Research Article

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2022.45.007164

Medusa Head Sign What is its Importance?

María José Coronado Assad[1]*, Diana Marcela Hernadez Muñoz[2], Jefferson David Jaimes Bautista[3], Daynela Margarita Navarro Barraza[4], Juan Sebastián Ramírez Marín[5], Carlos Andrés Berrocal Martinez[6], Mario René López Monzón[7] and Katherine Belen Ruíz López[8]

[1]*Epidemiologist, Universidad CES, Colombia,* ***ORCID:*** 0000-0001-7076-7977

[2]*General Physician, Universidad Industrial de Santander, Colombia,* ***ORCID:*** 0000-0002-9923-5840

[3]*General Physician, Universidad Industrial de Santander, Colombia,* ***ORCID:*** 0000-0001-5042-9663

[4]*General Physician, Universidad Libre, Barranquilla, Colombia,* ***ORCID:*** 0000-0001-6117-4416

[5]*General Physician, Fundación Universitaria Autónoma de las Américas, Colombia,* ***ORCID:*** 0000-0003-4606-7828

[6]*General Physician, Universidad del Sinú, Colombia,* ***ORCID:*** 0000-0002-8629-8527

[7]*General Physician, Universidad de San Carlos de Guatemala, Ecuador,* ***ORCID:*** 0000-0002-2228-2227

[8]*Intern Physician, Universidad Regional Autónoma de Los Andes, Ecuador*

***Corresponding author:** María José Coronado Assad, Epidemiologist, University CES, Colombia

ARTICLE INFO

Received: July 06, 2022

Published: July 15, 2022

Citation: María José Coronado Assad, Diana Marcela Hernadez Muñoz, Jefferson David Jaimes Bautista, Daynela Margarita Navarro Barraza, Juan Sebastián Ramírez Marín, et al., Medusa Head Sign What is its Importance?. Biomed J Sci & Tech Res 45(2)-2022. BJSTR. MS.ID.007164.

Keywords: Developmental Venous Anomalies; Cavernomas; Medusa Head; Cerebral Venous Angioma

ABSTRACT

Background: Developmental venous anomalies are the most frequently encountered common form of vascular malformations, with a reported incidence of up to 2.6% in a series of 4,069 brain autopsies. Parenchymal veins are divided into superficial and deep draining veins. Cerebral cavernous malformations, also known as cavernomas or cavernous hemangiomas, are clusters of abnormal hyalinized capillaries with no intervening brain tissue. The medusa head sign is seen in a developmental venous anomaly, where multiple radially arranged tributaries drain into a larger vein.

Methodology: A narrative review was carried out through various databases from January 2007 to February 2022; the search and selection of articles was carried out in journals indexed in English. The following keywords were used: developmental venous anomalies, cavernomas, medusa head, cerebral venous angioma.

Results: Developmental venous anomaly (DVA), also known as cerebral venous angioma, is a congenital malformation of the veins that drain the normal brain. The etiology of developmental venous anomalies remains uncertain but may be related to developmental arrest of venous structures. Isolated developmental venous anomalies do not require treatment. The jellyfish head sign is seen in a developmental venous anomaly (DVA), where multiple radially arranged tributaries drain into a larger vein.

Conclusion: This review offers precise information on the location of developmental venous anomalies, the studies available for their diagnosis, and the therapeutic approach offered, as well as other differential diagnoses.

Introduction

The developmental venous abnormality is the most frequent configuration of vascular malformations, it has an incidence of 2.6% in a series of 4069 brain autopsies. In 1967, Wolf et al. reported on the first patient who died from intracranial hemorrhage due to a lesion that was diagnosed as a venous angioma [1]. Since then, various terminologies have been used, such as venous malformation, venous angioma, and medullary venous malformation, implying that they were considered rare lesions, involving a high risk of bleeding [2]. Parenchymal veins are divided into superficial and deep draining veins. Superficial draining veins include pial veins, intracortical veins, subcortical veins, and superficial medullary veins. The deep draining veins are deep and form the four venous convergence zones on their way to the subependymal veins [3]. Cerebral cavernous malformations, which are also known as cavernomas or cavernous hemangiomas, are a class of capillaries that present hyaline alteration in the absence of intervening brain tissue. Consequently, to recurrent microhemorrhages and thrombosis, they are surrounded by deposits of hemosiderin and gliosis [2,3]. The jellyfish head sign is very common in developmental venous anomalies, where many radial inflows drain into a larger vein. Although this sign by itself does not have any clinical importance, since it does not represent any compromise for the patient's health, just as it is a rare sign [4,5]. The importance of the jellyfish head sign lies in the association that it could have with other vascular malformations that, if they can compromise the patient's health, so this sign, if present, is very suggestive that it may be associated with other pathologies [6,7]. For this reason, it was decided to carry out this work, in order to provide precise information on the location of developmental venous anomalies, the studies available for their diagnosis, and what is the therapeutic approach that is offered, as well as other diagnoses. differentials.

