. FLAIR MR image showing resolution of bithalamic hyperintensities (**E**). Right lateral common carotid angiogram showing complete obliteration of the dAVF (**F**).

an upper motor neuron pattern. Brain MRI demonstrated asymmetrical bilateral thalamic FLAIR hyperintensities and moderate heterogeneous bithalamic gadolinium enhancement (Fig. 5A–C). MR venography suggested a vas-

FIG. 4. Case 4. **A–D:** Images obtained at the time of diagnosis. Coronal T1-weighted MR image without gadolinium showing right thalamic hypointensity (A). Coronal T1-weighted MR image with gadolinium showing right greater than left bithalamic enhancement (B). FLAIR MR image showing right greater than left bithalamic hyperintensities (C). Right lateral common carotid angiogram, demonstrating a Modified Borden-Shucart Type 3S dAVF (arrow) deriving arterial supply from branches of the right middle meningeal and occipital arteries (arrowhead). Venous drainage occurred via an enlarged superior vermian vein, refluxing through the vein of Galen into both internal cerebral veins (block arrows, D). **E and F:** Images obtained 12 weeks after endovascular embolization of the dAVF. FLAIR MRI showing resolution of bithalamic hyperintensities (E). Right lateral common carotid angiogram showing complete obliteration of the dAVF (F).

cular malformation in the posterior fossa including occlusion of the proximal straight sinus and enlarged cortical veins over the right parietal region. Catheter angiography demonstrated a modified Borden-Shucart Type 3S torcular dAVF with arterial supplies from branches of the occipital and middle meningeal arteries bilaterally (Fig. 5D).

Retrograde venous drainage occurred via a tortuous superior vermian vein with reflux into the vein of Galen as well as into the right parietal cortical veins. No filling of the straight sinus was seen, consistent with occlusion. Transarterial Onyx embolization was performed twice, though vermian CVD persisted after both procedures. A midline suboccipital craniotomy was then performed for clip ligation of the vermian CVD. Intraoperative angiography demonstrated complete dAVF obliteration (Fig. 5F). Two days after surgery the patient exhibited minor symptomatic recrudescence, and brain MRI demonstrated several small embolic infarcts in the supratentorial regions bilaterally. The etiology of these embolic events was unclear. The patient's condition improved following this event, and he was discharged to a rehabilitation facility.

Brain MRI performed 3 months after treatment demonstrated complete resolution of thalamic edema (Fig. 5E), though several residual punctate areas of susceptibility-weighted signal intensity were noted within both thalami. MR venography also showed recanalization of the straight sinus and normalization of venous drainage. Clinical examination demonstrated improvement on the Kokmen Short Test of Mental Status to 34/38 (–1 attention, –3 recall). His family observed that he had undergone a remarkable recovery back to his cognitive baseline and near complete resolution of his left hemiparesis.

Some description of this patient's condition and course were included in a prior report.⁵² Further details of this patient's cerebral angiogram before and after treatment, pre- and postintervention neurological condition, and MRI findings are included here to increase comparability between multiple patients exhibiting the same symptoms and etiology.

Review of 19 Cases of dAVF-Induced Thalamic Dementia

Using multiple electronic databases (PubMed, Ovid, and EBSCO), a comprehensive review of the international literature (case reports, series, and reviews) was performed using the key phrases “dural arteriovenous fistula,” “dAVF,” “arteriovenous malformation,” “thalamic dementia,” “progressive dementia,” “cortical venous drainage,” “cortical venous hypertension,” “bithalamic edema,” and “bilateral thalamic edema.” Bibliographies of relevant publications were examined to identify additional cases. Through this comprehensive search, 15 previously published cases of dAVF-induced thalamic dementia were identified for comparison with our 4 cases. Characteristics of these 19 cases are compared in Tables 2 and 3.

