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The Management of Symptomatic Cerebral Developmental Venous Anomalies: A Clinical Experience of 43 Cases

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Background: Developmental venous anomalies (DVAs) are rare vascular diseases becoming more frequently diagnosed. Most patients with DVAs have no clinical symptoms with the exception of a few patients with epilepsy, intracranial hemorrhage, or neuro-function deficit. There is still controversy with respect to treatment strategies for symptomatic DVAs.

Material/Methods: Forty-three cases of symptomatic DVAs from January 2006 to October 2015 were retrospectively reviewed and the imaging characteristics of DVAs by CT, MRI, and DSA and the treatment modalities for DVAs were studied.





Results: Typical imaging characteristics of symptomatic DVAs were wedge or umbrella-shaped collections of dilated medullary veins converging in an enlarged subependymal or transcortical collecting vein, draining to the superficial or deep vein system. Based on location and draining vein features, symptomatic DVAs were tentatively classified into six different subtypes. Of the 43 cases, 19 were treated by surgical methods and 24 were treated conservatively.

Conclusions: We concluded that the rate of accompanying abnormalities in cases of symptomatic DVAs was high. Intracerebral hemorrhage was usually attributed to associated CMs or AVMs. The associated lesions and the branches responsible for bleeding could be resected while preserving the collecting vein as far as possible.

MeSH Keywords: **Central Nervous System Venous Angioma • Intracranial Hemorrhages • Neurosurgery**

Abbreviations: **AVM** – arteriovenous malformation; **CM** – cavernous malformation; **CT** – computed tomography; **DSA** – digital subtraction angiography; **DVA** – developmental venous anomaly; **MRI** – magnetic resonance imaging

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Background

McCormick [1] classified cerebral vascular malformations into venous angiomas, arteriovenous malformations (AVMs), capillary telangiectasia, and cavernous malformations (CMs). In 1986, Lasjaunias et al. [2] first used the term “developmental venous anomaly (DVA)” to depict venous angiomas. They believed there existed anastomotic channels, so-called medullary veins, between the deep and superficial venous systems. Superficial and deep venous systems reach equilibrium through these channels. When one system is occluded due to venous anomaly, tumor compression, intravenous thrombosis, or arterial-venous shunt, the medullary veins open and dilate to compensate the blood draining to the alternative system. Studies suggest that most patients with DVAs have no clinical symptoms except for a few patients with epilepsy, intracranial hemorrhage, or neurofunction deficit [3–7]. DVAs were once thought to be rare lesions, however, with the advance of imaging techniques, especially the widespread use of magnetic resonance imaging (MRI), the detection rate for DVAs has increased dramatically [8,9]. DVAs have a benign natural course and good prognosis [4,6,10,11]. However, there is still controversy with respect to treatment strategies for symptomatic DVAs. In the present study, 43 cases with symptomatic DVAs were included, from January 2006 to October 2015, and the clinical characteristics and surgical interventions were studied.

Material and Methods

Patient population

The present study included 43 consecutive patients from January 2006 to October 2015 with symptomatic DVAs in our Department of Neurosurgery. The diagnosis of DVA was confirmed by radiological imaging and/or histological characteristics. The patient medical records, radiological imaging, and videos of surgical procedures were retrospectively reviewed. Institutional review board approval was obtained at Chinese PLA General Hospital for this retrospective review of the data, and informed consents were signed by all patients.

Patient evaluation

All patients underwent radiological assessment of their lesions by computer tomography (CT) and/or MRI and/or digital subtraction angiography (DSA). DVA localizations and accompanying abnormalities were recorded. All patients received follow-up neuroimaging (MRI or DSA) and neurological examinations. The outcomes of patients who underwent surgery were assessed according to the modified Rankin Scale (mRS) scores at the time of final follow-up.

Results

Patient and lesion characteristics

The patient characteristics and treatment details are summarized in Table 1. Of the 43 patients, 19 (44%) were female and 24 (56%) were male; the mean age was 35 years (range 2 to 58 years). Eleven patients presented with seizures, 16 with intracranial hemorrhages, 16 with headaches, and 4 with facial hemangiomas.

