Unilateral contrast-enhancing pontomedullary lesion due to an intracranial dural arteriovenous fistula with perimedullary spinal venous drainage: the exception that proves the rule

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A large spectrum of possible diagnoses must be taken into consideration when a contrast-enhancing lesion of the pontomedullary region is found on MRI. Among these diagnoses are neoplastic, inflammatory, and infectious, as well as vascular pathologies. The authors report a rare case of an intracranial dural arteriovenous fistula (DAVF) with perimedullary spinal venous drainage (Cognard Type V) that initially presented as a unilateral contrast-enhancing pontomedullary lesion mimicking a brainstem neoplasm in a 76-year-old man. Following occlusion of the DAVF by transarterial embolization that resulted in clinical and radiological improvement, the fistula recurred 10 months later and was finally cured by a combined endovascular and surgical approach that resulted in complete occlusion. Clinical symptoms and MRI findings gradually improved following this treatment. A literature review on the MRI findings of Cognard Type V DAVF was performed. Centrally located medullary or pontomedullary edema represents the typical imaging finding, while unilateral edema as seen in the authors' patient is exceptionally rare. The hallmark imaging finding suggestive of DAVF consisting of perimedullary engorged vessels may not always be present or may only be very subtly visible. Therefore, the authors suggest performing contrast-enhanced MR angiography or even digital subtraction angiography in the presence of an unclear edematous brainstem lesion before scheduling stereotactic biopsy.

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DURAL arteriovenous fistulas (DAVFs) are abnormal communications between dural arteries and dural venous sinuses, meningeal veins, or cortical veins. They account for 10%–15% of intracranial arteriovenous malformations.^{12,33} Grading of DAVF is based on the venous drainage pattern according to the classification schemes of Borden et al. and Cognard et al.^{4,8} Venous cortical or perimedullary drainage and dilation are the hallmarks of high-grade DAVF and strongly correlate with increased risk of intracranial hemorrhage and nonhemorrhagic neurological deficit. The exact etiology of DAVFs is unknown. They are predominantly idiopathic, though they may also be congenital lesions or found in association with dural venous sinus thrombosis, trauma, previous

craniotomy, and tumors. While spinal DAVF is an established cause of vascular myelopathy, intracranial DAVFs with perimedullary spinal venous drainage (Cognard Type V) are rare disorders and often represent a diagnostic and therapeutic challenge. They have been mainly described in case reports or small series that reported a typical clinical and radiological presentation of progressive myelopathy, central cord edema, and engorged perimedullary veins.

Here, we report a case of an intracranial DAVF with exclusive perimedullary spinal venous drainage that presented initially with a very unusual MRI appearance suggestive of a right lateral brainstem neoplasm. After delayed diagnosis, the DAVF was completely occluded by transarterial embolization and the patient's symptoms gradually improved.

ABBREVIATIONS DAVF = dural arteriovenous fistula; DSA = digital subtraction angiography; MRA = MR angiography.

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However, the fistula recurred after 10 months. Curative fistula treatment was achieved through a combined approach of partial transarterial embolization followed by surgical fistula ligation via a suboccipital craniotomy.

Case Report

A 76-year-old man was referred to our institution with a 3-week history of nausea and vomiting, inability to walk, and blurred vision. The patient's medical history included prostatectomy for prostate carcinoma 7 years earlier, resection of several benign colorectal adenomas 6 months earlier, goiter, recently diagnosed intermittent atrial fibrillation, arterial hypertension, and hypercholesterolemia. There was no history of trauma. Neurological examination showed severe truncal and limb ataxia accentuated on the right side, postural instability, dysphagia, right-sided ptosis, and mild facial asymmetry. Brain MRI revealed a contrast-enhancing lesion of the right pontomedullary junction extending to the right inferior cerebellar peduncle with slight mass effect (Fig. 1A–C). Suspicion of a neoplastic lesion (brainstem glioma or lymphoma) or inflammatory lesion (i.e., tumefactive demyelinating lesion) was raised. CSF analysis showed an elevated total protein level

of 1010 mg/L and elevated CSF/serum albumin ratio of 14.5, indicative of blood-brain barrier disruption. There was no local synthesis of immunoglobulins. The cell count was normal (1/ μ l) with 90% lymphocytes and 10% monocytes. Analysis for B- and T-cell clonality was negative. Findings of further laboratory tests were normal except for a nonspecific elevation of antinuclear antibody (ANA) of 1:3200 (reference titer < 1:50, Hep2-cells). The pontomedullary lesion showed a mildly increased metabolism on FDG-PET (not shown) that was not very indicative of cerebral lymphoma.

