

Hybrid surgery for an arteriovenous malformation fed by an accessory middle cerebral artery and drained by a developmental venous anomaly: A case report and literature review

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Abstract. An accessory middle cerebral artery (AMCA), which mainly acts in the collateral circulation of the middle cerebral artery (MCA), is a rare anatomic malformation. Similar to other intracranial vessels, cerebrovascular disease can occur in the AMCA. However, the development of an arteriovenous malformation (AVM) in the AMCA is very rare, especially in conjunction with developmental venous anomalies (DVAs). Here, a rare case of an AMCA combined with an AVM and a DVA was reported. The patient was a 47-year-old female with intracranial hemorrhage at symptom onset. CT and MRI showed lesions in the left Sylvian fissure and insula accompanied by hemorrhage. DSA suggested a left AMCA; an AVM of the AMCA was located in the deep Sylvian fissure. The AVM was diffusely developed and drained into the DVA. The operation was performed in a hybrid operating room. The major feeding artery of the AVM, which was derived from the AMCA, was clipped, then the AVM and DVA were subsequently removed. Intraoperative DSA showed that the AVM and DVA were radically removed. A pathological examination confirmed the presence of an AVM. The patient recovered well and was discharged. Therefore, as highlighted in this case report, rare AVMs can be found in AMCAs and can even occur simultaneously with a DVA. Hybrid surgical treatment can be used to remove AVMs and can lead to an improved prognosis.

Introduction

An accessory middle cerebral artery (AMCA) is a relatively uncommon anomaly of the middle cerebral artery (MCA) in which the AMCA arises from the anterior cerebral artery (ACA) that coexists with the main trunk of the MCA (1,2). The angiographic incidence of AMCA is 0.32% (3). Anatomically,

an AMCA mainly acts as collateral circulation for the MCA. Similar to other intracranial vessels, the AMCA is involved in many diseases, such as aneurysms and cerebral infarction (4-6). Although arteriovenous malformations (AVMs) can theoretically occur in an AMCA, this finding is extremely rare (7). Most AVMs drain through the veins of the brain, and the drainage vein has, in some patients, been found to be enlarged or stenosed or to exhibit tortuosity (8). However, AVMs accompanied by developmental venous anomalies (DVAs) are rare, and only approximately 20 relevant articles have been published in the literature to date (7). A DVA is a benign and mostly silent condition without clinical symptoms (9). However, when a DVA merges with an AVM, due to the high blood flow in the AVM, drainage of blood from the DVA is difficult, resulting in drainage resistance. This feature creates complications within the AVM and makes DVAs prone to bleeding (10). Cases in which an AMCA was accompanied by an AVM and a DVA are rarely reported. However, the treatment of one such case is reported in this paper, and the relevant literature is reviewed.

Case report

A 47-year-old woman was admitted due to a headache accompanied by nausea and vomiting for 1 h. The patient had always been healthy without any history of hypertension or diabetes. This disease was not induced by any other condition. The patient developed a sudden and severe headache at rest and subsequently developed nausea and vomiting accompanied by the inability to use her right limb and dysphasia. The patient was admitted to our hospital after symptom onset. At the time of presentation, the patient was alert with no seizure. An admission examination showed the following. The patient was alert but exhibited incomplete motor aphasia. The left limbs moved freely, while the right limbs were reflexive (muscle strength grade III). The Babinski sign was positive in the right lower limb. A head CT and MRI after admission showed lesions in the left Sylvian fissure and insular lobe. CT showed high and slightly high mixed densities. The signals were confounding on MRI. An abnormally thickened vein extended from the brain surface to the deep lesions (Fig. 1). Further DSA revealed a vascular malformation in the left MCA and an AMCA that originated from the anterior communicating artery (Fig. 2A and B). The vascular malformation in the

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AMCA was seen in the deep Sylvian fissure (Fig. 2C and D). The vascular malformation was diffuse, and a small number of fine branching arteries served as its blood supply. It drained into the DVA (Fig. 3). The patient was diagnosed with a left frontal hemorrhage with an AMCA that merged with an AVM and a DVA. AVM resection was scheduled to be performed in a hybrid operating room.

