Dural arteriovenous fistula as a treatable dementia

Ikponmwosa Enofe, MBBS, MPH, Ike Thacker, MD, and Sadat Shamim, MD

Dementia is a chronic loss of neurocognitive function that is progressive and irreversible. Although rare, dural arteriovenous fistulas (DAVFs) could present with a rapid decline in neurocognitive function with or without Parkinson-like symptoms. DAVFs represent a potentially treatable and reversible cause of dementia. Here, we report the case of an elderly woman diagnosed with a DAVF after presenting with new-onset seizures, deteriorating neurocognitive function, and Parkinson-like symptoms.

In this case, we highlight how familiarity with symptoms of dural arteriovenous fistulas (DAVFs) can reduce the incidence of misdiagnosis of dementia syndromes and lead to early treatment and better outcomes (1).

CASE PRESENTATION

An 82-year-old woman presented with a 1-year history of reduced limb control and difficulty finding words. She had been on carbidopa-levodopa without significant improvement. One month prior, she had a cluster of generalized unprovoked seizures with loss of consciousness with initiation of levetiracetam. Subsequently, she had progressive loss of memory. In the distant past, she had dural sinus thrombosis, recurrent deep venous thrombosis, and pulmonary embolism, for which she was on warfarin. She also complained of a constant loud whooshing sound. She had a nonfocal neurological exam with the following exceptions: speech was fluent, appropriate, but slow; her Mini-Mental State Examination was 21 with poor recall and attention and an inability to draw overlapping pentagons or write words; and her clock drawing test showed marked visuospatial abnormalities *(Figure 1a)*.

Magnetic resonance imaging of the brain revealed a chronic thrombus in the right transverse sinus with enlarged cerebral veins and multiple right-sided leptomeningeal collaterals. There were hyperintense signal changes in the periventricular white matter on the T2-weighted sequence. A fluid-attenuated inversion recovery sequence showed a hyperintense signal over a portion of the right transverse sinus with the loss of normal flow void. Cerebral angiography revealed a hypervascular DAVF predominantly at the left skull base midline parietal calvarium. Vessels from the external carotid, vertebral, and subclavian arteries supplied the DAVF, which drained into the torcular Herophili, superior sagittal sinus, and left and right transverse-sigmoid sinuses. Occlusion or near occlusion of the right transverse sinus was noted. Marked venous hypertension was present in the right cerebral hemisphere, deep basal ganglia, and posterior fossa. Cortical veins were enlarged. There was prolonged drainage bilaterally from the cerebellar hemispheres *(Figure 2a)*.

The patient underwent a right partial DAVF transarterial endovascular embolization *(Figure 2b)* with marked improvement in her clinical symptoms on repeat evaluation 14 weeks later. Her Mini-Mental State Examination score increased from 21 to 25. On the clock drawing test, she was able to fill in numbers correctly into a predrawn circle. Her ability to draw overlapping pentagons improved, and she could write legible and comprehensible sentences *(Figure 1b)*. Her whooshing sounds nearly resolved.

DISCUSSION

DAVF is an abnormal connection between arteries and veins. It is rare, accounting for about 10% to 15% of vessel malformations in the brain (1). Blood is supplied to DAVFs mainly through branches arising from the external carotid artery. Vessels from the internal carotid and meningeal arteries can also be involved (2).

The exact mechanism of DAVF development is unknown. A percentage of it is believed to develop from acquired causes that result in increased pressure in the dural sinuses. The presence of a transverse sinus thrombus in our patient corroborates previous reports identifying thrombosis in the venous sinuses as a possible trigger for developing DAVF (3–6). DAVF usually presents in the fifth or sixth decades of life (3, 6). It occurs with a female-to-male ratio of 1:1.65 (1). No family history or hereditary pattern predisposes or increases the risk of developing a DAVF (3).

Patients with DAVFs present with a wide array of clinical symptoms. Common symptoms range from the less severe

From the Department of Radiology (Thacker) and the Division of Neurology, Department of Internal Medicine (Shamim), Baylor University Medical Center at Dallas.

Corresponding author: Sadat Shamim, MD, 3600 Gaston Avenue, Wadley Tower Suite 1155, Dallas, TX 75246 (e-mail: sadat.shamim@bswhealth.org).

