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Adult Infratentorial Pial Arteriovenous Fistula Treated with Detachable Coils: A Case Report and a Review of Literature

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Abstract

An appropriate therapeutic option for pial arteriovenous fistula (PAVF) can vary according to the angioarchtecture of the lesion. We present a case of adult infratentorial PAVF treated by transarterial coil embolization. A 26-year-old man was referred to our institution for an asymptomatic intracranial vascular lesion. Cerebral angiograms revealed PAVF fed by three arteries in the right cerebellomedullary cistern. The feeding arteries were accurately identified by three-dimensional rotational angiography and were successfully embolized using coils while normal arterial flow was preserved. This case report suggests that stepwise transarterial coil embolization can cure PAVF under detailed evaluation of its angioarchitecture.

Keywords: pial arteriovenous fistula, vascular malformation, endovascular treatment

Introduction

Pial arteriovenous fistula (PAVF) is a vascular disorder of the brain, that directly connects arteries and veins without the nidal component.[1,2] The best treatment modality for each PAVF case varies according to its location and angioarchitecture.[3] Little is known regarding the safety and effectiveness of the endovascular treatment for PAVF because of its rarity.[4] Here, we report a case of PAVF successfully treated with transarterial coil embolization. We also reviewed the literature of infratentorial PAVF.

Case Report

A 26-year-old man with a history of hepatoblastoma and cryptorchidism underwent head magnetic resonance imaging (MRI) because of dizziness. There were no demonstrable cranial nerve deficits. He had no family history or hereditary disease. Dizziness improved spontaneously, but MRI showed a cluster of dilated vessels occupying the right cerebellomedullary cistern (Fig. 1a). Six-vessel cerebral angiography revealed a high-flow arteriovenous shunt within the right cerebellomedullary cistern (Fig. 1b-e). The shunt was supplied by the arteries branching from the right vertebral artery (VA) and the right posterior inferior cerebellar artery (PICA). The feeding arteries anastomosed each other, and they flowed into multiple drainage routes that drained into bilateral internal jugular veins and the right superior petrosal sinus. Varix formed on the drainage route behind the medulla oblongata. There was no nidal component and dural supply. On the basis of these findings, the patient was diagnosed with a PAVF. We suggested transarterial embolization to prevent future development.

While the patient is under general anesthesia, a guiding catheter was positioned within the right VA. Before the embolization procedure, a balloon microcatheter (Scepter C; MicroVention, Tustin, CA, USA) was threaded into each feeder, and superselective angiography was performed with balloon occlusion. Arterial supplies to normal nervous tissues were not observed from each feeding artery. Then, all three supplying arteries were occluded in turn using detachable coils through two microcatheters. After two of the feeders from the VA were occluded, the feeding artery that branched from the PICA was occluded just proximal to the

