Moyamoya disease concurrent with dural arteriovenous fistula: A case report and literature review

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Abstract. Moyamoya disease (MMD) and dural arteriovenous fistula (DAVF) are two distinct types of intracranial lesion that share different pathogenic mechanisms. Under rare circumstances, patients with MMD have been reported to have concurrent DAVF. The present case study reports on a 47-year-old male admitted due to sudden headache. Head CT revealed hemorrhage of the right thalamus with ventricular extension. On CT angiography, the normal vasculature in the anterior and posterior circulation disappeared and was replaced by moyamoya-like vessels. The patient received conservative management and was discharged 3 days later. After three months, the patient was readmitted for acute cerebellar hemisphere infarction. Angiogram indicated that the DAVF (Cognard classification I) was supplied by the left middle meningeal artery, occipital artery and posterior meningeal artery and drained into the transverse-sigmoid sinus and occipital sinus. Conservative management of the DAVF was adopted. The patient was stable and lived independently during a 4-year follow-up. A literature review of the reported cases was also performed to further characterize this rare entity. The management of DAVF concurrent with MMD depends on its clinical presentation and invasiveness. For patients with symptoms or cortical venous drainage, endovascular intervention should be performed. For asymptomatic DAVFs or those without cerebral venous drainage, close follow-up is a reasonable option.

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Abbreviations: BAVM, brain arteriovenous malformation; DAVF, dural arteriovenous fistula; MMA, middle meningeal artery; MMD, moyamoya disease; MMS, moyamoya syndrome; OA, occipital artery

Introduction

Moyamoya disease (MMD) is an idiopathic chronic disease characterized by progressive steno-occlusive alteration of the internal carotid artery terminal and the beginning of the anterior cerebral artery and middle cerebral artery (1). Dural arteriovenous fistula (DAVF) is an uncommon vascular malformation that is characterized by abnormal connections between meningeal arteries and dural venous sinuses, meningeal veins or cortical veins (2).

MMD is currently considered a genetic disease (1). For DAVF, progressive stenosis or thrombosis of the dural venous sinus are thought to have a pivotal role in the genesis of DAVF (2). In general, there is no relationship between MMD and DAVF; however, in extremely rare circumstances, patients with MMD have been reported to have concurrent DAVF (3,4). In the present study, another case of MMD concurrent with DAVF was reported. In addition, a literature review of the reported cases was also performed to further expound this rare scenario.

Case report

A 47-year-old male was admitted was admitted to The First Hospital of Jilin University (Changchun, China) on Sep 27th 2015 due to sudden onset of headache. The patient was generally healthy and denied a history of hypertension, diabetes or any other chronic diseases. Physical examination was unremarkable except for mild neck stiffness. Head CT indicated hemorrhage of the right thalamus with ventricular extension (Fig. 1A and B). CT angiography (CTA) revealed that the normal vasculature in the anterior and posterior circulation disappeared and was replaced by moyamoya-like vessels (Fig. 1C and D). A diagnosis of hemorrhagic MMD was made. The patient received conservative management, including analgesic (flurbiprofen, 50 mg/bid), antemetic (tropisetron hydrochloride, 5 mg/bid) and fluid infusion (normal saline and 5% glucose solution) and was discharged 3 days later.

