

Developmental Venous Anomalies with Obstructive Venous Drainage Disorder Related Hemorrhage-is Endovascular Treatment Feasible? A Case Report and Literature Review

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1. Abstract

Developmental venous anomalies (DVAs) are widely deemed as anatomic variants of normal venous drainage with low hemorrhage risk. Hence, DVAs are deemed as benign and conservative management of DVA is strongly preferred. For the patients requiring surgical intervention, operation should focus on resection of the hematoma or cavernous angioma, with the DVAs protected. However, DVAs are associated with potential weaknesses due to their angioarchitectural characteristics. A case of DVA patient was followed and analysed. The present case illustrates catastrophic hemorrhagic venous infarct of DVA with obstructive venous drainage, and, to some extent, argues against the assumption that DVA is benign in nature. As a result, we propose that patients with "symptomatic" DVA should undergo MRI and DSA to exclude underlying pathology and clarify the specific structure of DVA, identifying the risk factors that might need intervention in advance, and the final treatment choice should be based on the specific characteristics of different DVA lesions.

2. Introduction

Developmental venous anomalies (DVAs), previously known as cerebral venous malformations, cerebral venous angiomas or cerebral venous medullary malformations, are the most common type of cerebral vascular malformation via autopsy studies (up to 60%

of all CVM) [1]. DVAs are one type of cerebrovascular malformation (CVM), sharing category with arteriovenous malformations (AVM), cavernous malformations (CM) and capillary telangiectasias. In general, Neurological symptoms or abnormal inspection results of DVAs are rare and usually attributed to hemorrhagic complications of coexisting cavernous malformations [2]. DVAs are considered to provide normal venous drainage for the cerebral territory where they reside [3], and are often accompanied by cavernous malformations or other vascular abnormalities, which might cause brain hemorrhage. It is widely believed that surgical excision or endovascular obliteration of DVAs carries high risk of venous infarction. Thus, up till now, for patients with DVAs, conservative management is the recommended choice [4]. Herein, we presented a DVA case with brain hemorrhage very likely caused by obstructive venous reflux disorder, owing to venous stenosis or thrombosis. And new idea about the treatment choice of DVA are put forward in this essay.

3. Case Description

A 20-year-old man presented to the emergency department with sudden severe headache without vomiting or limb dysfunction for 2 hours. PE: clear-minded, fluent in language and there were no positive neurological signs. CT imaging demonstrated hemorrhage in right lateral fissure area (Figure 1). And CT angiography

(CTA) revealed the existence of cerebrovascular malformation (Figure 2). Hence, brain digital subtracting angiography (DSA) was performed, showing that the venous phase of the right carotid artery exhibited a deep-type large DVA with typical "jellyfish head" changes (Figure 3), draining the right frontal-parietal area. The venous blood flow was drained to the right lateral sinus via the dilated drainage vein with multiple stenosed segments (Figure 3 B and D, white arrow). And the drainage vein was obviously narrow at the entering point of the lateral sinus (Figure 3 B, white arrow). Moreover, a thrombus or plaque could be seen in the curl of the drainage vein (Figure 3 B, red arrow), and obvious blood stasis was observed in the whole DVA. Given the lesion location of this DVA (central sulcus area) and the treatment principle (conservative treatment is recommended except the patients with com-

plications of hematoma or cavernous angioma). We recommended conservative treatment for this patient after communicating with his family, and magnetic resonance imaging (MRI) scan was recommended for him. But his family transferred the patient to another hospital. However, the patient suffered from sudden disturbance of consciousness the next day. CT scan showed that the right temporal occipital lobe hemorrhage increased significantly, broking into bilateral ventricle. And cerebral hernia occurred (Figure 4). Then bilateral ventricular drilling and drainage was performed in emergency, and craniotomy was performed. During the operation, no other types of accompanied cerebrovascular malformation was found, but the hemostasis of the bleeding DVA area was difficult. Finally, the patient died of excessive bleeding.