CT scans were performed for all patients, MRI for 43 patients, and DSA for 34 patients. Two patients had associated AVMs, 13 had CMs, and one patient had cortical dysplasia. The DVAs were located in the cerebral hemisphere in 27 patients, in the cerebellar hemisphere in six patients, and in the brain stem in six patients. Non-contrast CT scans revealed brain stem hemorrhages in six patients, parenchymal hematomas in 9 patients, and intraventricular hemorrhages in one patient. Contrast-enhanced CT scans showed numerous linear or dot-like enhancing foci converging in a single enlarged tubular draining vein. Stellate configuration around an emanating transcortical vein was a typical image characteristic of DVAs in MRI (Figure 1A, 1B). Angiographic findings included normal circulation time and normal arterial and capillary phases. The pathognomonic angiographic appearance was visualized on the late venous phase and consisted of wedge or umbrella-shaped collections of dilated medullary veins converging in an enlarged subependymal or transcortical collecting vein (Figure 1C, 1D).

Classification of symptomatic DVAs

Based on location and draining vein features, we tentatively classified symptomatic DVAs into six different subtypes (Table 2, Figure 1): Type A DVAs were located in the cerebral hemisphere and drained into the superficial cortical vein or dura sinus (12 cases, Figure 1A–1D); Type B DVAs were located in the cerebral hemisphere and drained into cerebral internal veins (7 cases, Figure 1E–1G); Type C DVAs were located in the cerebral hemisphere and drained into subependymal vein (8 cases, Figure 1H–1I); Type D DVAs were located in the cerebellum (6 cases, Figure 1K); Type E DVAs were located in the brain stem (6 cases, Figure 1L, 1M); Type F DVAs were subcutaneous sinus pericranii (4 cases, Figure 1N–1P).

Treatment strategies

Thirteen patients with associated CMs, two with associated AVMs, and two with pure DVAs and intracranial hematomas were resected with preservation of collecting veins of DVAs. In our initial surgical experience, one draining vein of DVA with associated CM was occluded by accident, which caused significant intraoperative brain swelling, requiring performance of

Table 1. Characteristics of patients with symptomatic DVAs.

Characteristics	n=43	
Demographics		
Age (yr) (mean)	34.6±10.8	
Sex male/female	24 (56%)/19 (44%)	
Major symptoms and signs		
Seizure	11	(26%)
Headache	12	(28%)
Hemorrhage	16	(37%)
Facial haemangioma	4	(9%)
Accompanying abnormality		
CM	13	(30%)
AVM	2	(5%)
Cortical dysplasia	1	(2%)
DVA localizations		
Cerebral	27	(63%)
Cerebellar	6	(14%)
Brain stem	6	(14%)
Localizations of DVA caputs		
Deep	13	(30%)
Subcortical	10	(23%)
Juxtacortical	15	(35%)
Mixed	5	(12%)
Subtypes		
Type A	12	(28%)
Type B	7	(16%)
Type C	8	(19%)
Type D	6	(14%)
Type E	6	(14%)
Type F	4	(9%)
Treatment strategies		
Open surgery	19	(44%)
Antiepileptic medicine treatment	10	(23%)
Observed and followed up	14	(33%)

AVM – arteriovenous malformation; CM – cavernous malformation; DVA – developmental venous anomalies; yr – year.

extended frontal lobe decompression. The mRS score was 2 at last follow-up (32 months postoperatively). In all other cases of open surgery, the branches responsible for bleeding were resected while the main collecting veins and normal branches were kept intact. With the paramount tenet of protecting and preserving collecting veins, all the later cases had good outcomes, and bleeding did not occur. The mRS score at last follow-up (mean, 24 months,) was 0 or 1.

Of the 11 patients with epilepsy, one patient with cortical dysplasia and seizure underwent frontal lobe resection (Figure 1E–1G) while the remaining patients with seizures were treated conservatively by antiepileptic medicine with seizure-free control in six patients and resolution of seizure attack in four patients.

One case with ventricular hemorrhage underwent ventriculopuncture and was drained until the clot disappeared (Figure 1H, 1I).

The other 14 patients were followed with a mean follow-up period of 13 months (range, 6 to 34 months). The patients remained neurologically intact.