Materials and Methods

A systematic review was carried out, in which a search was carried out in various databases, which we highlight PubMed, Scielo and ScienceDirect. The compilation and selection of articles was carried out in journals indexed in English from the years 2007 to 2022. Keywords, the terms were carried out according to the DeCS and MeSH methodology: Developmental venous anomalies; cavernomas; jellyfish head; cerebral venous angioma. 58 original and review publications associated with the theme were identified, only 28 articles met the established inclusion requirements, such as articles that were in a range not less than the year 2007, that were full text articles and that reported on the developmental venous anomalies and the medusa head sign. As exclusion criteria, we found articles that did not have sufficient information and that did not present the full text at the time of review.

Results

Developmental Venous Anomaly

Developmental venous disorder (DVA), known as cerebral venous angioma, is a congenital malformation of the veins that drain the normal brain. They were once considered rare before cross-sectional imaging, but are now the most common cerebral vascular malformations, accounting for 55% of all such lesions [8]. A VAD is characterized by the caput medusae sign of veins draining into a single larger collecting vein, which in turn drains into a dural sinus or deep ependymal vein. It is shaped like a palm tree [9]. This type of malformations can be visible in an MRI with contrast, they present a prevalence of approximately 2.5 to 9% [9]. Developmental venous anomalies are usually incidental findings. However, patients may present intracranial hemorrhage (1-5%) or also ischemic stroke and epilepsy [10]. The cause of these alterations remains uncertain but may be associated with the arrest of the development of venous structures. Histologically, abnormally thickened veins with normal feeding arteries and capillaries are evident. In Table 1 we can identify the most frequent locations where this type of injury occurs [9-13].

Table 1.

Localization	Frequency of Presentation	Drainage
Frontoparietal Region	36-64%	Usually draining into the front horn of the lateral ventricle
Cerebellar Hemisphere	14-27%	Drains into the fourth

Most Frequent Locations of Developmental Venous Abnormalities:

a) Angiography

Angiographically, the appearance of caput medusae (a collection of dilated medullary veins converging into an enlarged transcortical or subependymal collecting vein) is pathognomonic and seen only in the venous phase. The arterial phase appears normal, although there may be late capillary flushing. No bypass present [14].

b) Magnetic Resonance Imaging

Developmental venous abnormalities are often visible on most sequences but can be subtle and are more easily seen on post-contrast T1-weighted images and susceptibility-weighted images (SWI). If there is an associated cavernous hemangioma, susceptibility-weighted sequences will be more sensitive to this component [15]. Additionally, in approximately 10% of cases, a high T2/FLAIR signal can be seen in the surrounding white matter; the etiology is unclear, but may be due to gliosis, edema, or leukoaraiosis [16]. SWI is the

preferred sequence in venous abnormalities and has been shown to have better detectability of venous structures than conventional T2*-weighted images. Signals in SWI are not compromised by low-velocity venous flow [16]. Thus, SWI has successfully demonstrated low-flow vascular formations such as DVA. The signal intensity of the veins will be low on SWI images, but will vary on phase images depending on the provider. However, it will be the same as other veins and the opposite of calcification [17].

c) Handling

Isolated developmental venous anomalies do not require treatment. However, informing the surgeon of the presence of a VAD is essential, since cauterization of the collecting vein can cause venous infarction of the draining brain parenchyma [18]. When isolated, developmental venous anomalies have a very low complication rate (0.15% per year), primarily from spontaneous thrombosis of the collecting vein leading to venous infarction and hemorrhage [15,18]. A more recent study has shown that, in extremely rare cases, VAD can become symptomatic due to various vascular complications. The authors suggested identification of the underlying pathological mechanism by magnetic resonance imaging for proper management. The importance of preserving the integrity of the DVA itself remains valid [18,19].

d) Differential Diagnosis

Table 2.

