

Case Report

Hemorrhagic presentation without venous infarction caused by spontaneous thrombosis of developmental venous anomaly and angiographic change after treatment

Hyun Woo Kim, Cheol Young Lee*

Department of Neurosurgery, Konyang University Hospital, Daejeon, South Korea

ARTICLE INFO

Keywords:

Cerebral hemorrhage

Developmental venous anomaly

ABSTRACT

Developmental venous anomalies (DVAs) have been known benign anatomical variations and generally do not cause symptoms. Although several cases of venous infarction caused by DVA thrombosis have been reported, it is not common in itself. But only hemorrhagic presentation in DVA is extremely rare. We describe a case with large intracerebral hemorrhage (ICH) without venous infarction caused by DVA thrombosis and angiographic change of successful treatment.

1. Introduction

We have known about various vascular anomalies of the central nerve system including arteriovenous malformations (AVMs) and Developmental venous anomalies (DVAs) are a fairly commonly reported entity. However, they have been known as mostly benign anatomical variations that generally cause no symptoms [1]. DVAs occur during the developmental process of the venous system, by retention of the primitive embryological medullary vein [2]. It is well known that it represents the shape of the caput medusa, composed by one longer drainage vein. Although venous infarction caused by DVA is uncommon, several cases have been reported and most cases have been known to be associated with venous thrombosis of the draining vein [3]. But presentation with only hemorrhage in DVA is extremely rare. We are reporting a case of large intracerebral hemorrhage (ICH) without venous infarction caused by DVA thrombosis and would like to discuss use of anticoagulant in treatment of symptomatic DVA thrombosis with ICH.

2. Case report

A 27-year-old woman without past any remarkable medical history or medication was admitted with a general tonic clonic seizure and sudden onset decreased consciousness. Neurological examination showed drowsy consciousness with language and motor impairments. Computerized tomography (CT) scan revealed ICH in the left frontal lobe with intraventricular hemorrhage (IVH) (Fig. 1A). CT angiography showed venous dilatation in the anterior portion of ICH (Fig. 1B).

Conventional and 3D rotational angiogram defined a large DVA collecting numerous small veins, contributing to the caput medusae shape in the venous phase. But the main draining vein of DVA did not drain into the superior sagittal sinus (SSS) and thrombosis was found in several disconnected areas regarding the SSS (Fig. 2A). Evaluation for hypercoagulable state was performed, the blood laboratory studies for thrombosis were either negative or within normal range: antiphospholipid, antinuclear and anticardiolipin antibodies, antithrombin III, lupus anticoagulant, factor V, protein C and S activity, and homocysteine. The patient had no history of oral contraceptive drugs. Immediate anticonvulsant therapy and intracranial pressure control were initiated. Due to the acute ICH, use of anticoagulants was of major concern. But with the assumption that without worsening of thrombosis, additional hemorrhage would not be a problem, systemic heparinization was started. This decision is based on the concept that anticoagulant therapy may be considered for venous sinus thrombosis with intracranial hemorrhage based on case series, although it has not been established anticoagulant is appropriate treatment for DVA thrombosis. Ten days later, follow-up cerebral angiography revealed the much decreased thrombosis and good drainage to the DVA (Fig. 2B). During use of anticoagulant, follow-up CT scan was performed at intervals of several days. Fortunately, the size of the hematoma was not increased and additional hemorrhage was not occurred (Fig. 3). The patient's neurological symptoms were gradually improved and she was discharged without any neurologic deficits. Anticoagulants and anticonvulsants were maintained for 6 months after discharge and neurologic changes or new symptoms were not observed after drug withdrawal. Initial brain magnetic resonance imaging (MRI) showed

* Corresponding author.

E-mail addresses: leecy009@hanmail.net, milelon@kyuh.ac.kr (C.Y. Lee).