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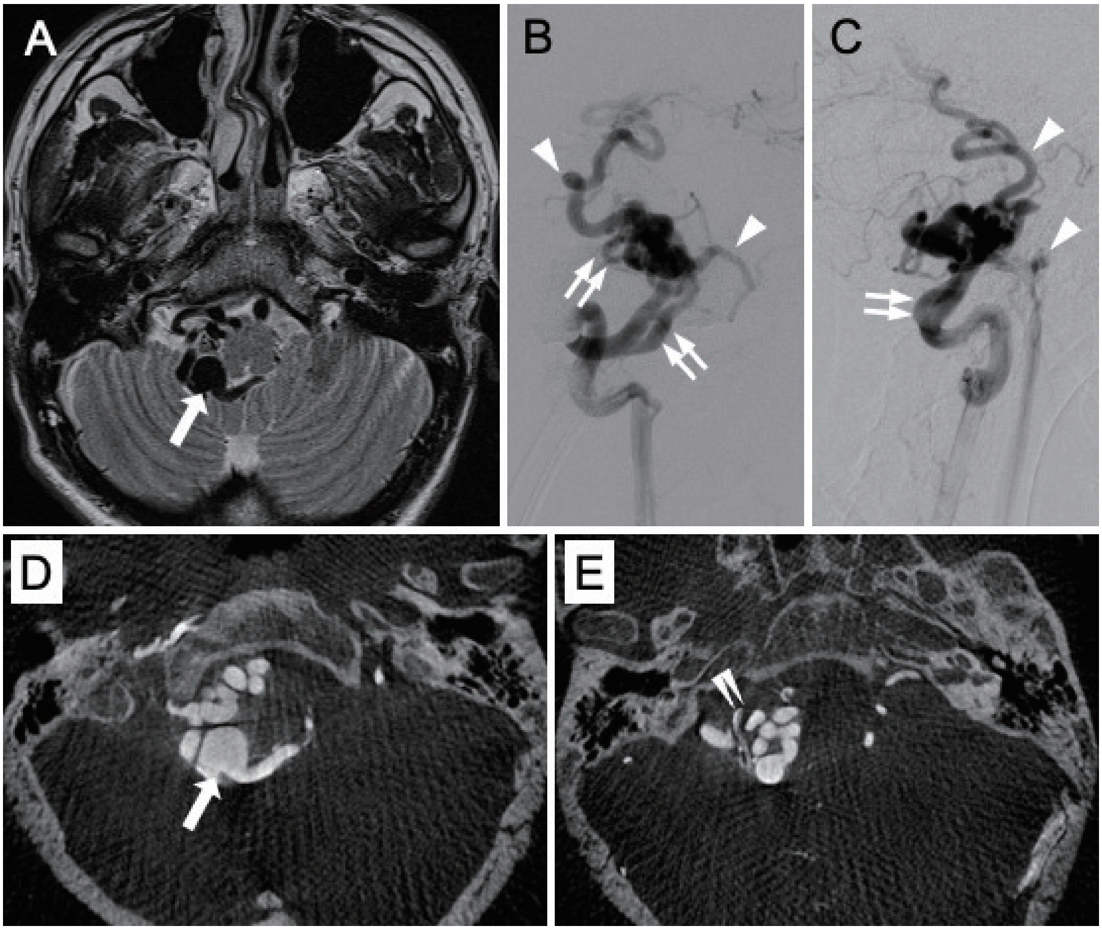

Fig. 1 T2-weighted magnetic resonance imaging (A) shows dilated and tortuous vessels in the right cerebellomedullary cistern. The varix (arrow) is observed behind the medulla oblongata. Anterioposterior view (B) and lateral view (C) of the right vertebral angiogram revealed pial arteriovenous fistula. Arrowheads indicate the early filling of veins, and double arrows indicate feeders branched from the vertebral artery and posterior inferior cerebellar artery. Axial views of slab MIP images (D and E) of the right vertebral angiogram show the detailed structure of the lesion. Note that vessels are distinguishable from each other and that it is easy to chase their connections. Double arrowheads indicate the right posterior inferior cerebellar artery.

varix, which was estimated as the shunt point. Immediate occlusion of the fistula that preserved the VA and the PICA flow was achieved. The postoperative course was uneventful, and the patient did not present any neurological deficits following treatment. The follow-up angiography performed 9 months after treatment showed no residual shunt (Fig. 2).

Discussion

PAVF was once recognized as a subtype of arteriovenous malformation (AVM), but it is currently considered a distinct entity from AVM and dural AVF in terms of angioarchitecture and clinical course. It is important to differentiate PAVF and other shunt disorders because the therapeutic goal of these vascular diseases can be different.[5] PAVF is composed of a single arterial feeder or multiple arterial feeders in direct connection to venous drainage without intervening nidal vessels.[6] The cause of PAVF in infants and children is often congenital,[7] whereas PAVF in adults is usually caused by an acquired factor, such as trauma or venous thrombosis.[8,9] In our case, there was no family history or possible cause of the shunt.

We searched PubMed using the keyword “pial arteriovenous fistula” and “non-galenic arteriovenous fistula” from the inception of the databases in December 2022. We excluded articles without clear descriptions regarding angiographic finding, treatment choice, and outcome. We also excluded galenic PAVF, secondary PAVF, cases with dural arterial supply, and untreated cases. Among 266 PAVF cases reported since 1977, 35 lesions (13.2%) were located in the posterior cranial fossa.[4,10-33] Those cases and the present case were reviewed (Table 1).

