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CASE REPORT

Non-haemorrhagic Pontine Venous Infarct due to Thrombosed Cerebellar Venous Angioma

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ABSTRACT

Developmental venous anomaly (also known as venous angioma or venous malformation) is one of the 4 main vascular malformations. Developmental venous anomaly is usually benign and clinically silent. This report is of a patient with cerebellar developmental venous anomaly with draining vein thrombosis, complicated by non-haemorrhagic venous infarct of the midbrain.

Key Words: *Brain infarction; Central nervous system venous angioma; Thrombosis*

INTRODUCTION

Cerebral vascular malformations have been classified by McCormick into 4 major pathologic types: arteriovenous malformation, cavernous angioma, capillary telangiectasia, and venous angioma, which is also known as developmental venous anomaly (DVA).[1] Venous angioma is the most common cerebral vascular malformation, comprising 63% of vascular malformations in 2 large autopsy studies.[2,3] The reported incidence is 2.56%.[3] Venous angioma is found incidentally and is usually clinically silent. Headache, seizure, focal neurological deficit, dizziness, and ataxia can be symptoms of venous angioma.[4] Complications of venous angioma include bleeding, thrombosis, and venous infarct. Non-haemorrhagic venous infarction of venous angioma is an exceptionally rare complication, that was first mentioned in the literature in 1986.[5] Since then, only 17 patients have been reported.[5-19] This report is of a patient with DVA in the cerebellum, complicated by brain stem venous infarct.

CASE REPORT

A 45-year-old previously healthy woman presented in February 2009 with sudden onset of severe vertigo and vomiting. She had seen her physician 10 days previously for irregular menses, and was prescribed oral contraceptive pills. At physical examination, she had nystagmus on left gaze. Her limb power, sensation, and Glasgow coma score were all normal.

Initial unenhanced urgent computed tomography (CT) of the brain showed a subtle hypodense area in the midbrain, but no mass effect or acute haemorrhagic focus was identified (Figure 1). Urgent magnetic resonance imaging (MRI) of the brain was performed later on the same day to rule out posterior fossa vascular insult.

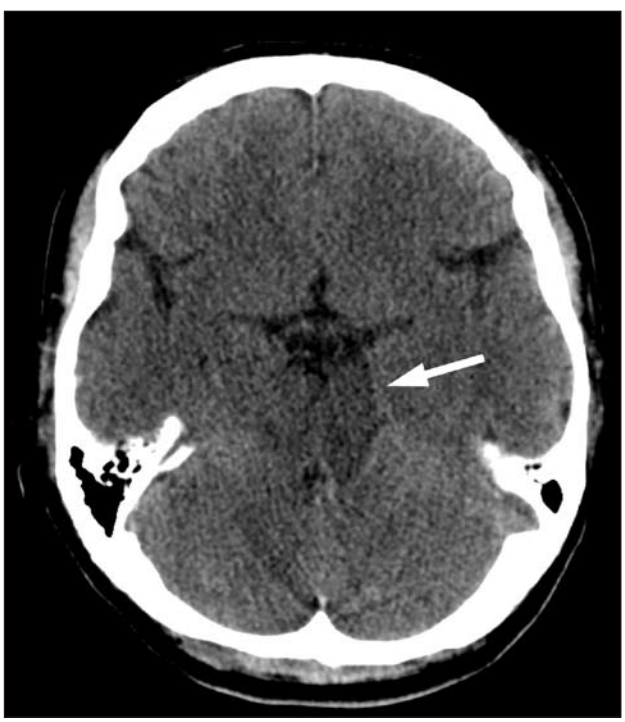

Figure 1. Axial computed tomography of the brain. Subtle hypodensity is noted at the left midbrain (arrow).