<https://doi.org/10.1016/j.inat.2018.01.004>

Received 31 August 2017; Received in revised form 9 January 2018; Accepted 20 January 2018

2214-7519/ © 2018 The Authors. Published by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

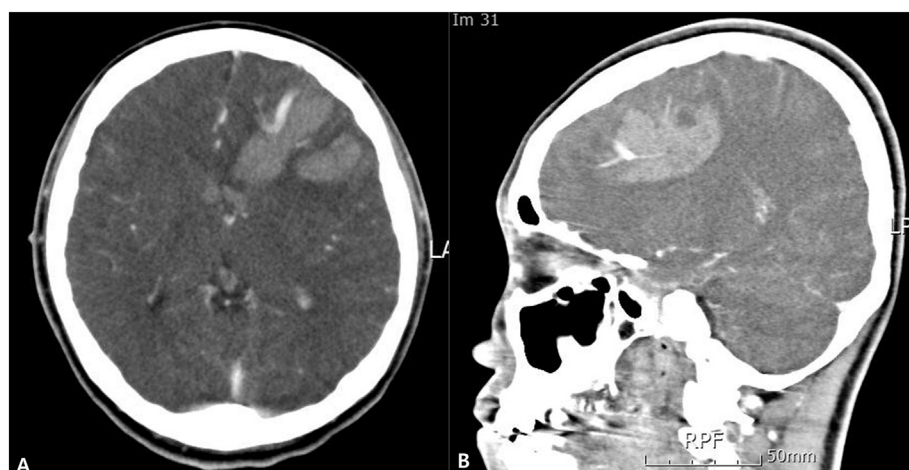


Fig. 1. A. Initial CT scan revealed ICH in the left frontal lobe and IVH.
B. CT angiography showed venous dilatation in anterior portion of ICH.

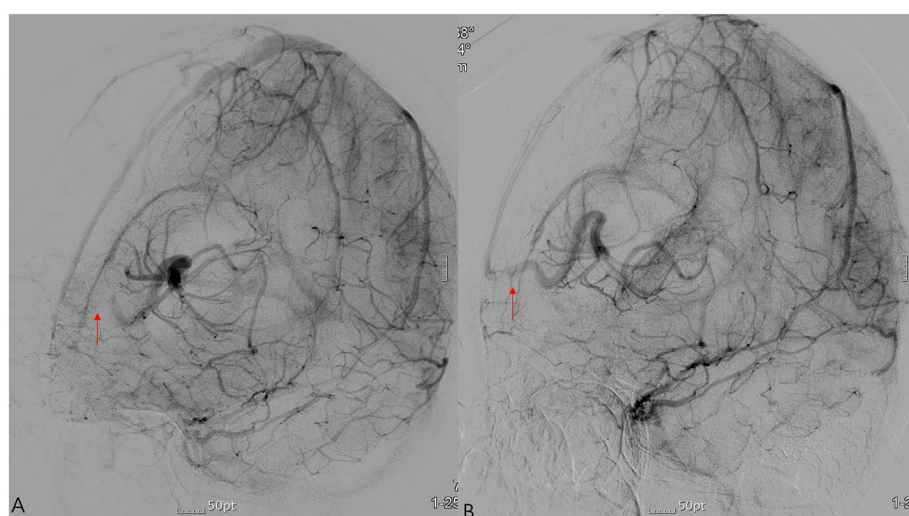


Fig. 2. A. Conventional and 3D rotational angiogram defined a large DVA collecting numerous small veins, contributing to the caput medusae shape in the venous phase. But the main draining vein of DVA did not drain into the superior sagittal sinus (SSS) and thrombosis was found in several disconnected areas regarding the SSS (red arrow).
B. Ten days later, follow-up cerebral angiography revealed that thrombosis in DVA was much decreased and DVA was connected to SSS and blood flow was well drained through DVA (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)



Fig. 3. Five days later after systemic heparinization, follow-up CT showed hematoma was not increased and additional hemorrhage was not occurred.

the same hemorrhagic findings as CT (Fig. 4A and B). There was no definite restriction, high signal in diffusion image (Fig. 4C). Although absence of restricted diffusion, venous infarction cannot be completely ruled out as a possible etiologic component of dominant hemorrhagic pathology. But we assumed that, if the hemorrhage had been secondary to transformation of the initial infarction, there would have been signs of infarctions in the original site, but in this patient, there was no such evidence. The opinion of the radiologist was the same as ours. In CT after 5 months, only small cerebromalatic change was visible in previous hematoma area and there was no evidence of cerebral infarction (Fig. 4D). If the hemorrhage were secondary to cerebral venous infarction, there would have been a broader cerebromalatic change involving cortical and subcortical area in follow up CT after 5 months. These findings suggest that it was a pure ICH without venous infarction rather than hemorrhagic transformation in infarcted area.

