DOI:10.4274/tnd.65928
Turk J Neurol 2017;23:234-238

Medusa-like Atypical Intracerebral Vein: A Rare Case of Symptomatic Developmental Venous Anomaly

Medusa Benzeri Atipik İntraserebral Ven: Nadir Görülen Bir Semptomatik Gelişimsel Venöz Anomali Olgusu

Aloysius Ebi Ligha[1], Faith Owhabel Robert[2]

[1]Texila American University, Guyana/Niger Delta University Faculty of Basic Medical Sciences, Nigeria
[2]Niger Delta University Faculty of Medicine, Department of Human Anatomy, Amassama, Nigeria

Abstract

Developmental venous anomaly (DVA) is an extreme variation of the normal transmedullary vein, which is necessary for the normal venous drainage of an area of the brain. It occurs in as many as 2% of individuals. In most cases, it is considered to be an incidental finding, however it can be symptomatic under very rare circumstances. The possible pathomechanism of symptomatic DVA is broadly divided into mechanical and flow-related. The flow-related pathomechanism accounts for the majority of cases. Susceptibility weighted magnetic resonance imaging study is the imaging modality of choice in all vascular anomalies because it can detect even very remote vascular abnormalities and its superiority over conventional imaging can never be over emphasized. Irrespective of the pathomechanism of DVA, conservative management remains the treatment of choice. This case of symptomatic DVA is presented to buttress the fact that though most symptomatic venous anomalies are associated with hemorrhage from cavernoma, in rare cases, symptoms may arise from DVAs that are not associated with other vascular malformations.

Keywords: Developmental venous anomaly, susceptibility-weighted images, perfusion magnetic resonance imaging

Öz

Gelişimsel venöz anomali (GVA) beynin bir bölgesinin normal venöz drenajı için gerekli olan normal transmedüller venin sıradışı bir varyasyonudur. Normal popülasyonda %2 oranında görülür. Çoğu olguda bu durum tesadüfi bir bulgu olarak gözlenmiştir, ancak çok nadir durumlarda semptomatik olabilir. Semptomatik GVA'nın muhtemel patomekanizması genel olarak mekanik ve akım ilişkili olarak ayrılmıştır. Olguların çoğunu akım ilişkili patomekanizma oluşturmaktadır. Manyetik rezonans incelemede 'susceptibility weighted imaging' sekansı tüm vasküler anomalilerde tercih edilen görüntüleme yöntemidir, çünkü çok uzak vasküler anormallikleri bile tespit edebilir ve konvansiyonel görüntülemeye göre üstünlüğü asla göz ardı edilemez. GVA'nın patomekanizması ne olursa olsun konservatif yaklaşım halen tercih edilen tedavi durumundadır. Bu semptomatik GVA olgusu, semptomatik venöz anormalliklerin çoğunun kavernoma bağlı kanama ile ilişkili olmasına rağmen, nadir durumlarda, diğer vasküler malformasyonlar ile ilişkili olmayan GVA'lara bağlı semptomların ortaya çıkabileceği gerçeğini desteklemek için sunulmuştur.

Anahtar Kelimeler: Gelişimsel venöz anomali, difüzyon ağırlıklı görüntüleme, perfüzyon manyetik rezonans görüntüleme

Address for Correspondence/Yazışma Adresi: Aloysius Ebi Ligha MD, Texila American University, Guyana/Niger Delta University Faculty of Basic Medical Sciences, Nigeria
Phone: 63(02)9540228 E-mail: docligha@yahoo.com ORCID ID: orcid.org/0000-0002-2265-6153
Received/Geliş Tarihi: 26.12.2016 **Accepted/Kabul Tarihi:** 01.04.2017

Introduction

Developmental venous anomalies (DVAs), also known as venous angiomas, represent congenital anatomically variant pathways in the normal venous drainage of an area of the brain. They were once thought to be rare, but are now considered to be the most common vascular malformation in the central nervous system (1,2). Angiograms show a "hydra" or caput medusae appearance as a result of smaller radial veins converging on a central draining venous vein. Though computed tomography (CT) scans may also show a rounded enhancing area, magnetic resonance imaging (MRI) has sufficient resolution to demonstrate the 'hydra' or caput medusae form.

