Case Report

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대한영상의학회지

Intracerebral Hemorrhage Caused by Thrombosis of a Developmental Venous Anomaly with an Unusual Structure: A Case Report

특이한 구조의 뇌정맥발달기형 내 혈전증에 의해 생긴 뇌출혈: 증례 보고

Sunhyang Lee, MD[1], Dae Yoon Kim, MD[1]*, Mi Kyung Kim, MD[2], Hyun Jin Kim, MD[1]

Departments of [1]Radiology and [2]Neurosurgery, Daejin Medical Center Bundang Jesaeng General Hospital, Seongnam, Korea

Developmental venous anomalies (DVAs) are common intracranial vascular malformations and they are generally do not cause clinical complications. In cases showing DVA and hemorrhage, the hemorrhage is usually associated with adjacent cavernous malformations. Very few cases of intracerebral hemorrhage (ICH) caused by thrombosis in DVA have been reported in the literature. In this case report, we present an interesting case of a large ICH caused by thrombosis within a DVA with an unusual structure that may have potentiated the thrombosis.

Index terms Cerebral Venous Angioma; Intracerebral Hemorrhage; Thrombosis; Stenosis; Magnetic Resonance Imaging

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***Corresponding author**
Dae Yoon Kim, MD
Department of Radiology, Daejin Medical Center
Bundang Jesaeng General Hospital,
23 Hwangsaeul-ro 341beon-gil,
Sungnam 13590, Korea.

Tel 82-31-779-0000
Fax 82-31-779-0062
E-mail hapkitar@naver.com

ORCID iDs
Sunhyang Lee
https://orcid.org/0000-0002-4817-0231
Dae Yoon Kim
https://orcid.org/0000-0001-5415-2614
Mi Kyung Kim
https://orcid.org/0000-0002-8824-0505
Hyun Jin Kim
https://orcid.org/0000-0002-3411-3690

INTRODUCTION

Developmental venous anomaly (DVA), previously known as cerebral venous angioma, is a common congenital intracranial vascular malformation that is observed in up to 2.5% of the general population (1). Despite the high incidence of DVAs, they rarely cause complications (2). Hemorrhage is often correlated with combined cavernous malformations rather than with DVA itself (3). Herein, we present a case of a large intracerebral hemorrhage (ICH) caused by thrombosis within a DVA.

CASE REPORT

A 48-year-old female presented to the emergency department with a sudden onset of severe headache, nausea, and a lowered consciousness level. Neurological examination showed grade 2 weakness in the right upper and lower extremities. Laboratory test results were within normal range, including coagulation factors, antiphospholipid antibody, prothrombin time and partial thromboplastin time.

CT revealed an approximately 6-cm ICH at an unusual site adjacent to diffuse low attenuation portions in the left occipital lobe, mimicking a left posterior cerebellar artery territory infarction (Fig. 1A). Subsequent MRI revealed different stages of hematomas in the left occipital lobe with perilesional edema, which corresponded with the CT's low attenuation change (Fig. 1B-D). There was no diffusion restriction within the lesion (data not shown). Herniation of the left lingual gyrus was observed. Additionally, parallel thin T2 signal voids formed by the congested veins traversed into the swollen left occipital lobe. Contrast-enhanced T1 weighted MRI also revealed dilated medullary veins, intermediate collector veins, and filling defects within it. We hypothesized that the thrombus within the DVA resulted in venous congestion, secondary cerebral edema, and hemorrhage. There were no other possible causes of hemorrhage around the ICH.

Two hours after hospitalization, the patient experienced acute neurological deterioration and underwent decompressive craniotomy, followed by evacuation of the hematoma without surgical resection of the DVA.