After three months, the patient was readmitted due to dizziness and gait disturbance. Diffusion-weighted MRI indicated acute infarction in the left cerebellar hemisphere (Fig. 2A) and encephalomalacia in the right thalamus (Fig. 2B). The cerebral blood volume (Fig. 2C and D) and cerebral blood flow (Fig. 2E and F) maps on perfusion-weighted MRI suggested relatively normal blood perfusion in the bilateral hemispheres.
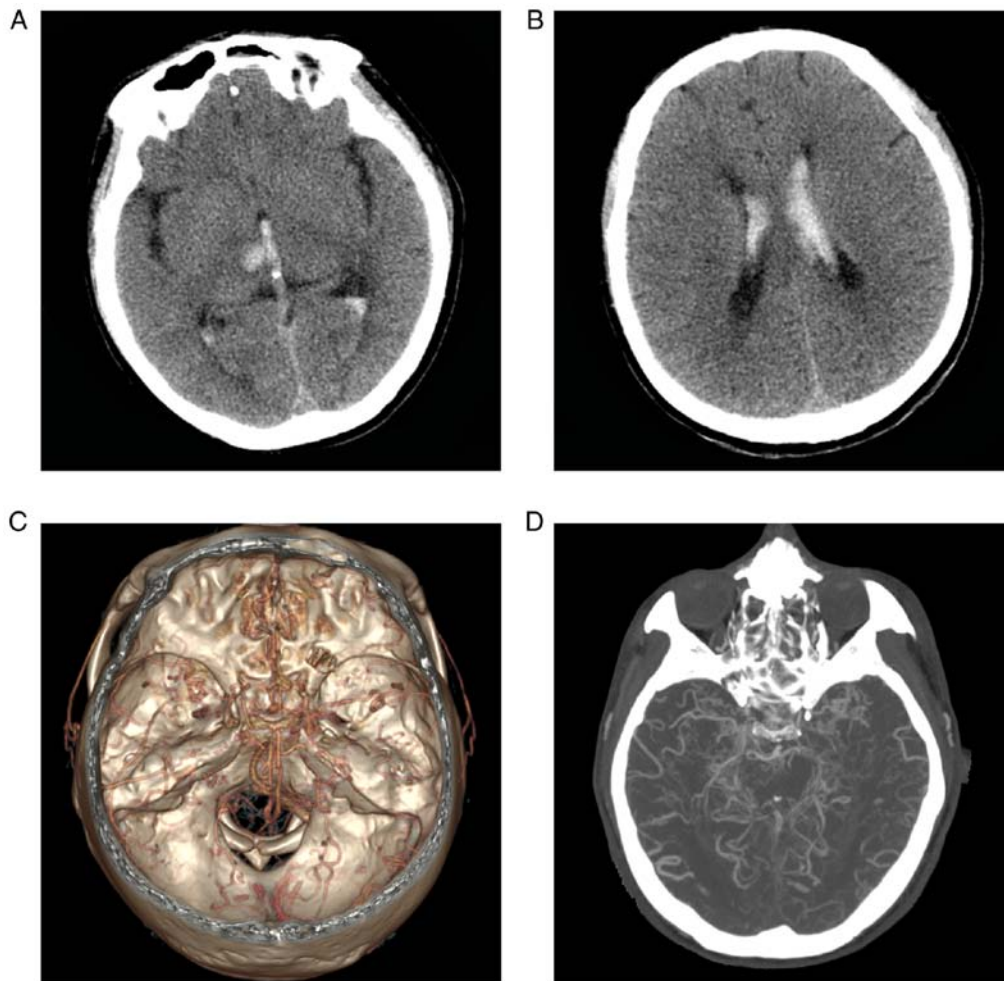
Figure 1. Head CT indicates (A) right thalamus hemorrhage with (B) ventricular extension. (C) Volume rendering and (D) maximum intensity projection of CT angiography reveals steno-occlusive alteration of the internal carotid artery terminus and posterior cerebral artery origin, and the normal vasculature in the anterior and posterior circulation is replaced by moyamoya-like vessels.

Conventional angiography of the internal carotid arteries and vertebral arteries (Fig. 3) confirmed the findings from previous CTA. An angiogram of the external carotid arteries indicated that the middle meningeal artery (MMA) and occipital artery (OA) had formed efficient collaterals with the brain vasculature (Fig. 4). A DAVF was also noted. The DAVF (Cognard classification I) was fed by the left MMA, OA and posterior meningeal artery (PMA). It drained into the transverse-sigmoid sinus and occipital sinus (Fig. 5).

As no retrograde blood flow or cortical venous drainage was noted, conservative management and follow-up were proposed for the DAVF. Oral aspirin (100 mg, QD) was prescribed. The patient was discharged 1 month later with no neurological deficit. He was stable and lived independently during a 4-year follow-up. However, the patient refused re-examination of the digital subtraction (DSA) and MR perfusion due to economic factors during the follow-up period. It was not possible to produce any further radiological evidence for the evaluation of the development of MMD and DAVF.

Discussion

MMD is an idiopathic steno-occlusive disease that mainly affects the anterior circulation. In a small proportion of patients, the posterior circulation may also be involved (1). As a consequence of an insufficient blood supply across the involved brain tissue, collateral vessels may arise from the cranial base perforators, which leads to the characteristic moyamoya-like vasculature in the cranial base (5).

