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CASE REPORT



Angioarchitecture of the hemorrhagic developmental venous anomaly with stenosis of the collecting vein and cavernous malformation: a case report

Yu lida^a, Kentaro Mori^a, Yosuke Kawahara^a, Issei Fukui^a, Ryotaro Yamashita^b, Mutsuki Takeda^b, Tatsu Nakano^b, Hiroki Taguchi^c and Motohiro Nomura^a

^aDepartment of Neurosurgery, Yokohama Sakae Kyosai Hospital, Yokohama, Japan; ^bDepartment of Neurology, Yokohama Sakae Kyosai Hospital, Yokohama, Japan; ^cDepartment of Neurosurgery, Taguchi Neurosurgery Clinic, Yokohama, Japan

ABSTRACT

We herein report a case of developmental venous anomaly (DVA) with venous congestion caused by stenosis of the collecting vein that presented with intracerebral hemorrhage (ICH). A 74-year-old woman was referred to our hospital a few days after the onset of motor aphasia. Computed tomography (CT) and magnetic resonance imaging (MRI) showed ICH in the left frontal lobe. Angiography revealed DVA in the left frontal lobe in the late venous phase. Stenosis of the collecting vein of DVA at the entrance to the superior sagittal sinus was detected and accompanied by cavernous malformation (CM) beside DVA. Cone-beam CT revealed the absence of the left septal vein and hypoplastic transverse caudate veins. The patient was treated by blood pressure management and no additional neurological symptoms were detected. DVA develops to compensate for the absence of pial or deep venous systems, and generally benign and clinically asymptomatic. However, the outflow restriction of DVA causes chronic venous hypertension and the formation of CM. These abnormalities are considered to occur during post-natal life and may result in ICH. The risk of hemorrhage needs to be considered in cases of DVA with restricted venous outflow or CM.

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Developmental venous anomaly; deep; medullary; cavernous malformation; hemorrhage

Introduction

Developmental venous anomalies (DVA) are the most common vascular malformations of the brain and are commonly identified in routine radiological examinations. DVA, which were previously called venous angiomas, consist of converging dilated medullary veins that drain into a transcerebral collector vein, which opens into the superficial subcortical or deep subependymal veins.¹ DVA are generally benign, and asymptomatic.² However, some DVA cause clinical symptoms, which may be due to coexisting cavernous malformation (CM) or DVA themselves. We recently encountered a case of DVA with stenosis in the collecting vein and CM, which presented with intracerebral hemorrhage (ICH). We herein discuss the etiology, angioarchitecture, association with CM, and clinical presentation of this case.

Case presentation

A 74-year-old woman with hyperlipidemia developed motor aphasia and visited our hospital a few days later. The patient had no neurological deficits other than motor aphasia. Computed tomography (CT) and T2-weighted (T2-WI) magnetic resonance imaging (MRI) showed ICH surrounded by edema in the left frontal lobe (Figure 1(A,B)). T2-WI also revealed a flow void medial to ICH (Figure 1(B)). MR angiography did not detect any abnormal vessels. Contrast-enhanced T1-WI MRI showed an enhanced vessel beside and multiple nodular lesions within ICH. These nodular lesions were suspected to be CM (Figure 1(C)). Angiography of the left common carotid artery revealed that

DVA, which formed the so-called caput medusa, was not present in the early venous phase, but appeared in the late venous phase (Figure 2(A–D)). Three-dimensional rotational angiography in the late venous phase showed stenosis in a collecting vein of DVA that entered the superior sagittal sinus (SSS) and right unilateral hypoplasia of the rostral end of SSS (Figure 2(E)). Unilateral hypoplastic rostral SSS with compensatory drainage through the parasagittal superior frontal vein was observed on the right side (Figure 2(A,B)). Hemi-SSS was detected on the left side; however, cortical veins were not identified (Figure 2(F)). The superficial drainage system on the left side drained through DVA as a compensatory route. Moreover, cone-beam CT revealed the absence of the left septal vein and hypoplastic transverse caudate veins. Dilated deep medullary veins (DMV) converged into DVA (Figure 3(A–C)). Based on these angiographic findings, this compensatory route through DVA drained the venous flow of both the superficial and deep venous systems in the left frontal region.