Surgical treatment was performed in a hybrid operating room. Femoral artery puncture was performed, and an arterial sheath was inserted. A left expanded pterional approach was adopted during surgery. The AMCA originating from the anterior communicating artery could be seen at the chiasmatic cistern after the Sylvian fissure on the left side was opened (Fig. 4A). The AVM was located on the surface of the insular lobe of the deep Sylvian fissure. The major feeding artery of the AVM, which originated from the AMCA, was clipped. The AVM and the DVA were subsequently removed from the AVM, which resulted in obsolete hemorrhage in the AVM. The AVM was diffuse, and the venous end drained into the DVA (Fig. 4B). The final pathological examination after removal showed an irregular vascular shape that consisted of various types of expanded and transparent veins and abnormal muscularized arteries. An abnormal arteriovenous anastomosis was also observed along with brain tissues between vessels and was diagnosed as an AVM (Fig. 4C and D). DSA was performed after AVM resection, which showed that the AVM and DVA were completely removed (Fig. 5). The patient recovered well within one week of the surgery. A one-month follow-up CT scan showed good recovery in the brain tissue (Fig. 6). The patient was in good condition, had grade IV muscle strength in the right limb, and could take care of herself.

Discussion

In 1973, Teal *et al* (11) proposed using the term 'AMCA' to describe an anomalous artery arising from the ACA. The origin of an AMCA can be the proximal or distal ACA, and the vessel that arises from the ACA then runs in the Sylvian fissure along with the main trunk of the MCA to supply part of the MCA territory (11,12). The angiographic incidence of AMCA is 0.32% (3). Manelfe *et al* (13) classified AMCA into three types: Type 1 is an anomalous vessel that arises from the internal carotid artery at a point proximal to its bifurcation (the duplicated MCA in Teal's classification), type 2 originates from the proximal portion of the ACA, and type 3 originates from the distal portion of the first segment of the ACA (A1) near the anterior communicating artery (14). The AMCA in this patient originated from the anterior communicating artery and was classified as Manelfe type 3. Typically, an AMCA serves as the collateral blood supply to the MCA territory. However, the clinical significance of AMCAs remains obscure (15).

Similar to normal MCAs, AMCAs can also be associated with many diseases, such as intracranial aneurysms (4), cerebral infarction (5), and moyamoya disease (6). However, an AMCA combined with an AVM is very rare, especially with the vein of the AVM draining into the DVA, as in this case. Due to differences in the development of the AMCA, such arteries can vary in diameter (16), and there are large differences in the feeding area distribution of its arteries (17).

For example, when the blood supply to the AMCA is extensive, the consequences are more severe if the AMCA undergoes an acute infarction (5,18). Moreover, when the AMCA is large with abundant blood flow, the artery is also prone to aneurysms (19,20). Therefore, the development of an AMCA may be associated with diseases. Although an AVM combined with an AMCA is relatively rare, we report such a case in this article.

AVMs are a type of congenital cerebrovascular condition which, along with capillary telangiectasia, cavernous malformation, and DVA, represent the 4 most commonly recognized subgroups of cerebrovascular malformations (21). An AVM is a collection of arteries and veins that form without an intervening capillary bed (22). However, a DVA is usually a collection of radiating veins that converge on a large and centrally located draining vein (23). This is classically described as having a fan-shaped 'caput medusae' appearance, according to a large prospective autopsy series, DVA is the most common type of vascular malformation in the brain. It is now accepted that DVA generally follow a benign clinical course (24).