According to previous studies, MMD may coexist with intracranial aneurysms, brain arteriovenous malformation (BAVM) and primitive carotid-basilar anastomosis (6). In extremely rare circumstances, transdural collaterals may anastomose directly with intracranial venous structures, leading to the formation of DAVFs (3,4). In a literature review of studies published in the English language, only 6 cases of AVF concurrent with MMD or moyamoya syndrome (MMS) were identified (Table I) (3,4,7-10). Among the 7 reported cases (including the case of the present study), 6 were DAVFs and 1 was a pial AVF. A total of 5 patients had concurrent MMD and 2 patients had concurrent MMS.

As a result of its rarity and the lack of research, the mechanisms underlying DAVF formation during MMD progression have remained elusive. It requires to be further investigated whether an association exists between DAVF and MMD. The specific location of a DAVF concurrent with MMD also remains to be studied. In clinical practice, progressive stenosis or occlusion of the dural venous sinus have a
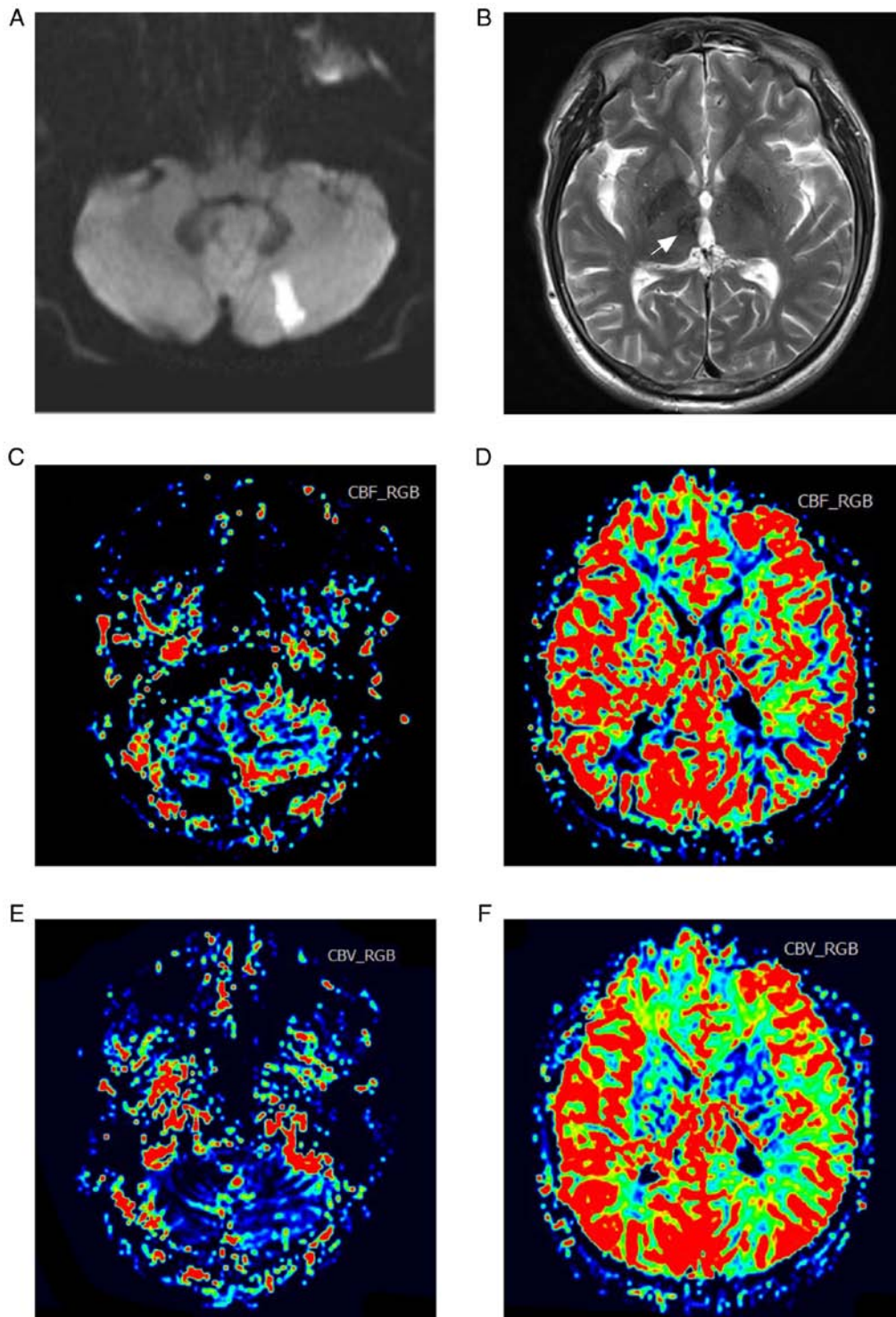
Figure 2. MRI with (A) diffusion-weighted imaging and (B) T2-weighted imaging indicates acute infarction of the left cerebellar hemisphere and old malacia in the right thalamus. The (C and D) cerebral blood volume and (E and F) cerebral blood flow maps on perfusion-weighted MRI indicated relatively normal blood perfusion in the bilateral hemispheres.