Follow-up transfemoral cerebral angiography 2 weeks after the craniotomy showed delayed appearance of the large collector veins of the DVA and partial filling defect (Fig. 1F). DVA was unveiled to have a peculiar architecture with several intermediate collector veins

Fig. 1. A 48-year-old female showing a large ICH with thrombosis within a developmental venous anomaly.
A. Axial non-contrast brain CT scan obtained on admission. Note the extensive ICH (black arrows) in the left occipitotemporal lobe. A segmental low-attenuation area (empty arrows) mimics left posterior cerebral artery infarction. The multifocal linear hyperdense areas (right image, arrowheads) were confirmed as thrombosed collector veins on MR angiography (MRI; black arrowheads in D).
ICH = intracerebral hemorrhage

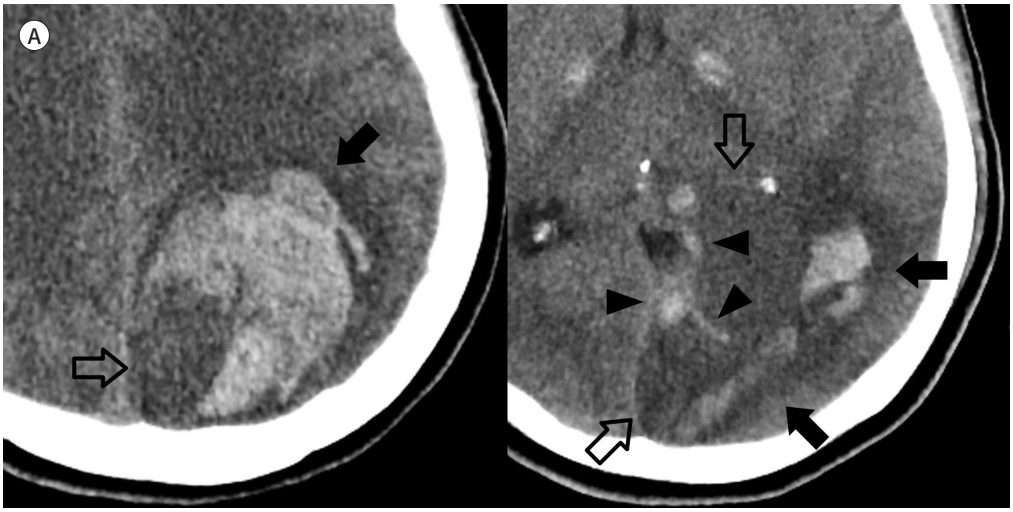
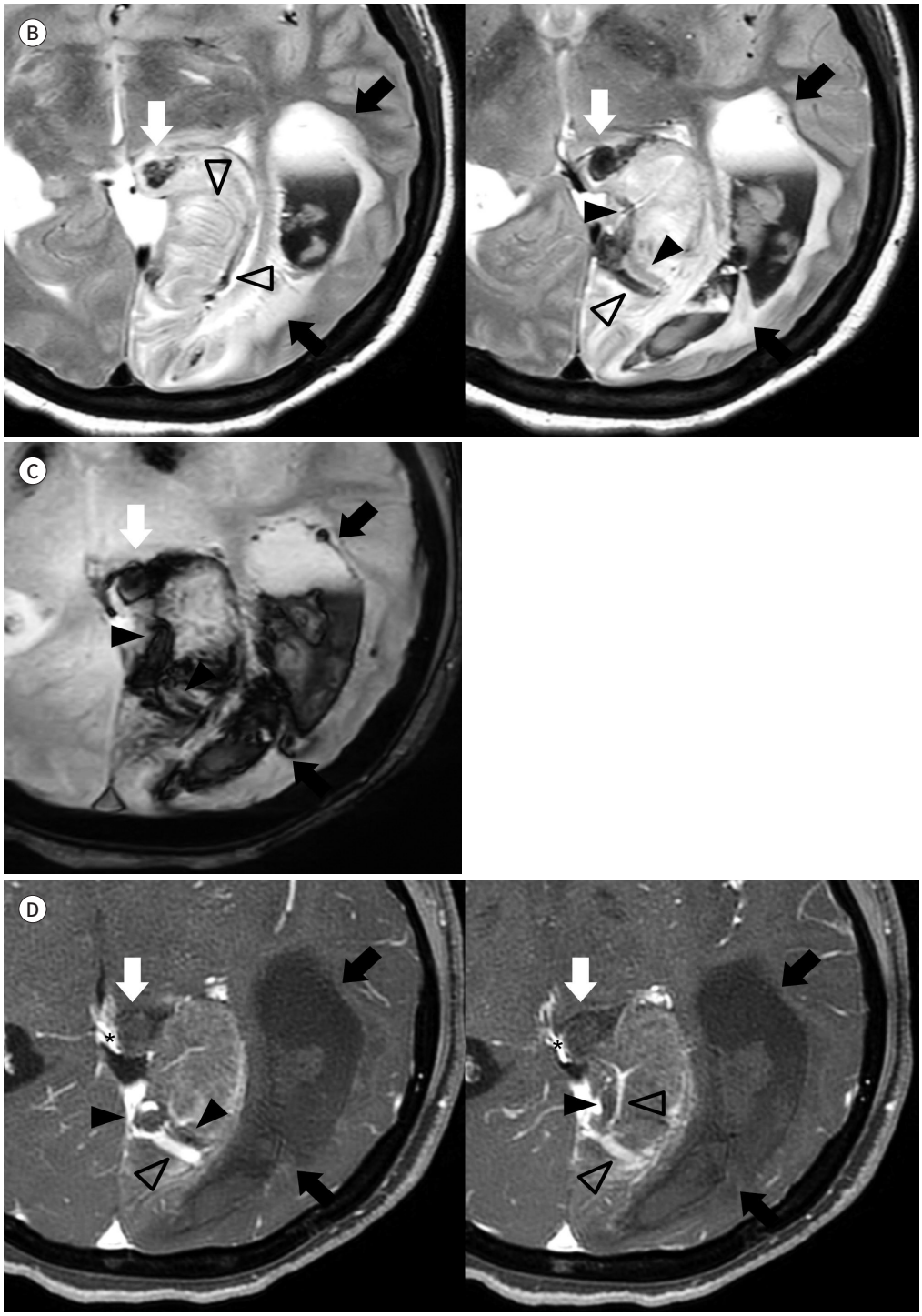
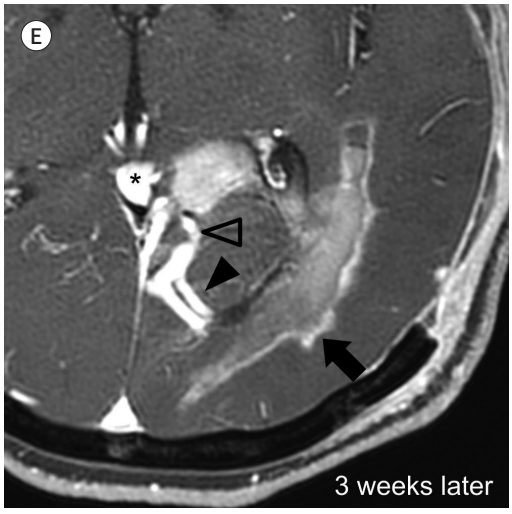