Therefore, during embryonic development, an AVM and a DVA may occur on an AMCA, as is the case in the patient described in this paper. It is possible that the AVM formed first, and then venous hypertension around the draining vein occurred, and the disturbed normal venous drainage due to venous hypertension caused by preexisting arteriovenous shunting may have promoted the development of a DVA (25). The first patient with a coexisting AVM and DVA was reported by Huang *et al* (26) in 1984, and a literature review of 22 cases was published by Zhang *et al* (7) in 2017. However, no reports have described an AVM accompanied by both a DVA and an AMCA.

When an AVM is accompanied by a DVA, the risk of hemorrhage is as high as 68%, and these patients are more prone to bleeding than are those with a single AVM (38%) (7). This finding may be related to the tenuous and slow-adapting terminal angioarchitecture that could lead to suboptimal venous drainage in the DVA (27,28). Therefore, patients with both an AVM and a DVA must be treated when rupture and bleeding occur. Microsurgical resection is an effective method for treating a ruptured AVM, especially when the AVM is shallow and is located in a non-relocating area (29). Whether the DVA needs to be removed during AVM resection must be determined. Despite the risk imposed by the aberrant drainage, successful neurosurgical management of these transitional lesions should target the AVM and retain a tissue-dependent DVA with low hemorrhagic risk. This prevents the DVA from draining normal brain tissue after removal and prevents catastrophic venous infarction (7).

The patient described in our paper had a diffusely developed AVM with unclear boundaries. We found bulky veins in the DVA, and the AVM around the DVA was gradually removed. Intraoperative DSA showed that the AVM was completely removed, and the DVA was not visible. We adopted a hybrid operation procedure for this patient because it ensured the complete clearance of the AVM. Intraoperative DSA can also be performed to accurately determine the position of the AVM (30,31). No serious complications occurred after the operation, and the patient's physical activity gradually recovered. However, a one-month postoperative follow-up CT

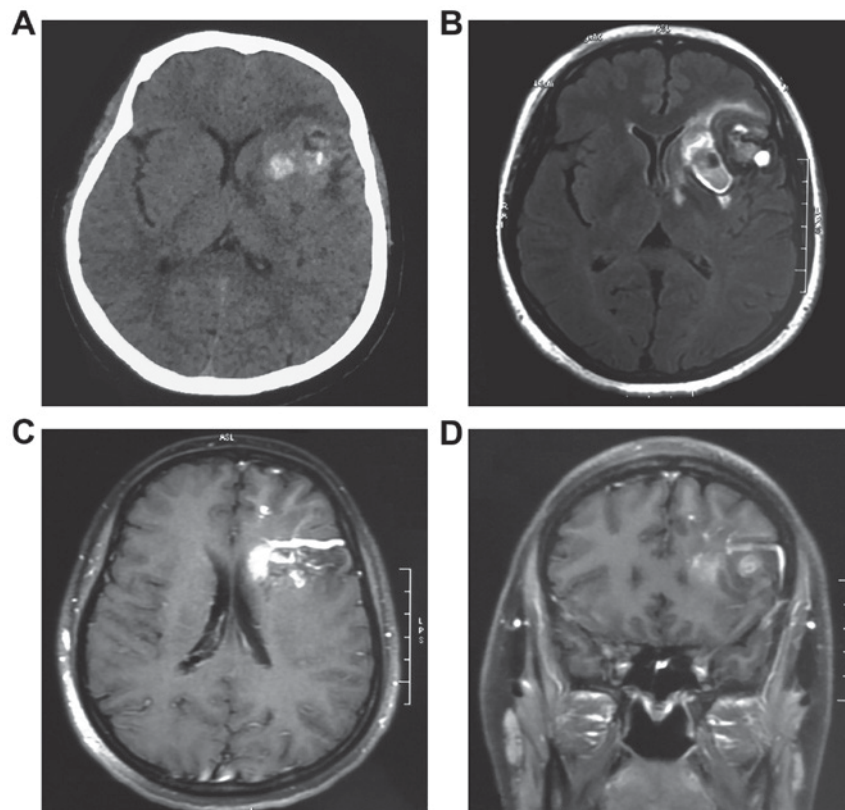


Figure 1. Preoperative CT and MRI images. (A) CT scan showing lesions in the deep left Sylvian fissure and the insular lobe with uneven, high and slightly high mixed density. (B) A flair MR image showing a confounding signal located in the deep Sylvian fissure that involved the insular lobe and part of the island surface. (C) Axial and (D) coronal MRI-enhanced T1 WI images showing an abnormally thick vein running from the surface of the brain to the deep lesion.