It was not possible to clarify whether hemorrhage was induced by venous congestion due to DVA with stenosis in the collecting vein or CM. We speculated that hemorrhage was associated with DVA. The hematoma slightly expanded during hospitalization, but then gradually decreased in size. The patient was conservatively treated by blood pressure management, and no additional neurological symptoms were detected. Since higher brain dysfunction persisted, the patient was transferred to another hospital for rehabilitation on the 39th day.

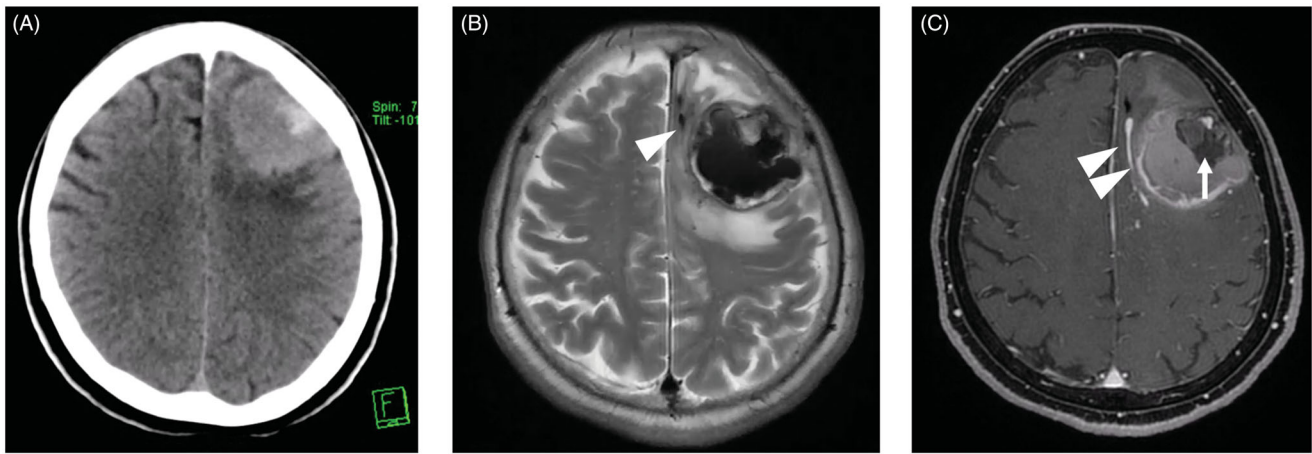


Figure 1. (A) Computed tomography (CT) showing intracerebral hemorrhage (ICH) with surrounding edema in the left frontal lobe. (B) T2-weighted (T2-WI) magnetic resonance imaging (MRI) revealing a flow void beside ICH (arrowhead). (C) Contrast-enhanced T1-WI MRI revealing an enhanced vessel around ICH (double arrowheads) and cavernous malformation within ICH (arrow).