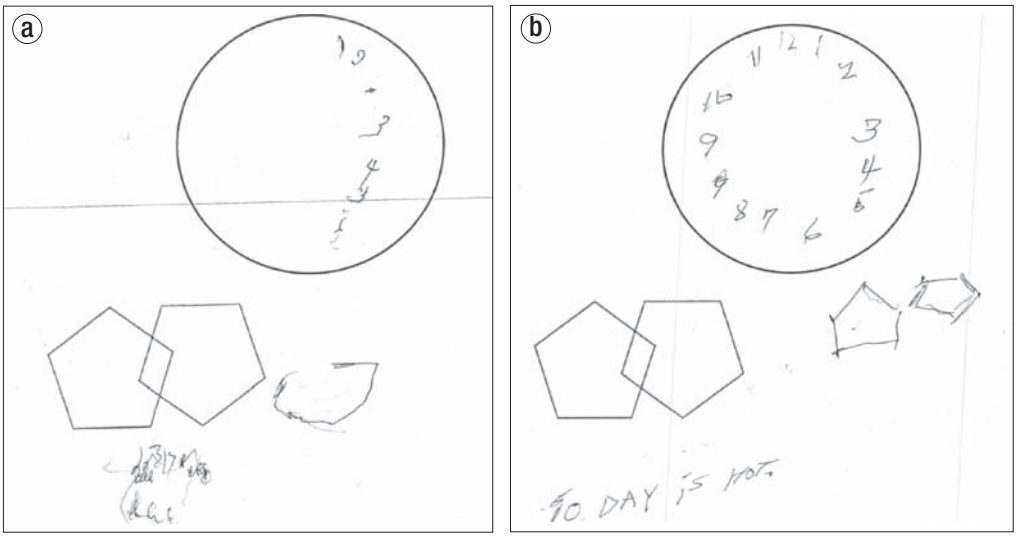

Figure 1. Mini-Mental State Examination **(a)** 12 days prior to partial embolization of the dural arteriovenous fistula, with a score of 21/30, and **(b)** about 2.5 months after partial embolization, with a score of 25/30.

presentations of headache, orbital bruit, pulsatile tinnitus, and ophthalmoplegia to more severe presentations of neurological deficits and acute intracranial hemorrhage (1, 3). There are only a few reports of DAVFs presenting with dementia syndrome, a decline in neurocognitive function, and parkinsonism (3, 7). The clinical presentation of DAVFs depends on their location and pattern of cerebral venous drainage (1, 3, 6). DAVFs with isolated dural sinus drainage have a benign clinical course. Most cases present with pulsating tinnitus and visual symptoms, depending on the proximity of the dural venous sinus to adjacent organs.

DAVFs with isolated dural sinus drainage to the cavernous sinus commonly present with ophthalmoplegia, ptosis, chemosis, retroorbital pain, and decrease in visual acuity. Pulsating tinnitus is the most frequent presentation of a DAVF with increased drainage to the transverse and sigmoid dural venous sinuses. Clinical presentation is best explained by the proximity of the sigmoid sinus to the auditory apparatus (3). In our case, increased drainage was seen in the transverse and sigmoid dural venous sinus, which explains the presence of pulsating tinnitus at the time of her presentation.

Cortical venous hypertension presents with more severe symptoms and carries a higher risk of future neurological events in comparison to a more benign clinical course seen in patients with isolated dural sinus drainage (3). Presentation of severe cortical venous hypertension includes but is not limited to intracranial hemorrhage and nonhemorrhagic congestive neurological deficit such as progressive dementia, parkinsonism, seizures, cerebellar symptoms, and other focal neurological deficits (3).

The exact pathogenesis for developing dementia in patients with DAVFs remains largely unknown. The occurrence of ischemia involving the frontal and temporal lobe is associated with the development of dementia (2, 8). Ischemia occurs from the downstream effect of arterialization of cortical venous drainage from cortical venous hypertension, leading to cerebral venous congestion (3). Some reports have suggested that patients with DAVFs involving the superior sagittal sinus have a higher rate of presenting clinically with dementia (9).

The mechanism by which DAVFs cause parkinsonism is also unclear (8). Hypoperfusion of the frontal lobe due to venous hypertension is thought to be responsible for developing parkinsonism in patients with DAVFs (2, 8). Another possible explanation is from hypoperfusion of the basal ganglia due to impaired drainage of the deep internal veins (8).

