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Treatment of Dural Arteriovenous Fistula Presenting as Typical Symptoms of Hydrocephalus Caused by Venous Congestion

—Case Report—

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Abstract

A 76-year-old woman presented with a dural arteriovenous fistula (DAVF) manifesting as typical symptoms of normal pressure hydrocephalus such as progressive dementia, gait disturbances, and urinary incontinence. The cerebrospinal fluid pressure during lumbar puncture was 120 mmH_2O. Computed tomography and magnetic resonance imaging showed ventricular dilation and diffuse white matter changes, which were consistent with the symptoms of hydrocephalus. Cerebral angiography revealed a DAVF in the transverse-sigmoid sinuses with severe cortical venous reflux into the superior sagittal sinus. Transarterial embolization of the feeding arteries reduced the venous flow from the cortical veins into the superior sagittal sinus. Her symptoms improved with reduction in ventricle size. However, she suffered recurrence of the same symptoms several months later. Computed tomography and magnetic resonance imaging demonstrated ventricular dilation associated with hydrocephalus. Angiography revealed a fistulous channel in the left transverse-sigmoid junction. Transvenous embolization was performed resulting in complete obliteration of the fistula. Magnetic resonance image findings such as ventricular dilation and diffuse white matter disappeared and the symptoms of hydrocephalus improved. Although DAVFs often present as venous hypertensive encephalopathy, this case presented with ventricular dilation and diffuse white matter changes, which are the typical neurological signs of normal-pressure hydrocephalus. Venous hypertension associated with the DAVF in the transverse-sigmoid sinuses may have been caused by normal pressure hydrocephalus.

Key words: dural arteriovenous fistula, dementia, hydrocephalus, venous hypertension, leukoencephalopathy

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Introduction

Dural arteriovenous fistulas (DAVFs) are arteriovenous shunts within the wall of the dural venous sinuses and represent 10% to 15% of cerebrovascular malformations. The commonly recognized initial symptoms of DAVF are tinnitus, headache, decreased cognitive function, and neurological deficits associated with intracranial hemorrhage.[4] DAVF may also present with progressive dementia as the initial symptom caused by venous hypertension.[2,8] We report a case of DAVF presenting with dementia, gait disturbance, and incontinence, associated with ventricular dilation and diffuse white matter changes on magnetic resonance (MR) imaging.

Case Report

A 76-year-old woman had a 3-year history of pulsatile tinnitus in her left ear and had suffered from progressive dementia, gait disturbance, and urinary incontinence for more than 1 year. On admission to our hospital, bruit was detected in the left mastoid area, but she did not complain of pulsatile tinnitus. Neurological examination revealed that cognitive function for recent memory and orientation were severely impaired, with mini-mental status examination (MMSE) score reduced to 16. She had magnetic gait and frequent urinary incontinence. Therefore, she exhibited the 3 primary symptoms of normal pressure hydrocephalus. Cerebrospinal fluid (CSF) pressure measured at lumbar puncture was 120 mmH_2O. Computed tomography showed hydrocephalus with nonobstructive ventricular dilation (Evans index 0.32). MR imaging showed diffuse hyperintensity in the white matter (Fig. 1A). Flow voids were observed in the cortex of the left parieto-occipital lobe and right cerebellum, which represented cortical venous reflux (Fig. 1E). Left common carotid angiography demonstrated a DAVF in the left transverse-sigmoid sinuses. Multiple occlusive changes in the dural venous sinuses were also observed, and severe retrograde filling of the superior sagittal sinus, left transverse-sigmoid junction, superior petrosal sinus, and left cortical veins was prominent (Fig. 2). In addition, angiography showed that the right internal and external carotid arteries supplied the DAVF at the left transverse-sigmoid junction (Fig. 3).

Staged endovascular procedures were planned to treat the DAVF. Selective transarterial embolization of the feeder arteries, which were to the left of the middle meningeal and occipital arteries, was first performed. Angiography after embolization showed reduction in cortical venous reflux and disappearance of the retrograde filling into the superior sagittal sinus. Her gait disturbance and urinary incontinence disappeared within 1 week, and cognitive function improved (MMSE score 23) in a few weeks. Two months later, her dementia had disappeared completely (MMSE score 30) and MR imaging showed the ventricular dilation with periventricular lucency had disappeared (Fig. 1B, F). We recommended transvenous embolization as the second procedure to obtain an anatomic cure, but the patient and her family were satisfied with the improvement of the symptoms and refused additional treatment.