Demographics and Presentation

Dural AVF-induced thalamic dementia is almost exclusively seen in men (18 of 19; 95%) and is most common in the 5th–7th decades of life (mean age 60 ± 10 years; range 43–77 years). All patients (19 of 19; 100%) presented with progressive cognitive dysfunction that included deficits in executive function, attention, memory, and disorientation. Fourteen of 19 patients (74%) had additional neurological deficits including ataxia (5 of 19; 26%), aphasia (5 of 19; 26%), confabulation (3 of 19; 16%), hemiparesis (1 of 19; 5%), third cranial nerve palsy (1 of 19; 5%), upper-extremity tremor (1 of 19; 5%), myoclonus (1 of 19;

FIG. 5. Case 3. Histopathology. **A and B:** H & E biopsy sections at medium power. Intermixed white and gray matter with reactive gliosis and prominent small vessels with reactive endothelium (solid arrow, A), occasional acute petechial hemorrhages (open arrow, A), focal microlesions with foamy macrophages (open arrowheads, B), numerous neuroaxonal spheroids (closed arrowheads, B), and rare nuclear pyknosis/karyorrhexis (open arrow, B). **C and D:** Immunohistochemistry at medium power. Staining for glial fibrillary acid protein (GFAP) demonstrated reactive astrocytes (arrowhead, C). Staining for CD68 showed microglia and macrophages surrounding vessels in focal microlesions (arrowhead, D). Figure is available in color online only.

5%), and incontinence (1 of 19; 5%). Some patients had more than 1 additional neurological deficit. The mean duration of symptoms at the time of angiographic diagnosis was 87 days (range 3 days to 18 months).

Neuroimaging

Pretreatment CT and MRI. Head CT results were reported in 10 of 19 cases (53%), 4 of which included contrast (Table 2). Five pretreatment CT scans (5 of 10; 50%) demonstrated bithalamic hypoattenuation indicating edema. All CT scans in which contrast was given demonstrated enhancement (4 of 4; 100%). One CT scan (1 of 10; 10%) also demonstrated petechial hemorrhages. Pretreatment brain MRI results were reported in 17 of 19 cases (89%), 16 of which included gadolinium administration (Table 2). Bithalamic T2/FLAIR hyperintensities indicating edema were noted in all cases (17 of 17; 100%), 8 with symmetric edema and 9 with asymmetrical edema. Four MR images (4 of 17; 24%) showed edema extending beyond the thalami (cortex, splenium, basal ganglia, or internal capsule). Ten scans (10 of 16; 62%) demonstrated gadolinium enhancement, which was typically bilateral, mild, and patchy, though occasionally appeared avid and homogeneous. No MRI scan (0 of 17; 0%) demonstrated any significant thalamic diffusion restriction.

Pretreatment Catheter Angiography. The majority of dAVFs producing thalamic dementia are located near the tentorial edge (14 of 19; 74%), though some were located

more peripherally. These locations included near the torcular herophili (1 of 19; 5%), the transverse sinus (1 of 19; 5%), the transverse-sigmoid sinus junction (1 of 19; 5%), a cerebral parasagittal sinus near the vertex (1 of 19; 5%), and near a persistent posterior falcine sinus (1 of 19; 5%) (Table 3). Reflux through the vein of Galen was angiographically demonstrated in all 19 cases, though venous routes to the vein of Galen were variable. Fourteen fistulas (14 of 19; 74%) were associated with one or more thrombosed sinuses (9 straight sinuses, 3 sigmoid sinuses, 2 transverse sinuses, and 1 superior sagittal sinus). All 19 dAVFs were located posteriorly: supratentorial, tentorial, or infratentorial. Arterial supply was often derived from multiple vessels (13 of 19; 68%). Regardless of nidus location, feeder vessels were most frequently derived from a middle meningeal artery (47%), posterior meningeal artery (37%), and/or occipital artery (37%).

Pretreatment Metabolic and Perfusion Imaging. Five of the 19 patients underwent cerebral emission imaging (3 SPECT, 1 Xenon CT, and 1 PET), and 1 received MR spectroscopy.^{32,73,76–78} Bithalamic hypoperfusion (4 of 4; 100%) and bithalamic congestion (2 of 2; 100%) were found each time these investigations were performed. PET results demonstrated bithalamic increase in fluorodeoxyglucose uptake in the single patient who underwent (Case 3). MR spectroscopy was also performed in a single patient and demonstrated bithalamic decrease in *N*-acetylaspartate, increase in choline, and increase in lactate.⁷³

TABLE 2. Published cases of dAVF-induced thalamic dementia syndrome: clinical characteristics

