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Case Report

Idiopathic cerebellar hemorrhage in a patient with isolated developmental venous anomaly: A case report

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Received: 08 December 2024
Accepted: 10 February 2025
Published: 07 March 2025

DOI
10.25259/SNI_1056_2024

Quick Response Code:

ABSTRACT

Background: Developmental venous anomalies (DVAs) are benign anatomical variations in venous angioarchitecture. They are considered low-flow malformations and are often incidental and clinically insignificant. Hemorrhagic complications from isolated DVAs are extremely rare, typically occurring due to coexisting cavernous malformation.

Case Description: A 33-year-old female presented with severe headaches and vertigo, progressing from balance issues and dizziness. Misdiagnosed initially as Meniere's disease, her symptoms included left oculomotor nerve palsy, left-sided hemidysmetria, dysdiadochokinesis, and positive Romberg's sign. Cranial computed tomography revealed an acute hemorrhage in the cerebellar vermis. Contrast-enhanced magnetic resonance imaging and angiography identified an isolated DVA. The absence of substantial mass effect or obstructive hydrocephalus prompted conservative management with steroids and analgesics, leading to full recovery.

Conclusion: This case underscores the importance of considering isolated DVA in cerebellar hemorrhage etiology in young adults and conservative treatment is recommended to prevent secondary complications, given the DVA's role in normal venous drainage.

Keywords: Cerebellar hemorrhage, Developmental venous anomaly, Developmental venous anomaly-related hemorrhage, Vascular malformation

INTRODUCTİON

A developmental venous anomaly (DVA) is a benign anatomical variation in the venous angioarchitecture, consisting of clusters of small veins draining into a larger collector vein that either drains into the deep or superficial cerebral venous system.[11] DVAs are commonly described as incidental findings without clinical significance and are considered low-flow malformations.[13] A distinctive "caput medusae" sign, seen on enhanced radiologic examination, is characterized by dilated medullary veins conversing on a transcortical collecting vein.[3] Since hemorrhagic complications are most likely due to a coexisting cavernous malformation (CM) or arteriovenous malformation (AVM), it is very rare for an isolated DVA to hemorrhage.[1,7] In addition, bleeding has mostly been reported as intra-parenchymal in most cases.[5] Here, we present an extremely rare case of symptomatic isolated DVA-related cerebellar hematoma with fourth ventricle extension, along with its management.

CASE REPORT

A 33-year-old female patient was admitted to our emergency department with severe headaches and vertigo. Her symptoms began a week ago with balance problems and dizziness, followed by headaches. Her condition worsened after receiving treatment for vertigo, which had started 3 days ago following a misdiagnosis of Meniere's disease.

Upon admission, she exhibited left oculomotor nerve palsy. Her visual acuity was normal, and no retinal hemorrhage, venous dilatation, or fundus tortuosity were observed. Examination of the cerebellum revealed left-sided hemidysmetria and dysdiadochokinesis with prolonged duration and increased oscillation, yet she was still able to reach the target. Romberg's sign was positive, indicating primarily anterioposterior instability.

Cranial computed tomography (CT) showed an acute hematoma at the cerebellar vermis, measuring 18 × 24 × 35 mm, with minimal extension into the fourth ventricle [Figure 1a]. Contrast-enhanced magnetic resonance imaging (MRI) of the brain revealed a cerebellar DVA in the posterior aspect of the hematoma with the "caput medusae" sign [Figure 1b]. The collector veins of the DVA from the fastigium cerebelli drain through a large vein within the fourth ventricle into the Galenic system. No other vascular malformation or CM was detected on digital subtraction angiography (DSA) [Figure 1c].

Considering the patient's clinical status, the absence of substantial mass effect or obstructive hydrocephalus, and the direct draining pattern of DVA into the main deep cerebral venous system, we decided against surgical intervention. The patient was hospitalized, and symptomatic treatment was initiated immediately with steroids and analgesics. Her symptoms gradually improved over the next 10 days, and she fully recovered. At the follow-up examination, the only radiological change detected was leukomalacia after 1 year [Figure 1d].