Symptoms of hydrocephalus recurred at 1 year 8 months after transarterial embolization. T_2-weighted MR imaging again showed periventricular hyperintensity and increased cortical veins (Fig. 1C, G). Angiography demonstrated DAVF development around the left transverse and

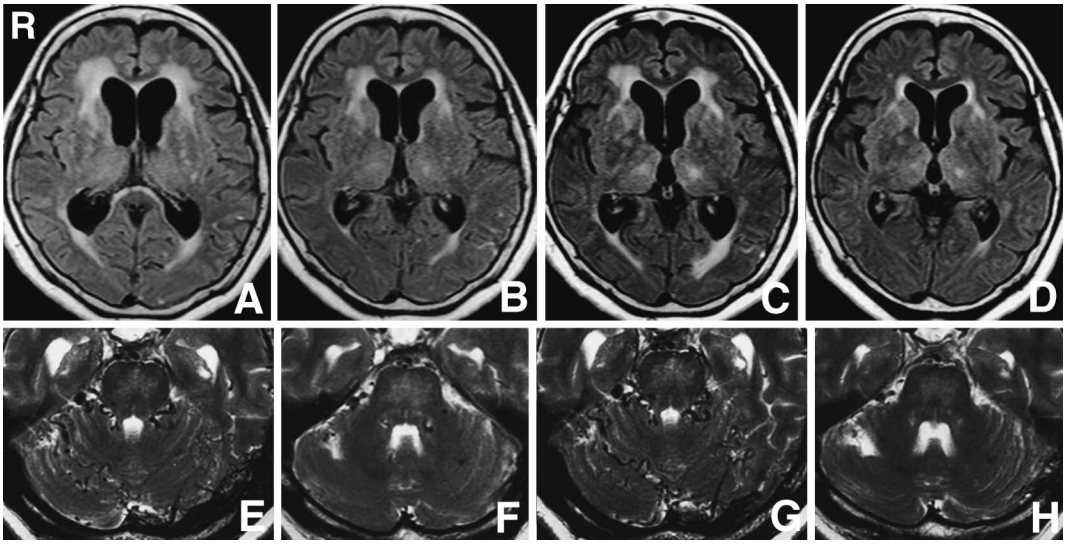

Fig. 1 Axial fluid-attenuated inversion recovery (A–D) and T_2-weighted (E–H) magnetic resonance images. A, E: Dilation of the ventricular systems and periventricular hyperintensity area, and abnormal flow voids in the cerebellum bilaterally are shown before transarterial embolization. B, F: Ventricle size and hyperintensity of periventricular area are reduced after transarterial embolization. C, G: After recurrence of the symptoms of hydrocephalus, increased hyperintensity changes in the periventricular area and cortical venous reflux are seen. D, H: Diffuse hyperintensity changes in the white matter improved and the cortical veins disappeared after transvenous embolization.

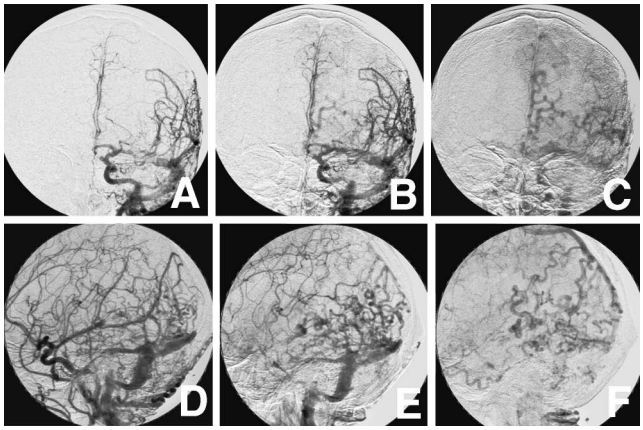

Fig. 2 Left common carotid angiograms, anteroposterior view (A–C) and lateral view (D–F), before transarterial embolization showing the left occipital and middle meningeal arteries supplying the dural arteriovenous fistulas, and severe retrograde filling of the superior sagittal sinus and left transverse sigmoid junction.