3. Discussion

DVAs represent a dilatation of a cerebral draining vein, which acts as a collector for small venules; DVAs are often found incidentally on examination and are generally considered as benign congenital vascular malformations, present in approximately 2.6% of the general population [4]. Generally, DVAs occur during the developmental process of the venous system, by retention of the primitive embryological medullary vein [5]. DVAs present a characteristic appearance on CT and MR studies, and seldom require cerebral angiography [6]. DVAs are rarely found in CT scans without contrast. After administration of

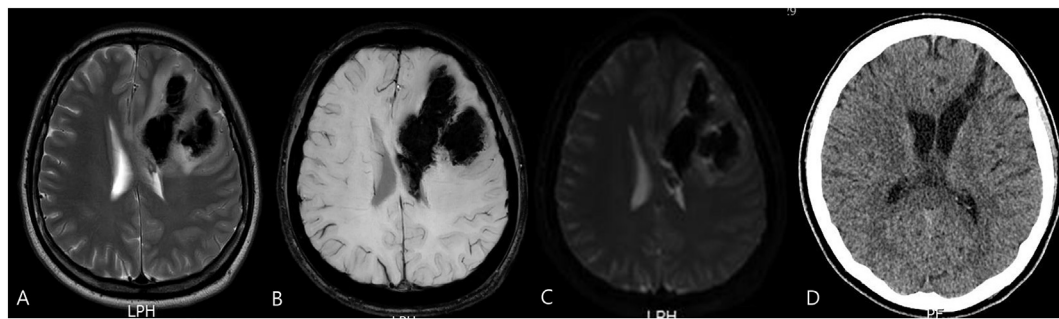


Fig. 4. A and B. T2 and susceptibility weighted imaging of magnetic resonance imaging showed the same hemorrhagic findings as CT.

C. There was no definite restriction, high signal in diffusion weighted image.

D. Follow-up CT after 5 months, only small cerebromalatic change was visible in previous hematoma area and there was no evidence of cerebral infarction.

contrast, the draining veins appear as linear focuses of enhancement. On brain MRI sequences, DVAs are displayed as flow void tubular images on both T1 and T2 weighted images. After intravenous administration of contrast, marked enhancement of the draining and medullary veins occurs, forming the typical caput medusae appearance [7]. On angiography, DVAs are found at the time of normal venous phase and acts as a vein that drains blood flow in normal brain and the underdevelopment of the usual draining pattern is clearly visible in most cases. DVAs may drain a cortical area toward the deep venous system or a deep area toward the superficial venous system [5]. Infarction of the territory drained by the DVA supports the fact that the DVA is essential for the venous drainage of the brain and that surgical resection should generally be avoided [8]. Thrombosis of DVA is a very rare phenomenon, and symptoms are rarely present. Most of symptomatic cases are associated with other vascular malformations, such as cavernomas or arteriovenous malformations (CMs or AVMs) or arterialized DVA [9]. Many authors suggest that prior to concluding that the cause of hemorrhage is DVA, it should first be looked through to see if there is any associated anomalies or other lesions such as CM, which may be more evident first on imaging studies. Another recent hypothesis suggests that most cavernous angiomas represent old hematomas secondary to a DVA, that could show autonomous growth by way of some angiogenic factors or repeated hemorrhages [8]. In our patient, there was no other associated vascular malformations such as CM or AVM within the territory of the DVA and hemorrhage. All the venules contributing to the caput medusae were located in, or near the hematoma. The above hypotheses are not always applicable. Stenosis at the dural opening of a DVA has been reported and may cause venous hypertension with secondary rupture or stagnation leading to thrombosis. Generally, infarction may be preceded by thrombosis and lead to hemorrhagic infarctions [3]. Infarction associated with DVA thrombosis is often reported, but pure cerebral hemorrhage is rarely reported without cerebral infarction. We have postulated that when the main venous anomalies were too large to hold the pressure during venous stasis in small venules, large ICH may occur before venous infarction. According to this mechanism, anticoagulation could be suggested for symptomatic DVAs despite ICH. But considering the rarity of this clinical presentation and the presence of another lesion potentially at risk for brain hemorrhage, anticoagulation therapy is not always appropriate. Guidelines for the treatment of DVA have not yet been established. In general, it is known that asymptomatic DVA does need treatment. Some authors recommend surgical removal if repeated hemorrhage has occurred [10]. But surgical resection of DVA will interfere with drainage of blood from adjacent brain parenchyma. Therefore, we consider surgical treatment will be dangerous, as fatal postoperative cerebral infarction has been reported [4]. A number of case series has reported favorable outcomes after anticoagulation therapy in acute infarction, particularly in instances of the draining vein associated with thrombus [3]. The use of anticoagulants in hemorrhagic lesions has a more favorable view and it should be judged appropriately according to the hemorrhage tendency,

the pathophysiology and the condition of the patient.

