Reprinted Intracerebral Hemorrhage from Developmental Venous Anomaly Alone

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Developmental venous anomalies (DVAs) are hemodynamically low flow, low resistance vascular malformations without clinical significance. Although most DVAs are asymptomatic and are found incidentally, sometimes they can be symptomatic with intracerebral hemorrhage, many of which are usually caused by associated cavernous malformations (CMs) rather than the DVAs themselves. Only a few cases have been reported in the literature where an intracerebral hemorrhage has been caused by a DVA alone. This report describes a case of an intracerebral hemorrhage due to DVA alone with review of the literature.

KEY WORDS : Developmental venous anomaly · Cavernous malformation · Intracerebral hemorrhage.

INTRODUCTION

Developmental venous anomalies (DVAs), also known as venous angiomas or venous malformations, are the most common form of intracranial vascular malformations. They represent anatomically disordered but physiologically essential venous drainage of surrounding normal cerebral tissue within a functionally normal arterial territory. In one third of patients with DVAs, other coexisting cerebral vascular malformations are found, predominantly cavernous malformations (CMs). DVAs are clinically silent lesions, of which the symptomatic hemorrhage rate is very low, with an estimated incidence of 0.22-0.34% per year.7,17 Furthermore, most hemorrhages are usually caused by associated CMs rather than DVAs alone. This report describes an intracerebral hemorrhage caused by a DVA without associated CMs.

CASE REPORT

A 16-year-old young man presented with increasing headache intensity. He had been admitted to our hospital two months previously because of seizure. At that time, computed tomography (CT) and magnetic resonance imaging (MRI) of the brain revealed a homogenous, subacute staged hematoma localized in the right frontal region (Fig. 1A-D). Cerebral angiograms demonstrated the typical appearance of a DVA in the right frontal area (centrally located caput medusae of the lesion) without a cavernous malformation or any other associated vascular malformations (Fig. 2). He was treated conservatively because of the small volume of hematoma and the lack of neurological signs and symptoms. However, the follow-up brain imaging showed the increasing volume of hematoma with mass effect. The heterogeneous, multiple-staged hematoma signal intensity at the same site suggested repeated hemorrhage of DVA (Fig. 1E-H). During the surgery for hematoma removal, neither a cavernous malformation nor any other associated vascular malformations were seen. The histological studies disclosed dilated thin-walled vessels diffusely distributed in normal white matter with an organized old hemorrhage (Fig. 3). His symptoms were relieved after surgery. In postoperative CT, the hematoma was near totally removed (Fig. 4). The postoperative course was satisfactory and he was discharged 17 days after operation without sequelae.

DISCUSSION

Developmental venous anomalies (DVAs) are relatively
benign lesions with little clinical significance and develop in compensation during embryogenesis after occlusion or maldevelopment of the normal medullary veins by some accidents such as an intrauterine ischemic event\(^2\)). These anomalous venous channels may be structurally weaker than normal cerebral veins and it is plausible to postulate that DVAs have a high bleeding potential. There are some reports of intracerebral hemorrhage caused by a DVA itself\(^4,9,11,14,15,18\)). However, many authors assert that DVAs alone are rarely if at all bled\(^3,7,13,16\)).

In fact, the bleeding risk associated with DVAs is lower than other vascular malformations (approximately 0.22-0.34% per year\(^7,17\)) except DVAs in the posterior fossa or during pregnancy\(^13,18\)). Several authors insist that in the case of bleeding, the hemorrhage in the context of a DVA is likely to be related to an associated CM rather than the DVA itself\(^7,13,17\). Abdulrauf et al.\(^11\) compared the clinical profile of patients harboring CMs with or without associated DVAs and found that the risk of hemorrhage associated with a DVA was extremely low. They suggested that, in cases of hemorrhage, small associated