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T2-weighted and fluid attenuation inversion recovery (FLAIR) images showed increased signal intensities in the bilateral midbrain and left thalamus (Figure 2). Diffusion-weighted images and apparent diffusion coefficient (ADC) map showed non-restricted diffusion over the midbrain. T1-weighted postcontrast images showed an enhanced slow-flow vascular lesion in the bilateral cerebellar hemispheres and cerebellar vermis. A larger central draining vein was noted in the quadrigeminal cistern that was connected to the great vein of Galen and straight sinus. The central draining vein was not opacified in postcontrast images, and showed internal T1-weighted isointense and T2-weighted hypointense signal, suggestive of acute thrombosis.

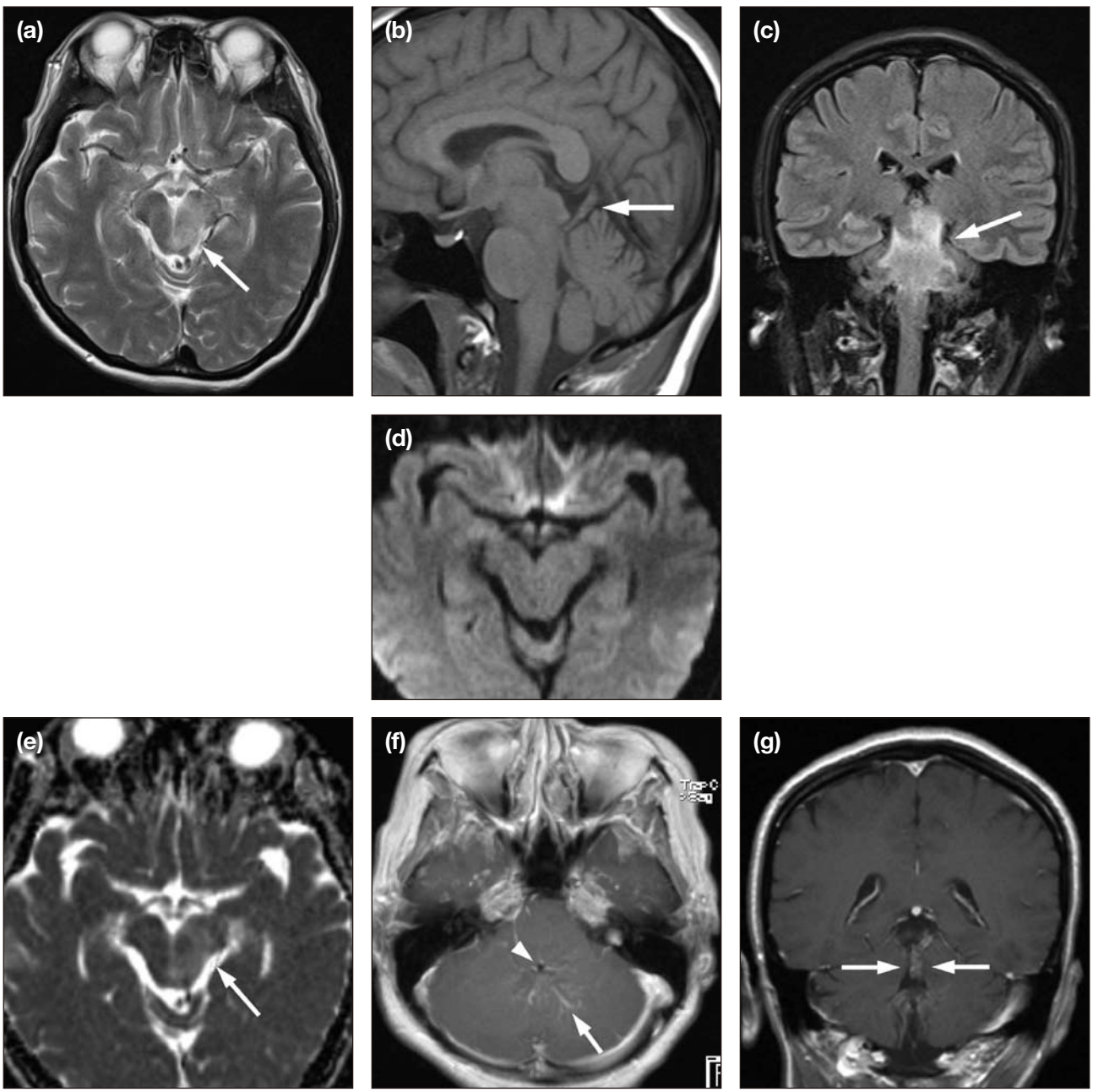

Figure 2. Magnetic resonance images of the brain. (a) Axial T2-weighted image showing hyperintense signal in the brainstem (arrow); (b) sagittal T1-weighted image showing no abnormal signal or haemorrhagic focus in the brainstem — an isointense tubular structure suggestive of a thrombosed draining vein is seen within the quadrigeminal cistern (arrow); (c) coronal fluid attenuation inversion recovery image of the brain showing increased signal intensity along the bilateral midbrain, pons, and cerebellar peduncles (arrow); (d) axial diffusion-weighted image (b = 1000 s/mm^2) of the brain showing no high-signal area; (e) axial apparent diffusion coefficient map of the brain showing hyperintense signal suggestive of vasogenic oedema in the left midbrain (arrow); (f) axial T1-weighted image with Gadolinium contrast showing 'caput medusae' enhancement of a venous angioma (arrow) and a non-enhanced draining vein (arrowhead); and (g) coronal T1-weighted image with Gadolinium contrast showing the non-enhanced thrombosed draining vein of the venous angioma (arrows).