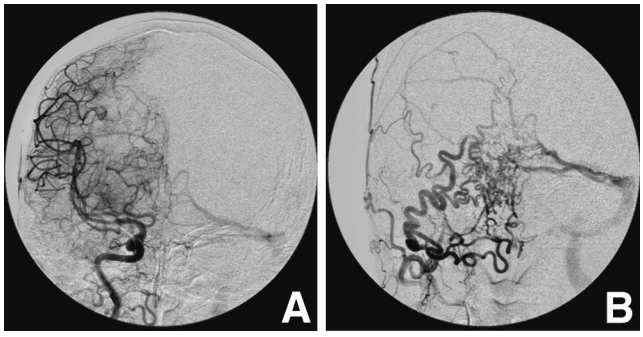

Fig. 3 Right internal (A) and external carotid (B) angiograms showing the dural arteriovenous fistula draining to the left transverse-sigmoid junction.

sigmoid sinuses. The DAVF showed severe retrograde drainage into the left superior petrosal sinus and transverse-sigmoid junction with cortical venous reflux. A fistulous channel was observed in the left transverse-sigmoid junction. Transvenous embolization for the left transverse-sigmoid junction was performed using detachable coils (Fig. 4A, D). Angiography showed complete obliteration of the DAVF in the left transverse and sigmoid sinuses (Fig. 4B, C, E, F). T_2-weighted MR imaging showed the ventricular dilation and periventricular lucency had disappeared with reduced cortical venous reflux (Fig. 1D, H). Gait disturbance, urinary incontinence, and cognitive function disappeared within 1 month. She was able to perform activities by herself and was discharged.

Discussion

In our case, the DAVF was detected in the transverse-sigmoid sinuses, which manifested as the 3 typical symptoms

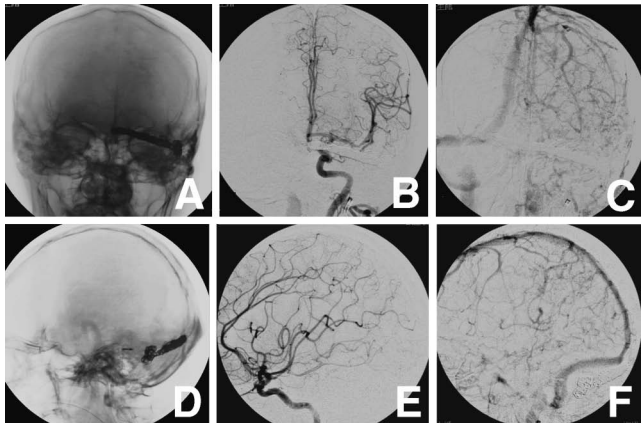

Fig. 4 Left carotid angiograms, anteroposterior view (A–C) and lateral view (D–F), after transvenous embolization of the transverse-sigmoid junction using detachable coils showing disappearance of the dural arteriovenous fistula and no abnormal venous filling.

of normal pressure hydrocephalus. Several patients with DAVFs have presented with progressive dementia.[2,7–9] These patients exhibited venous congestive encephalopathy caused by venous hypertension. The deep white matter in cerebral hemispheres is most vulnerable to venous congestion. Hyperintensity on T_2-weighted MR imaging may be reversible after treating venous congestion.[8,9] Five of 40 patients with DAVF suffered from encephalopathy, and remission of cognitive symptoms occurred in each patient following selective embolization.[2] One previous patient with DAVF of the superior sagittal sinus presented with the symptoms of hydrocephalus.[6]

The mechanism of nonobstructive hydrocephalus caused by venous hypertension is unclear. The conventional view of CSF transport is that CSF is absorbed from the subarachnoid compartment through the arachnoid villi or granulations that project into the venous sinuses of the dura mater on the convexity of the brain.[3] The hydrostatic pressure difference between the CSF compartment and venous sinuses may be the driving force for absorption.[3] Cerebral venous hypertension resulting from direct arteriovenous shunting and/or obstruction of the sinuses may cause problems with the CSF dynamics. If the pressure increases in the sinuses, especially the sagittal sinus, the hydrostatic pressure difference is reduced and CSF absorption is impaired.[5,6] Hydrocephalus secondary to venous hypertension may be observed in young patients with high flow fistulae and vein of Galen malformation, as well as in adults with unruptured arteriovenous malformations without ventricular obstruction.[1,7] Therefore, hydrocephalus may occur secondary to venous hypertension in the sinuses.[5–7] Therefore, ventricular dilation may be the final result of encephalopathy due to venous hypertension.

The present case of DAVF presented with the 3 typical symptoms of idiopathic normal-pressure hydrocephalus including dementia, gait disturbance, and urinary incontinence, corresponding with the specific findings on MR

imaging which indicated hydrocephalus. This case demonstrated increased pressure in the superior sagittal sinus because of severe retrograde filling similar to the previous reported case.[6] These symptoms and the changes on MR imaging disappeared after the treatment of DAVF in both cases. We believe that the occurrence of venous hypertension, especially in the sagittal sinus, due to DAVF is attributable to normal pressure hydrocephalus.

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