Authors & Year	Age (yrs), Sex	Presentation	Imaging Findings	Treatment	Outcome
Nakada et al., 1985	63, M	RPD, ataxia	BTE by CT	Surgery	Death
Ito et al., 1995	49, M	RPD, ataxia, aphasia	BTE by CT/MRI; bilthalamic vascular congestion by SPECT	TAE followed by surgery	Symptomatic/radiographic resolution
Greenough et al., 1999	62, M	RPD, ataxia	BTE by MRI	Surgery	Symptomatic/radiographic improvement
Tanaka et al., 1999	77, M	RPD	BTE by CT; bilthalamic hypoperfusion by Xenon scan	Multiple TAEs	Symptomatic/radiographic near resolution
Bernstein et al., 2003	69, M	RPD	Normal CT	TAE	Symptomatic/radiographic improvement
Tamamoto et al., 2003	67, F	RPD, incontinence	BTE by CT/MRI; bilthalamic vascular congestion by SPECT	TAE	Symptomatic/radiographic improvement
Tominaga et al., 2003	73, M	RPD, ataxia	BTE by MRI; bilthalamic vascular congestion by SPECT	Surgery	Symptomatic improvement, radiographic resolution
Gonçalves et al., 2008	43, M	RPD	BTE by MRI	TAE	Symptomatic/radiographic resolution
Matsumura et al., 2008	73, M	RPD	BTE by MRI	TAE	Symptomatic improvement, radiographic resolution
Racine et al., 2008		RPD	BTE by MRI	TAE followed by surgery	Symptomatic improvement, radiographic resolution
Wilson et al., 2008	48, M	RPD, aphasia, ataxia	BTE by CT/MRI	TAE	Symptomatic/radiographic resolution
Sugrue et al., 2009	51, M	RPD	BTE by MRI	TAE	Symptomatic resolution, radiographic improvement
Yamamoto et al., 2010	51, M	RPD	BTE by MRI	TAE	Symptomatic improvement, radiographic resolution
Geraldes et al., 2011	64, M	RPD, ataxia	BTE by MRI	TAE	Symptomatic improvement, radiographic resolution
Santillan et al., 2011	50, M	RPD	BTE by CT/MRI	TAE	Symptomatic/radiographic resolution
Present study	53, M	RPD, aphasia	BTE by MRI	TAE	Symptomatic improvement, radiographic resolution
Present study	59, M	RPD, aphasia	BTE by MRI	TVE	Death from unrelated medical condition
Present study	60, M	RPD	BTE by MRI	TAE followed by surgery	Symptomatic/radiographic resolution
Present study	71, M	RPD, hemiparesis	BTE by MRI	TAE followed by surgery	Symptomatic improvement, radiographic resolution

BTE = bilthalamic edema; OCSL = open craniotomy and surgical ligation; RPD = rapidly progressive dementia; TAE = transarterial embolization; TVE = transvenous embolization.

Posttreatment Imaging. All but one case reported posttreatment imaging findings. Fifteen of the 19 cases reported posttreatment catheter angiography and 5 of these were performed in delayed fashion. Eight of the 10 immediate posttreatment angiograms demonstrated dAVF obliteration (8 of 10; 80%). The remaining 2 showed dramatic fistula flow reductions.^{5,77} All 5 delayed angiograms showed no evidence of dAVF recurrence (5 of 5; 100%). Three of the 19 cases reported posttreatment CT imaging, 2 of which showed resolution of bilthalamic hypoattenuation (2 of 3; 67%) and the other demonstrated dramatic improvement (1 of 3; 33%). Posttreatment MRI results were available for 13 of 19 cases (68%). Bilthalamic T2 hyperintensities resolved in all cases (13 of 13; 100%). Bilthalamic enhancement resolved in all cases that reported pre- and posttreatment MRI with gadolinium administration (6 of 6; 100%). Of the 3 cases reporting both pre- and posttreatment perfusion imaging, all demonstrated cerebral blood flow normalization (3 of 3; 100%). In general, posttreat-

ment imaging revealed dramatic improvement or complete resolution of pretreatment abnormalities.

Delays in Diagnosis/Treatment

A delay in diagnosis and/or treatment of the offending dAVF was specifically noted in 4 of the 19 cases (21%)—3 from the published literature^{21,53,73} and 1 from the present series (Case 3). In 3 of these 4 cases (including our Case 3), precise time data were provided;^{21,53} the average delay from presentation to angiographic diagnosis was 23 days. In addition, treatment following angiographic diagnosis was delayed further in one of these cases, as the lesion was initially felt to be incidental.⁵³ Finally, 2 patients (including our Case 3) underwent invasive stereotactic biopsy prior to catheter angiography⁷³—both of which were nondiagnostic and one led to an asymptomatic thalamic hemorrhage (in our Case 3).