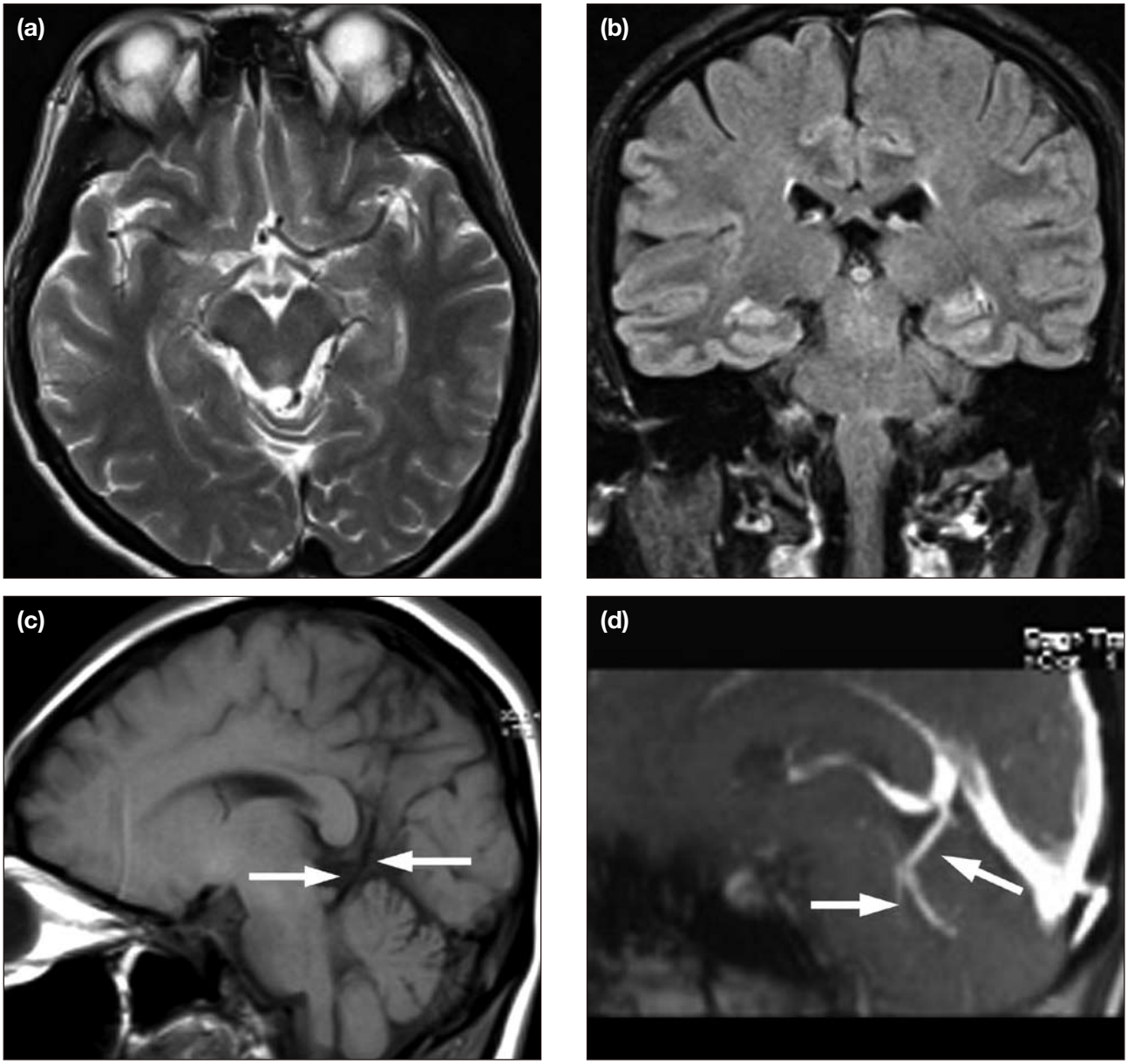

Figure 3. Magnetic resonance images of the brain. (a) Axial T2-weighted image showing subsidence of the T2 hyperintensity in the brainstem; (b) coronal fluid attenuation inversion recovery image of the brain showing the absent T2-weighted hyperintense signal along the brainstem; (c) sagittal T1-weighted image showing recanalisation of the previously thrombosed draining vein flow void (arrows); and (d) sagittal venogram demonstrating the venous angioma draining into the great vein of Galen (arrows).

Low–molecular weight heparin (enoxaparin 0.4 mL by subcutaneous injection) was started, and her symptoms gradually subsided 3 days after the onset of treatment. Heparin was substituted by warfarin 2 mg and the patient was discharged 6 days after admission.

Follow-up MRI was performed 1 week after the patient was discharged. The T2-weighted and FLAIR images (Figure 3) showed complete resolution of the midbrain signal. The thrombosed central draining vein showed contrast opacification and recanalisation in a magnetic resonance venogram.

DISCUSSION

Lasjaunias et al first classified venous angioma into DVAs, and proposed that these lesions are not true vascular malformations, but arrested medullary vein development at the time when normal arterial development is nearly completed.