Since the imaging diagnosis of this lesion remained unclear and a neoplastic lesion had not been ruled out, a stereotactic biopsy via a left frontal trajectory was performed. Histomorphology and immunohistochemistry showed edematous brain tissue with mild astrogliosis and microgliosis. There were no signs of a neoplastic, inflammatory, or infectious process.

An empirical treatment with prednisolone was initiated, and the patient's neurological symptoms gradually improved over a period of 3 months. The patient was able to walk, his dysphagia was markedly improved, and the previously implanted percutaneous gastrostomy tube could be removed. Follow-up MRI showed a slight de-

FIG. 1. A–C: MR images obtained at initial presentation. Coronal FLAIR (A), axial T2-weighted (B), and axial MPRAGE T1-weighted (C) images showing a contrast-enhancing right pontomedullary lesion (arrows) with minimal extension into the right inferior cerebellar peduncle and mild mass effect. **D:** Follow-up MR image obtained 3 months after the initial presentation following steroid treatment, showing complete resolution of lesional contrast enhancement (arrow in D). **E and F:** Follow-up MR images obtained 8 months after initial presentation following clinical deterioration, revealing enlargement of the lesion with extension to the left side of the medulla on T2-weighted images (arrow in E). Prominent engorged veins are visualized on the anterior cord aspect at the cervicomedullary junction suggestive of DAVF (arrowheads in F). Unchanged mixed hyper-/hypointense signal alteration on T2-weighted and postcontrast T1-weighted images within the right sigmoid sinus indicates chronic venous sinus thrombosis (black arrowheads in B–E).

crease in the size of the lesion and complete resolution of contrast enhancement (Fig. 1D). Prednisolone was tapered to a dose of 20 mg per day.

However, 8 months after the initial presentation, the patient's symptoms worsened again and were now accompanied by hiccups. On MRI, enlargement of the lesion now extending from the pontomedullary junction to the C-3 level of the cervical cord, and notably, to the left side of the medulla, was demonstrated (Fig. 1E). Newly contrast-enhancing engorged veins on the anterior aspect of the medulla were found to be suspicious for a DAVF (Fig. 1F). In addition, findings suggestive of a partially recanalized chronic thrombosis of the left sigmoid sinus were noted for the first time, but in retrospect had been present without change on the earlier MRI studies (Fig. 1B–E). Subsequent digital subtraction angiography (DSA) (Fig. 2) demonstrated a DAVF with feeding arteries originating from the right middle meningeal, ascending pharyngeal, posterior auricular, and occipital arteries and draining into a lateral medullary and then subsequently into the anterior perimedullary/perispinal veins (Cognard Type V). The DAVF was completely obliterated by transarterial embolization with Onyx (ev3) (Fig. 3). The patient experienced a transient worsening of his ataxia and conjugate gaze palsy to the right side. His neurological condition improved slowly within the next 6 months. Gait ataxia persisted but the patient was able to walk short distances without assistance. He reported only mild diplopia and

dysphagia. Follow-up MRI at 6 months showed a complete regression of the medullary edema and contrast enhancement. The prominent anterior spinal vessels were no longer visible, and MRI findings were otherwise normal except for a small area of right medullary gliosis (Fig. 2F).

Ten months following treatment, bulbar symptoms recurred and MRI demonstrated recurrent central medullary edema and engorged anterior spinal veins. Cerebral angiography showed recurrence of the previously occluded DAVF fed by small branches of the right occipital artery, posterior auricular artery, and neuromeningeal division of the ascending pharyngeal artery, again with exclusive perimedullary drainage. The fistulous point was exactly outlined by CT angiography via a microcatheter injection from the right external carotid artery, which was located posterior to the jugular foramen at the dura of the occluded right sigmoid sinus draining on a lateral medullary bridging vein (Fig. 3C). A second transarterial Onyx embolization using a double-lumen balloon catheter (Scepter XC, MicroVention) resulted in occlusion of occipital artery feeders. Small transmastoid feeders of the ascending pharyngeal artery and posterior auricular artery were not embolized because of the potential risk of cranial nerve dysfunction upon their sacrifice. A right retrosigmoid craniotomy was performed, and the remaining feeding arteries of the fistula were identified (Fig. 3A) and surgically interrupted (Fig. 3B). The feeding artery pedicle was secured by a vascular clip. Successful occlu-