The etiology and mechanism of DVAs are unknown, but it is currently accepted that they act like a compensatory system of cerebral parenchyma venous drainage due to either early failure, abnormal development, or an intrauterine occlusion of normal small transcerebral veins (3). They may occur in as many as 2% of individuals (4). Although for several years DVAs were commonly called venous angiomas, the newer term DVA has been recommended as more appropriate because the involved vessels are not abnormally formed, but rather merely dilated. In recent publications, the majority of DVAs found incidentally were asymptomatic, but there are isolated reports of patients with symptoms secondary to hemorrhage or thrombosis of DVAs (5). Symptoms such as dizziness and ataxia have been claimed to be associated with DVAs, but attributing such generalized symptoms to this lesion is difficult (4). Symptoms that are directly related to a DVA most often involve thrombosis and/or adjacent hemorrhage (6,7).

DVAs are classified as deep (i.e., draining into deep subependymal veins and the galenic system) or superficial (i.e., draining into cortical veins). In about 70%, the superficial pattern is present, and deep drainage is found in 20%. A third category, which is a combination of both, occurs in 10% (3,8). Apart from the hydra appearance, complex DVAs can have multiple collector veins, associated with both deep and superficial drainage and usually drains a larger area. In general, 36-56% of DVAs occur most often in the frontal lobe, followed by the parietal lobe (12% to 24%). Occipital lobe and temporal lobes account for 4% and 2-19%, respectively. The cerebellum 14-29%, the basal ganglia 6%, and the thalamus and brainstem make up less than 5% (3).

Pereira et al. (9) reported a review of all possible pathomechanisms of symptomatic DVAs, which were divided into mechanical and flow-related causes. More than 92.7% of accurate symptomatic DVAs exhibit either of these mechanisms. In the mechanical-related pathomechanism, the venous trunk of the DVA may compress intracranial structures, especially if well dilated and is in close relation with vulnerable structures. The neurologic symptoms caused by mechanical compression accounts for 32.7% of all cases of symptomatic DVAs, and the most common presenting symptoms are hydrocephalus, tinnitus, facial hemispasm, and trigeminal neuralgia (10,11,12). The flow-related pathomechanism can either be due to increased or decreased flow. In increased flow, a slightly early venous filling can be present, which has been described as ″microshunts″ (3,13). In their experience, an increased medullary blush is not considered to be a real shunt, but rather it demonstrates a rapid transit time because medullary veins are enlarged. Restriction of flow, on the other hand, can occur in two pathomechanisms; by an obstacle to the normal flow or by a "functional" obstacle that can be caused by an increase in venous pressure due to a distant dural arteriovenous shunt. The restriction of outflow can produce a wide range of clinical presentations ranging from venous congestive edema to hemorrhage, just like sinus and cortical venous thrombosis (14,15).

Case Report

A 28-year-old male presented to our outpatient department with a two-week history of a ringing sensation and fullness in the right ear, insomnia, and headache. A physical examination and otoscopy revealed a hyperemic right ear canal with ipsilateral congested turbinates. There was no facial asymmetry. An audiogram showed normal pure-tone average with mild hearing loss at 800 Hz (Figure 1). Five months later, the patient presented to the outpatient clinic with a subjective symptom of headache, pain in the periorbital area, and dizziness. Neurological examination revealed loss of balance with a positive Romberg sign. MR imaging showed a lesion in the right frontoparietal region of the cerebral hemisphere (Figure 2). An axial T2-weighted MRI showed an abnormal hypo-intense dilated draining vein in the right frontoparietal area, which was enhanced on the image sequence obtained following gadolinium contrast administration. This vein was connected to an intracerebral ″hydra″ or medusa-like DVA. There was neither mass effect, acute infarct, hemorrhage, midline shift, nor hydrocephalus. The brainstem and cerebellum were preserved. Cranial CT angiography was subsequently requested to eliminate any coexisting arteriovenous malformation. The result