In the first case of delay, the initial discovery of the

TABLE 3. Published cases of dAVF-induced thalamic dementia syndrome: angiographic findings

Authors & Year	Mod. Borden-Shucart Grade	Location	Arterial Feeders	Venous Outflow Pattern
Nakada et al., 1985	3S	Tentorial	MMA	Vein of Galen, thrombosed straight sinus
Ito et al., 1995	2S	Tentorial	Bilat PMA & OccA	Straight sinus, inferior sagittal sinus, vein of Galen, basal vein of Rosenthal
Greenough et al., 1999	Straight sinus	Tentorial	NA	Vein of Galen, internal cerebral veins, thrombosed straight sinus
Tanaka et al., 1999	2S	Tentorial	Bilat OccA, rt APA, bilateral MMA	Straight sinus, vein of Galen, thrombosed transverse sinus
Bernstein et al., 2003	3S	Torcula	NA	Vein of Galen, internal cerebral veins, thrombosed sigmoid sinus
Tamamoto et al., 2003	3S	Tentorial	Lt STA, rt MMA, lt PMA	Vein of Galen, basal vein of Rosenthal, internal cerebral veins
Tominaga et al., 2003	2S	Transverse-sigmoid sinus junction	Lt ECA, lt PMA	Straight sinus, vein of Galen, thrombosed sigmoid sinus
Gonçalves et al., 2008	2S	Tentorial	Meningeal branches of rt ICA, rt ECA, rt OccA	Straight sinus, vein of Galen, internal cerebral veins
Matsumura et al., 2008	2S	Tentorial	NA	Vein of Galen
Racine et al., 2008	3S	Torcula	Rt OccA, lt PMA	Vein of Galen, thrombosed straight sinus
Wilson et al., 2008	2S	Tentorial	NA	Straight sinus, vein of Galen, thrombosed sigmoid sinus
Sugrue et al., 2009	2S	Superior sagittal Sinus	Bilat MMA, PMA, marginal TentA	Posterior falcine sinus, vein of Galen, basal vein of Rosenthal, internal cerebral veins, thrombosed straight sinus
Yamamoto et al., 2010	3S	Tentorial	Bilat ICA	Vein of Galen, internal cerebral vein, thrombosed straight sinus
Geraldes et al., 2011	2S	Torcula	Bilat PMA, rt MMA	Vein of Galen, thrombosed transverse and sagittal sinus
Santillan et al., 2011	3S	Tentorial	Medial TentA, MMA	Vein of Galen, basal vein of Rosenthal, internal cerebral veins, thrombosed straight sinus
Present study	2S	Tentorial	Bilat MMA, inferolateral trunks of ICAs	Straight sinus, vein of Galen, internal cerebral veins
Present study	3S	Tentorial	Lt OccA, bilat PMA	Superior vermian vein, vein of Galen, internal cerebral veins, partially thrombosed straight sinus
Present study	3S	Torcula	Rt OccA	Vein of Galen, internal cerebral veins
Present study	3S	Torcula	Bilat OccA, bilat MMA	Superior vermian vein, vein of Galen, thrombosed straight sinus

APA = ascending pharyngeal artery; ECA = external carotid artery; MMA = middle meningeal artery; NA = not available; OccA = occipital artery; PMA = posterior meningeal artery; STA = superficial temporal artery; TentA = tentorial artery.

dAVF via catheter angiography occurred relatively early in the course of the illness but was considered an incidental finding.⁵³ As the patient's cognitive function continued to deteriorate (eventually progressing to coma), repeat head CT was performed, demonstrating low attenuation of the diencephalon. The diagnosis of dAVF-induced thalamic dementia was then made, and the dAVF underwent surgical obliteration. Unfortunately, the patient remained in a coma after surgery and died 21 days later. In the second case of delay, a lack of white matter changes on brain MRI initially altered the authors' differential diagnosis away from dAVF.⁷³ During this delay, the patient suffered a capsulolenticular hematoma, and his neurological condition progressed to coma. Nine days later, catheter angiography was performed, revealing the offending dAVF. The patient