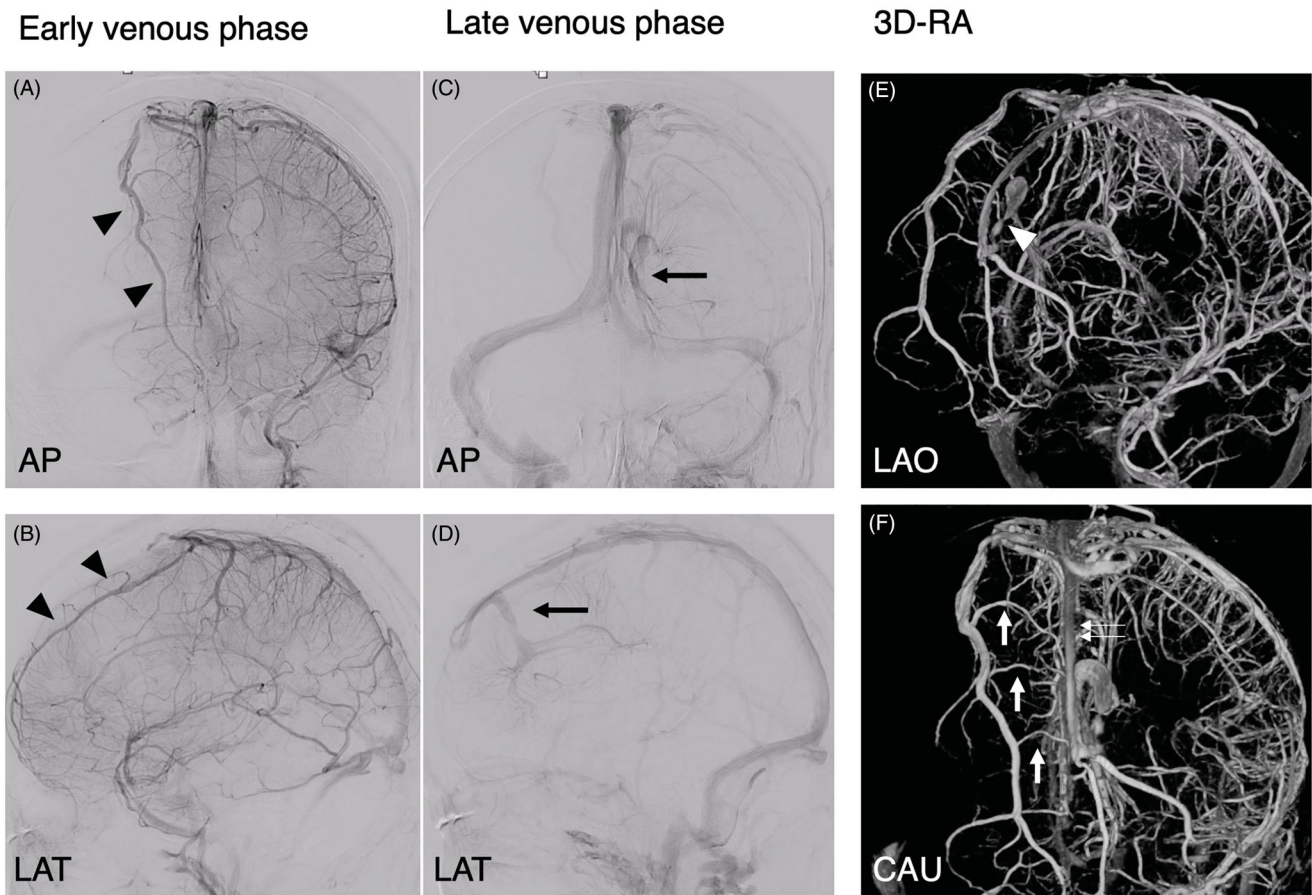


Figure 2. Angiography of the left common carotid artery in the early venous phase in anteroposterior (AP) (A) and lateral (LAT) projections (B) showing the right unilateral hypoplastic rostral superior sagittal sinus (SSS) with a compensatory enlarged parasagittal superior frontal vein (arrowheads). In the late venous phase (C, D), the developmental venous anomaly (DVA) appeared and drained into rostral SSS, and venous stagnation was observed in DVA (arrow). Three-dimensional rotational angiography (3D-RA) in the left anterior oblique (LAO) view (E) showing stenosis of the collecting vein at the SSS entrance (arrowhead). The caudal view (CAU) (F) showing the enlarged parasagittal superior frontal vein with cortical veins (arrows) on the right side. Hemi-SSS (double arrows) was observed on the left side, whereas cortical veins were not.