A summary of the reviewed cases is shown in Table 2. Of the total of 36 patients whose infratentorial PAVF was treated, the most common presentation symptom was fo-

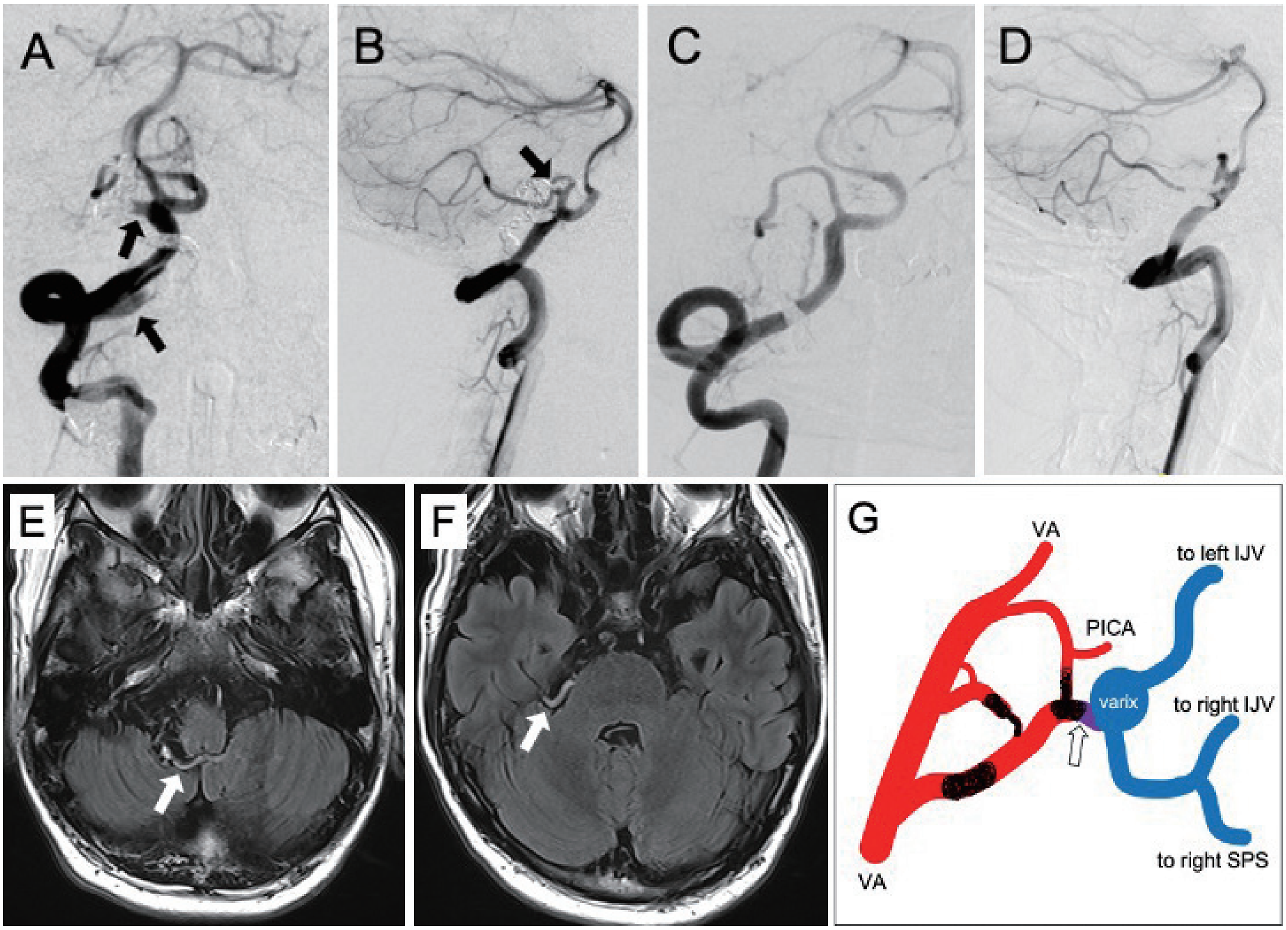

Fig. 2 Anterioposterior view (A) and lateral view (B) of the right vertebral angiogram immediately after coil embolization. The right vertebral angiogram 9 months after treatment (C and D) showed no residual sunt. Note that the origins of each feeder (arrows in A and B) are no longer shown in C and D. Fluid attenuated inversion recovery images 3 months after treatment (E and F) show obstructed varix and veins (arrows).
G: A simplified schematic drawing of the angioarchitecture. The open arrow indicates the fistula. The sites trapped by coils are also depicted. IJV, internal jugular vein; PICA, posterior inferior cerebellar artery; SPS, superior petrosal sinus; VA, vertebral artery

cal neurological deficit (12 cases, 33.3%), which might be caused by venous congestion or the mass effect of dilated vessels. Hemorrhage was observed in six patients, and only two patients had an asymptomatic lesion. Thirteen patients (36.1%) received treatment in their adulthood. Few lesions (9 cases, 25.0%) were fed by multiple arteries. A total of 29 lesions received endovascular treatment, of which 14 were occluded only with detachable coils and other 14 were occluded with liquid embolic agents. Most cases (33 cases, 91.7%) achieved a favorable outcome (Glasgow Outcome Scale 4 or 5).