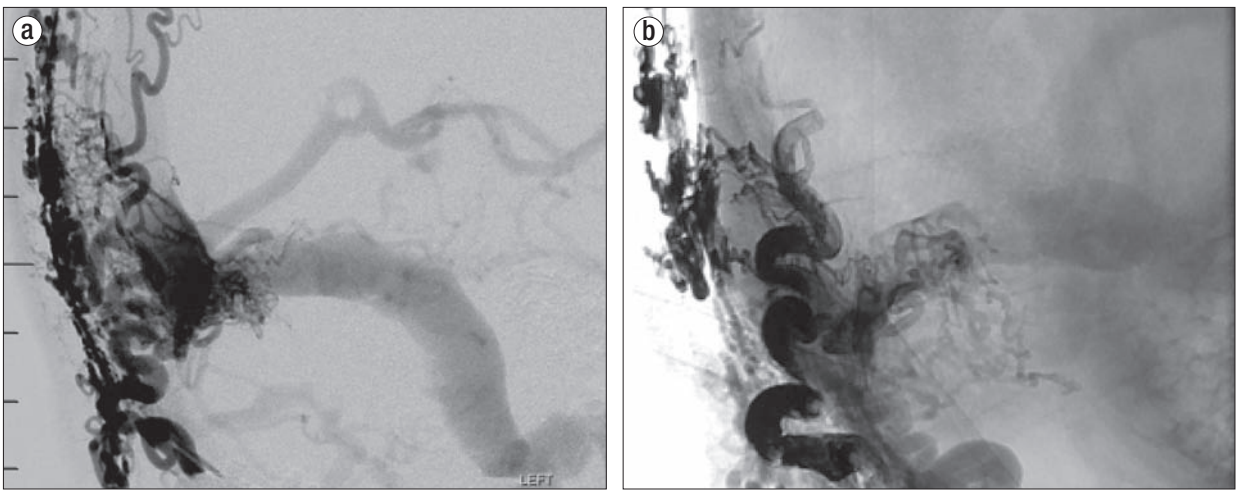

Figure 2. (a) Digital subtraction angiogram of the direct left occipital artery injection. An extensive rete of arterial pedicles supplying the fistula is densely radiopaque. The ipsilateral transverse sinus exhibits early opacification. Note the retrograde drainage via the straight sinus. **(b)** Unsubtracted completion of the left occipital angiogram shows hyperdense embolization material throughout feeding vessels, with diminished shunt physiology.

Previous reports on patients presenting with dementia and parkinsonism showed frontal lobe, temporal lobe, and basal ganglia ischemia. Ischemia was a direct result of the connection between the DAVF and superior sagittal sinus. It has been hypothesized that this leads to the development of an arterial steal phenomenon believed to be responsible for the ischemia. Local venous congestion was also involved in the development of hypoxia. Dementia was attributed to frontal lobe ischemia, while damage to the frontal white matter and basal ganglia was hypothesized to be the cause of the parkinsonism (3, 10).

Other symptoms reported in our patient such as decreased speech output may be the result of decreased frontal lobe perfusion. Seizures could be attributed to global venous congestion (8). Hemorrhage in the basal ganglia has been reported to cause seizure-like tonic-clonic movements (5). It appears that the seizures seen in our patient were more from venous congestion than from hemorrhage. Reduced speech, as with dementia, showed significant improvement with postendovascular embolectomy.

Unlike most known causes of dementia syndromes, DAVFs do not have a single constant or defining symptom. The presentations of DAVFs are highly variable and may be reversible. DAVF represents one of the rare but reversible causes of dementia. Early diagnosis and treatment may have the potential of dramatically improving patients' clinical condition and minimizing long-term residual disability.

1. Henderson JB, Zarghouni M, Hise JH, Opatowsky MJ, Layton KF. Dementia caused by dural arteriovenous fistulas reversed following endovascular therapy. *Proc (Bayl Univ Med Cent)* 2012;25(4):338–340.
2. Ma C, Lu Q, Shi W, Su Z, Zhao Y, Li C, Liu Z. Diagnosis and treatment of a dural arteriovenous fistula presenting with progressive parkinsonism and dementia: A case report and literature review. *Exp Ther Med* 2015;9(2):523–526.
3. Zipfel GJ, Shah MN, Refai D, Dacey RG Jr, Derdeyn CP. Cranial dural arteriovenous fistulas: modification of angiographic classification scales based on new natural history data. *Neurosurg Focus* 2009;26(5):E14.
4. Netravathi M, Pal PK, Bharath RD, Ravishankar S. Intracranial dural arteriovenous fistula presenting as parkinsonism and cognitive dysfunction. *J Clin Neurosci* 2011;18(1):138–140.
5. Geraldes R, Albuquerque L, Ferro JM, Sousa R, Sequeira P, Campos J. Rapidly progressive cognitive impairment, ataxia, and myoclonus: an unusual presentation of a dural arteriovenous fistula. *J Stroke Cerebrovasc Dis* 2012;21(7):619.e3–619.e5.
6. Gandhi D, Chen J, Pearl M, Huang J, Gemmete JJ, Kathuria S. Intracranial dural arteriovenous fistulas: classification, imaging findings, and treatment. *AJNR Am J Neuroradiol* 2012;33(6):1007–1013.
7. Chahbazian K, Théaudin M, Lehmann P, Sachet M, Adams D, Saliou G. Reversible pseudo-Creutzfeldt-Jakob syndrome related to cerebral dural arteriovenous fistula. *J Am Geriatr Soc* 2014;62(10):2024–2026.
8. Fujii H, Nagano Y, Hosomi N, Matsumoto M. Dural arteriovenous fistula presenting with progressive dementia and parkinsonism. *BMJ Case Rep* 2014 Jun 2;2014. doi: 10.1136/bcr-2014-203921.
9. Labeyrie MA, Lenck S, Saint-Maurice JP, Bresson D, Houdart E. Dural arteriovenous fistulas presenting with reversible dementia are associated with a specific venous drainage. *Eur J Neurol* 2014;21(3):545–547.
10. Kajitani M, Yagura H, Kawahara M, Hirano M, Ueno S, Fujimoto K, Sakaki T, Taoka T, Nakagawa H, Kichikawa K. Treatable fluctuating Parkinsonism and dementia in a patient with a dural arteriovenous fistula. *Mov Disord* 2007;22(3):437–439.