Discussion

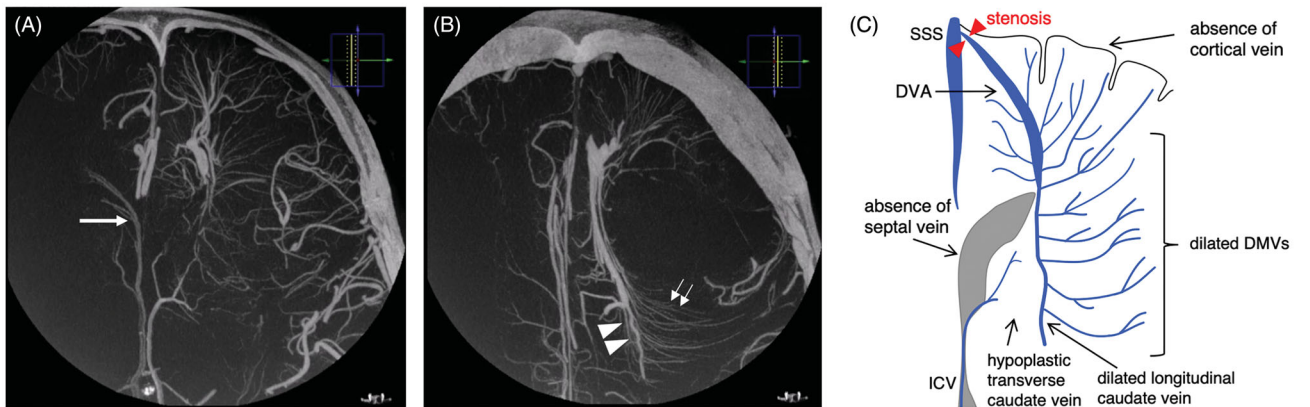
Normal structure of superficial and deep venous systems in the fronto-parietal lobe and striatum, and the angioarchitecture of DVA

DVA are the most frequently encountered intracranial vascular malformation with an incidence of 2.6% in autopsy and 6.4% on

CT or MRI.^{3,4} The etiology of DVA has not yet been elucidated in detail; however, they are considered to develop as a compensatory venous system for aplasia, hypoplasia, or the occlusion of various segments of the normal medullary, pial, or subependymal veins.⁵

Basic knowledge of the normal architecture of medullary veins provides a more detailed understanding of the etiology of DVA.