[20] DVAs most commonly occur at the periventricular region, around the frontal horn of the lateral ventricles,[21] while the second most common site is adjacent to the fourth ventricle. DVA is the most common cerebral vascular malformation, and most DVAs are solitary. One-third of DVAs are associated with cavernous angioma.[20]

Most DVAs are asymptomatic and are found incidentally, although sometimes DVA may present with headache, seizure, focal neurological deficit, dizziness, and ataxia.

DVAs have a characteristic appearance on CT, MRI, and angiographic studies. Non-enhanced CT scan is usually normal or shows a slightly hyperdense tubular structure with parenchymal hypodensity related to underlying vasogenic oedema if the DVA is thrombosed. In contrast, on enhanced CT scan, an area of branching linear enhancement in the white matter, near the angle of the ventricle, is visible. Surrounding oedema and mass effect are typically absent with non-complicated DVAs.[22]

MRI has a high sensitivity for detection of DVA, showing linear structures with flow voids, usually around the periventricular region. After Gadolinium injection, there is marked enhancement of the enlarged medullary tributaries and subependymal draining veins.[23] A 'caput medusae' appearance in postcontrast MRIs and the venous phase of a cerebral angiogram is a diagnostic appearance, as the dilated medullary veins 'medusa head' drains into the enlarged transcortical draining vein and the superficial venous system.[24]