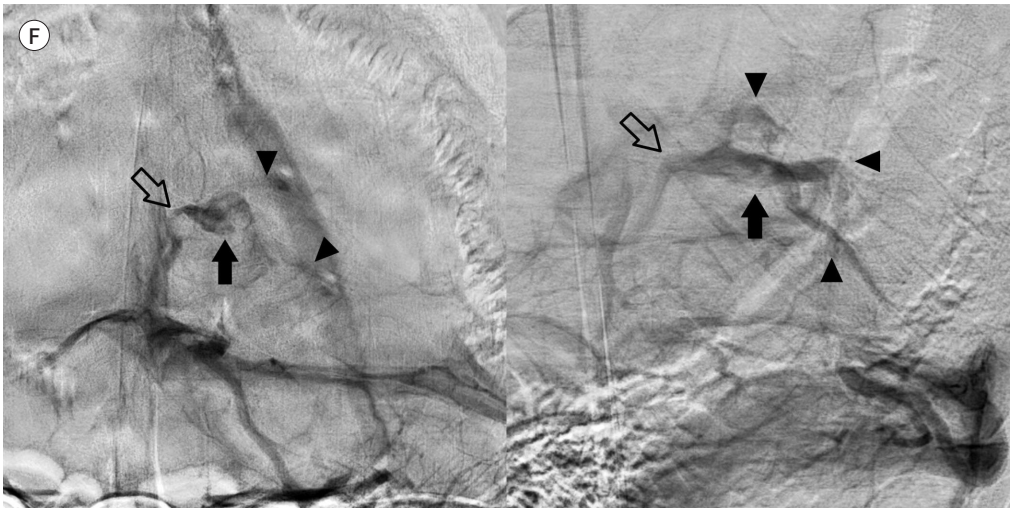
Fig. 1. A 48-year-old female showing a large ICH with thrombosis within a developmental venous anomaly.
B-D. Axial T2-weighted images (B), T2 fast-field echo image (C), and post-contrast images (D) on admission. Marked swelling and ICHs (black arrows) are seen in the left occipital lobe, correlating with the CT findings. Note the signal-void vascular structures of the medullary veins, intermediate collector veins, and a single confluent collector vein (empty arrowheads in B, D) traversing the swollen left occipital lobe and multifocal thrombosis (black arrowheads) in the collector veins as filling defects, causing occlusion and partial obstruction. The findings also show left lingual gyrus herniation (white arrows), leading to the collapse of the vein of Galen (asterisks in D).
ICH = intracerebral hemorrhage