underwent embolization of the dAVF, and his dementia slowly improved over 3 months with resolution of the associated myoclonus but not the hemiparesis. In the third case of delay, a malignant neoplasm was suspected, in part due to gadolinium enhancement noted on brain MRI.²¹ The patient underwent a stereotactic biopsy that was nondiagnostic, which prompted reevaluation of the differential diagnosis, performance of catheter angiography, and endovascular treatment of the offending dAVF. The patient's neurological condition fully resolved within 4 months. In the fourth case of delay (Case 3 of the present series), a malignant neoplasm was suspected, in part due to gadolinium enhancement noted on brain MRI. The patient underwent stereotactic biopsy that was nondiagnostic and complicated by an asymptomatic thalamic hemorrhage. Six weeks

of vitamin supplementation for the presumed diagnosis of Wernicke's encephalopathy ensued. Due to further deterioration in the patient's cognitive status, catheter angiography was performed, which identified the offending dAVF. The patient underwent combined endovascular and surgical obliteration of the dAVF, and his neurological condition improved dramatically over the ensuing 3 months.

Treatment

All 19 patients underwent dAVF treatment with surgery, endovascular therapy, or a combination. Sixteen of the 19 patients (84%) were treated initially with endovascular embolization, 15 via a transarterial approach, and 2 via a transvenous approach; 2 approaches were performed in 1 patient in a single procedure. Eleven of these patients (11 of 19; 69%) had elimination of the CVD—8 with complete obliteration of the dAVF and 3 with obliteration of the CVD alone. One patient required a second endovascular procedure with subsequent complete obliteration of the dAVF. Therefore, overall success (defined as angiographic elimination of the CVD) was achieved in 75% (12 of 16) of patients undergoing endovascular embolization. No procedural complications were reported.

The remaining 4 patients who initially underwent endovascular therapy required surgical intervention, all of which led to complete radiographic obliteration on postoperative imaging. Another 3 cases were treated exclusively with open surgery. One of these cases noted successful intraoperative obliteration of the dAVF, but did not report postoperative imaging findings.⁵³ The other 2 cases documented complete radiographic obliteration of the dAVF on postoperative imaging.^{23,78} Therefore, the overall cure rate for surgically treated dAVFs was 100% (7 of 7; 6 documented by postoperative imaging, 1 documented by surgeon impression). One surgical complication was reported in which hydrocephalus developed after surgery requiring shunt treatment.²³

Long-Term Patient Outcome

Eighteen of 19 cases had long-term patient outcomes reported. Of these, 6 patients (6 of 18; 33%) experienced complete radiographic and functional recovery following dAVF obliteration, all within 6 months of their treatment. Ten patients (10 of 18; 55%) experienced significant but incomplete improvement in their dementia and other neurological deficits in late follow-up (typically months from treatment). One patient (1 of 18; 6%—the earliest reported case) died in the hospital 21 days after surgery—likely the result of delayed treatment.⁵³ One patient (1 of 18; 6%—Case 2 in the present series) died of an unrelated cause (myocardial infarction) 3 months after surgery, although his dementia had been steadily improving since dAVF treatment. Most recoveries began soon after dAVF obliteration, though maximal recovery often took weeks to months. Only 4 cases reported on follow-up catheter angiography, all of which demonstrated continued obliteration of the treated dAVF.^{5,65,73,86} No recurrence of dAVF-related symptoms was reported in any of the 18 cases.

Discussion

The incidence of dementia as a presenting symptom of

high-grade dAVFs has been reported to be as low 0%⁷¹ and as high as 11%.⁷² The manner by which this occurs is fairly well established—arterialized venous reflux from the fistula produces regional venous congestion and parenchymal edema, leading to functional compromise of the affected brain regions.⁷⁹ Two types of dAVF-induced dementia have been described, each of which has its own distinct clinical and neuroimaging characteristics. Cortical dementia due to dAVF is characterized by rapidly progressive cognitive dysfunction including impairments in verbal fluency and language comprehension, apraxia, visuospatial discordance, and memory dysfunction that is frequently accompanied by focal cortical deficits including hemiparesis, somatic sensory disturbances, aphasia, alexia, and/or visual disturbances.^{13,16,18,28,31,36,37,41,54,61,69,80,81,84,85,87,89} These symptoms are associated with widespread and/or multilobar hyperintensities on FLAIR and T2-weighted imaging.^{31,41,54,82} Thalamic dementia due to dAVF is characterized by rapidly progressive cognitive dysfunction including disorientation, executive dysfunction, attention deficits, memory impairment, confabulation and disinhibition. Hereto, additional neurological deficits are frequent but relate to dysfunction of the thalamus itself (e.g., thalamic aphasia or ataxia) or nearby structures (e.g., internal capsule leading to hemiparesis, third cranial nerve leading to diplopia). These symptoms are accompanied by a more focused profile of hyperintensities on FLAIR and T2-weighted imaging primarily involving the bilateral thalami.^{21–23,32,34,48,49,53,62,65,73,76–78,86,88}

