
Reversibility of White Matter Changes and Dementia after Treatment of Dural Fistulas

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Summary: We describe two patients with dural fistulas who presented with dementia and diffuse white matter signal changes on MR that significantly improved after surgery. One patient had preoperative embolization.

Index Terms: Fistula, arteriovenous; Dementia; Interventional neuroradiology

Dural fistulas account for about 6% of supratentorial and 35% of infratentorial cerebral arteriovenous malformations (1). Dural fistulas of the posterior fossa, which constitute the majority of dural fistulas, were described in the 1930s by Tonnis (2) and Rottgen (3). Patients with dural fistulas can present with alterations in mental status (4–9) and abnormal magnetic resonance (MR) imaging studies (7, 10).

Case Report

Case 1

A 55-year-old right-handed man had severe dementia, global aphasia, and the ability to follow only simple commands. Four months before admission, bifrontal and occipital headaches began. Two months before admission, he started having intermittent seizures. In the month before admission, the patient lost the ability to walk because of increased spasticity of the lower extremities, and his speech deteriorated until he was only able to say single words.

An MR image showed diffuse white matter disease (Fig 1A). A cerebral angiogram showed arteriovenous shunting between the right ascending pharyngeal artery and the jugular bulb (Fig 1B). The right internal jugular vein and the left transverse sinus were occluded. There was significantly delayed venous drainage with retrograde filling of the right sigmoid, transverse, superior sagittal, and straight sinuses and their tributaries.

After all other causes of metabolic and anatomic dementia had been excluded, the patient had surgery, during

which the right sigmoid sinus and jugular bulb were exposed, skeletonized, and obliterated with muscle. The right external carotid artery was ligated. The postoperative angiogram showed significantly earlier venous drainage, no residual fistula, and drainage of the brain via cortical veins into the cavernous sinus and pterygoid plexus.

Postoperatively, there was gradual and continual improvement in the patient's clinical condition. Six months after surgery, he was able to ambulate independently with good balance, his memory and speech had improved dramatically, and he was fully conversant. There was only a minimal residual cognitive deficit. A follow-up MR image showed significant diminution of the white matter abnormalities (Fig 1C).

Case 2

A 56-year-old right-handed man had a 3-month history of memory loss, unsteadiness of gait, emotional lability, and transient disorientation with right-sided pulsating tinnitus and headaches.

An MR image showed diffuse white matter disease in the right hemisphere (Fig 2A). A cerebral angiogram showed arteriovenous shunting between meningeal branches of the right internal carotid artery (Fig 2B), multiple branches of the right external carotid artery (Fig 2C), and the left occipital and superficial temporal arteries. The right sigmoid sinus was partially occluded, and there was retrograde filling of the right sigmoid and transverse sinuses and the superior sagittal sinus.

The right external carotid feeders were embolized three times with polyvinyl alcohol over a period of 2 weeks with no apparent change in the patient's clinical status. When surgery was performed 2 weeks later, the right transverse and sigmoid sinuses were skeletonized and meningeal feeders coagulated. An intraoperative angiogram showed no residual arteriovenous shunting.

Postoperatively, the patient experienced a new transient sixth-cranial-nerve palsy and slight left hemiparesis that resolved significantly by 2 days and completely by 1 month after surgery. The patient's gait instability, memory

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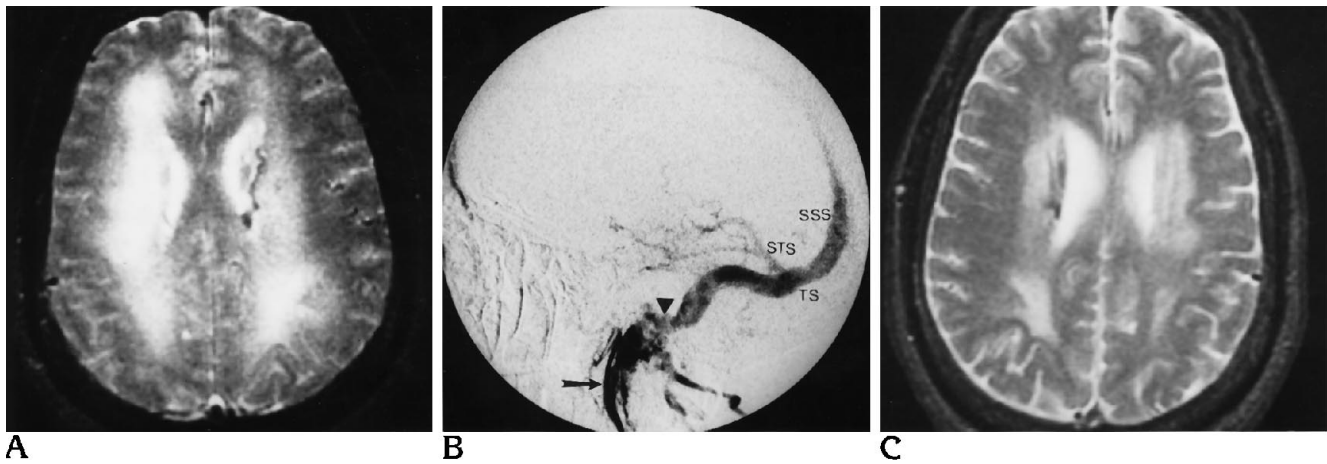


Fig 1. Case 1.