Complications of venous angioma are rare, and most are related to haemorrhage, with an estimated rate of 0.15% per lesion per year.[6] Among the haemorrhagic DVAs, 33% to 48% are due to neighbouring cavernous malformation.[6] Thrombosis of the draining vein of a DVA leading to venous brain infarction is a rare complication, and only a few patients have been reported in the literature.[5-19] Masson et al proposed that DVA are abnormal dilated veins that lack smooth muscle cells and elastic connective tissue, and have limited capacity for regulation and adaptation.[11] This results in haemodynamic disturbances and predisposes to thrombosis. The predisposing factors for DVA thrombosis are similar to those for dural sinus thrombosis, and include use of the oral contraceptive pill and hereditary hypercoagulable states, such as protein C or S deficiency or antithrombin deficiency.[11] De Bruijn et al showed that women using oral contraceptives have a 13-fold risk for dural sinus thrombosis.[25] For this patient, oral contraceptive pills were prescribed a few days before the onset of symptoms, and may have triggered thrombosis of an underlying DVA.

The MRI findings of a thrombosed DVA was clearly demonstrated for this patient. Abnormal T2 and FLAIR signal changes were noted in the brainstem. The thrombosed collector vein of the DVA showed T1 hyperintense signal and T2 hypointense signal, which corresponds with the presence of an acute intraluminal clot. Initial diffusion-weighted imaging and ADC map revealed non-restricted diffusion and increased ADC values in the parenchyma adjacent to the DVA, which suggested underlying vasogenic oedema. The pathophysiological mechanisms of venous thrombosis leading to venous infarct remain controversial. Some authors have described the initial event of venous infarction as an increase in venous pressure.[23] This leads to disruption of the blood-brain barrier, and leakage of fluid (vasogenic oedema) and haemorrhage into the extracellular space.[26] Persistent venous congestion leads to a decrease in cerebral blood flow and venous infarct (cytotoxic oedema). Due to the differences in the pathophysiology of venous thrombosis and arterial stroke, venous infarct may not have the same prognostic value as arterial infarct, as venous infarct may be completely reversible if successful anticoagulant therapy is given.[11]

In the area of venous infarct, proton magnetic resonance spectroscopy will show normal N-acetyl aspartate peak with elevation of lactate peak. The imaging findings may represent ischaemic tissue at risk of infarction and the potential reversibility of the condition.[27] Cerebral angiogram can demonstrate filling defects in the venous collector in the late venous phase.[9]

There is no consensus of definitive therapy for symptomatic or asymptomatic DVA. Most authors recommend conservative treatment and observation for symptomatic and asymptomatic DVA.[28,29] Some authors suggest surgical ligation of the common draining vein or radiosurgery.[28,29] However, DVA is a compensatory drainage route for the brain; blockage of the drainage pathway can produce venous infarct of underlying brain tissue, and can be fatal if it occurs in the posterior fossa.[30] Therefore, surgical management is usually reserved for patients with DVA presenting with extensive intracerebral haemorrhage. Removing the haematoma and underlying cavernoma preserves the associated venous angioma.[31,32]

Seventeen similar cases have been reported in the literature (Table 1).[5-19] The mean age at the time of DVA thrombosis is 37 years; men and women are equally affected. The haemorrhagic rate of thrombosed DVA is 33%. Ten of 18 patients (55%) were given anticoagulants. The neurological outcomes of the treated patients with non-haemorrhagic venous infarct are better than

Table 1. Literature reports of patients with developmental venous anomaly.