Dural AVF–Induced Thalamic Dementia: Presentation, Imaging, Treatment, and Outcome

Dural AVF–induced syndrome occurs almost exclusively in men in their 5th–7th decades of life, typically without prior history of neurological trauma. The most common symptoms of this syndrome are deficits in attention, memory, executive functioning, and disorientation. Other symptoms are variably present, including ataxia, aphasia, amnesia, and/or hemiparesis. The rapidity of onset is typically weeks to a few months (mean duration in our review 87 days). CT/MRI workup invariably demonstrates bilateral thalamic edema, not uncommonly associated with patchy enhancement and occasionally petechial hemorrhage. Diffusion restriction is not seen. Obvious vascular abnormalities such as dilated deep venous structures including the vein of Galen are often not appreciated on standard axial imaging; therefore, dedicated vascular imaging must be obtained to secure the diagnosis.

Most offending dAVFs are located at the tentorial edge, but more peripheral lesions with venous drainage that ultimately involves the vein of Galen can also produce this syndrome (e.g., at the torcular herophili). Once identified, dAVF treatment—either via endovascular or surgical means—carries low risk and is highly effective at disconnecting the CVD, the portion of the dAVF that is responsible for the dementia syndrome. Often complete radiographic obliteration of dAVF is achieved. In our review of the 16 cases treated with endovascular therapy, no procedural complications occurred, and CVD obliteration was achieved 75% of the time. In our review of the 7 cases treated with surgical therapy, one procedural

complication occurred (postoperative hydrocephalus requiring a shunt) and CVD obliteration was achieved 100% of the time (6 documented by postoperative imaging; 1 by surgeon impression). Both endovascular and surgical therapies are therefore excellent options, with decision making regarding which approach to take involving an assessment of the following factors: available endovascular access to the dAVF and CVD, age and comorbidities of the patient, and patient/family preference regarding level of invasiveness of the procedure versus procedural success rate. Symptom resolution invariably begins within days of elimination of the CVD with maximal neurological recovery being achieved over the ensuing months. In our review of the 18 cases reporting long-term clinical outcome, 33% of patients experienced complete resolution of their dementia symptoms, 55% experienced significant but incomplete resolution of their dementia symptoms, 6% died of unrelated causes, and 6% died due to progressive dAVF-induced symptoms. The latter was the first reported case of dAVF-induced thalamic dementia and included a substantial delay in initiating treatment. No recurrences of the offending fistula or the dementia symptoms have been reported, though most studies did not include follow-up vascular imaging to detect an asymptomatic recurrence.

Thalamic Dementia Syndrome: Differential Diagnosis and Workup

Dural AVFs are one of a number of conditions that can cause thalamic dementia. Classically, this term was used to describe the constellation of cognitive symptoms present in patients who develop thalamic ischemic stroke—especially those with bilateral thalamic involvement.^{12,38,40,59,67,74,75} These symptoms can manifest in several particular patterns owing to the various functions performed by the different thalamic nuclei. In the context of dAVFs, the hallmarks of the symptom complex—deficits in executive function, memory, learning, attention, disinhibition, and confabulation—correspond best to the anteromedial and central thalamic regions.^{12,20,21,38,43,51,56,59,62,65,74,85} Sensory and motor symptoms typically ascribed to posterolateral and inferior thalamic distributions tend to be uncommon or absent in patients with dAVFs. The term thalamic dementia syndrome has also been applied to patients having similar symptoms related to other pathological conditions affecting the thalami, including deep venous system thrombosis, bilateral diencephalic tumors, viral and prion diseases, osmotic myelinolysis, and toxic insults (see Table 4).