A, Preoperative axial spin-echo MR image (2951/100/1 [repetition time/echo time/excitations]) shows diffuse abnormal increased signal intensity throughout the white matter of the centrum semiovale.

B, Preoperative right ascending pharyngeal artery angiogram (lateral) shows arteriovenous shunting between the right ascending pharyngeal artery (*arrow*) and the jugular bulb (*arrowhead*). Note the retrograde filling of the sigmoid, transverse (TS), superior sagittal (SSS), and straight (STS) sinuses and their tributaries.

C, Axial spin-echo MR image (3000/100/1) 6 months after surgery shows significant improvement in the diffuse white matter signal changes.

deficit, and emotional lability were significantly improved, and he no longer had pulsating tinnitus and headaches. An MR examination 3 months after treatment showed significant improvement in the white matter changes (Fig 2D).

Discussion

Dural fistulas have been classified into four types on the basis of their venous drainage pattern: type I, drainage into a sinus (or a meningeal vein); type II, sinus drainage with reflux into cerebral veins; type III, drainage solely into cerebral veins; and type IV, association with supratentorial or infratentorial venous lakes (11). Both of our patients had type II fistulas.

The cause of dural fistulas is uncertain. Many believe they are acquired lesions that develop because of thrombosis of a dural sinus (12). Perhaps the initiating event is whole or partial occlusion of a dural sinus resulting in venous hypertension. This may then cause retrograde enlargement of the microscopic arterial supply of the wall of the adjacent dural sinus. Nishijima et al (13) confirmed histologically that dural fistulas are located within the wall of involved dural sinuses. These channels progressively enlarge, resulting in arteriovenous shunting. Eventually the shunting becomes significant enough to itself cause further obstruction of normal venous drainage.

Venous hypertension, whether it is caused by the initial sinus thrombosis or subsequent arte-

riovenous shunting, causes impeded cerebrospinal reabsorption in the arachnoid villi and increased intracranial pressure (7). The consequent passive congestion of the brain exists not only on the side of the dural fistula but also in the contralateral hemisphere (14) and causes the MR and clinical changes found in these patients. In our patients, the arteriovenous shunting was most likely the major contributor to the venous hypertension, as evidenced by the significant improvement in MR finding and clinical status after transection of the fistulous feeders without reopening the dural sinuses.

In our patients, it was interesting to note that there was relative sparing of the cortical regions by the white matter changes. We hypothesize this was because of the greater venous hypertension in the deep venous system than in the superficial venous system. The latter has a greater number of alternative pathways to the external jugular system, such as through the diploetic space. In case 2, a number of enlarged cortical veins were noted on the side opposite the white matter changes. We believe this was caused by the lower peripheral vascular resistance on the side opposite the fistula and partially occluded sinus, resulting in greater venous drainage on that side.

Dural fistulas of the transverse and sigmoid sinuses vary in both symptoms and prognosis (5). Initial complaints are often nonspecific and