Illustrative case

A 14-year-old boy presented with headache after a head injury. Preoperative CT and MRI showed hemorrhage in the left cerebellar hemisphere, and angiogram during the late venous phase showed venous branches converged and drained into the sigmoid sinus (Type D). Intraoperatively, the blood clot was evacuated, and the distal radicles of the DVAs, which were responsible for bleeding, were coagulated. No sign of venous thrombosis of the main collecting veins was seen, and these collecting veins were preserved. The MRI and DSA examinations showed draining veins were preserved. (Figure 2) Histological examination of the hematoma and the neighboring parenchyma specimens confirmed that no other vascular malformation (including cavernoma) was found. Postoperative, the patient had a good recovery.

Discussion

The imaging characteristics of symptomatic DVAs

DVAs cannot be detected by non-contrast CT scan. Parenchymal changes or calcifications may be the only detectable abnormalities. Numerous linear or dot-like enhancing foci converging in a single enlarged, tubular draining vein may be seen in enhanced CT scans [4–7]. MRI has been studied as an imaging diagnostic method for DVAs [9]. The typical imaging characteristics in MRIs are stellate configuration around an emanating

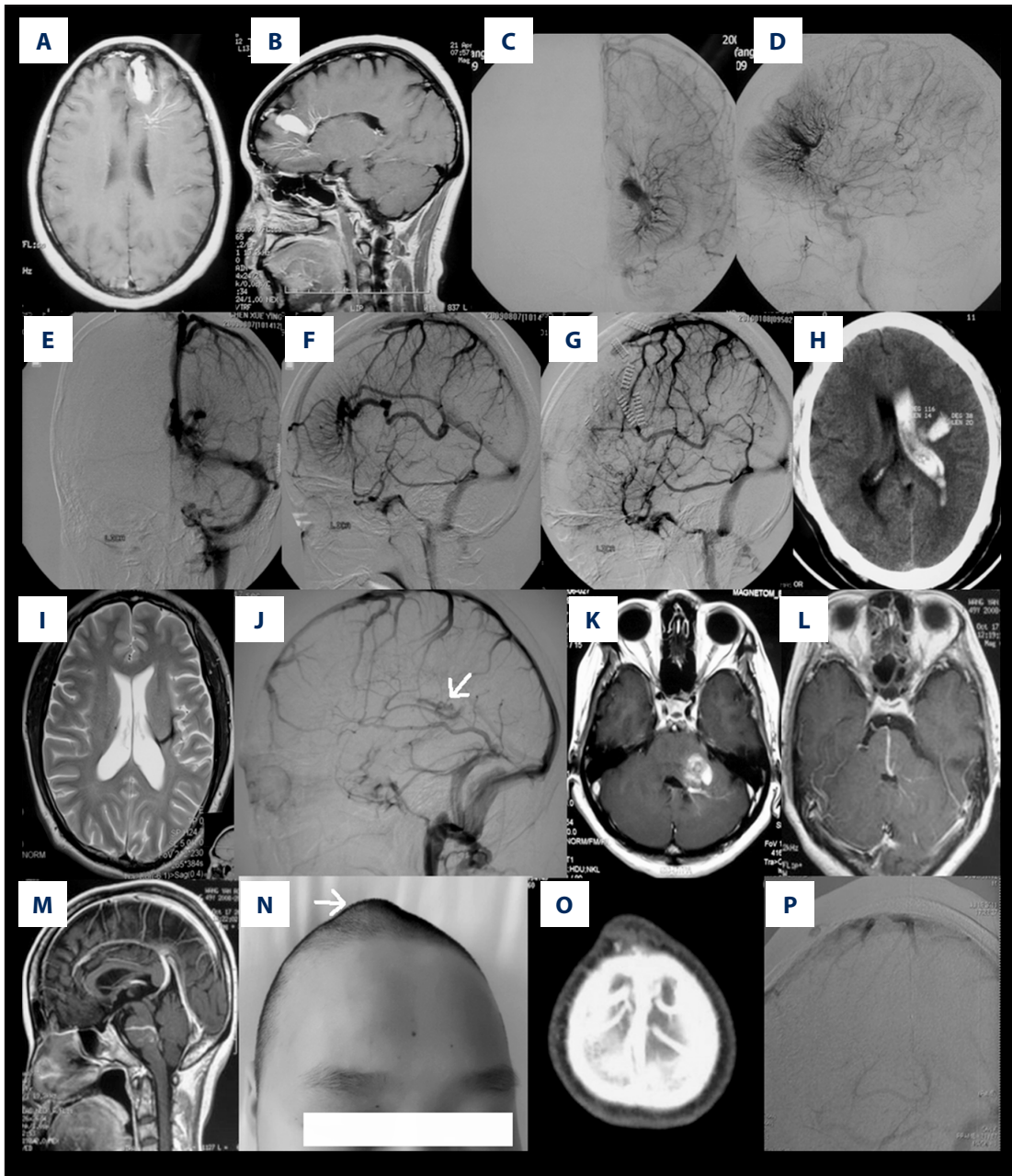


Figure 1. The morphological features of DVAs in different subtypes. (A–D) Demonstrate typical type A DVAs, axial and sagittal MR view (A, B) showing medullary veins arranged in stellate configuration and converged to an enlarged vein draining into a superficial cortical vein; anterior-posterior and lateral view of DSA (C, D) showing a “caput medusae”-like collections