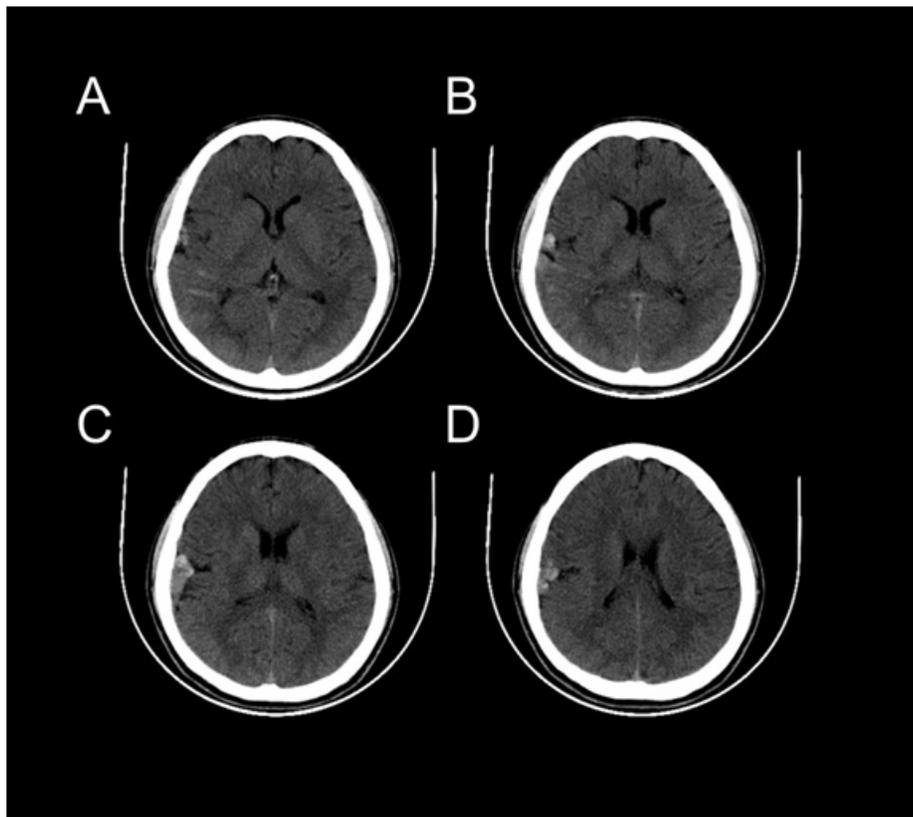


Figure 1: CT imaging demonstrated hemorrhage in the right frontal-parietal lobes

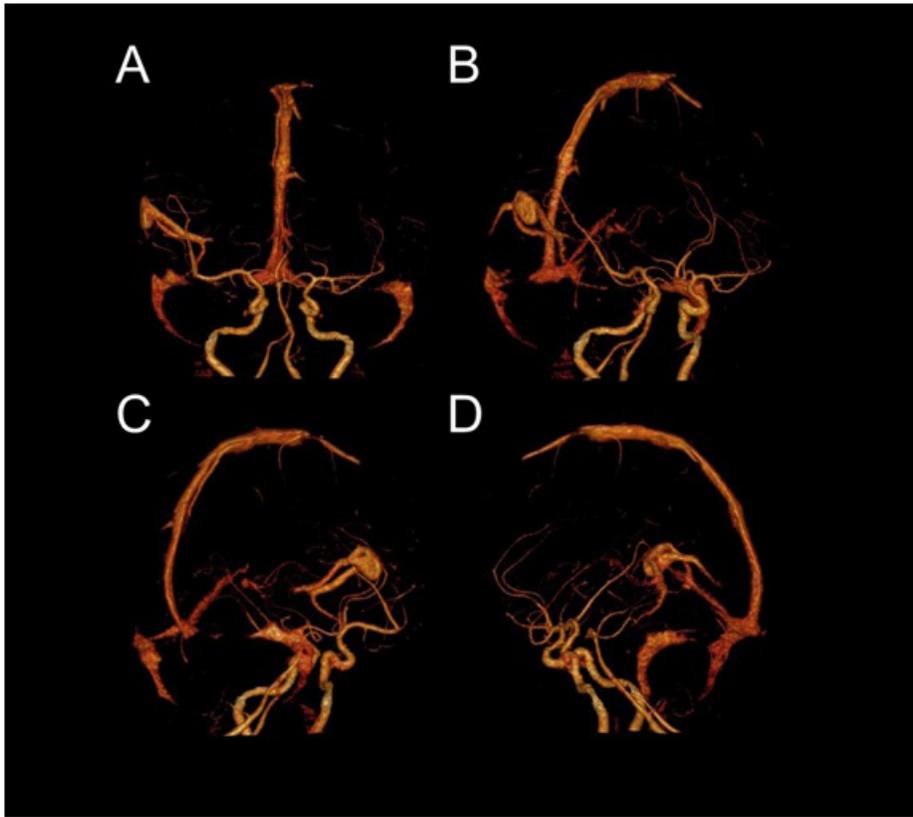


Figure 2: CTA revealed the existence of cerebrovascular malformed vessels draining to the lateral sinus

