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Letter to the Editor

The Multifocal Dural Arteriovenous Fistula Presumed to Present as a Progressive Cognitive Impairment

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Dear Editor,

A dural arteriovenous fistula (dAVF) is a pathologically high-flow connection between arteries and veins in the dura mater, bypassing the capillary bed and potentially leading to a variety of neurological complications.[1] Some dAVFs remain asymptomatic, but they can present with headache, pulsatile tinnitus, seizure, and focal neurological deficit. The clinical presentation of dAVFs is determined by the involved vasculature and related brain regions.[1] Persistent venous congestion/hypertension without hemorrhage involving the thalamus or diffuse white matter structures can manifest as rapidly progressive cognitive decline or parkinsonism mimicking neurodegenerative disorders.[1,2] Herein, we present a patient with rapidly progressive cognitive impairment due to dAVF.

A 67-year-old man presented with rapidly progressive cognitive decline and an altered personality. He had a 6-year education level and worked as a real estate agent. He was on anti-diabetic medication and new oral anticoagulants for deep vein thrombosis in his leg. He had quit smoking and drinking alcohol several years ago. He complained of easy forgetfulness, word-finding difficulty, and being prone to anger over the past three months. However, his ability to communicate with clients was not impaired and he did not exhibit other features such as changes in appetite, impulsivity, socially inappropriate behavior, or apathy. His spouse reported that these changes developed relatively acutely and persisted for 3 months. He denied headache, transient visual obscuration, pulsatile tinnitus, or dizziness but complained of brain fog. He also reported that his speech had become slurred, although his articulation seemed to be normal.

On neurological examination, there was no papilledema and his cranial nerves were normal. The motor and sensory system, gait, and cerebellar function tests were all within the normal limits. He underwent neuropsychological testing using the Seoul Neuropsychological Screening Battery.[3,4] It revealed 19/30 of the Mini-Mental State Examination, 0.5 of the global clinical dementia rating, and 3.5 of the sum of boxes. He scored significantly poorly (below −1.0 standard deviation) on the delayed recall of verbal (z=−2.53) and visual memory task (z=−1.40) of the memory function, the Korean version of Boston naming test (z=−2.23) of the language domain, controlled oral word fluency test (animal, z=−2.48; phonemic, z=−1.85), and the Stroop test (z=−2.45) of the executive domain. The Rey complex figure copy test of the visuospatial function was borderline (z=−0.95). The patient's impulsivity and aggressive behavior toward the examiner prevented him from completing the forward digit span test

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(attention domain). He also had difficulty concentrating during the cognitive tests and occasionally became angry with the examiner. Based on his preserved daily activities, he was diagnosed with mild cognitive impairment. Laboratory findings, including blood chemistry, thyroid function test, vitamin B6/B12 levels, and syphilis screening, were within the normal limits. Brain magnetic resonance images revealed a high-flow arteriovenous fistula, involving the vein of Galen, straight sinus, and left transverse sinus, as well as multifocal microbleeds in the frontal lobe (**Fig. 1**). The time-resolved 3D contrast-enhanced magnetic resonance angiography revealed prominent draining veins with abnormal flow-related signal intensities in the straight sinus and the right sigmoid sinus; however, no feeding artery was identified. These findings suggested venous congestion due to dAVF. The high-flow dAVF with diffuse cortical venous reflux was considered to be the cause of the patient's cognitive decline and neurobehavioral symptoms (aggressiveness). The fundoscopic examination and optical coherence tomography were normal. Digital subtraction angiography (DSA) was scheduled for further evaluation and treatment planning, but the patient transferred to another hospital for treatment.

dAVF accounts for 10%–15% of all intracranial vascular malformations.[1] dAVF becomes symptomatic if hemorrhage or venous hypertension/congestion occurs. dAVFs can present acutely with hemorrhage, or subacutely with venous hypertension/congestion without hemorrhage. In the latter cases, the neurological manifestations include pulsatile tinnitus, seizures, altered consciousness, and intracranial hypertension. Atypical presentations include dAVF cases mimicking transient global amnesia (TGA),[5,6], parkinsonism,[7,8]