Present case



Normal control

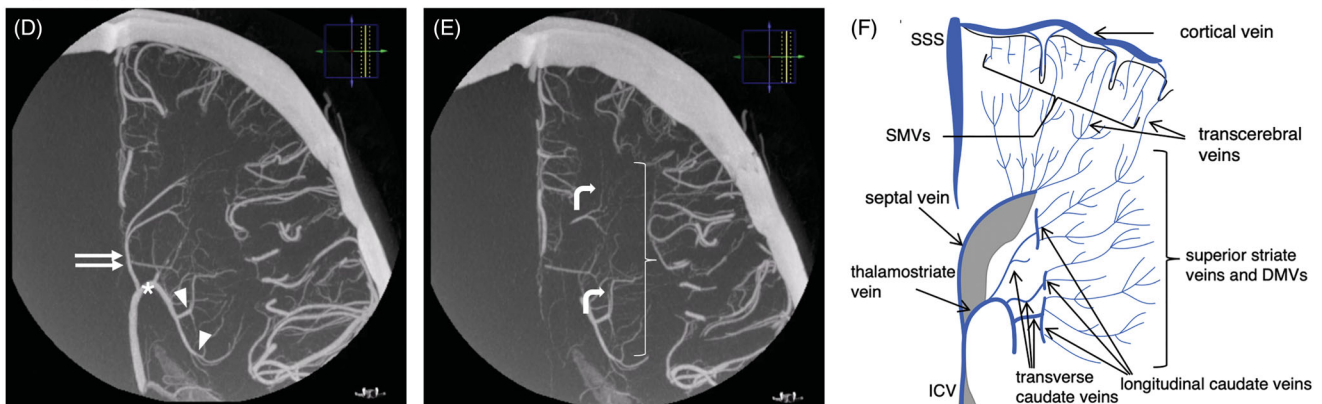


Figure 3. (A, B) Axial section of cone-beam CT of the present case clearly showing the right septal vein on the normal right side (arrow). On the other hand, the left septal vein was not detected, and deep medullary veins (DMVs) around the left anterior horn (double small arrows) converged on the developmental venous anomaly (DVA). Transverse caudate veins on the left side were not developed. Deep venous flow in the left front-parietal lobe did not drain through the septal or transverse caudate veins, it drained through DVA with a compensatory dilated longitudinal caudate vein (double arrowheads) and DMV (double small arrows). (C) Schematic representation of the venous angioarchitecture of the present case in the same view as (A) and (B). (D, E) Axial section of cone-beam CT in a normal case clearly showing the left septal vein (double arrows) and transverse caudate veins (arrowheads). Superior striate veins and DMV (bracket) flow into the longitudinal caudate veins (curved arrows), which converge on the thalamostriate vein (asterisk) through the transverse caudate veins and eventually reach the internal cerebral vein (ICV). (F) Schematic representation of the normal venous angioarchitecture in the same view as (D) and (E).

Supratentorial medullary veins are divided into two groups: superficial and deep medullary veins. Superficial medullary veins (SMV) function in venous drainage in the territory of the cerebral cortex and subcortical white matter. SMV drain into the pial veins and eventually into SSS. On the other hand, DMV play a role in venous drainage in the deep white matter and striate body. They converge on subependymal veins located along the lateral ventricles (LV) and eventually drain into the internal cerebral vein (ICV) and basal vein of Rosenthal. Anastomoses have also been reported between the pial and subependymal veins called transcerebral veins.⁶ Subependymal veins are distributed along with LV and receive the venous flow of DMV from the medial and lateral sides of LV. The septal and thalamostriate veins are subependymal veins located in the frontal horn and the mid-body of LV, which are tributaries of DMV of the front-parietal lobe and striatum. The septal vein, to which venous flow from DMV in the anterolateral region of the frontal horn converges, drains into ICV from the medial side of LV. Superior striate veins on the superior part of the striate body and DMV on the front-parietal lobe converge on the longitudinal caudate veins, which course along the anterolateral corner of the frontal horn and mid-body of LV. Longitudinal caudate veins converge on the thalamostriate vein through the transverse caudate veins

and eventually reach ICV from the lateral side of LV (Figure 3(D-F)).

It is considered that DVA are congenital and develop to compensate for the absence of the aforementioned normal venous system during intrauterine life.⁵ However, recent some studies indicate that DVA develop postnatally.^{7,8} They hypothesized that early postnatal venous occlusion could trigger remodeling of medullary veins, thus resulting in DVA formation. In fact, there is continued development of the cerebral venous system in postnatal life.⁹ Although whether the DVA are congenital or postnatal origin is controversial, changes in venous architecture and drainage patterns may contribute to the development of DVAs.

In the present case, the septal vein was well developed on the normal right side. On the other hand, the left septal vein was not detected; therefore, DMV around the left anterior horn converged on DVA, not the septal vein. Furthermore, the transverse caudate veins on the left side were not developed. Therefore, deep venous flow in the left front-parietal lobe was not drained through the septal and transverse caudate veins, but through DVA with compensatory dilated DMV and longitudinal caudate veins (Figure 3(A-C)).

Hypoplasia of the rostral third of SSS was detected on the right side in the present case. San Millán Ruíz *et al.*¹⁰ classified

four types of rostral SSS anatomies: (1) classic anatomy with fully developed rostral SSS; (2) duplication; (3) complete or bilateral hypoplasia; (4) unilateral hypoplasia. The present case corresponded to type 4 rostral SSS. Unilateral hypoplastic rostral SSS with compensatory drainage through the parasagittal superior frontal vein was observed on the right side (Figure 2(A,B)). Hemi-SSS was identified on the left side, whereas cortical veins were not. The superficial drainage system on the left side appeared to use a compensatory drainage route through DVA into left hemi-SSS (Figure 2). These radiological findings suggested that DVA developed in the present case to compensate for the superficial and deep venous systems of the front-parietal venous territory, resulting in aplasia, hypoplasia, or thrombosis of the subependymal and pial veins.