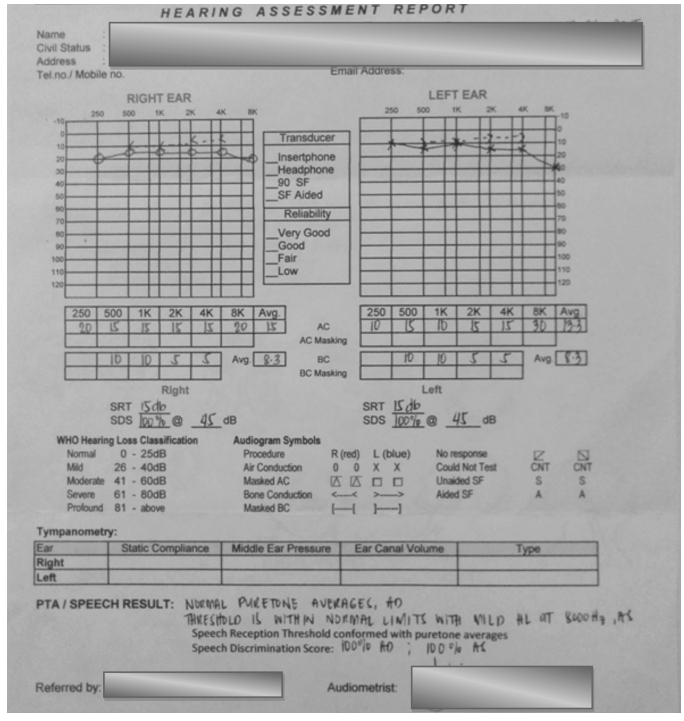

Figure 1. Audiogram showing normal puretone average with mild hearing loss at 800 Hz in the left.

showed no gross abnormality within the arteries comprising the Circle of Willis. A stellate vascular configuration with a dilated cortical vein was seen in the right frontal lobe. Major drainage was within the superior sagittal sinus with the size of the vein ranging from 4.9-5.9 mm. No gross intracranial hemorrhage, midline shift or hydrocephalus was noted (Figure 3). Although T2-weighted MR imaging and CT angiography are sensitive and able to detect small vascular structures, susceptibility-weighted angiography imaging, a high resolution 3D gradient-echo MR imaging technique with both phase and magnitude information with improved sensitivity to detect occult vascular abnormalities, which are invisible on conventional imaging, was performed and it revealed a better delineation and characterization of the medusa-like distinct deep medullary veins and draining vein of the DVA with no evident occult or remote vascular abnormalities (Figure 4). On perfusion MRI, there was a marked increase in cerebral blood flow (CBF), cerebral blood volume (CBV), mean transit time (MTT), and time-to-peak (TTP) of both the DVA and surrounding brain parenchyma (Figure 5). Conservative treatment was adopted, and the patient's condition remained stable after several months of follow-up.

Discussion

Most patients with DVA have no symptoms; the DVA is found incidentally during imaging investigations. In other words, DVAs are most often diagnosed in young adults who present with symptoms that are not in the real sense related to the DVA per se, as observed in this case. Previous reports showed a high incidence of intracranial hemorrhage associated with DVAs, and that has been currently attributed to the coexistence of a cavernous malformation (16). It is, therefore, pertinent to suggest that any DVA that presents with signs and symptoms that can be linked directly to the DVA itself requires vigorous investigation using MRI and or cranial CT angiography, to exclude the possibility of a coexisting cavernous malformation or any arteriovenous malformation.

Even though DVAs are considered benign, under rare circumstances they can be symptomatic. In a review of symptomatic case reports (9), most of the possible pathomechanisms of DVAs could be identified. Out of the 69 symptomatic cases, a mechanical (obstructive or compressive) pathomechanism was implicated in 20.3%, resulting in either hydrocephalus or nerve compression-related symptoms. The flow-related pathomechanism, on the other hand, accounted for 69%, which were further subdivided