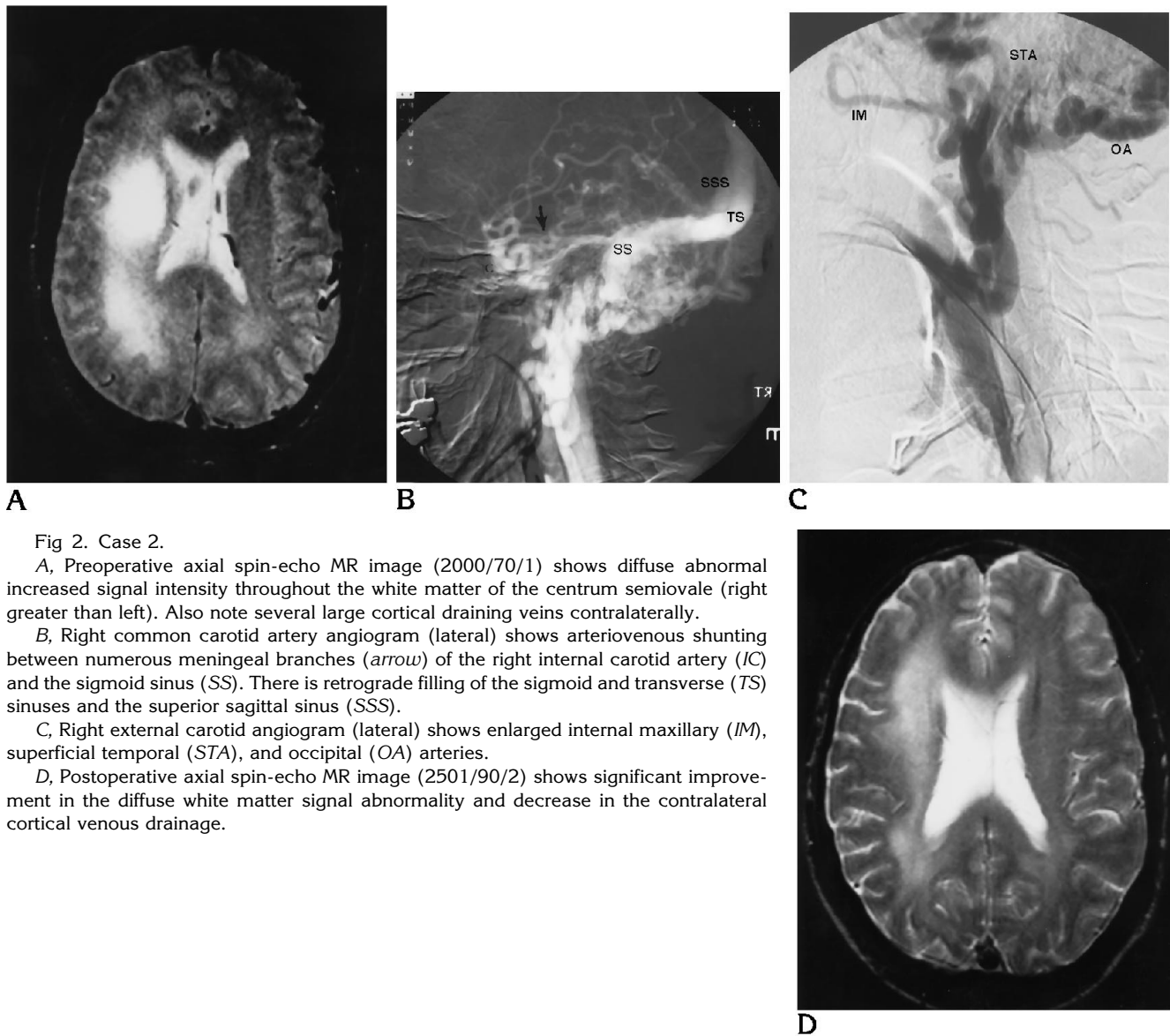


Fig 2. Case 2.

A, Preoperative axial spin-echo MR image (2000/70/1) shows diffuse abnormal increased signal intensity throughout the white matter of the centrum semiovale (right greater than left). Also note several large cortical draining veins contralaterally.

B, Right common carotid artery angiogram (lateral) shows arteriovenous shunting between numerous meningeal branches (*arrow*) of the right internal carotid artery (IC) and the sigmoid sinus (SS). There is retrograde filling of the sigmoid and transverse (TS) sinuses and the superior sagittal sinus (SSS).

C, Right external carotid angiogram (lateral) shows enlarged internal maxillary (IM), superficial temporal (STA), and occipital (OA) arteries.

D, Postoperative axial spin-echo MR image (2501/90/2) shows significant improvement in the diffuse white matter signal abnormality and decrease in the contralateral cortical venous drainage.

include pulse-synchronous tinnitus, bruit, headache, papilledema, seizures, hemorrhage, proptosis, visual disturbances, altered mental status, and transient or permanent neurologic deficits (12, 14).

Obrador et al (4) reviewed 96 cases of dural fistula and found that alteration of mental status was a prominent symptom in 12% of patients. In a review of 45 published cases of dural fistula of the transverse and sigmoid sinuses, Ishii et al (5) found dementia in 10 patients. Dementia was more commonly seen in patients with associated occlusion of the venous sinuses, as was present in both of our patients. There have been rare reports of improvement in dementia

after treatment of a dural fistula (5, 6), as was the case with our patients.

Diffuse white matter changes have been reported in association with a number of diseases including hydrocephalus, acquired immunodeficiency syndrome, multiple sclerosis, progressive multifocal encephalopathy, metabolic and neurodegenerative diseases, disseminated encephalomyelitis, and lymphoma. There have been a few reports describing diffuse (5) or focal (15, 16) abnormal decreased attenuation of the white matter on computed tomograms of patients with dural fistulas. Ishii et al (5) reported one patient in whom these changes became less prominent after treatment. Although abnor-

mal MR studies have been described in patients with dural fistulas (7, 10), we were unable to find a previous report of diffuse abnormal signal changes such as were seen in our patients.

Spontaneous regression of a dural fistula has been reported (17, 18) but is rare. Moreover, although these lesions have been regarded as benign, their natural history can be one of relentless progression producing severe neurologic deficits and death (4). To improve the prognosis, early therapy is essential. The goal of treatment is to improve cerebral hemodynamics, especially venous return, by interrupting the arteriovenous shunting or by recanalization of the sinus that is occluded. Current surgical approaches to dural fistulas of the transverse and sigmoid sinuses include complete excision and packing of the involved sinus (19, 20). Transarterial (20–22) or transvenous (21) endovascular therapy, with or without adjunct surgery, has also been successful in managing these difficult cases.

Mental deterioration represents a rare but devastating consequence of dural fistulas of the transverse and sigmoid sinuses. The MR studies of these patients can be striking. Successful treatment of the fistula can result in significant improvement in the white matter changes seen on MR and in the clinical status of these patients.

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