Patient number	Study	Age (years)	Sex	Site	Haemorrhage	Diffusion-weighted imaging	Anticoagulation	Neurological deficit
1	Bouchacour et al,[5] 1986	37	F	Frontal	No	No	No	Yes
2	Field and Rusel,[7] 1995	34	F	Frontal	Yes	No	No	Yes
3	Merten et al,[6] 1998	50	F	Frontal	Yes	No	Yes	No
4	Thobois et al,[8] 1999	25	F	Parietal	No	No	Yes	Yes
5	Konan et al,[9] 1999	31	M	Cerebellum	No	No	No	Yes
6	Herbreteau et al,[10] 1999	45	M	Parietal	No	No	No	No
7	Masson et al,[11] 2000	43	M	Parietal	No	No	Yes	Yes
8	Masson et al,[11] 2000	68	M	Frontal	No	No	Yes	Yes
9	Hammoud et al,[16] 2002	26	F	Frontal	No	Restricted	No	Yes
10	Peltier et al,[12] 2004	32	M	Cerebellum	Yes	No	No	Yes
11	Flacke et al,[14] 2006	49	M	Frontal	No	Non-restricted	Yes	No
12	Parker and Sabb,[19] 2007	22	F	Frontal	No	Restricted	Yes	No
13	Brasse et al,[15] 2008	26	M	Frontal	Yes	No	No	Yes
14	Walsh et al,[18] 2008	38	M	Frontal	No	Non-restricted	Yes	No
15	Walsh et al,[18] 2008	52	M	Parietal	Yes	Non-restricted	No	Yes
16	Gama et al,[17] 2008	19	F	Frontal	Yes	Restricted	Yes	No
17	Prasad et al,[13] 2009	16	F	Frontal	No	Non-restricted	Yes	No
18	This report, 2009	45	F	Cerebellum	No	Non-restricted	Yes	No

those of untreated patients (63% versus 25%). Diffusion-weighted images were performed for 8 patients. Most (80%) of the patients with non-restricted diffusion venous infarct had good neurological outcomes and prognoses. The presence of cytotoxic oedema does not indicate irreversible damage or poor clinical outcome; 2 of 3 patients with restricted diffusion venous infarct had good neurological outcomes.[33]

Of the 18 DVAs, 3 (27%) were located in the cerebellum. Two of the previously reported patients with thrombosed cerebellar DVAs did not receive heparin, and they both had neurological deficits.[9,12] Low–molecular weight heparin was given to this patient, and she had a good clinical outcome with no neurological deficit.

The good clinical outcome for this patient may be due to the absence of underlying tissue infarction (cytotoxic oedema) and haemorrhage. The conclusion of the effectiveness of anticoagulation therapy for the treatment of thrombosed DVA cannot be made solely by a few cases, and further large case series are required before drawing a definite conclusion.

REFERENCES

1. McCormick WF. The pathology of vascular ("arteriovenous") malformations. J Neurosurg. 1966;24:807-16.
2. Garner TB, Del Curling O Jr, Kelly DL. The natural history of intracranial venous angiomas. J Neurosurg. 1991;75:715-22.
3. Sarwar M, McCormick WF. Intracerebral venous angioma. Case report and review. Arch Neurol. 1978;35:323-5.
4. Saito Y, Kobayashi N. Cerebral venous angiomas: clinical evaluation and possible etiology. Radiology. 1981;139:87-94.
5. Bouchacour E, Carpena JP, Bories J. Accident ischemique par thrombose d'un angiome veineux: a propas d'un cas. J Radiol. 1986;67:631-5.
6. Merten CL, Knitelius HO, Hedde JP. Intracerebral hemorrhage from a venous angioma following thrombosis of a drainage vein. Neuroradiology. 1998;40:15-8.
7. Field LR, Rusel EJ. Spontaneous hemorrhage from a cerebral venous malformation related to thrombosis of the central draining vein: demonstration with angiography and serial MR. AJNR Am J Neuroradiol. 1995;16:1885-8.
8. Thobois S, Nighoghossian N, Mazoyer JF. Thrombophlébite corticale et anomalie veineuse du développement. Rev Neurol. 1999;155:48-50.
9. Konan AV, Raymond J, Bourqouin P. Cerebellar infarct caused by spontaneous thrombosis of a developmental venous anomaly of the posterior fossa. AJNR Am J Neuroradiol. 1999;20:256-8.
10. Herbreteau O. Auffray-Calvier E, Desal H. Angiome veineux symptomatique: a propos d'un cas. J Neuroradiol. 1999;26:126-31.
11. Masson C, Godefroy O, Leclerc X. Cerebral venous infarction following thrombosis of the draining vein of a venous angioma (developmental abnormality). Cerebrovasc Dis. 2000;10:235-8.
12. Peltier J, Toussaint P, Desenclos C. Cerebral venous angioma of the pons complicated by nonhemorrhagic infarction. Case report. J Neurosurg. 2004;101:690-3.
13. Prasad S, Hurst RW, Kasner SE. Postpartum thrombosis of a developmental venous anomaly. Neurology. 2009;72:92-3.
14. Flacke S, Stuer C, Stoffel M. Symptomatic developmental venous anomaly after spontaneous thrombosis of the collector vein. Clin Neuroradiol. 2006;16:131-3.
15. Brasse G, Stammel O, Siemens P. Thrombose eines venosen Angioms mit sekundarem Stauungsinfarkt. Nervenarzt. 2008;79:703-5.
16. Hammoud D, Beauchamp N, Wityk R. Ischemic complication of a cerebral developmental venous anomaly: case report and review of the literature. J Comput Assist Tomogr. 2002;26:633-6.
17. Gama RL, Nakayama M, Tavora DG. Thrombosed developmental venous anomaly associated with cerebral venous infarct. Arq Neuropsiquiatr. 2008;66:560-2.
18. Walsh M, Parmar H, Mukherji SK. Developmental venous anomaly with symptomatic thrombosis of the draining vein. J Neurosurg. 2008;109:1119-22