pivotal role in the formation of DAVF (2). Trauma, craniotomy, infection and venous sinus thrombosis are also responsible for a small proportion of DAVFs (11).

However, none of the previous case studies have reported thrombosis or stenosis of the venous system. A total of 3 patients developed AVF (1 case of pial AVF and 2 of DAVF) after extra-cranial-to-intracranial vascular bypass (3,4,8). No other risk factors were identified. However, concurrent AVF is an acquired disease rather than a congenital anomaly. This deduction is based on the following evidence. First, 4 of the reported AVFs were identified in a delayed fashion during the imaging follow-up of MMD or MMS (4,8,10). Furthermore, delayed development of BAVM has also been reported in patients with MMD or MMS (12,13). As the BAVM shares a similar vasculature with AVF, it may share a similar pathogenic mechanism with AVF in patients with MMD or MMS.
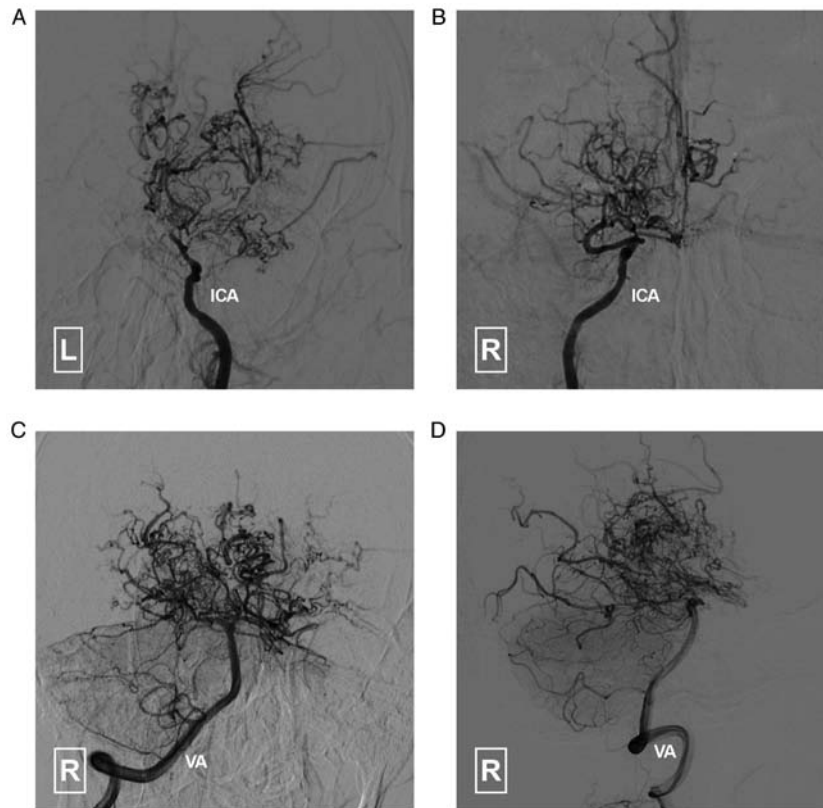
Figure 3. Angiogram of the bilateral ICAs (A and B) in AP view indicates steno-occlusive alteration of the ICA terminus. Angiogram of the right VA in (C) anteroposterior and (D) lateral views indicates steno-occlusive alteration of the proximal posterior cerebral artery. Extensive moyamoya-like vessels developed across the anterior and posterior circulation. ICA, internal carotid artery; VA, vertebral artery; L, left; R, right.
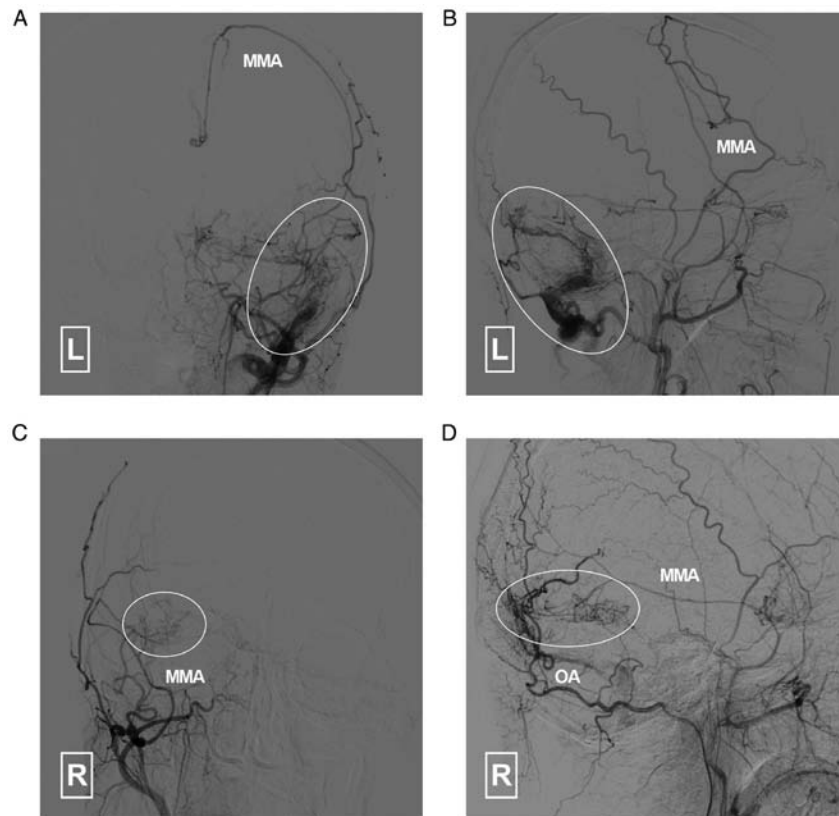