Differential Diagnosis	Synthesis
Arteriovenous malformations	Are a type of high-flow intracranial vascular malformation composed of enlarged feeding arteries, a nest of vessels closely associated with the cerebral parenchyma through which arteriovenous shunt and drainage veins occur.
Dural sinus thrombosis or dural arteriovenous fistula	Is a subset of cerebral venous thrombosis, which often coexists with deep vein or cortical thrombosis, and presents similarly, depending mainly on the affected sinus. Ural arteriovenous fistulas (aDFd) are a heterogeneous set of conditions that share arteriovenous leads of the dural vessels. They present variably with bleeding or venous hypertension and can be difficult to treat.
Sturge-Weber Syndrome	Sturge-Weber syndrome, or encephalotrigeminal angiomatosis, is a phacomatosis characterized by facial port wine spots and pyal angiomas. It is part of a broad spectrum of possible phenotypes included in craniofacial arteriovenous metameric syndrome.
Demyelination	Demyelination will often be incorrectly equated to multiple sclerosis, when in fact it is a generic pathological term that simply describes the loss of normal myelin around axons in the central nervous system. This should be distinguished from demyelination in which normal myelin formation does not exist.

Generally, appearances will be typical and no differential should be offered. In some cases, the appearances of the images may be atypical or confused with a concurrent pathology (e.g., hemorrhage). In Table 2 we can see the main considerations for a differential diagnosis [18-22].

Medusa Head Sign

It is recommended that gadolinium-enhanced T1W MRI be used to identify this sign. VADs, frequently established in the juxtacortical and periventricular regions, are commonly seen in the frontal and parietal lobes and in the brachium pontis [23,24]. VAD by itself is of no clinical importance. However, it can occur in conjunction with a cavernoma, and is seen in approximately 25-30% of cavernomas. In image 1 we can identify this type of radiological sign in the magnetic resonance [18,20,21,23,24]. Jellyfish head signal on T1W axial magnetic resonance imaging post brain contrast showing a large developmental venous abnormality with multiple radiant veins (arrows) draining into it (Figure 1).

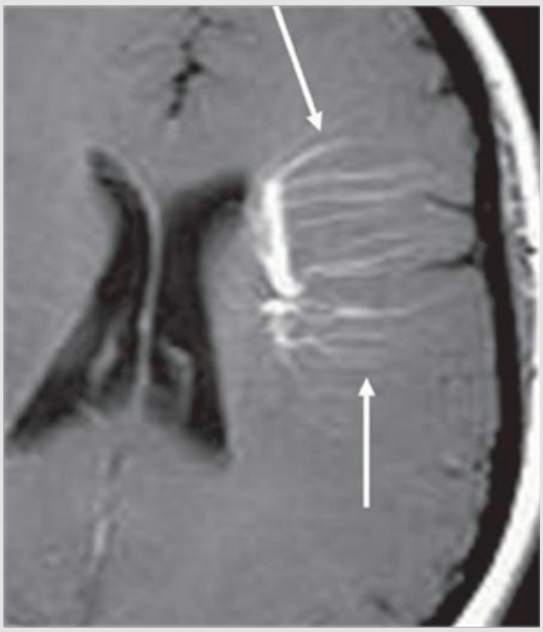

Figure 1.

Cavernomas and their Clinical Importance

Cerebral cavernous malformations, also known as cavernomas or cavernous hemangiomas, are clusters of abnormal hyalinized capillaries with no intervening brain tissue. Due to recurrent microbleeds and thrombosis, they are often surrounded by hemosiderin deposits and gliosis [24]. These alterations are characterized by a slow flow and low pressure, causing the average risk of rupture to be lower compared to other vascular malformations, such as arteriovenous malformations. The risk of rupture depends on the location of the cavernoma, the presence

of an associated developmental venous anomaly, and gender [25]. Asymptomatic familial cases have a higher annual bleeding rate than asymptomatic sporadic cases. These alterations can also arise de novo. They can grow, shrink, or remain stable over time [25]. Among the main options for management, we can find conservative management with images and serial observation, microsurgical resection and stereotactic radiosurgery. When considering treatment options, the risks of surgery must be carefully weighed against the risks of having an untreated cavernous malformation [26]. The natural history of cavernous malformations, clinical presentation, location of the lesion, number of bleeding events, and medical comorbidities help guide this discussion. Therefore, the importance of the medusa head sign lies in its association with the cavernoma, since this sign alone would not present any clinical importance [20,26].