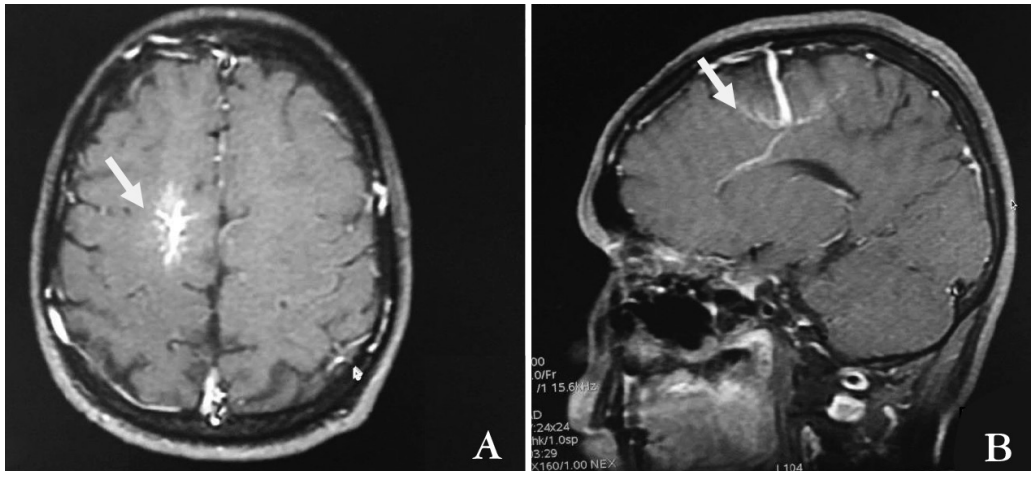

Figure 2. Gadolinium enhanced magnetic resonance imaging showing a lesion (caput medusae) in the frontoparietal region in A arrow and dilated draining vein in B arrow

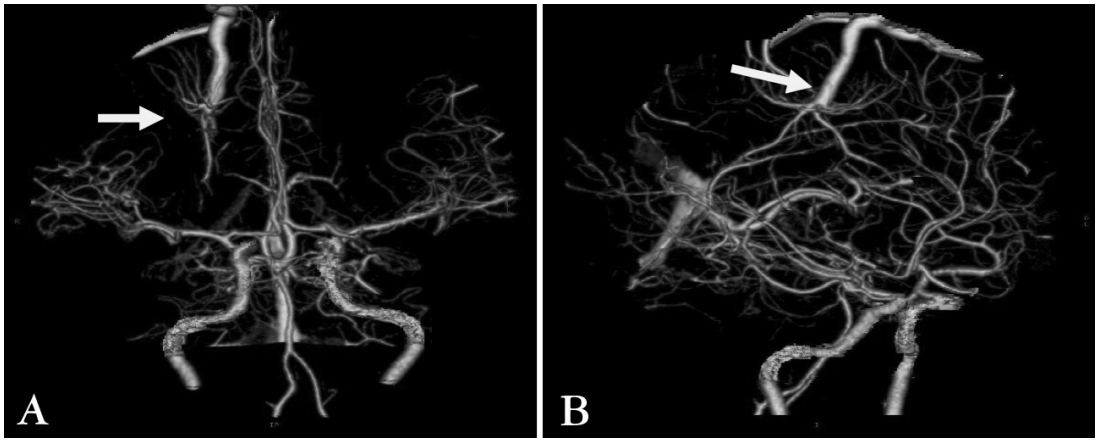

Figure 3. A computed tomography angiogram images showing smaller radial veins A, arrow converging on a central draining venous trunk, B arrow

into; complications resulting from an increase of flow into the DVA, accounting for 27.4% (due to an arteriovenous shunt using the DVA as the drainage route), and decrease of outflow was put at 37.6%. Lastly, a remote shunt with increased venous pressure accounted for 5.7%, leading to symptoms of venous congestion. The pathomechanisms of 93.7% of all symptomatic DVAs could be explained. However, in a fraction of cases, no clear pathomechanism could be identified and this has been categorized as idiopathic.