Figure 4. Angiogram of the left ECA in (A) AP and (B) lateral views indicate that the MMA has anastomosed with the pial artery at the midline. A dural arteriovenous fistula (white circle) is noted in the left occipital region. Angiogram of the right ECA in (C) AP and (D) lateral views indicates that the MMA and OA have anastomosed with the pial arteries (white circle). AP, anteroposterior; ECA, external carotid artery; MMA, middle meningeal artery; OA, occipital artery; L, left; R, right.
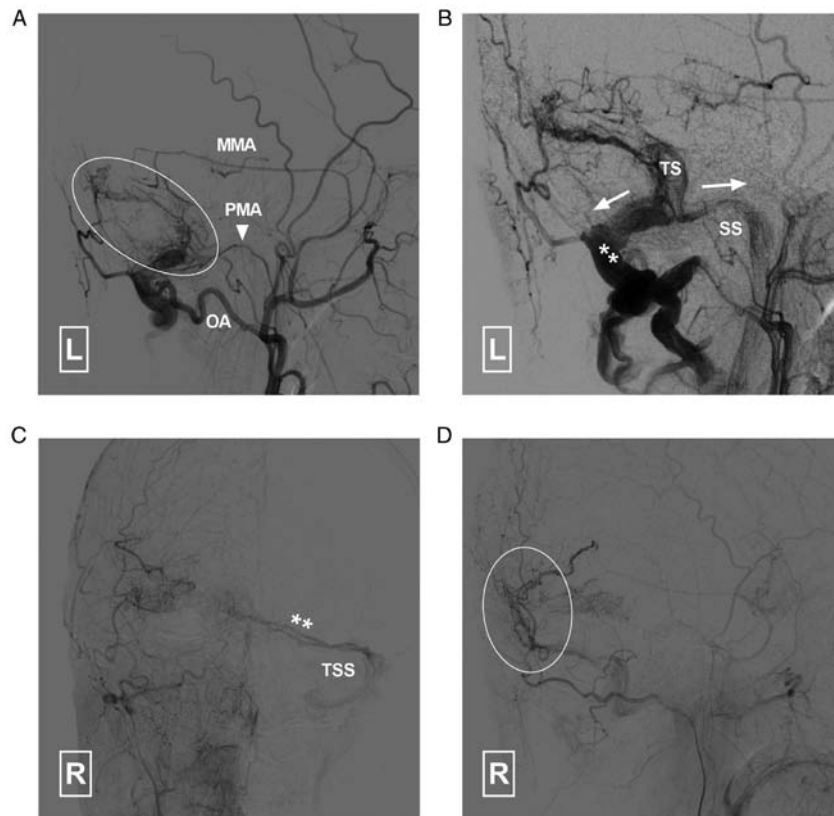
Figure 5. Angiogram of the left ECA in (A) arterial and (B) capillary phases indicates that the DAVF is supplied by the left MMA, PMA (arrowhead) and OA. It drained (white arrows) to the TS, SS and occipital sinus. Angiogram in (C) anteroposterior and (D) lateral views of the right ECA indicates that the DAVF (double asterisk and circle) is also supplied by the right ECA and drains to the left TSS. DAVF, dural arteriovenous fistula; ECA, external carotid artery; MMA, middle meningeal artery; PMA, posterior meningeal artery; SS, sigmoid sinus; TS, transverse sinus; TSS, transverse-sigmoid sinus; L, left; R, right.