Whether asymptomatic PAVF should be treated or not is controversial because a natural history of PAVF and risk of bleeding remains unclear because of its rarity. Yu et al. presented that a natural history of PAVF is unfavorable, especially in patients with multiple feeding arteries and high blood flow.[34)] Moreover, PAVF in adults can cause high-flow occlusive venopathy in a major sinus, resulting in the onset of symptoms.[35)] The present case also had deep venous drainage, which can cause cerebral venous congestion. Treatment for asymptomatic PAVF with the factors mentioned above might be reasonable.

The therapeutic options for PAVF include surgery, endovascular treatment, and their combination. For all options, the key procedure for radical cure is the interruption of feeders close to the fistula with the preservation of the venous drainage route.[28)] Total resection of the lesion is unnecessary. As a previous review of the literature revealed that surgical treatment offers a higher occlusion rate than that of endovascular treatment,[6,16)] surgery provides greater benefit for PAVF in areas of easy surgical access. By contrast, endovascular treatment is favorable when the lesion is deep-seated, surrounded by critical structures, or obscured by a dilated vein. For infratentorial complex PAVF

Table 1 Literature review of treated infratentorial pial arteriovenous fistula

Reference	Age	Sex	Presentation	Location	Feeder	Varix	Treatment	Complication	GOS
Vinuela *et al.*[10]	24	M	focal neurological sign	CMC	PICA	+	E, detachable balloon	transient lateral medullary syndrome	5
Vinuela *et al.*[11]	4	M	retardation, seizure, macrocephaly	CMC	PICA	+	S, clip	–	5
Smith *et al.*[12]	46	M	focal neurological sign	CPC	SCA	–	E, coil	–	4
Garcia-Monaco *et al.*[13]	11	F	headache, focal neurological sign	CMC	PICA	+	E, NBCA	–	5
Morimoto *et al.*[14]	38	M	focal neurological sign	CMC	VA	–	S, cauterization	–	5
Coubes *et al.*[15]	13	F	focal neurological sign	CMC	VA	+	E, NBCA	–	5
Hoh *et al.*[16]	57	M	hemorrhage	NA	VA	+	S, clip	transient dysphonia and dysphagia	5
	12	M	asymptomatic	NA	SCA	+	E, coil	–	5
Oya *et al.*[17]	60	M	hemorrhage	CPC	AICA	+	S, cauterization	transient facial nerve palsy	5
Yoshida *et al.*[18]	1	M	macrocephaly	PMC	BA	+	E, NBCA	–	1
	1	F	CHF, retardation, focal neurological sign	PMC	BA	+	E, NBCA	–	1
	6	M	headache, macrocephaly	CMC	PICA	+	E, NBCA	–	5
	3	F	headache, focal neurological sign	PMC	SCA	–	E, coil	–	5
Limaye *et al.*[19]	28	M	headache	PMC	SCA	+	E, NBCA	–	5
Passacantilli *et al.*[20]	54	F	hemorrhage	convexity	SCA, PICA	–	S, clip	surgical site infection	5
Izzo *et al.*[21]	0	F	macrocephaly	CMC	VA	+	E, coil	–	5
Masuoka *et al.*[22]	51	F	hydrocephalus	convexity	AICA	+	S, resection	–	5
Lv *et al.*[23]	9	F	headache	PMC	BA	+	E, coil	–	5
	17	M	focal neurological sign	CMC	PICA	+	E, coil	–	5
Guimaraens *et al.*[24]	2	M	focal neurological sign	CMC	PICA	+	E, coil	–	5
Newman *et al.*[25]	6	M	hydrocephalus, focal neurological sign	CPC	VA	+	E, Onyx	–	5
		M	hemorrhage	CPC	VA	NA	E, Onyx	–	4
Guerra *et al.*[26]	0	M	macrocephaly	CPC	VA	+	E, coil	–	5
Requejo *et al.*[27]	12	M	retardation	CMC	PICA	NA	E, coil	–	4
	8	F	headache	PMC	SCA	NA	E, coil, NBCA	–	5
	0	F	CHF	CMC	VA	–	E, coil	–	1
	10	M	retardation	CPC	VA	+	E, coil	–	4
Kanai *et al.*[28]	73	M	focal neurological sign	convexity	SCA	+	E+S, coil, resection	–	5
Lylyk *et al.*[29]	27	M	hydrocephalus	PMC	SCA	+	E, coil, NBCA	–	5
Ye *et al.*[30]	21	M	hemorrhage	NA	PICA	+	E, NBCA	–	5
Akamatsu *et al.*[31]	4	M	hemorrhage	convexity	SCA	+	E, NBCA	–	5
Wang *et al.*[32]	65	M	focal neurological sign	convexity	PICA	+	E, coil, Onyx	–	5
Yan *et al.*[33]	1	F	macrocephaly, hydrocephalus, retardation	convexity	SCA, PICA	+	E, coil, Onyx	subdural effusion	5
Jin *et al.*[4]	3	M	seizure	convexity	VA	+	E, coil	–	5
	8	M	headache	convexity	VA	+	E, coil	–	5
Present case	26	M	asymptomatic	CMC	VA, PICA	+	E, coil	–	5