Discussion

The study carried out by Nikolaos et al, in which a systematic review is carried out in different databases, reaching the conclusion that if cavernomas are not treated in a timely manner, they can lead to the patient suffering from intracerebral hemorrhage, seizures, focal neurological deficits or headaches, these can be identified by techniques such as Magnetic Resonance, being able to plan the surgical treatment, in this they analyze the available treatment methods, such as microsurgical resection, stereotactic radiosurgery and conservative management, according to the characteristics of the lesion [27]. Another study by Michael et al proposes that developmental venous anomalies are congenital lesions that are believed to arise from aberrations that occur during venous development, but these still continue to provide normal venous drainage to the brain territory in which they reside. Surgical or endovascular obliteration of developmental venous abnormalities is considered to develop a significant risk of venous infarction; therefore, conservative management is the one of choice for patients with these lesions [28]. The methodology is the strength of this study, with respect to the literature search, and steps in the selection of relevant articles, quality assessment and data extraction. Also, we identify limitations, within these are studies that present little information on the possible origin of this type of malformations and especially the jellyfish head sign, there is a lack of more clinical trial analysis studies that demonstrate the efficacy of therapeutic approaches available to date and the conservative management of this same radiological sign, so more studies are needed to answer these questions.

Conclusion

Developmental venous anomaly (DVA), also known as cerebral venous angioma, is a congenital malformation of the veins that drain the normal brain. They were thought to be rare before cross-sectional imaging but are now recognized to be the most common cerebral vascular malformations, accounting for approximately 55% of all such injuries. The etiology of developmental venous anomalies remains uncertain but may be related to developmental arrest of venous structures. Histologically they consist of a series of abnormally thickened veins with normal feeding arteries and capillaries. The most frequent locations are in the frontoparietal region and the cerebellar hemisphere. Isolated developmental venous anomalies do not require treatment. However, informing the surgeon of the presence of a VAD is essential, since cauterization of the collecting vein can cause venous infarction of the draining brain parenchyma. The jellyfish head sign is seen in a developmental venous anomaly (DVA), where multiple radially arranged tributaries drain into a larger vein. VAD is considered a non-pathologic variation of venous drainage and, by itself, is generally of no clinical significance. However, it can occur in conjunction with a cavernoma, and is seen in approximately 25-30% of cavernomas.

References

1. Santucci GM, Leach JL, Ying J, Leach SD, Tomsick TA (2008) Brain parenchymal signal abnormalities associated with developmental venous anomalies: detailed MR imaging assessment. Am J Neuroradiol 29(7): 1317 1323.
2. Ruiz DS, Delavelle J, Yilmaz H, Gailloud P, Piovan E, et al. (2007) Parenchymal abnormalities associated with developmental venous anomalies. Neuroradiology 49(12): 987-995.
3. Kroll H, Soares BP, Saloner D, Dillon WP, Wintermark M (2010) Perfusion-CT of developmental venous anomalies: typical and atypical hemodynamic patterns. J Neuroradiol 37(4): 239-242.
4. Akers A, Al Shahi Salman R, A Awad I, Dahlem K, Flemming K, et al. (2017) Synopsis of Guidelines for the Clinical Management of Cerebral Cavernous Malformations: Consensus Recommendations Based on Systematic Literature Review by the Angioma Alliance Scientific Advisory Board Clinical Experts Panel. Neurosurgery 80(5): 665-680.
5. Flemming KD, Link MJ, Christianson TJ, Brown RD (2012) Prospective hemorrhage risk of intracerebral cavernous malformations. Neurology 78(9): 632-636.
6. Al Shahi Salman R, Berg MJ, Morrison L, Awad IA (2008) Hemorrhage from cavernous malformations of the brain: definition and reporting standards. Angioma Alliance Scientific Advisory Board. Stroke 39(12): 3222-3230.
7. Fischer A, Zalvide J, Faurobert E, Albiges Rizo C, Tournier Lasserve E (2013) Cerebral cavernous malformations: from CCM genes to endothelial cell homeostasis. Trends Mol Med 19(5): 302-308.
8. Moore SA, Brown RD, Christianson TJ, Flemming KD (2014) Long-term natural history of incidentally discovered cavernous malformations in a single-center cohort. J Neurosurg 120(5): 1188-1192.
9. Al Shahi Salman R, Hall JM, Horne MA, Moultrie F, Josephson CB, et al. (2012) Scottish Audit of Intracranial Vascular Malformations (SAIVMs) collaborators. Untreated clinical course of cerebral cavernous malformations: a prospective, population-based cohort study. Lancet Neurol 11(3): 217-224.
10. Hanson EH, Roach CJ, Ringdahl EN, Wynn BL, DeChancie SM, et al. (2011) Developmental venous anomalies: appearance on whole-brain CT digital subtraction angiography and CT perfusion. Neuroradiology 53(5): 331-341.