According to the limited evidence, the ischemic environment and consequent angiogenesis have been suggested to have pivotal roles in the formation of AVFs. The levels of proangiogenic factors such as basic fibroblast growth factor and vascular endothelial growth factor are both elevated in the dura of patients with MMD or DAVF (3). For these patients, progressive arterial occlusion both in the anterior and posterior circulation may create a robust ischemic environment for collateral formation from the external carotid and vertebral arteries. A DAVF may form during this angiogenic process (14).

The management of MMD-associated AVF depends on its clinical presentation and invasiveness. Among the cases retrieved in the present literature review, 3 patients underwent successful transvenous and/or transarterial embolization of the AVFs; 2 of these patients were treated for persistent tinnitus or eye symptoms and 1 patient was treated for the presence of cortical venous drainage (3,4,9). A total of 4 patients were managed conservatively (7,8,10).

For the patient of the present study, a close follow-up strategy was also adopted. This conservative strategy was selected for the following reasons. First, the DAVF was incidentally detected and the patient was asymptomatic. The patient was first admitted to our hospital for thalamus hemorrhage and re-admitted for cerebellar infarction 3 months later. The hemorrhage and infarction have nothing to do with the DAVF. It may be proposed that both the hemorrhage and infarction resulted from the MMD, based on the following reasons:

i) This patient has no history of hypertension, which is the most common cause of cerebral hemorrhage in the thalamus or basal ganglion; ii) DAVF is a superficial cerebrovascular disease and cannot lead to deep parenchymal hemorrhage such as thalamus hemorrhage in this patient; iii) patients with MMD frequently develop slim and fragile vessels across the brain surface and paraventricular area and these fragile vessels and microaneurysms in the fragile vessels are prone to bleed (15); iv) catheter angiogram did not reveal any arteriovenous malformation or aneurysm around the hemorrhagic thalamus; v) the cerebellum is supplied by the three paired cerebellar arteries (superficial cerebellar artery, anterior inferior cerebellar artery and posterior inferior cerebellar artery). However, the DAVF in this patient was supplied by the MMA, OA and PMA. Hence, the DAVF was not responsible for the cerebellar infarction.

Furthermore, no cortical venous drainage was identified on angiogram. For patients with DAVF, cortical venous drainage is considered a risk factor of future hemorrhage. Those patients without cortical venous drainage would have a relatively benign natural course (2).

In addition, the perfusion-weighted MRI indicated relatively normal blood perfusion in the bilateral hemispheres, i.e. no severe blood insufficiency was noted across the bilateral hemispheres, which may have been compensated by the collaterals during the progression of the disease.