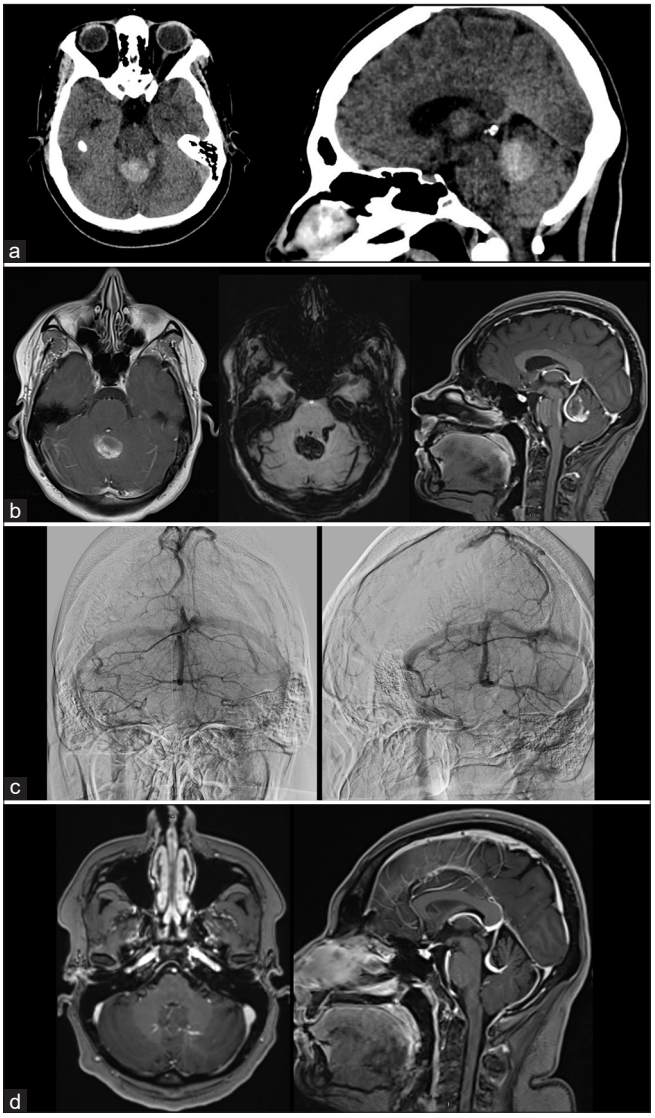

Figure 1: (a) Brain computed tomography scans in axial and sagittal sections showing an acute hematoma on the cerebellar vermis, measuring 18 × 24 × 35 mm, with minimal extension into the perimesencephalic cistern. (b) Magnetic resonance imaging (MRI) scans: Axial T1-weighted MRI with contrast enhancement and susceptibility-weighted imaging demonstrating several adjacent linear flow voids and a curvilinear collecting vein indicative of a developmental venous anomaly (DVA), known as the "caput medusae" sign. Sagittal T1-weighted MRI with contrast enhancement showing a dilated collector vein of DVA through into the vein of Galen. (c) Digital subtraction angiography images: Anteroposterior and oblique/lateral views revealing dilated collector veins with the absence of a feeding artery. The DVA drains from the fastigium cerebelli through a fourth ventricular vein into the vein of Galen, clearly shown in the delayed phase of the angiographic injection. (d) T1-weighted MRI with contrast enhancement in axial and sagittal sections, showing degenerative sequelae with hemorrhage-related signal changes manifesting as leukomalacia.

DISCUSSION

The most common localization of DVA is supratentorial, predominantly in the frontal region.[13] The risk of hemorrhage from an isolated DVA is considered extremely low unless there is a coexisting CM or AVM within the drainage territory of the DVA or venous outflow restriction of DVA.[2,8] Furthermore, in about 10–40% of cases, DVAs are frequently observed in conjunction with CM.[2,8] Therefore, neurologic symptoms or abnormal examination findings are usually attributed to DVA's complications, most likely from this coexisting CM.[1,7]

The risk of hemorrhage associated with DVAs has been reported as 0.22–0.68%/year, with acute spontaneous

intracerebral hemorrhages being the primary cause.[7] Compared to supratentorial DVAs, infratentorial DVAs have a higher risk of bleeding.[4,10] These hemorrhages are most commonly described as intra-parenchymal.[5]