19. Parker BJ, Sabb BJ. Developmental venous anomaly complicated by cerebral venous infarction. Radiol Case Rep. 2007;2:1-4.
20. Lasjaunias P, Burroes P, Planet C. Development venous anomalies (DVA): the so-called venous angioma. Neurosurg Rev. 1986; 4:233-42.
21. Wilms G, Demaerel P, Marchi G. Gadolinium-enhanced MR imaging of cerebral venous angiomas with emphasis on their drainage. J Comput Assist Tomogr. 1991;15:199-206.
22. Lotz PR, Quisling RG. CT of venous angiomas of the brain. AJNR Am J Neuroradiol. 1983;4:1124-6.
23. Wilms G, Marchal G, Vas Hecke P. Cerebral venous angiomas. MR imaging at 1.5 tesla. Neuroradiology. 1990;32:81-5.
24. Valavanis A, Wellauer J, Yasargil MG. The radiological diagnosis of cerebral venous angioma: cerebral angiography and computed tomography. Neuroradiology. 1983;24:193-9.
25. De Bruijn SF, Stam J, Koopman MM. Case-control study of risk of cerebral sinus thrombosis in oral contraceptive users and in carriers of hereditary prothrombotic conditions: the Cerebral Venous Thrombosis Study Group. BMJ. 1998;316:589-92.
26. Tsai FY, Wang AM, Maovich VB. MR staging of acute dural sinus thrombosis: correlation with venous pressure measurements and implications for treatment and prognosis. AJNR Am J Neuroradiol. 1995;16:1021-9.
27. Hsu LC, Liring JF, Fuh JL. Proton magnetic resonance spectroscopy in deep cerebral venous thrombosis. Clin Neurol Neurosurg. 1998;100:27-30.
28. Pak H, Patel SC, Malik GM. Successful evacuation of a pontine haematoma secondary to rupture of a venous angioma. Surg Neurol. 1982;18:193-202.
29. Malik GM, Morgan JK, Boulos RS. Venous angiomas: an underestimated cause of intracranial haemorrhage. Surg Neurol. 1988;30:350-8.
30. 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.
31. Buhl R, Hempelmann RG, Stark AM. Theurapeutical considerations in patients with intracranial venous angiomas. Eur J Neurol. 2002;9:165-9.
32. Nishizaki T, Tamaki N, Matsumoto S. Consideration of the operative indication for posterior fossa venous angiomas. Surg Neurol. 1986;25:441-5.
33. Peeters E, Stadnik T, Bissay F. Diffuseion-weighted MR imaging of an acute venous stroke: case report. AJNR Am J Neuroradiol. 2001;22:1949-52.