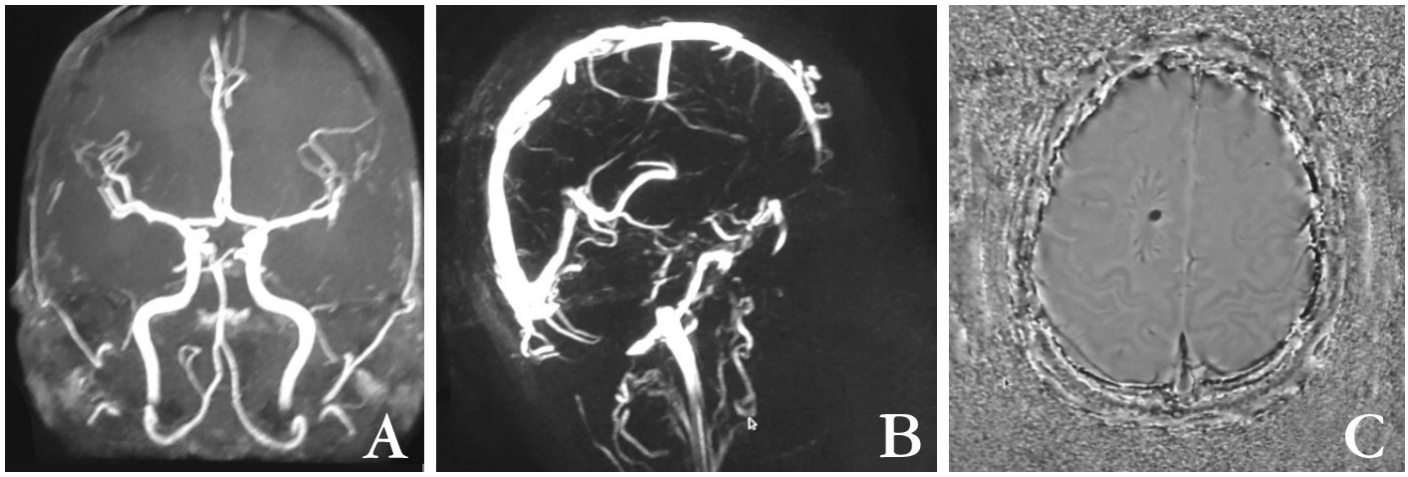

Figure 4. Susceptibility weighted angiogram (A), showing normal anterior and posterior arterial circulation, venogram (B), showing small medullary veins draining into a collector vein and (C) 3D susceptibility weighted image showing no ocult or remote hemorrhage.