AICA, anterior inferior cerebellar artery; BA, basilar artery; CHF, congestive heart failure; CMC, cerebellomedullary cistern; CPC, cerebellopontine cistern; E, endovascular; NA, not available; NBCA, n-butyl-2-cyanoacrylate; PICA, posterior inferior cerebellar artery; PMC, perimesencephalic cistern; S, surgical; SCA, superior cerebellar artery; VA, vertebral artery

Table 2 Summary of clinical characteristics in reported pial arteriovenous fistula

Presentation		
(One patient may have multiple symptoms)		
Focal neurological deficit		12
Headache		7
Symptom due to heamorrhage		6
Macrocephaly		6
Retardation		5
Hydrocephalus		4
Seizure		2
Asymptomatic		2
Location of the shunt		
Cerebellomedullary cistern		12
Cerebellar convexity		8
Perimesencephalic cistern		7
Cerebellopontine cistern		6
NA		3
Feeder		
(One patient may have multiple feeders)		
VA		12
PICA		12
SCA		10
BA		3
AICA		2
Patient with multiple feeders		9
Varix		
+		29
−		5
NA		2
Treatment		
Surgical		6
Endovascular	Coil only	14
	NBCA	10
	Onyx	4
	Balloon	1
Combined (Surgical+Endovascular)		1
Glasgow Outcome Scale		
5		29
4		4
1		3

AICA, anterior inferior cerebellar artery; BA, basilar artery; NA, not available; NBCA, n-butyl-2-cyanoacrylate; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery; VA, vertebral artery

like our case, the endovascular procedure is preferred to obliterate the shunt point precisely and safely. The currently mainstream embolic materials are liquid embolic agents.[23,34] They are useful particularly for PVAF with multiple feeders or fistulas as they can penetrate over the shunt point into the drainers or other feeders.[36] However, owing to the high-flow perfusion of PAVF, the distal migration of liquid agents is likely to occur.[5] Partial or total occlusion of the draining veins without reducing the arterial flow can lead to intra- or postoperative rupture.[36] Liquid embolic agents also have a risk of back-streaming to the parent vessel, especially when the feeding artery is a short branch of the parent artery. For these reasons, we chose detachable coils as embolic materials in the present case. Owing to the prior flow reduction by the occlusion of the proximal feeders, stable placement of the coils just proximal to the shunt point was achieved.

For the safe and complete interruption of a shunt, pretreatment selective angiography and three-dimensional rotational angiography are helpful in observing accurate angioarchitectural characteristics. Several reports in the cerebrovascular field showed the usefulness of slab maximum intensity projection (MIP) images derived from rotational angiography data.[37,38] This can be applied to understanding the angioarchitecture of PAVF. PAVF does not have nidus, and its affected vessels are often dilated. For these reasons, it is relatively easy in PAVF, compared with other shunt disorders, to chase and comprehend vascular connections by examining slab MIP images.

Conclusions

The present case suggests that transarterial coil embolization is an effective treatment modality for adult infratentorial PAVF. Understanding the detailed angioarchitecture is necessary for optimal treatment planning.

Patient Consent

The patient has consented to the submission of the case report to the journal.

Conflicts of Interest Disclosure

All authors declare no competing interests.

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