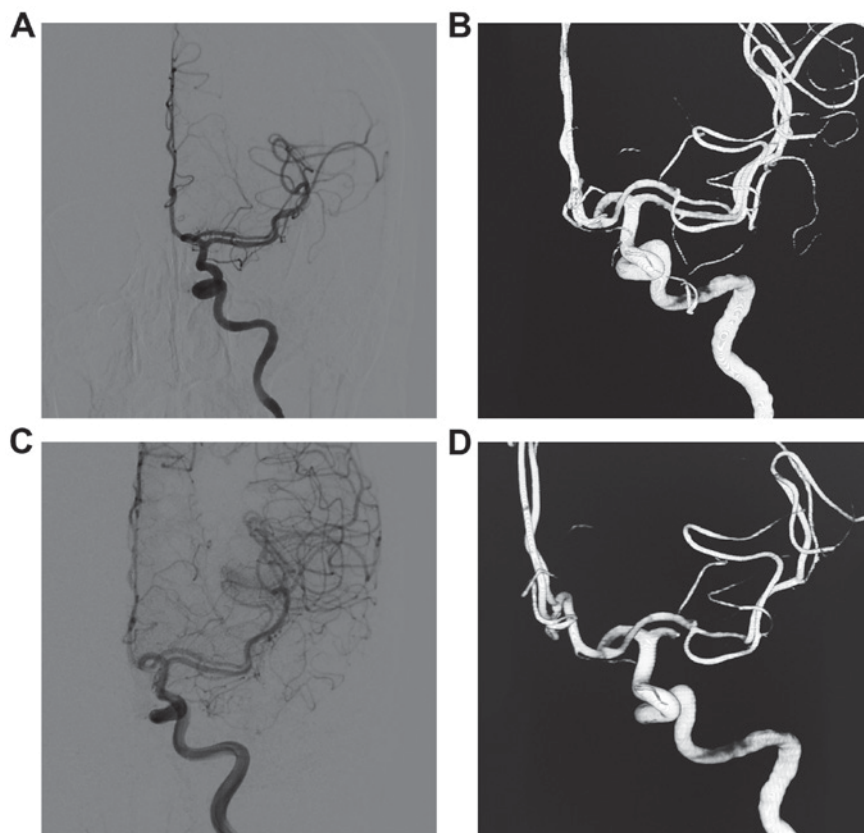


Figure 2. Preoperative DSA image. (A) 2D- and (B) 3D-reconstructed DSA images showing a vascular malformation in the middle cerebral artery along with an accessory middle cerebral artery that originated from the anterior communicating artery. (C) Vascular variations in the accessory middle cerebral artery. (D) The normal MCA was cut to locate the AMCA vascular malformation. MCA, middle cerebral artery; AMCA, accessory middle cerebral artery.