![Fig. 1. The upper four figures are computed tomography (CT) (A) and magnetic resonance (MR) images (B, C, and D) at first admission with symptoms such as headache and seizure. There is a subacute staged, homogenous round-shaped hemorrhage on the right frontal area. The lower four figures are CT (E) and MR images (F, G, and H) at second admission with increasing intensity of headache. There is a multiple staged, heterogenous hemorrhage on the same area.](image)

![Fig. 2. Cerebral angiograms demonstrate the typical appearance of a developmental venous anomaly on the right frontal area with centrally located caput medusae.](image)

![Fig. 3. Pathologic findings reveal the dilated thin-walled vessels diffusely distributed in normal white matter with lack of elastic fiber in vessel walls. A : H & E, × 40. B : H & E, × 100. C : Elastic fiber stain, × 400](image)
CMs might have been missed. Others have suggested the possibility that the hemorrhage destroyed the underlying CM, which could not be detected by pathologic examination.\(^7^,^{17}\) McLaughlin et al.\(^{17}\) asserted that the hemorrhage rate of a patient with a DVA was similar to the rates presented in previous reports for patients with CMs without previous symptomatic hemorrhage and it supported that the hemorrhage in patients with DVAs might be related to an underlying but not yet documented CMs. Kovácś et al.\(^{13}\) insisted that in cases of DVAs with bleeding, associated malformations should be thoroughly looked for. It is interesting that the CMs associated with a DVA have a more aggressive clinical course\(^{15,6,28}\) and are more likely to present with symptomatic hemorrhage than CMs alone\(^{30}\).

Abe et al.\(^2\) insisted that DVAs share a common origin and pathogenetic mechanism with CMs. It has been postulated that the abnormal hemodynamics of DVAs might induce the formation of CMs; certain DVAs produced local venous hypertension with resultant microhemorrhage, growth factor release, and creation of CMs\(^5,19\). Based on these hypotheses, DVAs can be regarded as the primary lesion leading to the occurrence of mixed vascular malformations. We believe these findings support the coexistence of a CM and a DVA which has been relatively commonly reported (2.1-23% of the occurrence rate)\(^21,22\). Although mixed vascular malformations of the brain have been thought to be uncommon, it can’t be elucidated that most hemorrhages are related with CMs rather than DVAs. Guclu et al.\(^8\) insisted that DVAs and CMs are two distinct entities with different pathogenetic mechanisms and that DVAs have a distinct biology and clinical behavior when compared to CMs using a gene study in a family whose members were affected by both disorders.

Because the risk of surgery appears to exceed that of the natural history of the disorder, many authors advocate conservative treatment. Kondziolkła et al.\(^{12}\) insisted that surgical removal or radiological obliteration of a DVA should not be considered unless a patient has a second life-threatening hemorrhage. Some authors advocate surgical resection for the treatment of surgically accessible DVAs, usually after intracerebral hemorrhage or for patients with recurrent hemorrhage\(^2,3,16\). Surgical resection of DVAs may frequently bring on a fatal venous infarction\(^{19}\). In order to prevent fatal complications, when surgical extirpation is needed, the DVA itself should be preserved intact, as the sparing of DVAs is not associated with rebleeding\(^{1,19}\). However, Lupret et al.\(^{15}\) reported that no severe cerebral edema was encountered despite the excision of converging medullary veins and the partial coagulation of the main draining veins.

In our case, magnetic resonance imaging (MRI) and cerebral angiograms revealed a repeated hemorrhage caused by the DVA without associated CMs. During the surgery, the small size of the DVA within the large hematoma had brought on the inevitable damage, but it did not lead to brain swelling or venous infarction. In spite of the thorough histological examination, no CMs or any other vascular anomalies were found.

**CONCLUSION**

Although DVAs very rarely cause intracerebral hemorrhage, we should keep in mind that DVAs might cause intracerebral hemorrhage without CM when an atypical intracerebral hemorrhage is diagnosed.

**References**


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*Fig. 4. Postoperative computed tomography scans revealing nearly total removal of the hematoma.*