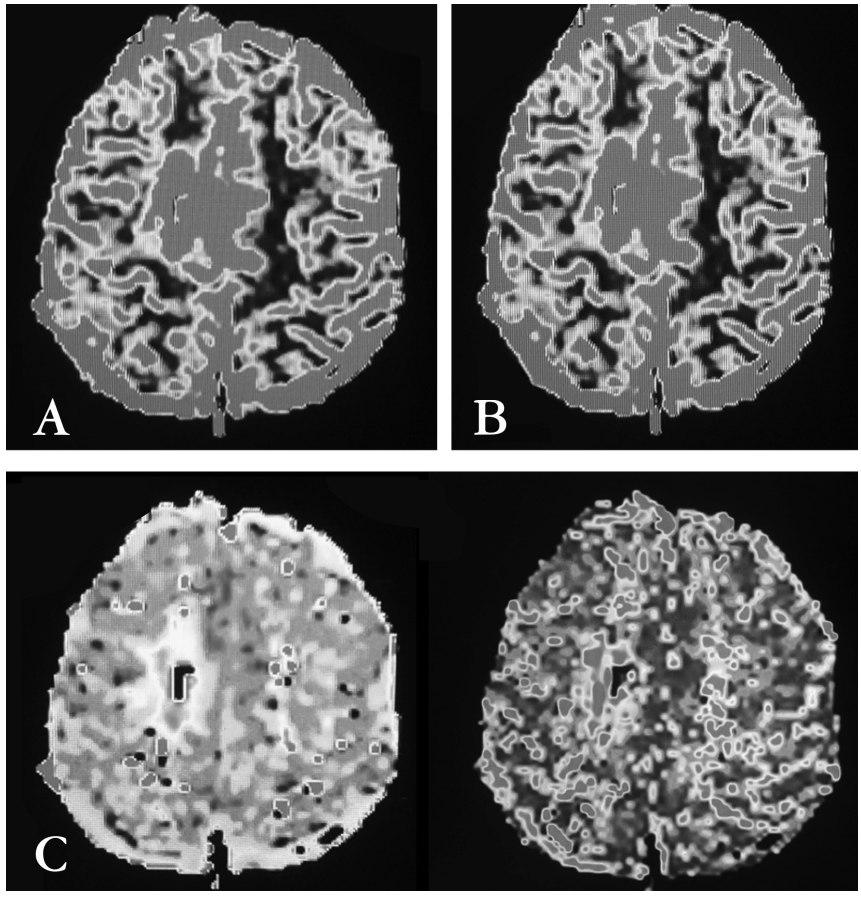

Figure 5. Perfusion magnetic resonance imaging maps demonstrate increased signal intensity in cerebral blood flow (A), cerebral blood volume (B), mean transit time (C) and time-to-peak of both developmental venous anomaly and surrounding parenchyma.

Although T2-weighted imaging and CT angiography are sensitive and able to detect small vascular structures, susceptibility-weighted imaging (SWI), has been reported to be the imaging modality of choice in the identification, characterization, and better delineation of medusa-like distinct deep medullary veins and the draining vein of DVAs and associated vascular malformations. These specific techniques have a unique advantage of precision in identifying the typical configuration with functional and anatomic information, which are not available with conventional imaging techniques, as was noted in our case, which is in conformity with a previous report by Reichenbach et al. (17). These techniques can be used safely with patients with renal failure because they do not require administration of a contrast agent.

In our case, signal intensity was observed around the parenchym of the DVA, which was in accordance with the review by Santucci et al. (18). The reason of the signal intensity changes remains unclear, but some possible implicated causes include edema, gliosis, demyelination, leukoaraiosis, ischemia, and glial metaplasia (18,19). In the MRI perfusion study, there was markedly increased CBV, CBF, TTP, and MTT of both the DVA and its vicinity. This is in agreement with work by Camacho et al. (20). The abnormally large drainage territory of the DVA may have resulted in relative volume overload with edema and/or congestion, which may be the underlying pathophysiology of the symptomatic DVA in this case.

In the treatment of isolated symptomatic DVAs, a conservative approach is the gold standard. Neurosurgical intervention is not a choice for the treatment of isolated DVAs because this on its own can lead to unacceptable consequences (7,21,22). Therefore, excision or ablation of DVAs should be avoided because they may form part of the normal venous drainage for adjacent normal neural tissue. In rare cases in which isolated symptomatic DVAs cause hemorrhage or thrombosis, surgical treatment should be targeted, not to the DVA, but the resultant complication such as cerebrospinal fluid diversion when there is ventricular outflow obstruction. Anticoagulation therapy may be an option in the uncommon event of DVA thrombosis and ischemic complications (16).

The actual incidence of symptomatic DVAs is unknown. DVAs should still be considered to be benign lesions, although in rare cases, they can be symptomatic according to the aforementioned conditions. Such cases should be vigorously investigated with more advanced imaging modalities such as SWI for possible coexisting malformations or complications arising from the DVA itself. The pathomechanism should be identified for proper management. The integrity of the DVA needs to be preserved irrespective of the treatment of choice. Excision or ablation of venous angiomas should be avoided because angiomas are mostly healthy neural tissue.