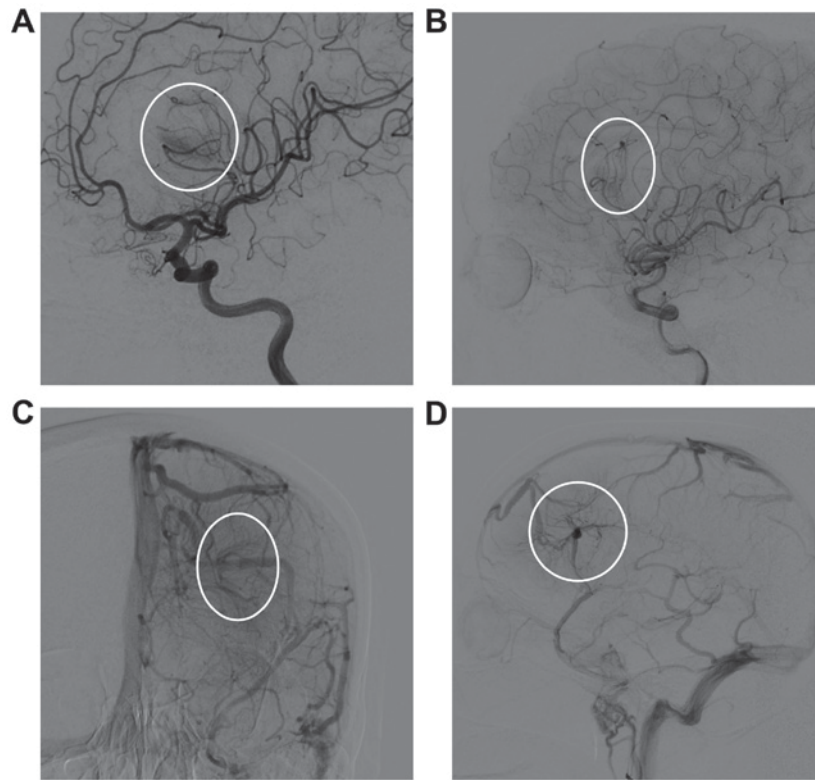


Figure 3. Details of the vascular malformation. (A and B) DSA in the arterial phase showing that the vascular malformation originated from the insular lobe of the accessory middle cerebral artery. The vascular malformation was diffuse. The artery serving as the blood supply was very small (see circle) and flowed into the DVA. (C and D) The DVA in the venous phase exhibited a DVA caput medusae (see circle). The vascular malformation accorded with the imaging for the AVM. DVAs, developmental venous anomalies; AVM, arteriovenous malformation.