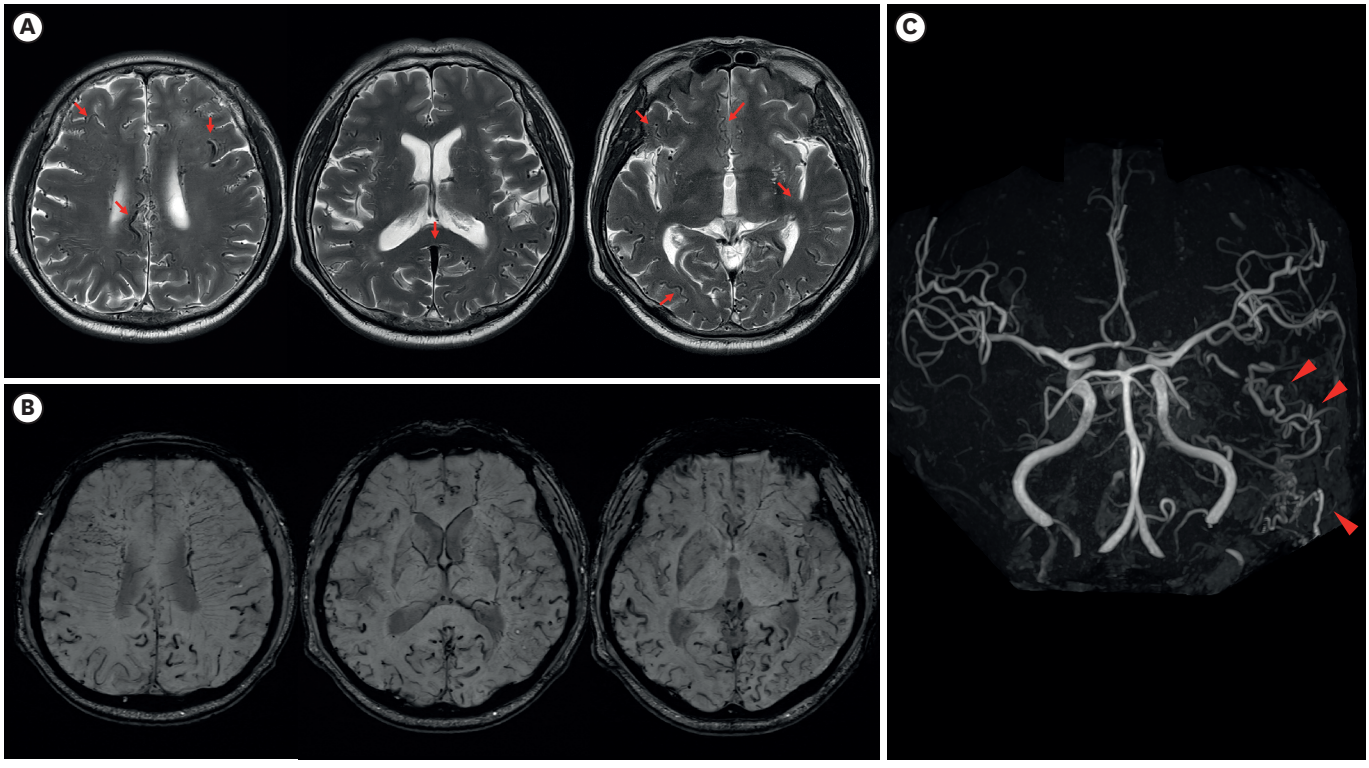

Fig. 1. (A) The T2-weighted images, (B) susceptibility-weighted images, and (C) TOF intracranial vessel images. (A) Prominent signal void structures were observed in the bilateral cerebral hemisphere (arrows). (B) Susceptibility-weighted images revealed abnormally conspicuous cortical veins in the bilateral hemispheres and multifocal microbleeds in the bilateral frontal lobes. (C) TOF angiography revealed abnormal flow-related signal intensities in the cortical veins in the left cerebral hemisphere (arrowheads).
TOF: time-of-flight.

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or unspecified rapidly progressive dementia.[2,9] dAVF patients presenting as TGA are characterized by temporal lobe involvement resulting in transient ischemia and venous congestion, causing transient cognitive impairment even in the absence of a definite brain parenchymal lesion.[5,6] dAVF that manifests as a progressive parkinsonism show a particular involvement of the transverse and straight sinus, various degrees of the basal ganglia and brainstem lesion, and even loss of presynaptic dopaminergic uptake.[7,8]