FIG. 2. Preembolization DSA images of right external carotid artery (A) and right vertebral artery (B) injections, lateral views. Superselective microcatheter injections of the petrosquamous branch of the right middle meningeal artery, anteroposterior view (C), and neuromeningeal division of the right ascending pharyngeal artery, oblique view (D), were obtained during transarterial embolization. A Cognard Type 5 DAVF with direct arteriovenous shunt on a cortical vein at the right posterior petrous ridge (long white arrow in A–D) with subsequent perimedullary spinal venous drainage into the anterior medullary/anterior spinal vein (short white arrows in A–D) is shown. Arterial feeders originate from the petrosquamous branch of the middle meningeal artery (white arrowheads in A and C), petrous branches of the posterior auricular artery (black arrowhead in A), and branches of the right occipital artery (black arrow in B) that arises from the C-1 collateral of the right vertebral artery, and branches from the neuromeningeal division of the ascending pharyngeal artery (long black arrow in D). After Onyx embolization of the middle meningeal artery and posterior auricular and ascending pharyngeal artery branches, the DAVF is completely occluded (E, right vertebral artery injection, lateral view). Follow-up MR image obtained 6 months postembolization reveals regression of pontomedullary edema with a small residual right medullary lesion (arrow in F).

FIG. 3. A and B: Intraoperative photographs obtained after right suboccipital craniotomy and elevation of the cerebellar tonsil. The feeding artery pedicle to the DAVF at the right posterior petrous ridge was identified (arrow in A) and transected. The proximal end was secured by a vascular clip (B). **C:** Angiographic CT image demonstrating the anatomical location of the DAVF at the posterior petrous ridge in the vicinity of the jugular foramen with a fine network of ascending pharyngeal artery feeders (neuromeningeal division, white arrowheads) draining into the lateral mesencephalic bridging vein (black arrow) and subsequently to the perimedullary veins (white arrow). Previous Onyx casts inside embolized feeders are visualized as high-density structures (black arrowheads). **D:** After surgery, complete fistula occlusion is demonstrated on DSA, external carotid artery injection, lateral view (arrow in D). Figure is available in color online only.

sion of the DAVF was confirmed by cerebral angiography (Fig. 3D). MRI performed 2 weeks later showed a near-complete resolution of the medullary edema. The patients' neurological status rapidly returned to the level before recurrence of the fistula and further improvement was noted. However, at the last follow-up 3 months later gait ataxia persisted.

Literature Review

We performed a literature review and identified 58 reported cases, including our case, of intracranial DAVF with perimedullary spinal venous drainage (Cognard Type V) in the English-language medical literature. Of these cases, 41 were documented with a description of MRI findings, and their imaging studies were reviewed. A summary is presented in Table 1. The typical imaging finding of a centrally located medullary or pontomedullary edema was present in 30 patients (73%). A unilateral lesion was seen in our patient and 2 other cases (7%).^{26,27} Contrast enhancement of the medullary lesions was present in 11 cases (27%), absent in 9 cases (22%), and not reported in 21 cases (51%). Prominent perimedullary flow voids on T2-weighted images were present in 18 cases (44%). Based on T1-weighted images, atypical perimedullary vessels were present in 17 cases, of which 14 were seen on contrast-enhanced T1-weighted images, and 3 on nonenhanced T1-weighted images. In 4 cases, atypical vessels were identified by contrast-enhanced MR angiography (MRA). Overall, atypical perimedullary vessels

were seen on standard MRI in 76% of cases (31 cases). Including MRA, the MRI detection rate of atypical vessels increased to 85% (35 cases).

Discussion

The reported case demonstrates the diagnostic challenge of an intracranial DAVF with perimedullary spinal venous drainage (Cognard Type V). This exceptionally rare type of DAVF can present with a wide array of symptoms due to the variable extent of venous congestive myelopathy.^{10,11,17,19,24} The onset can either be sudden or gradual, making a solely clinical diagnosis impossible.^{18,26,31} Therefore, clinicians confronted with such a lesion are frequently misdirected to a tentative diagnosis of brainstem and spinal cord infarction, Guillain-Barré syndrome, demyelination and inflammation, or—as in our case—a neoplasm.

As the preeminent MRI finding in Cognard Type V DAVF, a central medullary edema was present in 73% of previously reported cases.^{3,9} The clearly unilateral space-occupying lesion observed in our patient rather represents the exception that proves the rule as only 2 comparable cases have been described to date.^{26,27} Atypical perimedullary vessels are the second MRI hallmark of Cognard Type V DAVF. Haryu et al. recently reported the presence of perimedullary flow-related signal voids in only 37% of patients with Cognard Type V DAVF.¹⁵ They concluded that this might be the reason why diagnosis of this rare type of DAVF is frequently delayed and recommended MRA in cases of nonspecific myelopathy. We agree that MRA should be performed in these cases. However, Haryu et al. only reviewed the presence of flow voids on sagittal T2-weighted MRI. When both T2- and T1-weighted MRI, in particular, contrast-enhanced T1-weighted images, are carefully examined, the detection rate of atypical perimedullary vessels by MRI reaches 76%. Together with MRA, the detection rate was 85% in the pertinent literature.

