Galenic dural arteriovenous fistula: unusual clinical presentation and successful endovascular therapy

Case report

JOHN B. WEIGELE, M.D., PH.D., JOHN C. CHALOUPKA, M.D., AND WALTER S. LESLEY, M.D.

Interventional Neuroradiology Service, Department of Radiology, University of Iowa Hospital and Clinics, Iowa City, Iowa

The authors report a case in which the clinical and neuroimaging findings were initially considered diagnostic of a brainstem glioma. Angiography revealed a deep venous system (galenic) dural arteriovenous fistula causing brainstem interstitial edema. Successful endovascular surgery resulted in complete clinical recovery of the patient and resolution of the structural abnormalities that had been observed on magnetic resonance images. The neuroimaging and therapeutic significance of this case are discussed.

KEY WORDS • galenic dural arteriovenous fistula • deep venous system • pons • edema

Dur al arteriovenous fistulas represent 10 to 15% of all intracranial vascular malformations. They most commonly occur in the transverse, sigmoid, and cavernous sinuses. Galenic (deep venous system) DAVFs are rare and constitute the type of DAVF that is most frequently associated with aggressive clinical behavior, presenting the clinician with diagnostic and therapeutic challenges.

We report the case of a patient in whom diffuse swelling and edema of the pons, midbrain, and thalami were initially thought to represent an infiltrating brainstem tumor. The initial findings were actually due to a galenic DAVF that caused reversible interstitial edema at the brainstem. The neuroimaging evaluation and endovascular therapy are discussed.

Case Report

This 53-year-old man presented with a several-month history of dizziness, double vision, perioral numbness, and personality changes.

Examination. The patient displayed left fifth, bilateral sixth, and right seventh cranial neuropathies, decreased sensation to pinprick on the right side, and dysdiadochokinesia on the left side.

Magnetic resonance images of the brain obtained at another institution demonstrated swelling and an increased T₂ signal in the pons, midbrain, and thalami (Fig. 1 upper left and right, and lower left). An infiltrating brainstem glioma was diagnosed on the basis of the clinical and neuroimaging findings, and the patient was transferred to our medical center for further treatment. When the MR images were reviewed, a large, enhancing vessel was noted posterior to the brainstem, which was considered an unusual finding in a case of glioma (Fig. 1 lower right). Prompted by our observation, the patient underwent digital subtraction cerebral angiography, which demonstrated a Djindjian Type III galenic system DAVF supplied by petrosal branches of both middle meningeal arteries, the neuromeningeal branch of the right ascending pharyngeal artery, the right marginal artery, and vermian branches of both superior cerebellar arteries (Fig. 2 left). The venous drainage flowed retrograde through enlarged pontomesencephalic and anterior cortical veins into the superior sagittal sinus (Fig. 2 right). The straight sinus, vein of Galen, internal cerebral veins, and basal veins of Rosenthal were occluded.

Endovascular Surgery. A transarterial embolization of the DAVF was performed 3 days later. Microcatheters were advanced over a microguidewire into the distal petrosal branches of both middle meningeal arteries and the neuromeningeal branch of the right ascending pharyngeal artery. The arteries supplying the DAVF were embolized with a 3:1 iodized oil/N-butyl cyanoacrylate mixture with added tantalum powder. A follow-up angiogram obtained 3 days postoperatively confirmed that the DAVF was completely occluded.

Postoperative Course and Treatment. The patient was discharged the following day with his cranial neuropathies rapidly improving. Five days after discharge deep venous thrombosis and pulmonary emboli developed. He was
treated at another institution with intravenous administration of tissue plasminogen activator and heparin. An oral course of anticoagulation medication (Coumadin) was started. Acute changes in the patient’s mental status developed 4 days later. An emergency computerized tomography scan of the head revealed a 4-cm right occipital hematoma. The partial thromboplastin time in this patient was greater than 140 seconds and the international normalized ratio was 7.68 at that time. An MR image of the brain demonstrated petechial hemorrhages in the pons, mesencephalon, and thalami. The patient again was transferred to our institution where the course of intravenously administered heparin was discontinued and an inferior vena cava filter was placed. The man recovered uneventfully.

The galenic DAVF remained completely occluded on a follow-up angiogram obtained 6 months later. A repeated MR image revealed total resolution of the previous swelling and edema in the brainstem and thalami (Fig. 3 left and center). Residual hemosiderin persisted from the prior cerebral hemorrhage (Fig. 3 center). The vessel dorsal to the brainstem had decreased markedly in size (Fig. 3 right). The neurological examination performed at that time yielded completely normal findings.

Sources of Supplies and Equipment

Prowler 14 microcatheters were purchased from Cordis (Miami, FL) and the Fastdasher 14 microguidewire from Target Therapeutics (Fremont, CA). The iodized oil, Lipiodol, was acquired from Savage Laboratories (Melville, NY) and the N-butyl cyanoacrylate (Histoacryl) from B. Braun (Melsungen, Germany). The tissue plasminogen activator (Alteplase) was obtained from Genentech (San Francisco, CA), the heparin from Elkins–Sinn (Cherry Hill, NJ), and the Coumadin from Dupont (Wilmington, DE).