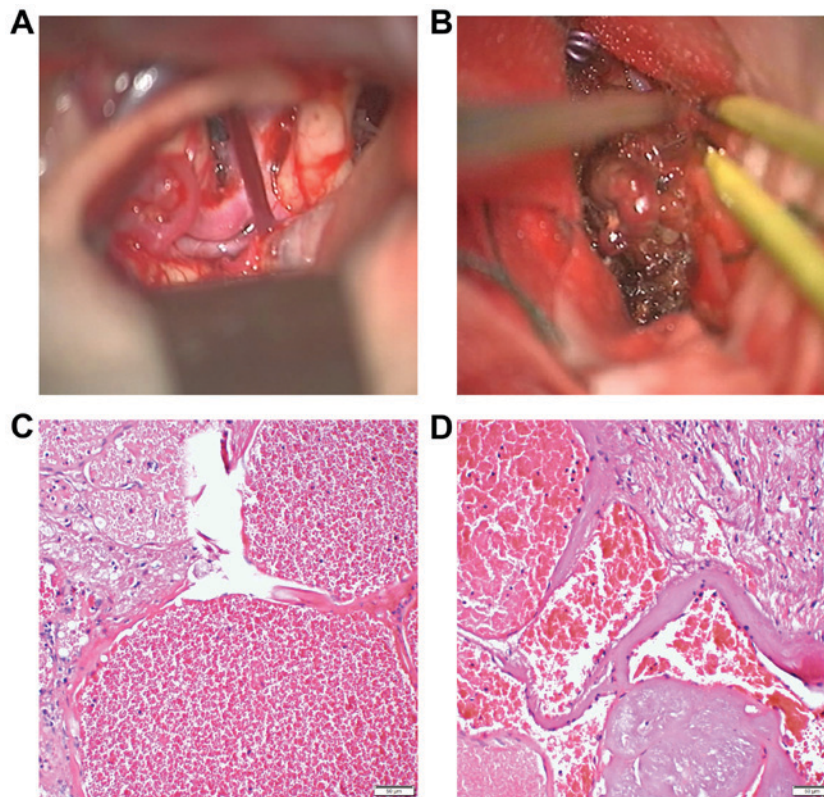


Figure 4. Intraoperative imaging and pathological images. (A) The AMCA originating from the anterior communicating artery could be seen at the left of the chiasmatic cistern. (B) The AVM was located on the deep Sylvian fissure. (C and D) H&E staining (magnification, x200) showed blood vessels with an irregular shape and size. Lesions consisting of various types of expanded and transparent veins and abnormal muscularized arteries were visible. Vascular structures of single fiber tissue, which are difficult to distinguish, as well as abnormal arteriovenous anastomosis, were visible. The brain tissue was visible among the vessels. AMCA, accessory middle cerebral artery; AVM, arteriovenous malformation.

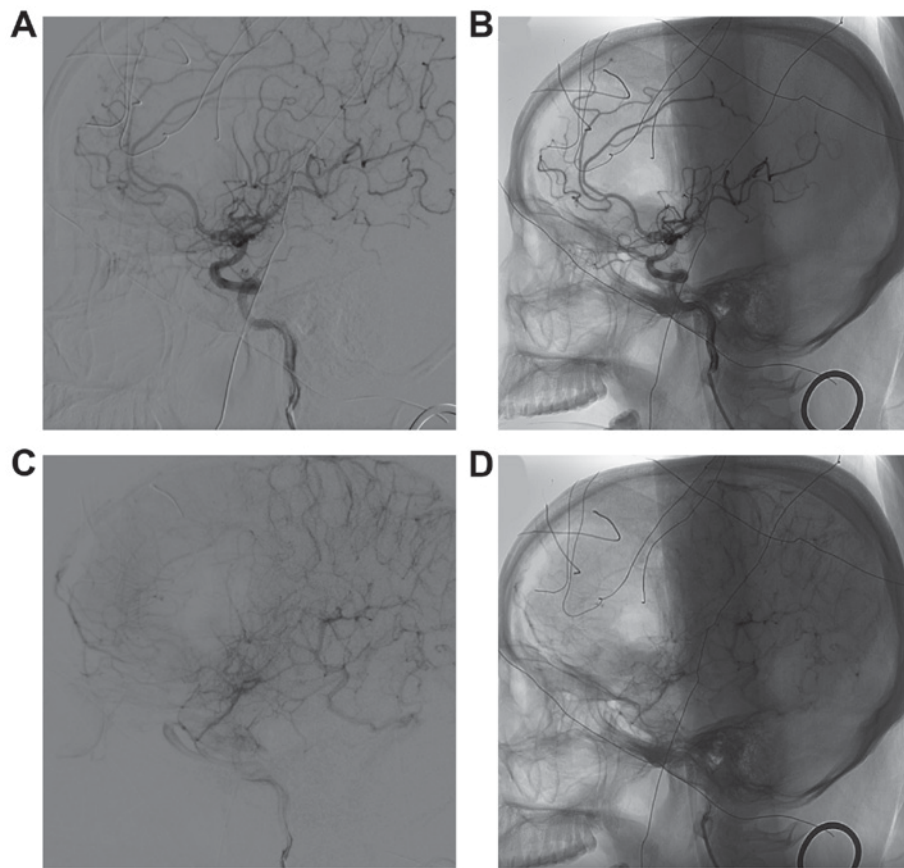


Figure 5. Postoperative DSA images. Postoperative DSA showing complete removal of the AVM. The AVM and drainage veins were not visible. (A) Early artery phase. (B) Early artery phase with mask image. (C) Venous phase. (D) Venous phase with mask image. AVM, arteriovenous malformation.