The diagnosis of the DAVF was delayed in our patient because of both the very unusual tumor-mimicking aspect of the initial unilateral contrast-enhancing pontomedullary lesion and the absence of abnormal perimedullary vessels on initial standard MRI, which resulted in the biopsy of the lesion. Intriguingly, both contrast enhancement and clinical symptoms temporarily responded to steroid therapy, underlining the possibility of an underlying inflammatory process. The presence of a partially recanalized, chronically occluded right sigmoid sinus may be another important hint to the diagnosis of a DAVF, which is known as a common sequela of intracranial venous sinus thrombosis.

In summary, this case demonstrates unusual imaging findings of a rare Cognard Type V intracranial DAVF with perimedullary venous drainage mimicking a brainstem neoplasm on initial MRI, which led to a delayed diagnosis and treatment of the lesion. The review of the literature underlines that the imaging hallmark of such DAVFs, consisting of perimedullary engorged vessels, may be present in only 76% of cases. Contrast-enhanced MRA or even DSA should be performed in cases of unclear edematous brainstem lesions.

TABLE 1. Published cases of Cognard Type V DAVF including MRI findings

Authors & Year	Age (yrs), Sex	Centromedullary Edema on Sagittal T2WI	Parenchymatous Enhancement	Flow Voids on T2WI	Atypical Vessels on T1WI or MRA	
					Atypical	Imaging Modality
Gobin et al., 1992	68, F	–	NR	NR	+	T1WI
Versari et al., 1993	50, M	–	NR	NR	+	T1WI+Gd
Bret et al., 1994	31, M	+	NR	–	–	
Ernst et al., 1997	71, M	+	–	+	+	T1WI+Gd
	47, M	+	NR	+	–	
	58, F	+	NR	–	–	
Chen et al., 1998	36, F	–	–	–	+	T1WI+Gd
	47, M	–	–	–	+	T1WI+Gd
Hähnel et al., 1998	67, M	+	–	+	+	T1WI+Gd
Trop et al., 1998	74, M	+	NR	–	+	T1WI+Gd
Bousson et al., 1999	36, M	+	+	+	–	
Hurst et al., 1999	54, M	+	+	–	+	T1WI+Gd
	50, M	+	+	+	–	
Oishi et al., 1999	62, F	+	+	+	–	
	53, M	+	–	–	+	T1WI+Gd
	40, F	–	–	–	+	T1WI
	75, F	+	–	–	+	T1WI
	51, F	–	–	–	–	
Wiesmann et al., 2000	46, M	–	NR	+	–	
Asakawa et al., 2002	64, M	+	+	–	+	MRA
Li et al., 2004	73, M	+	NR	–	–	
Pannu et al., 2004	42, M	+	–	–	+	T1WI+Gd
Satoh et al., 2005	38, F	–	NR	+	–	
Tanoue et al., 2005	70, M	+	NR	+	–	
Akkoc et al., 2006	45, M	+	NR	+	–	
Renner et al., 2006	58, M	+	NR	+	–	
Lagares et al., 2007	65, M	+	NR	–	–	
van Rooij et al., 2007	58, M	+	NR	+	–	
	65, M	+	NR	+	–	
	72, F	+	NR	+	–	
Sugiura et al., 2009	69, F	–	+	NR	+	T1WI+Gd
Patsalides et al., 2010	53, M	+	+	+	–	
Aixut Lorenzo et al., 2011	67, F	+	NR	+	–	
Kim et al., 2011	45, M	+	+	–	+	T1WI+Gd
Ogbonnaya et al., 2011	64, F	–	NR	NR	+	T1WI+Gd
Peltier et al., 2011	58, F	+	+	NR	+	T1WI+Gd
Foreman et al., 2013	59, M	+	NR	–	+	T1WI+Gd
Haryu et al., 2014	62, M	+	NR	–	+	MRA
	64, M	+	+	+	+	MRA
	68, M	+	–	+	+	MRA
Present study	76, M	–	+	–	–	

NR = not reported; T1WI = T1-weighted imaging; T2WI = T2-weighted imaging; + = present; – = absent.

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