Although the overall prevalence of dAVF is similar in both sexes, dementia due to dAVF is prevalent in middle- to older-aged (in their 60s) men.[2] The cognitive impairment due to dAVF is usually reported as thalamic edema[2,9] or diffuse white matter lesion.[10,11] The former is called thalamic dementia, which is caused by venous congestion and ischemia of the thalamus due to impaired deep venous drainage. It usually occurs from venous reflux to the vein of Galen and straight sinus. Cortical dAVF may induce medullary venous congestion due to venous hypertension of the draining sinus or direct reflux into the arterialized medullary veins.[1,2] In our case, the patient presented with a dAVF involving the cortical veins of both hemispheres, the vein of Galen, and the straight sinus. Previously reported asymptomatic dAVF cases among individuals with regular check-ups revealed focal involvement, particularly in the transverse sigmoid or tentorial sites.[12] In our patient, although there was no prominent white matter lesion or thalamic edema, microbleeds near the engorged cortical veins suggested venous congestion. Taken together, this mixed involvement may be the cause of insidious onset cognitive impairment, despite the lack of evidence of reversibility after treatment. The management of symptomatic dAVF includes endovascular embolization, surgical occlusion of the fistula, radiosurgery, or a combination of those treatments. Most patients showed an improvement with the appropriate treatment of dAVF.[2,9-11]

Our report has significant limitations. Among them, the absence of DSA results and the inability to definitively establish posttreatment symptoms improvement. Nonetheless, it highlights the potential correlation between DAVF and cognitive impairment. DAVF should be included in the differential diagnosis of subacute-onset cognitive decline, behavioral symptoms, and various neurological manifestations, such as parkinsonism, pulsatile tinnitus, and intracranial hypertension in middle-aged to older men.

REFERENCES

1. Wilson M, Enevoldson P, Menezes B. Intracranial dural arterio-venous fistula. Pract Neurol 2008;8:362-369.
PUBMED | CROSSREF
2. Brito A, Tsang AC, Hilditch C, Nicholson P, Krings T, Brinjikji W. Intracranial dural arteriovenous fistula as a reversible cause of dementia: case series and literature review. World Neurosurg 2019;121:e543-e553.
PUBMED | CROSSREF
3. Ryu HJ, Yang DW. The Seoul Neuropsychological Screening Battery (SNSB) for comprehensive neuropsychological assessment. Dement Neurocogn Disord 2023;22:1-15.
PUBMED | CROSSREF
4. Kang YJ, Na DL. Seoul Neuropsychological Screening Battery. 2nd ed. Seoul: Human Brain Research & Consulting Co., 2012.
5. Takahashi Y, Yamamoto T, Abe T, Tokutomi T, Yoshimura F, Yuge T, et al. Transient global amnesia and dural arteriovenous fistula of the anterior cranial fossa. Kurume Med J 1996;43:223-229.
PUBMED | CROSSREF
6. Johnson P, Ghodke B, Khot S. Dural arteriovenous fistula causing recurrent transient global amnesia. Neurohospitalist 2018;8:200.
PUBMED | CROSSREF

7. Kim HR, Lee JY, Kim YK, Park H, Kim HJ, Son YJ, et al. Dural arteriovenous fistula-associated reversible parkinsonism with presynaptic dopaminergic loss. J Mov Disord 2015;8:141-143.
PUBMED | CROSSREF
8. Tominaga A, Kiura Y, Kondo H, Takeshita S, Magaki T, Sakoguchi T, et al. Dural arteriovenous fistula-derived parkinsonism symptoms: a case report with literature review. J Neuroendovasc Ther 2022;16:257-264.
PUBMED | CROSSREF
9. Holekamp TF, Mollman ME, Murphy RK, Kolar GR, Kramer NM, Derdeyn CP, et al. Dural arteriovenous fistula-induced thalamic dementia: report of 4 cases. J Neurosurg 2016;124:1752-1765.
PUBMED | CROSSREF
10. Matsuda S, Waragai M, Shinotoh H, Takahashi N, Takagi K, Hattori T. Intracranial dural arteriovenous fistula (DAVF) presenting progressive dementia and parkinsonism. J Neurol Sci 1999;165:43-47.
PUBMED | CROSSREF
11. Mendonça N, Santos G, Duro D, Machado E, Goulão A, Santana I. Multiple dural arteriovenous fistulas presenting as rapidly progressive dementia. Neurologist 2012;18:130-132.
PUBMED | CROSSREF
12. Hirata K, Ito Y, Ikeda G, Uemura K, Sato M, Marushima A, et al. Detection rate and radiological features of asymptomatic intracranial dural arteriovenous fistula: analysis of magnetic resonance imaging data of 11745 individuals in the Japanese brain check-up system. J Neuroendovasc Ther 2021;15:787-792.
PUBMED | CROSSREF