of dilated medullary veins converging in superior sagittal sinus during the late venous phase. (E–G) Demonstrate typical type B DVAs, anterior-posterior and lateral view of DSA show umbrella-shaped medullary veins converged and drained into the internal cerebral vein and then into Galen vein (E, F). Postoperatively, abnormal vessels were resected, epilepsy disappeared and deep draining vein was preserved (G). (H–J) Demonstrate type C DVAs. CT scan showing intraventricular hemorrhage (H), MRI showing an enlarged vein drained into a subependymal vein (I). (J) Showing abnormal veins in the lateral view of DSA (white arrow). (K) Demonstrate typical type D DVAs, an enlarged vein drained into a subependymal vein in the fourth ventricle, associated with a cavernous malformation in the brachium pontis. (L, M) Demonstrate a draining vein collecting deep cerebellar blood and coursed as an anterior transpontine vein and drained into the pontomesencephalic vein (Type E). (N–P) Demonstrate typical type F DVAs, non-pulsive soft scalp swelling could be seen in the frontal midline area (N, white arrow). Subcutaneous sinus pericranii was shown in CTA (O). In the lateral view of DSA, abnormal vein was demonstrated (P).

Table 2. Classification of symptomatic DVAs.

Subtypes (n)	Major symptoms and signs				Accompanying abnormality			Surgery (n)
	Seizure (n)	Hemorrhage (n)	Facial haemangiom (n)	Headache (n)	CM (n)	AVM (n)	Cortical dysplasia (n)	
Type A (12)	6	3	–	3	3	–	1	4
Type B (7)	–	2	–	5	–	2	–	2
Type C (8)	5	1	–	2	–	–	–	1
Type D (6)	–	4	–	2	4	–	–	6
Type E (6)	–	6	–	–	6	–	–	6
Type F (4)	–	–	4	4	–	–	–	–

AVM – arteriovenous malformation; CM – cavernous malformation; DVA – developmental venous anomalie.