Distinguishing these various etiologies starts with a thorough history. Symptom onset can separate etiologies in many cases, with acute onset (minutes) suggesting arterial infarction, subacute onset (days to weeks) suggesting dAVF, deep venous thrombosis, or infection, and chronic (months or years) suggesting tumors or prion disease. A variable or recurring presentation would suggest a toxic or metabolic etiology. Other clinical clues that may help delineate the underlying condition include accompanying presenting symptoms (fever for infection) and medical history (oral birth control for hypercoagulable deep venous thrombosis; alcohol abuse for Wernicke encephalopathy; recent hyponatremia for osmotic myelinolysis; recent vector exposure for West Nile and Japanese encephalitis). Any of these presentations in conjunction with rapidly progress-

ing dementia should prompt neuroimaging with MRI to further assist in differentiating the possible causes.

MRI can be of great value in helping to determine the underlying etiology of the thalamic dementia syndrome. A comparison of the radiographic findings for the various conditions that cause thalamic dementia is provided in Table 4. The presence of diffusion restriction most strongly suggests arterial stroke, toxicity, infection, or, to a lesser degree, lymphoma or deep venous thrombosis.^{1,2,4,25–27,35,39,42,45,50,57,60,70,90} Conversely, diffusion restriction has not been found in patients with dAVF-induced thalamic dementia. Strong gadolinium enhancement is most consistent with a high-grade neoplasm;^{26,27,58} however, other conditions including dAVFs can also have some degree of gadolinium enhancement.^{62,76,77} Therefore, vigilance is required when considering the diagnosis of neoplasm, and additional diagnostic tests are often indicated prior to proceeding with stereotactic brain biopsy. The distribution of T2/FLAIR signal hyperintensities may also offer clues to the diagnosis. Arterial strokes will follow arterial distributions, while neoplasms are more inclined to circumscribe tissue planes.^{2,4,11,15,27,45,58,70} Deep AVFs and deep venous thromboses produce T2/FLAIR hyperintensities centered within the bilateral anteromedial thalamic regions, though extensions beyond the thalami are reported in a significant number of cases.^{5,18,21,22,76,77}

In many instances, a definitive diagnosis for the thalamic dementia syndrome cannot be reached after history, physical examination, and MRI. Due to the overlapping nature of the signal characteristics associated with many of the causative conditions, we advocate that patients receive a catheter angiogram prior to an invasive diagnostic procedure (e.g., stereotactic biopsy) or prolonged medical therapy (e.g., vitamin supplementation for suspected Wernicke encephalopathy) because of its ability to definitively delineate a vascular lesion from other etiologies. Although catheter angiography is presently the only way to conclusively diagnose this condition, some progress has been made with noninvasive vascular imaging modalities of dAVFs. Coley et al. have reported that a new technique called MR digital subtraction angiography (MR-DSA) is capable of producing time-resolved dynamic vascular imaging of the brain that was useful for the initial confirmation and subsequent monitoring of dAVFs, though they admit that catheter angiography is still necessary for detailed anatomical evaluation.¹⁴ Hori et al. evaluated the utility of time-resolved 3D MR digital subtraction angiography (Time-SLIP 3D MRDSA) for assessment of dAVFs.²⁹ In comparing intraarterial DSA to this technique, which does not require injection of contrast material, they reported accurate diagnosis and evaluations of hemodynamic information relating to 6 dAVFs. Jagadeesan et al. documented that MR susceptibility-weighted imaging is highly accurate for the detection of arteriovenous shunting in arteriovenous malformations of the brain, but this technique has yet to be applied to dAVFs themselves.³³

Conclusions

Our report emphasizes that unexplained rapidly progressing dementia, particularly in middle- to older-aged

TABLE 4. Differential diagnosis for patients with rapidly progressive dementia and bithalamic edema