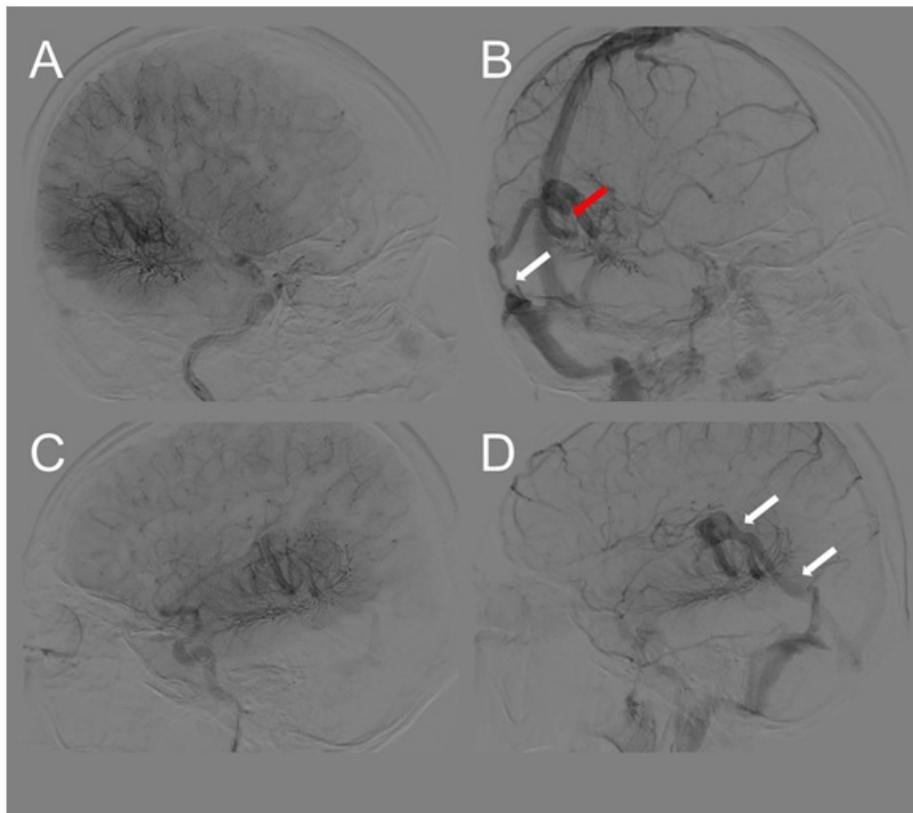


Figure 3: The venous phase of the right carotid artery showed a deep-type large DVA with typical "jellyfish head" changes, draining the right lateral fissure area. The venous blood flow was drained to the right lateral sinus through the drainage vein, which was dilated and with multiple stenosed segments(Figure 3 B and D, white arrow). The drainage vein was obviously narrow at the entering point of the lateral sinus (Figure 3 B, white arrow). And a highly suspicious thrombus can be seen in the curl of the drainage vein (Figure 3 B, red arrow). Obvious blood stasis was seen in the whole DVA.