Clinical presentation

DVA are generally benign, and asymptomatic.² They function in normal cerebral venous drainage. Catastrophic venous ischemic and hemorrhagic complications resulting from the surgical removal of DVA have been reported in some cases.^{11,12} Neural tissue damage or dysfunction within the DVA drainage territory is not typically observed.¹ However, San Millán Ruíz *et al.*¹³ reported parenchymal abnormalities, such as atrophy, white matter lesions, calcifications, and CM formation, within the drainage territory of DVA. They also noted that these abnormalities caused by chronic cerebral ischemia secondary to venous hypertension were acquired after birth. These abnormalities do not generally lead to neurological deficits; however, venous hypertension may induce ICH.

CM associated with DVA is detected in approximately 13% of cases.¹³ CM is currently considered to be responsible for the vast majority of symptoms previously attributed to DVA themselves.^{14,15} Previous studies suggested that the hemorrhagic incidence of CM associated with DVA was higher than that of isolated CM.¹⁶ DVA are considered to play an important role in the development and growth of CM. The probable pathogenesis of this development appears to result from hemodynamic changes in DVA. Chronically elevated blood pressure due to flow disturbances within the territory of DVA causes microhemorrhages around DVA. As a result, hemorrhagic angiogenesis leads to CM formation.¹⁷ In the present case, CM were detected in the white matter of the left frontal lobe within the DVA drainage territory. Chronic venous hypertension with DVA due to flow disturbances caused by stenosis of the collecting vein at the SSS entrance may contribute to the development of CM, which may, in turn, cause hemorrhage.

The risk of hemorrhage from DVA themselves is low. Garner *et al.*¹⁸ retrospectively evaluated the risk of hemorrhage associated with DVA as 0.22% per year, while McLaughlin *et al.*¹⁵ reported that it was 0.68% per year, although only 50% of their patients were symptomatic. Only a few case reports of DVA with ischemic or hemorrhagic infarctions or with focal neurological symptoms have been published to date.^{13,19} The pathophysiology of symptomatic DVA themselves includes increased inflow, decreased outflow, and mechanical compression.¹⁹ Increased inflow may result from upstream arteriovenous malformations or DVA with micro or diffuse arteriovenous shunt. Decreased outflow includes stenosis or thrombosis of the outflow vein. Mechanical compression of the outflow vein may result in obstructive hydrocephalus or neurovascular nerve compression syndrome.¹⁹ Pereira *et al.*¹⁹ summarized 8 cases from their database and 18 previous cases of obstructed or restricted DVA

outflow. Seven patients (27.6%) presented with hemorrhage (intraparenchymal or subarachnoid), whereas the major presentation was venous congestion with edema. Another 51.7% of cases had thrombosis of the venous collector, and 24.1% had stenosis at some point along the DVA drainage route. Mechanical obstruction led to venous congestion, resulting in hemorrhagic or venous infarction in all symptomatic patients. In the present case, tight stenosis of the collecting vein of DVA was detected at the point of dural passage into SSS. Angiography also showed venous stagnation in DVA. Therefore, hemorrhage may have been induced by venous congestion due to the stenotic collecting vein of DVA. Although it currently remains unclear whether hemorrhage was caused by venous congestion or CM, the risk of hemorrhage needs to be considered in cases with the outflow restriction of DVA and CM within the drainage territory of DVA.

Conclusion

DVA form to compensate for the absence of normal parenchymal veins. In the present case, the absence of septal and pial veins and hypoplasia of the transverse caudate veins were identified. We speculate that DVA developed in the present case to compensate for the superficial and deep venous systems, resulting in aplasia, hypoplasia, or thrombosis of the subependymal and pial veins. Although DVA follow a benign course, chronic venous hypertension in DVA may induce the formation of CM during post-natal life. Furthermore, the restricted outflow of DVA contributes to venous hypertension. Therefore, the risk of hemorrhage needs to be considered in cases of DVA with outflow restriction or CM.