Pereira *et al*. reported that 17 out of 80 presented with vascular complications directly attributed to an isolated DVA.[9] Kurt *et al*. documented a 21.8% hemorrhage rate (22/101) in the vicinity of DVAs. Among these hemorrhages, 45.5% (10/22) were caused by isolated DVAs, while the remaining 45.5% were associated with a CM, one with a hemangioma, and one with a tumor.[6] However, the study did not differentiate between supratentorial and infratentorial isolated DVA-related hemorrhages. Given these findings, careful long-term follow-up is crucial in patients with unexplained cerebellar hemorrhage due to DVA, even when initial imaging fails to reveal an associated CM, as delayed detection remains a possibility.[8]

Isolated DVA-related intracranial hemorrhage can occur due to increased inflow through arteriovenous micro shunts where the DVA acts like an AVM, decreased outflow due to obstruction or thrombosis of the collecting vein, or idiopathically.[9] In addition, some authors have defined arterialized DVAs, where a DVA appears on an angiogram during the arterial phase, accompanied by numerous arteriovenous communications with capillary blush, as exhibiting clinical behavior similar to AVMs, which carry a higher risk of hemorrhage than classic DVAs.[5] Our case, however, involves a nonarterialized DVA, although hemorrhage still occurred. We have classified our patient as an idiopathic case of DVA-related cerebellar hemorrhage. No CM or AVM was detected, and thrombosis of the DVA was ruled out through radiologic evaluations.

DVA-related intracranial hemorrhage typically occurs between the medullary veins and the collecting veins of the DVA, often following a benign clinical course.[7] It becomes symptomatic through persistent flow-related complications or mechanical compression of the outflow vein, resulting in obstructive hydrocephalus or neurovascular nerve compression syndrome.[9] In our patient, severe headaches and left oculomotor nerve palsy, along with cerebellar signs such as vertigo, left-sided hemidysmetria, dysdiadochokinesis, and a positive Romberg's sign, were attributed to the location of the hemorrhage.

Since DVAs serve as alternate compensatory venous drainage pathways for normal parenchyma in the absence of territorial habitual venous drainage, surgery should be reserved for life-threatening mass effects or raised intracranial pressure.[3] The primary aims of surgery are decompression and relieving mass effect, with all efforts made to preserve the main collecting vein to avoid venous infarction.[3,13] Therefore, we elected conservative management since no arteriovenous shunting was appreciated on DSA.

The primary cause of cerebellar hemorrhage in older adults is typically hypertension, whereas AVM is more common in younger individuals.[12] Other causes include trauma, coagulopathy, ischemic stroke with hemorrhagic transformation, cerebral amyloid angiopathy, septic embolism, encephalitis, vasculitis, sympathomimetic drugs, tumors, supratentorial surgery, aneurysm of the posterior cerebral circulation, and other vascular malformations such as dural arteriovenous fistula and CM.[12] While cerebral DVAs are generally considered benign, several studies have attempted to highlight the potential for isolated DVAs to hemorrhage and cause significant clinical symptoms.[6] An isolated DVA itself must be considered among the list of primary causes of intracranial hemorrhage.

CONCLUSION

Herein, we present a case of symptomatic DVA-related cerebellar hematoma associated with headaches, vertigo, and oculomotor nerve palsy without a coexisting CM. When considering the etiology of a cerebellar hemorrhage, especially in young adults, clinicians should suspect DVA despite its generally benign nature. Given the role of DVAs in venous drainage of the normal brain tissue, especially deep venous angioarchitecture, as in our case, conservative management can be recommended to avoid secondary complications.

Authors' contributions: Study conception and design: OB, NG. Data collection: OB, OM, NG. Analysis and interpretation of results: OB, TH. Draft manuscript preparation: OB, TH, KOM. Critical revision of the article: KOM, NG. Study supervision: NG. All authors (OB, TH, KOM, NG) reviewed the results and approved the final version of the manuscript.

Ethical approval: Institutional Review Board approval is not required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation: The authors confirm that they have used Artificial Intelligence (AI)-Assisted Technology for assisting in the writing or editing of the manuscript or image creations.

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How to cite this article: Baskurt O, Hiçdönmez T, Mınarecı K, Gazioglu N. Idiopathic cerebellar hemorrhage in a patient with an isolated developmental venous anomaly: A case report. Surg Neurol Int. 2025;16:82. doi: 10.25259/SNI_1056_2024