and a single confluent collector vein, as observed on MRI. A week later, MRI showed complete disappearance of the DVA thrombus, reappearance of the intermediate collector veins, and restoration of the outlet size of the confluent collector vein, which seemed to have an underlying segmental narrowing in the distal portion (Fig. 1E). The vein of Galen was also released from herniation of the lingual gyrus. There was also improvement in brain edema

Fig. 1. A 48-year-old female showing a large ICH with thrombosis within a developmental venous anomaly.
E. Axial post-contrast MR images obtained 3 weeks after hospitalization demonstrates complete loss of venous thrombosis, reappearance of the collector veins (black arrowhead, in comparison with the left image of D), and increased size of the DVA outlet, while still showing residual stenosis at the outlet of the confluent collector vein (empty arrowhead, in comparison with the right image of D). Note the sequential improvement of the ICH (black arrow, in comparison with C) and lingual gyrus herniation, resulting in recovery of the vein of Galen (asterisk).
F. Digital subtraction angiography scans in the anteroposterior (left image) and lateral (right image) views obtained 2 weeks after the craniotomy show delayed appearance of a couple of intermediate collecting veins (black arrowheads) and a fusiform confluent collector vein (black arrows) with a remnant filling defect. The intrinsic stenosis of the confluent collector vein outlet and superimposed collapse (empty arrows, corresponding MR image not shown here) by lingual gyrus herniation are visible. This DVA seems to drain venous flow to the vein of Galen. Note that the caudal end of the contrast bolus is passing the sigmoid sinus normally, suggesting a delayed venous phase.
DVA = developmental venous anomaly, ICH = intracerebral hemorrhage





and sequential evolution of the residual ICH.

The patient was started on anticoagulant treatment with once-daily administration of edoxaban (30 mg), 18 days after partial removal of the hematoma. Thirty days after the surgery, the patient fully regained motor functions of the right extremities; however, she had remaining right homonymous hemianopsia and motor aphasia. She had no recurrent ICH or further neurologic deterioration with physical therapy and ongoing anticoagulation therapy.

This case report was approved by the Institutional Review Board of our institution (IRB No. 2021-01-008) and the requirement for informed consent was waived.

DISCUSSION

DVAs are characterized by brain-parenchyma-traversing dilated medullary veins and draining collector veins, composed of mature thin-walled venous channels (4). A recent meta-analysis showed that the hemorrhagic risk of DVA was less than 0.7% (5). Another population-based study of 93 adults with incidental DVAs demonstrated no symptomatic hemorrhages or infarcts in 492 person-years of follow-up (2).