11. Pearl MS, Chen JX, Gregg L, San Millàn D, Belzberg A, et al. (2012) Angiographic detection and characterization of "cryptic venous anomalies" associated with spinal cord cavernous malformations using flat-panel catheter angiotomography. Neurosurgery 71(1): 125-132.
12. Yamgoue Tchameni YT, Messerer M, Zerlauth JB, Levivier M, Daniel RT (2014) Isolated developmental venous anomaly of the pons with transpontine drainage: case report. Clin Neuroradiol 24(1): 77-81.
13. Dammann P, Wrede KH, Maderwald S, El Hindy N, Mueller O, et al. (2012) The venous angioarchitecture of sporadic cerebral cavernous malformations: a susceptibility weighted imaging study at 7 T MRI. J Neurol Neurosurg Psychiatr 84(2): 194-200.
14. Pereira VM, Geibprasert S, Krings T, Aurboonyawat T, Ozanne A, et al. (2008) Pathomechanisms of symptomatic developmental venous anomalies. Stroke 39(12): 3201-3215.
15. Ruiz DS, Yilmaz H, Gailloud P (2009) Cerebral developmental venous anomalies: current concepts. Ann Neurol 66(3): 271-283.
16. Su IC, Krishnan P, Rawal S, Krings T (2013) Magnetic resonance evolution of de novo formation of a CM in a thrombosed developmental venous anomaly: a case report. Neurosurgery 73(4): E739-E744.
17. Hong YJ, Chung TS, Suh SH, Park CH, Tomar G, et al. (2010) The angio architectural factors of the cerebral developmental venous anomaly; can they be the causes of concurrent sporadic cavernous malformation? Neuroradiology 52(10): 883-891.
18. Sharma A, Zipfel GJ, Hildebolt C, Derdeyn CP (2013) Hemodynamic effects of developmental venous anomalies with and without cavernous malformations. Am J Neuroradiol 34(9): 1746-1751.
19. Tomycz ND, Vora NA, Kanal E, Horowitz MB, Jovin TG (2010) Intracranial arterialized venous angioma: case report with new insights from functional brain MRI. Diagn Interv Radiol 16(1): 13-15.
20. Oran I, Kiroglu Y, Yurt A, Ozer FD, Acar F, et al. (2009) Developmental venous anomaly (DVA) with arterial component: a rare cause of intracranial haemorrhage. Neuroradiology 51(1): 25-32.
21. Erdem E, Amole AO, Akdol MS, Samant RS, Yaşargil GM (2012) Developmental venous anomaly coexisting with a true arteriovenous malformation: a rare clinical entity. J Neurointerv Surg 4(4): e19.
22. Im SH, Han MH, Kwon BJ, Ahn JY, Jung C, et al. (2008) Venous-predominant parenchymal arteriovenous malformation: a rare subtype with a venous drainage pattern mimicking developmental venous anomaly. J Neurosurg 108(6): 1142-1147.
23. Gross BA, Batjer HH, Awad IA, Bendok BR, Du R (2013) Brainstem cavernous malformations: 1390 surgical cases from the literature. World Neurosurg 80(1-2): 89-93.
24. Atwal GS, Sarris CE, Spetzler RF (2017) Brainstem and cerebellar cavernous malformations. Handb Clin Neurol 143: 291-295.
25. Yeon JY, Kim JS, Choi SJ, Seo DW, Hong SB, et al. (2009) Supratentorial cavernous angiomas presenting with seizures: surgical outcomes in 60 consecutive patients. Seizure 18(1): 14-20.
26. Lu XY, Sun H, Xu JG, Li QY (2014) Stereotactic radiosurgery of brainstem cavernous malformations: a systematic review and meta-analysis. J Neurosurg 120(4): 982-987.
27. M Niholaos, C Nohra, C Ameet (2015) Management of Cerebral Cavernous Malformations: From Diagnosis to Treatment. Scientific World Journal 2015: 808314.
28. Michael, M Joseph (2017) Developmental venous anomalies. Handb Clin Neurol 143: 279-282.

ISSN: 2574-1241

DOI: 10.26717/BJSTR.2022.45.007164

María José Coronado Assad. Biomed J Sci & Tech Res