Diagnosis	T1-Weighted	T1-Weighted w/ Gadolinium	T2/FLAIR	DWI/ADC
High-grade glioma*	Hypointense	Irregular or rim-enhancement, central hypointensity (necrosis)	Rim hyperintense, often central hypo- or isointense	Hyperintense DWI, hypointense ADC
Lymphoma†	Hypo- or isointense	Enhancement (90% homogeneous in non-AIDS cases, 75% ring-enhancement in AIDS cases)	Iso- or hyperintense	Hyperintense DWI, hypointense ADC
Wernicke's encephalopathy‡	Hypointense	Variable enhancement	Symmetric periaqueductal and paraventricular hyperintensities	Weakly to moderately hyperintense DWI, variable ADC intensities
Carbon monoxide poisoning§	Hypointense	Patchy enhancement	Hyperintense, especially GP and cortical white matter	Hyperintense DWI, hypointense ADC
Hepatic encephalopathy¶	Hyperintense, especially GP (manganese)	No enhancement	Patchy white matter hyperintensities, may require fast FLAIR to detect	Hyperintense DWI, hypointense ADC
Osmotic myelinolysis**	Patchy hypointensities	Rare enhancement	Hyperintense, especially central pons	Hyperintense DWI, hypointense ADC
West Nile Virus/Japanese encephalitis**	Hypointense	Variable enhancement	Hyperintense, BG, midbrain & sulci (leptomeningeal inflammation)	Hyperintense DWI (posterior limb of internal capsule, corona radiata), ADC hypointense
Prion disease**	Hypointense	No enhancement	Hyperintense, especially pulvinar (pulvinar sign in variant CJD)	Hyperintense DWI, BG and cortex, hypointense ADC
Arterial infarction (bithalamic infarction; top of the basilar syndrome)**	Hypointense	Variable enhancement	Hyperintense, BG & midbrain ("V" sign)	Hyperintense DWI, hypointense ADC
Deep venous system thrombosis (hypercoagulable; sickle cell; idiopathic)**	Hypointense thalami, possible hyperintensity (clot in sinus)	Patchy enhancement	Hyperintense	Heterogeneously hyperintense DWI, hypointense ADC
dAVF	Hypointense	Variable, patchy enhancement	Hyperintense, possible extension beyond thalami	Normal DWI & ADC

ADC = apparent diffusion coefficient; BG = basal ganglia; CJD = Creutzfeldt-Jakob disease; DWI = diffusion-weighted imaging; GP = globus pallidus.

* References 27, 45, 58, and 70.

† References 26 and 70.

‡ References 25, 35, 45, 46, 63, 70, 83, 92, and 93.

§ References 57 and 70.

¶ References 41, 45, 70, and 90.

** Reference 70.

men, should be evaluated with MRI and likely also catheter angiography. MRI signal characteristics most consistent with dAVF are bilateral anteromedial thalamic hyperintensities on T2/FLAIR sequences without evidence for gadolinium enhancement, petechial hemorrhages, or restricted diffusion. However, variability in these signal characteristics exist, and extension of the T2-weighted/FLAIR hyperintensities beyond the thalami, gadolinium enhancement (typically mild to moderate in intensity and patchy in appearance), and petechial hemorrhage has been reported in the context of dAVF-induced thalamic dementia. Therefore, we recommend evaluation with catheter angiography prior to stereotactic brain biopsy (which likely carries increased risk in dAVF patients due to the hyperemic nature of the involved tissue) and/or prolonged medical treatment (which delays dAVF treatment and subject patients to the potential of rapidly progressive symptoms) unless a definitive diagnosis of a nonvascular etiology has

been reached through other means. Once a dAVF is identified as the cause of the thalamic dementia syndrome, prompt treatment should be initiated, given that rapid neurological decline in the days following initial dAVF diagnosis has been reported in several cases. Treatment with either endovascular or surgical techniques carries a low risk of complications and is associated with a high degree of success, often leading to dramatic resolution of the presenting symptoms and resolution of the MRI abnormalities.

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Disclosures

Dr. Derdyn reports that he is a consultant for Penumbra, MicroVention, and Silk Road, and has direct stock ownership in Pulse Therapeutics. Dr. Moran reports that he is a consultant for Medtronic Neurovascular. Dr. Lanzino reports that he is a consultant for Covidien and Medtronic.

Author Contributions

Conception and design: Zipfel, Holekamp, Derdeyn, Rich. Acquisition of data: Holekamp, Mollman, Kolar, Kramer, Derdeyn, Moran, Lanzino. Analysis and interpretation of data: Zipfel, Holekamp, Mollman, Murphy, Kolar, Perrin, Rich. Drafting the article: Holekamp, Mollman. Critically revising the article: Zipfel, Holekamp, Murphy, Derdeyn, Moran, Perrin, Rich, Lanzino. Reviewed submitted version of manuscript: Zipfel, Holekamp, Mollman, Murphy, Derdeyn, Moran, Perrin, Rich, Lanzino. Approved the final version of the manuscript on behalf of all authors: Zipfel. Statistical analysis: Holekamp, Mollman. Administrative/technical/material support: Zipfel. Study supervision: Zipfel.

Correspondence

Gregory J. Zipfel, Department of Neurological Surgery, Washington University, Campus Box 8057, 660 S. Euclid Ave., St. Louis, MO 63310. email: zipfelg@wustl.edu.