4. Conclusions

Most DVAs are benign lesions and are known to cause almost no symptoms. However, if DVA plays a major drainage role in the surrounding brain tissue, symptoms may occur if the drainage is interrupted, such as thrombosis. In few cases with thrombosis, it has been reported venous infarction occurs, which often result in secondary hemorrhagic formations. But if the main draining vein is too large, hemorrhage as main pathologic state can occur firstly because the venules cannot capacitate the pressure. Even with this hemorrhage, if the hemorrhage was caused by thrombosis of large drainage vein, we think the use of anticoagulants may be considered as treatment option. But further large studies are needed to assess the efficacy and safety and the optimal timing of systemic anticoagulation therapy for thrombosed DVAs.

Conflict of interest

None.

Sources of funding

None.

Acknowledgement

None.

References

- [1] J. Peltier, P. Toussaint, C. Desenclos, D. Le Gars, H. Deramond, Cerebral venous angioma of the pons complicated by nonhemorrhagic infarction. Case report, *J. Neurosurg.* 101 (2004) 690–693, <http://dx.doi.org/10.3171/jns.2004.101.4.0690>.
- [2] R. Töpper, E. Jürgens, J. Reul, A. Thron, Clinical significance of intracranial developmental venous anomalies, *J. Neurol. Neurosurg. Psychiatry* 67 (1999) 234–238, <http://dx.doi.org/10.1136/jnnp.67.2.234>.
- [3] C.L. Merten, H.O. Knitellus, J.P. Hedde, J. Assheuer, H. Bewermeyer, Intracerebral haemorrhage from a venous angioma following thrombosis of a draining vein, *Neuroradiology* 40 (1998) 15–18, <http://dx.doi.org/10.1007/s002340050530>.
- [4] S.K. Rammos, R. Maina, G. Lanzino, Developmental venous anomalies: current concepts and implications for management, *Neurosurgery* 65 (2009) 20–29, <http://dx.doi.org/10.1227/01.NEU.0000347091.06694.3E>.
- [5] A.V. Konan, J. Raymond, P. Bourgoin, J. Lesage, G. Milot, D. Roy, Cerebellar infarct caused by spontaneous thrombosis of a developmental venous anomaly of the posterior fossa, *Am. J. Neuroradiol.* 20 (1999) 256–258.
- [6] A. Zouaoui, J.C. Maillard, V. Ganthier, G. Chedid, S. Dangeard, Modern imaging in cerebral vein angioma, *J. Neuroradiol.* 22 (1995) 86–102.
- [7] C. Lee, M.A. Pennington, C.M. Kenney, MR evaluation of developmental venous anomalies: medullary venous anatomy of venous angiomas, *Am. J. Neuroradiol.* 17 (1996) 61–70.
- [8] P.W. McCormick, R.F. Spetzler, P.C. Johnson, B.P. Drayer, Cerebellar hemorrhage associated with capillary telangiectasia and venous angioma: a case report, *Surg. Neurol.* 39 (1993) 451–457, [http://dx.doi.org/10.1016/0090-3019\(93\)90030-5](http://dx.doi.org/10.1016/0090-3019(93)90030-5).

- [9] M. Walsh, H. Parmar, S.K. Mukherji, A. Mamourian, Developmental venous anomaly with symptomatic thrombosis of the draining vein, *J. Neurosurg.* 109 (2008) 1119–1122, <http://dx.doi.org/10.3171/JNS.2008.109.12.1119>.
- [10] G.M. Malik, J.K. Morgan, R.S. Boulos, J.I. Ausman, Venous angiomas: an underestimated cause of intracranial hemorrhage, *Surg. Neurol.* 30 (1988) 350–358, [http://dx.doi.org/10.1016/0090-3019\(88\)90197-8](http://dx.doi.org/10.1016/0090-3019(88)90197-8).