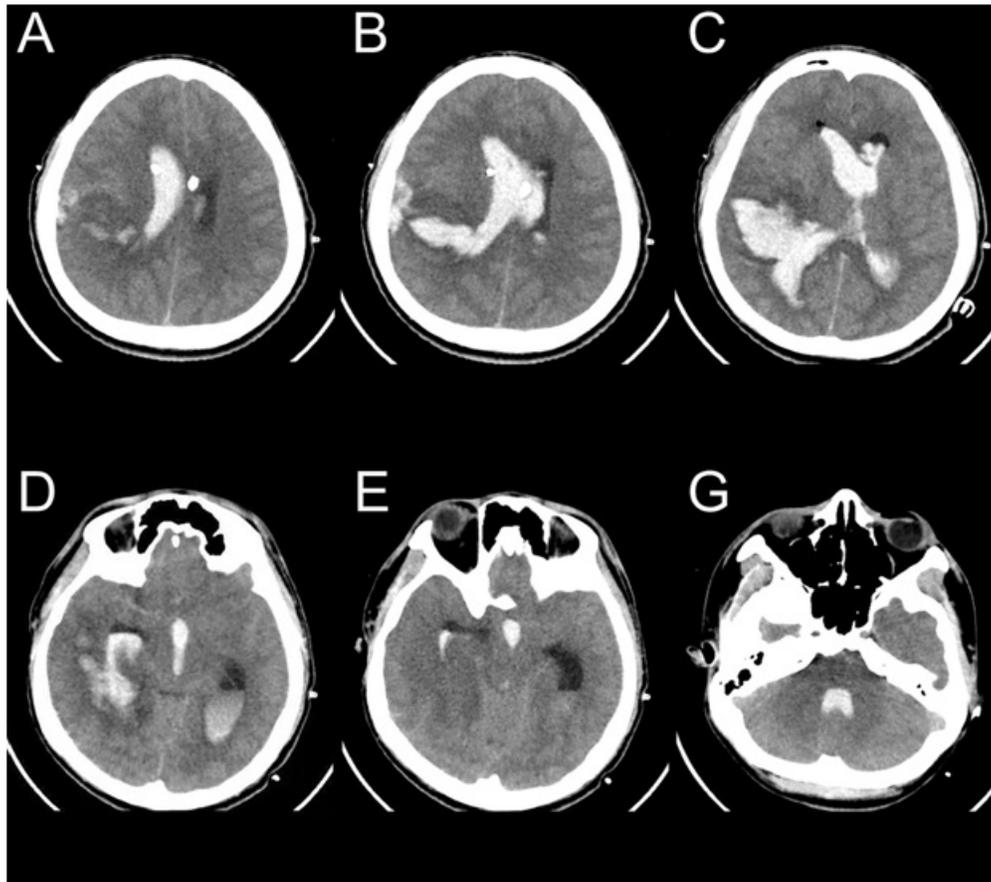


Figure 4: The CT scan showed that the right temporal occipital lobe hemorrhage increased significantly with a large area of infarction around the hematoma. And the hemotoma broke into ventricle, inducing cerebral hernia. Bilateral ventricular drilling and drainage was performed, but the decompression effect of intracranial pressure was unsatisfactory.

4. Discussion

DVAs are the most common form of brain vascular malformations with a reported incidence of up to 2.6% in 4069 brain autopsies, much more frequently encountered than AVMs [5]. Wolf, et al. reported the first patient who died of intracranial hemorrhage diagnosed as a venous angioma in 1967 [6]. Since then, venous lesions, e.g., the venous malformation, venous angioma, and medullary venous malformation came into our view. In 1986, Lasjaunias, et al. first suggested that, different from venous angiomas and vascular malformation, this kind of venous malformation should be deemed as normal anatomic variants, naming this variation as DVA. DVAs is consisted of radially oriented and converging dilated medullary veins, which drain centripetally into superficial subcortical or subependymal veins, forming the so-called caput medusae⁷. And the reported annual bleeding rate of 0.2% to 0.3% is likely due to bleeding from coexisting cavernous malformations. In this case, the CT scan on admission revealed intracranial hemorrhage in the right tempora. Moreover, the brain CTA manifested the cerebrovascular malformation of the same location. And DSA confirmed the cerebral venous malformation with blood stasis.

With the widely use of MRI and CTA, DVAs are now more frequently discovered⁸, although DVAs were usually discovered incidentally. DVAs are deemed as benign variation. But DVAs are

often accompanied by other types of cerebrovascular malformations, particularly cavernous angioma [9,10]. As a result, patients with "symptomatic" venous malformation should undergo MRI to ascertain underlying pathology. Meanwhile, it is believed that such associated pathology be treated independently and conservative management of DVA is strongly preferred. For patients requiring surgical intervention (e.g., patients with hemorrhage or cavernous angioma), operation should focus on resection of the hematoma or cavernous angioma, protecting the DVAs [2]. However, DVAs are more or less associated with potential weaknesses due to their angioarchitectural characteristics¹, hence there are still disputes remaining on its clinical management and prognosis.