Finally, there were multiple feeders supplying the DAVF. Embolization of the feeders may pose a risk of compromising

Table I. Clinical data of the patients with MMD/MMS and concurrent DAVFs reported in the literature.

First author/year	Age/sex	Presenting symptom	Imaging findings on admission	Concurrent diseases	MMD or MMS	Type of AVF	Detection of MMD/MMS and AVF	Side of AVF	Supplying arteries of AVF	Draining veins of AVF	Cognard classification	Treatment for MMD/MMS	Treatment for AVF	Outcome (mRS)	(Refs.)
Killory/2008	44/M	Headache and tinnitus	IVH	NA/NM	MMD	DAVF	Simultaneously	R	OA, MMA, PAA, APhA	TS	I	STA-MCA bypass	TAE, TVE	0	(3)
Zaletel/2011	71/M	Urinary retention, cognitive decline	SAH	Hypertension	MMS	DAVF	Simultaneously	R	OA	TS	I	No	Conservative	4	(7)
Hanaoka/2011	45/F	Hemiparesis	CI	NA/NM	MMD	DAVF	Delayed	L	NA/NM	T-SS	IIB	STA-MCA bypass	TVE	NA/NM	(4)
Feroze/2015	51/F	Transient aphasia, progressive extremity weakness, headache	NA/NM	NA/NM	MMD	Pial AVF	Delayed	Bilateral	Branch of ICA	Vein of Trolard	NA/NM	STA-MCA bypass	Conservative	0	(8)
Liu/2016	52/F	Proptosis and chemosis of right eye, tinnitus	NA/NM	NA/NM	MMD	CDAVF	Simultaneously	R	MHT	SOV, IPS	NA/NM	No	TVE via direct cannulation of SOV	0	(9)
Koduri/2019	14/F	Tonic-clonic seizure, facial droop, hemibody weakness	CI	Down syndrome, hypothyroidism	MMS	DAVF	Delayed	Bilateral	Right MMA, Left MMA, OA, and muscular branches of VA	Left SS, Right SS	Left I, Right I	Bilateral pial synangiosis	Conservative	NA/NM	(10)
Present case	47/M	Dizziness and gait disturbance	Cerebellar infarction	No	MMD	DAVF	Delayed	Left	MMA, OA, PMA	Left T-SS, OS	I	No	Conservative	1	/

APhA, ascending pharyngeal artery; AVF, arteriovenous fistula; CDAVF, cavernous dural AVF; CI, cerebral infarction; F, female; ICA, internal carotid artery; IPS, inferior petrosal sinus; IVH, intraventricular hemorrhage; L, left; M, male; MCA, middle cerebral artery; MHT, meningohypophyseal trunk; MMA, middle meningeal artery; MMD, moyamoya disease; MMS, moyamoya syndrome; mRS, modified Rankin Scale; NA/NM, not applicable/not mentioned; OA, occipital artery; OS, occipital sinus; PAA, posterior auricular artery; R, right; SAH, subarachnoid hemorrhage; SOV, superior ophthalmic vein; SS, sigmoid sinus; STA, superficial temporal artery; TAE, transarterial embolization; TS, transverse sinus; T-SS, transverse-sigmoid sinus; TVE, transvenous embolization; VA, vertebral artery; PMA, posterior meningeal artery.

the transdural collaterals. Furthermore, evidence that bypass surgery is superior to medical therapy is not convincing in adult patients at present (16). Hence, for asymptomatic DAVFs or those without cerebral venous drainage, close follow-up is a reasonable option. However, for those patients concurrent with high-grade DAVFs, aggressive management through the endovascular route or open surgery is recommended to avoid future catastrophic intracranial hemorrhage. However, re-examination of the DSA and MR perfusion was refused by the patient of the present study during follow-up. The radiological progression of the MMD and DAVF remained undetermined for this patient, which limits the universality of the conservative treatment for this patient.

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Availability of data and materials

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

Authors' contributions

KH and YZ designed the study and drafted the manuscript. XC, KX and JY collected and analyzed of the clinical data. JY critically revised the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Ethics approval is not required for case reports at our institution. Informed consent for participation in the study or use of the medical data was obtained from the patient.

Patient consent for publication

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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