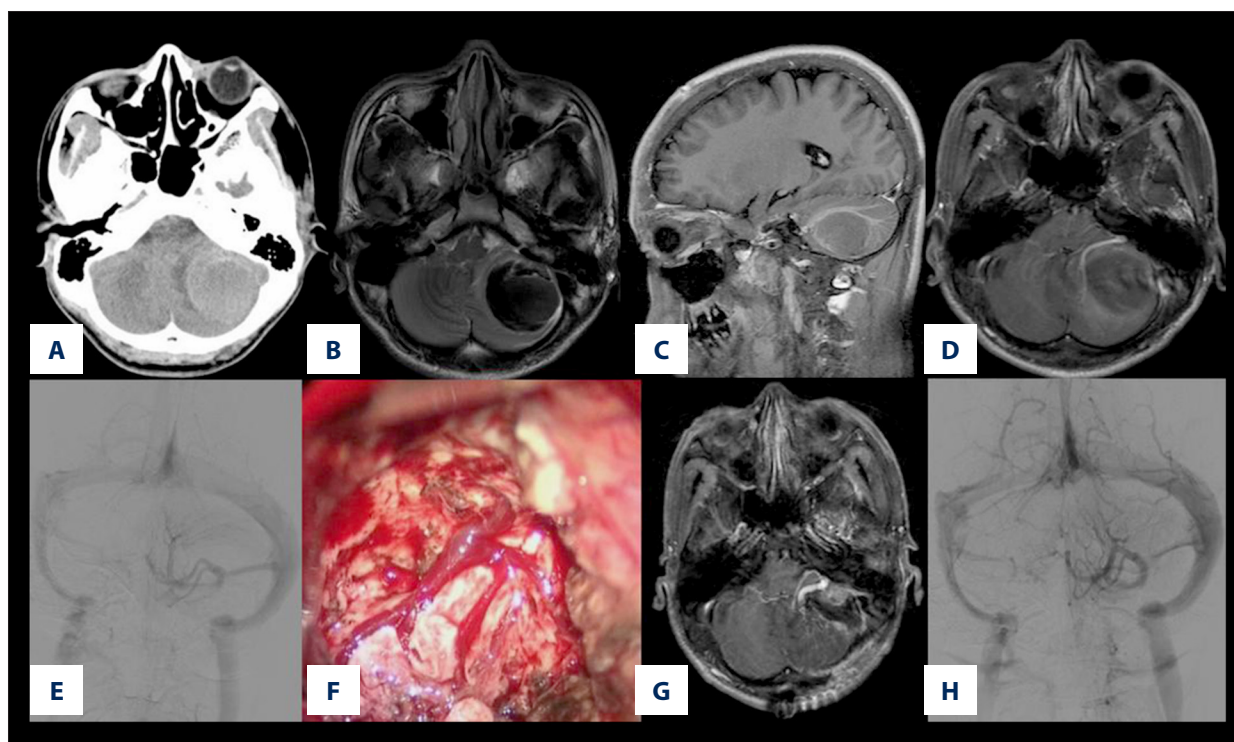


Figure 2. Illustrative case of a typical DVA with bleeding (Type D). (A, B) Preoperative CT and MRI showing hemorrhage in left cerebellar hemisphere. (C, D) Preoperative MRI showing DVAs in left cerebellar hemisphere. (E) Preoperative angiogram anteroposterior view showing branches converged and draining into sigmoid sinus. (F) Intra-operative image showing the clot was evacuated and draining veins were preserved. (G, H) Postoperative MRI and DSA showing draining veins were preserved.

transcortical vein. MRI findings of parenchymal abnormalities surrounding DVAs may be related to edema, gliosis, or leuko-araiosis secondary to altered hemodynamics in the drainage area [8]. The angiographic appearance of these anomalies is that a series of small deep veins converge towards a larger collector. One or more of these veins follow a transhemispheric course before draining into a normal deep or superficial vein [2,6].

Association with other vascular malformations

In our study, predominant symptoms and signs of symptomatic DVAs included headaches and seizures (62.8%). Sixteen patients (37.2%) had other associated abnormalities

The most common and clinically significant entity that has been associated with DVAs is CMs. DVAs with one or more CMs are typically located in the region of the DVA's caput medusae [4–6,12,13]. Based on MRI findings, Wurm et al. [14] reported 15 of 58 patients (25.9%) with cerebral, cerebellar, or brain stem CMs also had associated DVAs. A similar percentage was reported by Zhang et al. [15], with this type of association in 11 of 41 (26.8%) cerebellar CMs patients. Gökçe et al. [9] reported CM in 17.3% patients with DVA (if the DVA was not localized in the drainage region, it was only 11%). In our study of patients with symptomatic DVAs, CMs were found in 13 patients (30.2%), with three in the frontal lobe, four in the cerebellar hemisphere, and the remaining six in the brain stem.

DVAs with AVMs are relatively rare [5,16]. In our study, there were two cases associated with AVMs, one located in the frontal lobe and the other in the occipital lobe, both presenting with parenchymal hemorrhage. In both cases, the AVMs were resected and DVAs were preserved.

Sinus pericranii were found in four children who presented with nonpulsatile frontal subcutaneous angioma when body position changed. Sinus pericranii is a rare congenital vascular abnormality characterized by abnormal connections between the intracranial and extracranial venous systems and is usually found in children [5,6]. In most instances, a sinus pericranii presents as a soft scalp swelling that appears with the patient in the recumbent position and disappears in the erect position. Sinus pericranii represents a communication between intracranial and extracranial venous drainage pathways in which blood may circulate directionally through dilated veins of the cranium. It can be considered the extracranial counterpart of DVAs; supporting the hypothesis of a common venous malformation disorder [17]. Treatment options in symptomatic patients include surgical resection or a transvenous endovascular approach. Gondolfo et al. [18] recommended assessing the drainage pattern of sinus pericranii. If sinus pericranii is the dominant drainage of usual brain venous outlets, then prudent treatment should be adopted. In our study, the four patients with sinus pericranii were observed and followed.