Disclosure statement

No potential conflict of interest was reported by the author(s).

References

1. Lasjaunias P, Burrows P, Planet C. Developmental venous anomalies (DVA): the so-called venous angioma. *Neurosurg Rev* 1986;9:233–42.
2. Fujii K, Matsushima T, Inamura T, Fukui M. Natural history and choice of treatment in forty patients with medullary venous malformation (MVM). *Neurosurg Rev* 1992;15:13–20.
3. Sarwar M, McCormick WF. Intracerebral venous angioma. Case report and review. *Arch Neurol* 1978;35:323–5.
4. Gökçe E, Acu B, Beyhan M, *et al.* Magnetic resonance imaging findings of developmental venous anomalies. *Clin Neuroradiol* 2014;24:135–43.
5. Okudera T, Huang YP, Fukusumi A, *et al.* Micro-angiographical studies of the medullary venous system of the cerebral hemisphere. *Neuropathology* 1999;19:93–111.
6. Huang YP, Okudera T, Fukusumi A, *et al.* Venous architecture of cerebral hemispheric white matter and comments on pathogenesis of medullary venous and other cerebral vascular malformations. *Mt Sinai J Med* 1997;64:197–206.
7. Brzegowy K, Kowalska N, Solewski B, *et al.* Prevalence and anatomical characteristics of developmental venous anomalies: an MRI study. *Neuroradiology* 2020; Nov 23. [Online ahead of print]. doi: 10.1007/s00234-020-02612-8
8. Horsch S, Govaert P, Cowan FM, *et al.* Developmental venous anomaly in the newborn brain. *Neuroradiology* 2014;56:579–88.
9. Padgett DH. The cranial venous system in man in reference to development, adult configuration, and relation to the arteries. *Am J Anat* 1956;98:307–55.
10. San Millán Ruíz D, Fasel JH, Gailloud P. Unilateral hypoplasia of the rostral end of the superior sagittal sinus. *AJNR Am J Neuroradiol* 2012; 33:286–91.

11. Senegor M, Dohrmann GJ, Wollmann RL. Venous angiomas of the posterior fossa should be considered as anomalous venous drainage. *Surg Neurol* 1983;19:26–32.
12. Yamamoto M, Inagawa T, Kamiya K, *et al.* Intracerebral hemorrhage due to venous thrombosis in venous angioma-case report. *Neurol Med Chir* 1989;29:1044–6.
13. San Millán Ruíz D, Delavelle J, Yilmaz H, *et al.* Parenchymal abnormalities associated with developmental venous anomalies. *Neuroradiology* 2007;49:987–95.
14. Töpfer R, Jürgens E, Reul J, Thron A. Clinical significance of intracranial developmental venous anomalies. *J Neurol Neurosurg Psychiatry* 1999;67:234–8.
15. McLaughlin MR, Kondziolka D, Flickinger JC, *et al.* The prospective natural history of cerebral venous malformations. *Neurosurgery* 1998;43:195–200.
16. Aoki R, Srivatanakul K. Developmental venous anomaly: benign or not benign. *Neurol Med Chir* 2016;56:534–43.
17. Idiculla PS, Gurala D, Philipose J, *et al.* Cerebral cavernous malformations, developmental venous anomaly, and its coexistence: a review. *Eur Neurol* 2020;83:360–8.
18. Garner TB, Del Curling O, Jr, Kelly DL, Jr, Laster DW. The natural history of intracranial venous angiomas. *J Neurosurg* 1991;75:715–22.
19. Pereira VM, Geibprasert S, Krings T, *et al.* Pathomechanisms of symptomatic developmental venous anomalies. *Stroke* 2008;39:3201–15.