Discussion

Dural AVFs are pathological arteriovenous shunts located in the dura mater, the cause of which is unknown. Mounting evidence suggests that they are acquired rather than congenital lesions. A number of proposed causes remain unproven and do not explain all cases. An attractive unifying theory suggests that DAVFs are caused by a neoangiogenesis disorder of the dura that may be triggered by any one of a number of inciting events such as dural sinus thrombosis, thrombophlebitis, infection, or inflammation. The pathological sequelae of DAVFs appear to be caused primarily by the arterIALIZATION of dural sinuses and cortical veins, rather than by an arterial steal due to the arteriovenous shunting. The venous hypertension resulting from arterIALIZATION may cause interstitial edema, parenchymal ischemia, infarction, and intracranial hemorrhage.7

Dural AVFs are typically classified by their location in relation to a dural sinus and their pattern of venous drainage (that is, according to the Djindjian classification).5 These lesions occur most commonly in the sigmoid and transverse sinuses, followed by the cavernous sinus.3 Deep venous DAVFs are quite rare, representing only 8.3% of all DAVFs in one large series.7

Galenic DAVFs are a heterogeneous group defined by drainage into the deep venous system at various sites.3 Reported presentations of galenic DAVFs include cata-

FIG. 1. Magnetic resonance images of the brain obtained at the time of the patient’s initial presentation. Edema and swelling in the pons and thalami are evident in the upper left, upper right, and lower left panels. Upper Left and Right: Axial fluid-attenuated inversion-recovery images (TR 8350 msec, TE 126 msec, time interval 1800 msec, number of excitations [NEX] 1) demonstrating a hyperintense, enlarged pons (upper left) and hyperintense, enlarged thalami (upper right). Lower Left: Axial T2-weighted image (TR 6500 msec, TE 96 msec, NEX 2) demonstrating a hyperintense, enlarged pons. Note the partial effacement of the prepontine cistern (arrow). Lower Right: Axial gadolinium-enhanced three-dimensional spoiled-gradient echo image (TR 26 msec, TE 7 msec, flip angle 60°, NEX 1) demonstrating a large enhancing vessel located dorsal to the brainstem (arrow).

FIG. 2. Superselective digital subtraction angiograms revealing a deep venous system (galenic) DAVF. Left: Angiogram depicting the petrosal branch of the right middle meningeal artery before embolization. Right: Retrograde deep venous drainage flow into the pontomedullary veins and an anterior cortical vein.

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Galenic dural arteriovenous fistula

The MR imaging appearances of DAVFs are quite varied. The brain parenchyma may be normal or demonstrate edema, infarction, and/or hemorrhage. Subarachnoid and subdural hemorrhages are common complications. The presence of dilated cortical veins without a visible parenchymal nidus on MR images indicates the presence of a DAVF with associated venoocclusive disease. A few descriptions of localized supratentorial white matter edema, cerebellar edema, and bithalamic edema caused by DAVFs have been published. Rare cases of brainstem edema caused by cavernous sinus DAVFs have been reported.

This case represents an unusual presentation of a galenic DAVF. The clinical features of slowly progressive cranial neuropathies and cerebellar signs, associated with the MR imaging findings of diffuse edema and swelling in the pons, midbrain, and thalami, led to the initial diagnosis of a brainstem glioma. The unexplained presence of a prominent vessel located dorsal to the mesencephalon on the MR image prompted us to obtain an arteriogram in the patient, which revealed a Djindjian Type III galenic DAVF with associated venoocclusive disease. The brainstem and thalamic swelling and edema completely resolved after successful endovascular obliteration of the DAVF. The reversibility of the imaging findings suggests that they represented interstitial edema due to venous hypertension, which was corrected by the endovascular surgery, rather than the presence of an infarct.

The treatment of galenic DAVFs is challenging, often requiring combined transarterial–transvenous embolization and open surgery. In our case, a complete cure was possible by using transarterial embolization alone. This was achieved by very distal placement of the microcatheters, close to the fistulous arteriovenous connections, and the use of a slow, well-controlled injection of a liquid adhesive for which the polymerization rate of the embolic mixture had been adjusted to allow for distal permeation into the fistulas, accomplished using a high iodinized oil/cyanoacrylate ratio.

The findings in this report indicate that the differential diagnosis proposed for MR imaging findings of brainstem swelling and edema should include a galenic DAVF, in addition to previously described diagnostic considerations including glioma, metastasis, lymphoma, germinoma, chordoma, progressive multifocal leukoencephalopathy, multiple sclerosis, acute disseminated encephalomyelitis, mesenchymoblastoma, acute infarction, and central pontine myelinolysis. Authors of some reports have argued that the MR imaging features of brainstem glioma are so characteristic that histological confirmation is unnecessary in the appropriate clinical setting. It is particularly important, therefore, to consider a potentially curable deep venous system DAVF in the differential diagnosis of brainstem edema found on MR images.

Disclaimer
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References
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