It has been reported that brain parenchymal abnormalities and cerebral varix occurred surrounding DVA [1]. These phenomena is considered to be closely related to the venous hypertension associated with DVAs, which has been confirmed by perfusion study, demonstrating that some DVAs have venous congestion. For the lack of normal drainage veins, DVAs might have potential venous hypertension and be vulnerable to hemodynamic changes [10]. Moreover, DVA is frequently accompanied by other types of cerebrovascular malformations, which stretches the limit of this less flexible venous anomaly and result in intracerebral hemorrhage or infarction [10-12]. In this case, the venous blood flow was drained

to the right lateral sinus. Multiple venous stenosis could be observed at the turns of the dilated drainage vein (Figure 3 B and D, white arrow), especially at the entering point of the lateral sinus (Figure 3 B, white arrow). Moreover, a thrombus or plaque can be seen in the curl of the drainage vein (Figure 3 B, red arrow). and the patient's DSA showed a poor drainage in the late stage of the vein, and the contrast agent was retained.

There are some reports in support of our assumption. Kim, P. et al. reported cerebral venous malformation complicated by spontaneous thrombosis [13]. Kathryn Sepelyak, et al. recorded patient with thrombosis of a DVA with hemorrhagic venous infarction [14]. Kyung Sik Yi, et al. also reported a similar case, and combined the previous research [14,15] they believe that the venous congestion caused by thrombotic obstruction of DVA and draining vein should be the pathologic mechanism of the venous infarction and parenchymal hemorrhages [16]. Actually, when discussing this case, some of our chief physicians mentioned that the drainage vessel of DVA is tortuous and contains multiple stenosis, resulting in poor drainage and venous congestion. Interventional operation (e.g., balloon dilatation or stent implantation) or anticoagulation therapy should be considered to relieve venous stenosis and improve blood flow, reducing the risk of congestion bleeding. The second cerebral hemorrhage imaging showed that the right temporal lobe hemorrhage increased significantly and broke into the bilateral ventricle, unlike cavernous vascular malformation hemorrhage (Figure 4), although the MRI scan was absent. Moreover, there were large areas of infarction around the hematoma (Figure 4). And other type of accompanied cerebrovascular malformation was not found during the operation. As a result, it is high likely that the stenosis of the drainage vein resulted in cerebral infarction hemorrhage.

Therefore, it is necessary to discuss the treatment choice of this case. Surgery is only indicated in patients with massive intracerebral hematoma, aiming at resection of the hematoma or cavernous angioma and with CVMs preserved [2]. Moreover, since DVAs drain normal brain tissue, radiosurgery is contraindicated. Is it suitable for us to perform endovascular treatment (e.g., balloon dilatation or stent implantation) to mitigate vascular stenosis and relieve blood stasis? We hold the opinion that the final choice should be based on the specific characteristics of different DVA lesions. For instance, for the DVAs that rarely cause obstructive hydrocephalus, Claudio Cavallo, et al. demonstrated ETV as an effective treatment in the management of obstructive hydrocephalus [17]. Dayna Griffiths, et al. reported a case of a thrombosed DVA with venous congestion and pontine hemorrhage that improved after anticoagulation treatment [18]. After the study of 6 cases, R. Assadsangabi, et al. believe that early recognition of thrombosed DVA as the underlying cause of the venous infarction and intracranial hemorrhage could lead to the appropriate anticoagulation therapy [19]. Moreover, Yukio Seki shared the same opinion with

us: venous malformations occasionally become symptomatic due to thrombosis of the draining vein, and the stenosis in the draining route might result in thrombus formation, venous congestion, or even catastrophic hemorrhagic venous infarct [20]. However, the current diagnostic and therapeutic principles consider it a benign variant, which does not need treatment itself. Therefore, for this case, our department did not carry out endovascular treatment or anticoagulation therapy without supportive treatment guideline.

5. Conclusion

DVAs are widely deemed as anatomic variants of normal venous drainage and the hemorrhage risk is low. Hence, in general, they are benign and conservative management of DVA is strongly preferred. For the patients requiring surgical intervention, operation should focus on resection of the hematoma or cavernous angioma, with the DVAs protected. However, DVAs are associated with potential weaknesses due to their angioarchitectural characteristics, hence we propose that the final treatment choice should be based on the specific characteristics of different DVA lesions. The present case illustrates a potential complication of DVA and, to some extent, argues against the assumption that DVA is benign in nature. As a result, patients with "symptomatic" DVA should undergo MRI and DSA to exclude underlying pathology and clarify the specific structure of DVA, identifying the risk factors that might need intervention in advance. Certainly, more research is necessary to refine the principles of treatment.

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