Most cases of hemorrhage and DVA are related to adjacent cavernous malformations (3). Our literature review showed that the cause of hemorrhagic DVAs without associated vascular malformations is related to acute thrombosis of the DVA (5-8). Thrombosis can be caused spontaneously or by stenosis of the outlet collector vein (9). Theoretically, Virchow's triad describes three factors that contribute to the development of venous thrombosis: 1) hypercoagulability, 2) stasis, and 3) endothelial injury; deep vein thrombosis is usually caused by a combination of these factors.

Idiculla et al. (5) explained the pathophysiology of hemorrhagic DVAs as an imbalance between inflow and decreased outflow likely because of sudden thrombus formation with or without outlet stenosis of the DVA, that results in a rapidly increased internal pressure of the collector veins and consequent hemorrhage. They showed that venous drainage disturbance caused cerebral edema and herniation, that led to further compromise of the intracerebral and extracerebral veins, which exacerbated venous drainage. This vicious circle between venous edema and venous compromise seemed to have played a critical role in DVA hemorrhage in our case. In our patient, the vein of Galen and parenchymal veins were compromised and displaced by lingual gyrus herniation and cerebral edema, respectively.

DVAs may have a vulnerable anatomic architecture for thrombosis (10). DVA usually consists of medullary veins that converge into a collecting vein (5). In contrast, the DVA structure in our case was peculiar, in that medullary veins were gathered into a couple of intermediate collector veins that converged into a confluent collector vein. Segmental narrowing was observed in the confluent collector vein. Venous flow stasis due to this vulnerable structure and combined stenosis of the distal collector vein of the DVA potentially led to an increased risk of venous thrombosis.

As specified by the triad of Virchow, another independent but possible mechanism for DVA thrombosis is microscopic degenerative endothelial changes in the DVA. Rammos et al. (10) reported that secondary changes in the thickened and hyalinized vessel walls could lead to a propensity for thrombosis, although most DVAs are inherently composed of normal endothe-

lium with mature thin-walled venous channels. DVAs contribute to normal venous drainage of the corresponding area as they traverse the brain parenchyma. In our case, because the large DVA covered most of the territory of the left occipital lobe, it could result in a large amount of ICH.

In our case, anticoagulation therapy was effective for thrombus reduction within DVA. The venous thrombosis tendency of DVA remained in terms of the peculiar architecture with multiple intermediate collecting veins and outlet confluent collector vein stenosis. Considering the normal venous drainage role of DVAs, DVA resection should be avoided to prevent catastrophic venous drainage disturbances (9).

Although most DVAs show benign clinical courses and DVAs themselves rarely cause ICH, we need to keep in mind that DVAs with vulnerable architecture or outlet stenosis can increase the potential risk of thrombosis and lead to subsequent ICH. Especially in the case of thrombosis in a large DVA, which is generally responsible for a wide range of normal venous drainage, the ICH could be quite catastrophic.

Author Contributions

Conceptualization, all authors; investigation, K.D.Y., L.S.; project administration, K.D.Y., K.M.K., L.S.; supervision, K.D.Y., K.M.K., K.H.J.; visualization, K.D.Y., L.S.; writing—original draft, K.D.Y., K.M.K., L.S.; and writing—review & editing, K.D.Y., K.M.K., L.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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특이한 구조의 뇌정맥발달기형 내 혈전증에 의해 생긴 뇌출혈: 증례 보고

이선향[1] · 김대윤[1*] · 김미경[2] · 김현진[1]

뇌정맥발달기형은 일반적으로 증상을 유발하지 않는 흔한 두개 내 혈관 기형이다. 뇌정맥발달기형과 관련된 출혈은 동반된 해면상 기형이 원인인 경우가 대부분인 것으로 알려져 있으며, 뇌정맥발달기형 내 혈전증이 뇌출혈을 일으킨 경우는 극히 드물게 보고되어 있다. 저자들은 혈전증을 유발할 수 있을 것으로 보이는 특이한 구조의 뇌정맥발달기형을 가진 환자에서 혈전증과 큰 뇌출혈이 생긴 1예를 경험하였기에 컴퓨터단층촬영 소견과 자기공명영상 소견을 보고하고자 한다.

대진의료재단 분당제생병원 [1]영상의학과, [2]신경외과

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