Dementia caused by dural arteriovenous fistulas reversed following endovascular therapy

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We describe two patients who presented with dementia as the primary neurological manifestation of their dural arteriovenous fistula (dAVF). Although dementia is not the most common presentation for a dAVF, these cases show that obliterating the fistula can eliminate this dreadful manifestation. This awareness may facilitate the diagnosis of dementia in cases caused by dAVF and allow for a minimally invasive treatment that restores cognitive function back to baseline.

Dural arteriovenous fistula (dAVF) is an acquired lesion that develops following dural sinus thrombosis and subsequent recanalization of the sinus leading to a direct communication between small sinus wall arteries and the sinus lumen. dAVFs represent 10% to 15% of all cerebrovascular malformations and may result in an array of signs and symptoms. The most common presentation is pulsatile tinnitus, headaches, focal neurological deficit, and visual disturbances. We report two cases in which the patients presented with dementia-type symptoms and were found to have dAVFs.

CASE 1

A 53-year-old woman who had a history of Von Hippel-Lindau disease and resection of a retinal hemangioblastoma as a child presented with a 3- to 4-month history of dysarthria, derangements in taste sensation, gait instability, and memory disturbance. She could no longer remember the names of her children and had to carry a notebook with pictures and names as a reminder. Her symptoms became so severe that she was unable to maintain her job as a phlebotomist. On initial computed tomography (CT), she had a large partially calcified thrombus in the distal portion of the superior sagittal sinus in addition to dilated deep collateral veins and mild ventriculomegaly.

An angiogram revealed an enlarged ophthalmic artery with multiple ophthalmic branches supplying a high-flow AVF in the middle cranial fossa draining directly into the sphenoparietal sinus and multiple enlarged cortical veins (*Figure 1a*). Transvenous endovascular coil embolization at the fistula site resulted in near complete occlusion of the dAVF (*Figure 1b*).

Because of a small recurrent dural fistula, the patient subsequently underwent neurosurgical intervention. A left pterional craniotomy was performed. Using a microscope and bipolar electrocautery, the residual fistula was cauterized. Intraoperative and follow-up angiograms revealed complete cure of the left temporal and sphenoid dAVF. Clinical follow-up 5 months later revealed that the patient's memory dysfunction and dysarthria had nearly resolved. The patient did have residual ataxia, generalized weakness, and blurred vision; however, these symptoms

Figure 1. Patient 1 (a) before and (b) after treatment. The pretreatment left internal carotid artery angiogram shows an enlarged ophthalmic artery (white arrow) with multiple branches supplying a high-flow arteriovenous fistula (black arrow) involving the middle cranial fossa. The posttreatment angiogram shows closure of the fistula and complete resolution of early venous drainage (arrow).

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had improved significantly since presentation.

CASE 2

A 74-year-old man presented with complaints of dizziness, altered mental status, and visual disturbances described as bright flashes. His son reported a noticeable decrease in memory and intellectual capacity over the preceding 6 months. Magnetic resonance imaging (MRI) at an outside facility revealed a right-sided “arteriovenous malformation.” The patient had no focal neurological deficits. Pretreatment angiographic images revealed dilatation of bilateral middle meningeal artery branches and transosseous superficial temporal artery branches supplying an AVF of the superior sagittal sinus (*Figure 2a*). The patient underwent two staged endovascular embolization procedures with a liquid agent. Posttreatment angiographic images revealed successful occlusion of the dural AVF centered on the superior sagittal sinus (*Figure 2b*). At 6-month clinical follow-up, the patient’s dizziness, altered mental status, and visual disturbances had significantly improved. According to his son, the patient’s memory and intellectual capacity had returned to baseline.

DISCUSSION

dAVFs are acquired lesions from the cerebral arterial vasculature to dural sinuses (1). They represent 10% to 15% of cerebrovascular malformations and most commonly develop in the region of the transverse and sigmoid sinuses (2). It is postulated that the lesions occur due to increased pressure within the dural sinuses, most commonly caused by sinus thrombosis (2–5). Symptoms generally do not develop unless there is increased venous sinus pressure from an increased volume via the dAVF or from sinus obstruction (4). In this situation, the detrimental effects on brain parenchyma become apparent. With the amount of venous hypertension being the variable, the resulting sequelae range from mild reversible symptoms to fatal intracranial hemorrhage (1, 4–6).

The common presenting symptoms of dAVFs range in severity from simple pulsatile tinnitus and focal neurological deficits to acute intracranial hemorrhage. The symptoms also vary depending on the location of the dAVF. For example, a carotid-cavernous fistula may present with proptosis, visual loss, and ophthalmoplegia, while a transverse sinus dAVF may lead to pulsatile tinnitus or parenchymal hemorrhage. Rarely, patients with dAVF present with progressive dementia due to chronic hypoperfusion, and this cognitive impairment has the potential for complete resolution following treatment of the fistula. Cases of cognitive decline with more insidious onset, in addition to the more common presentations, have been reported. In one such case, the patient progressed

Figure 2. Patient 2 (a) before and (b) after treatment. The pretreatment right external carotid artery angiogram shows dilatation of a right middle meningeal artery branch (black arrow) supplying a superior sagittal sinus arteriovenous fistula (white arrow) of the superior sagittal sinus. The posttreatment angiogram shows a resolved superior sagittal sinus dural arteriovenous fistula (arrow) without detectable arteriovenous shunting.

from baseline cognitive function to dementia within 1 year (1, 4–7). dAVFs may also mimic well-known cognitive disorders such as Parkinson’s disease (1) and normal-pressure hydrocephalus (5).

A dAVF can be suspected based on physical examination and history but is ultimately diagnosed with various types of imaging. Noncontrast CT images are usually normal but may reveal evidence of subarachnoid hemorrhage. Contrast-enhanced imaging may show tortuous feeding arteries and dilated cortical draining veins. MRI may reveal evidence of thrombosed sinus and associated T1/T2WI flow voids. On fluid-attenuated inversion recovery (FLAIR) images, an isointense thrombosed sinus along with adjacent edema may be seen if venous congestion or ischemia is present. Digital subtraction angiography (DSA) usually reveals predominance of vascular supply from meningeal arteries. Additionally, cortical veins and retrograde venous drainage may be seen.

Endovascular approaches are generally recommended as the initial treatment in most dAVFs (5). Depending on the anatomy, a transarterial, transvenous, combined transarterial/transvenous, or transorbital approach can be used to cure a dAVF endovascularly (8–11). Multiple embolic agents can be used; however, liquid embolic agents and coils tend to be the primary materials (12). These techniques have been shown to eliminate the symptoms as well as offer an anatomic cure (5). If the patient continues to experience symptoms, a combined endovascular and neurosurgical approach is recommended to obliterate any remaining fistula (13).

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