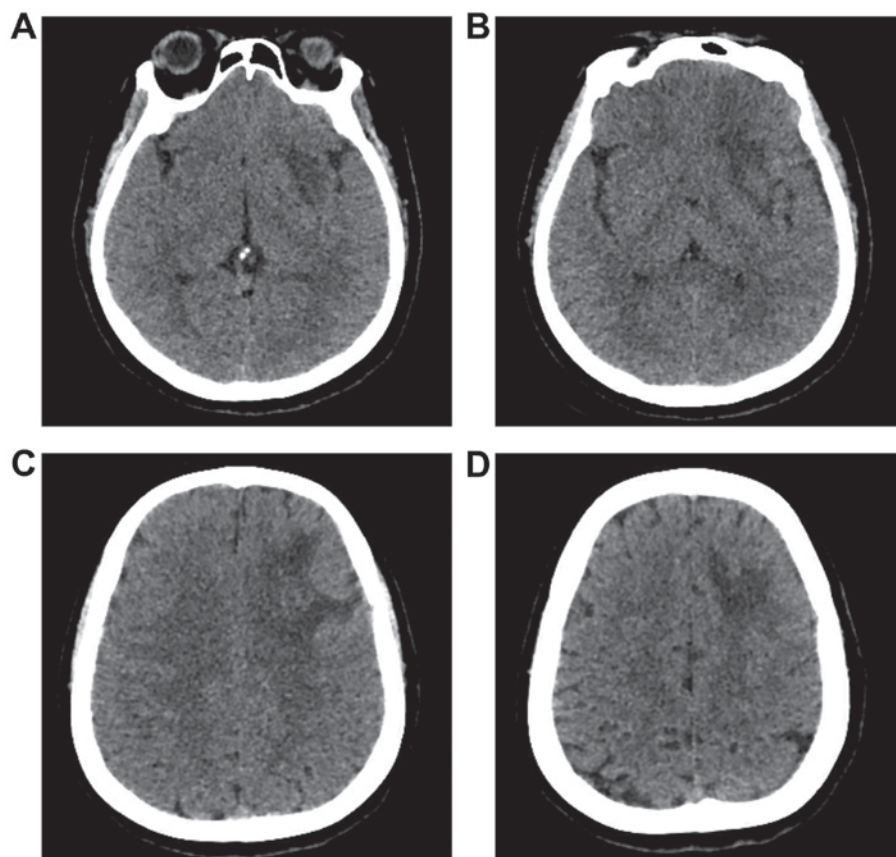


Figure 6. CT images at 6 months postoperative. (A-D) Postoperative CT images showing that the brain tissue had recovered, with mild edema.

scan showed that cerebral edema persisted, and this may have been related to the removal of the DVA during the operation.

In addition to microsurgical resection of AVMs and DVAs, radiosurgery, embolization and combination therapy are effective treatments (7). In 2017, Zhang *et al* (7) reviewed 22 cases of coexisting AVMs and DVAs. Of these, stereotactic radiosurgery was performed in 7 cases, embolization was performed in 6 cases, surgical resection was performed in 4 cases, and multimodal therapy was performed in 5 cases. AVMs were treated by targeted embolization, and the DVAs were carefully preserved (32). When an AVM is accompanied by a DVA and no evidence of bleeding is present, radiotherapy is also an effective treatment. Aksoy *et al* (10) performed radiosurgery in a patient with both an AVM and a DVA and reported a good patient outcome at the 6-month follow-up evaluation. However, as the effect of radiation is generally delayed until 2–4 years, whether the patient remained well at a longer-term follow-up is unclear (10). Nevertheless, it is important to note that only the AVM should be obliterated by radiosurgery, whereas the DVA should be completely preserved (9).

Therefore, as reported in this rare case, AVMs can be found in an AMCA and can also be simultaneously observed with a DVA. Hybrid surgical treatment can be used to remove AVMs, and this approach can improve the patient's prognosis.

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Availability of data and materials

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

Authors' contributions

KL analysed the patient data and wrote the initial draft. YG, LQ and BX collected the images of the patients and analyzed the literature. JY and KX were the surgeons who treated the patient. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Ethics Committee of the First Hospital of Jilin University and written informed consent form was obtained from the patients. All procedures performed were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Patient consent for publication

Not applicable.

Competing interests

The authors have declared that they have no competing interests.

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