Ethics

Informed Consent: Inform consent was filled out by all participants.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.E.L., Concept: A.E.L., F.O.R., Design: A.E.L., F.O.R., Literature Search: A.E.L., F.O.R., Writing: A.E.L., F.O.R.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Zimmer A, Hagen T, Ahlhelm F, Viera J, Reith W, Schulte-Altedorneburg G. [Developmental venous anomaly (DVA)]. Radiologe 2007;47:870-874.
2. Jones BV, Linscott L, Koberlein G, Hummel TR, Leach JL. Increased Prevalence of Developmental Venous Anomalies in Children with Intracranial Neoplasms. AJNR Am J Neuroradiol 2015;36:1782-1785.
3. Okamura T, Kurokawa Y, Ikeda N, Abiko S, Ideguchi M, Watanabe K, Kido T. Microvascular decompression for cochlear symptoms. J Neurosurg 2000;93:421-426.
4. Kovács T, Osztie E, Bodrogi L, Pajor P, Farsang M, Juhász C, Szirmai I. Cerebellar developmental venous anomalies with associated vascular pathology. Br J Neurosurg 2007;21:217-223.
5. Fenzi F, Rizzuto N. Ataxia and migraine-like headache in a girl with a cerebellar developmental venous anomaly. J Neurol Sci 2008;273:127-129.
6. Vieira Santos A, Saraiva P. Spontaneous isolated non-haemorrhagic thrombosis in a child with development venous anomaly: case report and review of the literature. Childs Nerv Syst 2006;22:1631-1633.
7. Brasse G, Stammel O, Siemens P, Töpper R. [Thrombosis of developmental venous anomaly and consecutive venous infarction]. Nervenarzt 2008;79:703-705.
8. Awad IA, Robinson JR Jr, Mohanty S, Estes ML. Mixed vascular malformations of the brain: clinical and pathogenetic considerations. Neurosurgery 1993;33:179-188.
9. Pereira VM, Geibprasert S, Krings T, Aurboonyawat T, Ozanne A, Toulgoat F, Pongpech S, Lasjaunias PL. Pathomechanisms of symptomatic developmental venous anomalies. Stroke 2008;39:3201-3215.
10. Watanabe H, Yasaki S, Horiuchi M, Takahashi Y. A case of cerebral venous angioma with paresis of the left arm and face. Nihon Ronen Igakkai Zasshi 2005;42:450-452.
11. Yagmurlu B, Fitoz S, Atasoy C, Erden I, Deda G, Unal O. An unusual cause of hydrocephalus: aqueductal developmental venous anomaly. Eur Radiol 2005;15:1159-1162.
12. Shim HJ, Song DK, Lee SW, Lee DY, Park JH, Shin JH, Kim S. A case of unilateral sensorineural hearing loss caused by a venous malformation of the internal auditory canal. Int J Pediatr Otorhinolaryngol 2007;71:1479-1483.
13. Komiyama M, Yamanaka K, Iwai Y, Yasui T. Venous angiomas with arteriovenous shunts: report of three cases and review of the literature. Neurosurgery 1999;44:1328-1334.
14. Eggermont JJ. On the pathophysiology of tinnitus: a review and a peripheral model. Hear Res 1990;48:111-123.
15. Forbes KP, Pipe JG, Heiserman JE. Evidence for cytotoxic edema in the pathogenesis of cerebral venous infarction. AJNR Am J Neuroradiol 2001;22:450-455.
16. Rammos SK, Maina R, Lanzino G. Developmental venous anomalies: current concepts and implications for anagement. Neurosurgery 2009;65:20-29.
17. Reichenbach JR, Jonetz-Mentzel L, Fitzek C, Haacke EM, Kido DK, Lee BC, Kaiser WA. High-resolution blood oxygen-level dependent MR venography (HRBV): a new technique. Neuroradiology 2001;43:364-369.
18. Santucci GM, Leach JL, Ying J, Leach SD, Tomsick TA. Brain parenchymal signal abnormalities associated with developmental venous anomalies: detailed MR imaging assessment. AJNR Am J Neuroradiol 2008;29:1317-1323.
19. Jung HN, Kim ST, Cha J, Kim HJ, Byun HS, Jeon P, Kim KH, Kim BJ, Kim HJ. Diffusion and perfusion MRI findings of the signal-intensity abnormalities of brain associated with developmental venous anomaly. AJNR Am J Neuroradiol 2014;35:1539-1542.
20. Camacho DL, Smith JK, Grimme JD, Keyserling HF, Castillo M. Atypical MR imaging perfusion in developmental venous anomalies. AJNR Am J Neuroradiol 2004;25:1549-1552.
21. Aoki R, Srivatanakul K. Developmental Venous Anomaly: Benign or Not Benign. Neurol Med Chir (Tokyo) 2016;56:534-543.
22. Yu XG, Wu C, Zhang H, Sun ZH, Cui ZQ. The Management of Symptomatic Cerebral Developmental Venous Anomalies: A Clinical Experience of 43 Cases. Med Sci Monit 2016;22:4198-4204.