Associated cerebral regional abnormalities

In a series of 84 consecutive DVAs confirmed by MRI and CT, San Millán Ruíz et al. [19] found that 65% of DVA cases were associated with parenchymal abnormalities (other than CMs) within the drainage territory. In our study, only one patient presented with refractory seizure, with MRI revealing an enlarged vein and cortical dysplasia in the left frontal region. Cerebral angiography showed DVAs draining into the great cerebral vein. The patient subsequently underwent a complete resection of the venous malformation with preservation of draining vein to internal cerebral vein and the cortical dysplasia was

partially resected [20]. This patient was seizure-free with no postsurgical neurological deficits (Figure 1E–1G).

Classification of symptomatic DVAs

We divided symptomatic DVAs into six different subtypes based on location and draining vein features. Type A DVAs were located in the cerebral hemisphere and drained into the superficial cortical vein or dura sinus (e.g., superior sagittal sinus, transverse sinus). This type of DVA was accompanied by high morbidity of epilepsy (6/12, 50.0%). In patients with refractory epilepsy or recurrent bleeding in a non eloquent brain area, the lesion could be removed. However, it is noteworthy that the brain tissue within the scope of the draining vein should be resected as much as possible. Type B DVAs were located in cerebral hemisphere and drained into cerebral internal veins. Operation should be avoided in this type. Type C DVAs were located in the cerebral hemisphere and drained into the subependymal vein. Most patients with this type also presented with epilepsy (5/8, 62.5%) and intraventricular hemorrhage occasionally occurred. Type D and E were infratentorial DVAs, the former ones located in cerebellum while the latter were located in brain stem. Infratentorial DVAs have been associated with a high ratio of CM compared with supratentorial DVAs (10/12 vs. 3/27, $p=0.0258$) and bleeding often occurred (10/12, 83.3%) [21,22]. Infratentorial cavernous hemangioma could be resected, and these collecting veins should be preserved. Type F DVAs were rare, subcutaneous sinus pericranii. To our knowledge, this is the first report classifying DVAs.

Surgical treatment strategies

To the best of our knowledge and review of the literature, DVAs have a benign natural course and good prognosis [4,6,10,11]. The majority of DVAs are incidental findings on MRI without any symptoms [4]. Most patients need no specific aggressive treatment except for those patients presenting with refractory epilepsy, repeat bleeding, or headache, and those patients suffering from cosmetic concern because of facial hemangioma [6,23,24]. The surgical resection in our study of symptomatic DVAs seemed very high (19/43, 44%). As for surgical techniques, some authors [4–7] have suggested that DVAs should not be resected in cases with potentially disastrous consequences occurring after occlusion of venous draining pathways while some authors [12] proposed a strategy of coagulation and division of the collecting vein of the DVA without leading to any obvious complications. However, in our initial surgical experience, one draining vein was coagulated and brain swelling occurred intraoperatively, requiring performance of an extended frontal lobe decompression. In our view, this strategy might increase the risk of serious postoperative brain edema due to compromise of venous drainage of normal brain, specifically in posterior fossa venous anomalies. Posterior fossa

venous anomalies often involve major cerebellar and brainstem venous drainage, and the disruption of venous anomaly is often catastrophic and might even cause cerebellar tonsillar herniation. In our study, two patients suffered from intracerebral hemorrhage with mass effect. Intraoperatively, the hemorrhage was evacuated and abnormal veins were found adjacent to the clot cavity. The veins were followed along to the ventricular wall and drained to the subependymal vein, and these collecting veins were preserved. This strategy has also been successfully reported by Zhang et al. [13]

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Conclusions

In our study, the rate of an accompanying abnormality in symptomatic DVAs was high. Intracerebral hemorrhage and epilepsy was usually attributed to associated CMs or AVMs. We found that the associated lesions could be resected and the draining vein should be preserved as far as possible to avoid catastrophic consequence. Therefore, when the epilepsy or the intracerebral hemorrhage was attributed to DVAs, the DVAs could be resected.

Competing interests

The authors declare that they have no competing interests.