



Unusual localization of Cavernous Malformation Associated with Developmental Venous Anomaly: A Case Report

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ABSTRACT

Central nervous system cavernomas are usually supra tentorial. Cerebellar localization is rare. Imaging, especially MRI can detect these malformations and look for other associated vascular malformations. If cavernous angioma is associated with a developmental venous anomaly, the venous anomaly should be spared due to the considerable risk of venous infarction following the abolition of cerebral venous drainage. We report the case of a 39 year-old man presenting with chronic headaches indicating brain MRI which showed cerebellar cavernoma associated to developmental venous anomaly.

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Introduction

Cavernomas are vascular malformations often associated with developmental venous anomalies. According to some other authors, the coexistence of these malformations results of a common pathogenesis. Imaging, especially MRI, detects and characterize these malformations which is helpful to guide therapeutic management [1,2].

Case report

A 39-year-old man, without particular medical history, was referred to our department for etiological assessment of chronic headaches evolving for 7 months without fever or traumatic context. General clinical examination was normal and did not reveal any focal neurological signs. Brain MRI was performed showing two rounded well-circumscribed right cerebellar lesions with central high intensity signal on T2WS and signal loss with blooming on GE sequence, corresponding to cavernomas (figure 1:a,b). There was also a developmental venous anomaly with collecting vessel and radial veins enhancing after gadolinium administration, corresponding to caput medusae sign (figure 2:c,d).

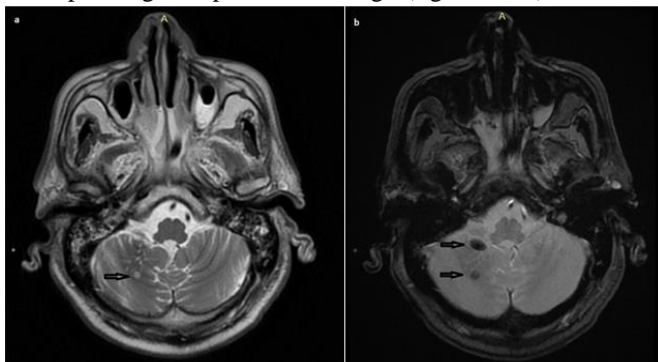


Figure 1. Brain MRI on T2 (a) and GE (b) sequences showing two rounded well-circumscribed right cerebellar lesions with central areas of high intensity signal on T2WS, signal loss blooming on GE sequence without surrounding edema (arrow).

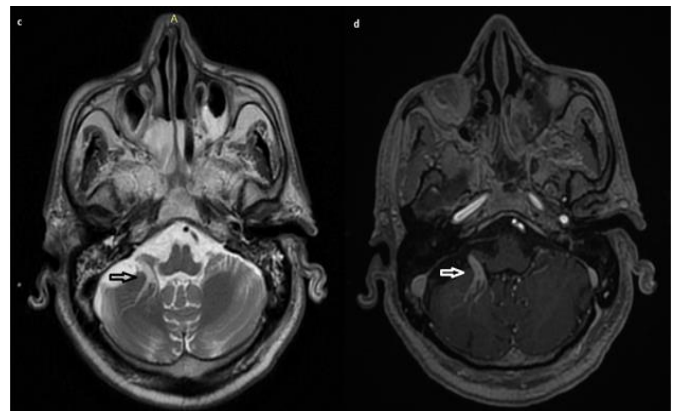


Figure 2. c. Brain MRI on T2 WS showing a right cerebellar large collecting vein with radical venous branches having phase-shift artifact and flow voids (arrow) d. MRI with gadolinium showing marked enhancement of the radial veins and the main collecting vessel (caput medusae sign) corresponding to developmental venous anomaly.

No neurosurgical treatment was performed for these two malformations due to the risk of collapsing the anomalous vein of the angioma and venous infarction. The patient received medical treatment based on codeine and paracetamol for 7 days. The evolution was marked by the regression of headaches. Clinical monitoring was recommended. MRI was performed 1 year later showing a stable appearance of the cavernoma.

Discussion

Cavernomas are vascular malformations belonging to hamartomas, as the arteriovenous malformations, telangiectasia and venous angiomas. They are formed of blood cavities with fibrous walls, united to each other without interlaced nerve tissue.

They are usually supratentorial but can be localized in the posterior cranial fossa, especially in the brainstem and the protuberance. The cerebellum is rarely affected.

Cavernomas are often associated with developmental venous anomaly (DVA, formerly known as venous angiomas) which were first described by Cushing and Bailey in 1928. They are composed entirely of veins surrounded by some elements of smooth musculature, connective tissue as well as nervous tissue, peculiarity that distinguishes them from cavernous angiomas [2,3].

The coexistence of these two vascular malformations, according to some authors, results from a common pathogenesis. DVA may result from aplasia, hypoplasia, or early occlusion of normally developing veins.

Clinically, cavernoma can cause epileptic seizures, headaches, dizziness, dysarthria, or hemorrhagic stroke. In the other hand, developmental venous abnormality, are usually asymptomatic. Headaches, epileptic seizures and, or hemorrhagic stroke were rarely described. A cerebral hemorrhage manifesting in a patient known for a developmental venous abnormality should look for a concomitant cavernous angioma [4].

Imaging aspect of these vascular malformations is characteristic. DVAs are visualized at angiography as a cluster of small medullary veins that converge to a single transcerebral vein draining into a superficial or deep venous efferent [1,3].

On the CT-scan, the collecting vein appears as a linear or curvilinear density, enhancing after contrast injection. Brain MRI with T1 post contrast and GE sequences shows the Caput Medusae sign with a large collecting vein and radial venous branches [1, 3, 5].

Cavernomas are better individualized at MRI. CT scans lack specificity and detect especially cavernoma complicated by hemorrhage, appearing as round well circumscribed or speckled lesion. On MRI, cavernoma has a central hypersignal area on the T1 and T2-weighted sequences which is due to the presence of methaemoglobin in the caverns with a peripheral hyposignal especially visible in T2 in fast imaging. MRI distinguishes 4 types of cavernoma:

Type 1: cavernoma with hyperintensity on T1 and T2 WS due to subacute hemorrhage

Type 2: classic cavernous angioma having mixed signal core and a surrounding hypointense rim giving « popcorn » appearance.

Type 3: hypointense cavernoma on T1 and T2 WS due to chronic resolved hemorrhage

Type 4: cavernoma presenting small punctate hypointense foci on GE sequence [1, 5].

Therapeutic management of cavernoma and DVA is different. Given the risk of devastating venous infarction, the treatment of DVA will be abstentionist. Moreover, its responsibility in the occurrence of a hemorrhage is often doubtful and is often linked to an associated cavernoma. For cavernoma, surgery is indicated if it is symptomatic and surgically accessible. In case of asymptomatic lesion, regular follow-up by MRI is recommended, usually an MRI is performed after 1 year and then every 2 years [1,5,6].

Conclusion

Cavernous hemangioma and developmental venous anomaly are benign embryologic vascular malformations that are usually associated. Their imaging features are characteristic. MRI should systematically look for these associations because the therapeutic management is different.

References

1. P.S. Idiculla, D. Gurala, J. Philipose, K. Rajdev, P. Patibandla. Cerebral Cavernous Malformations, Developmental Venous Anomaly, and its coexistence: A review. *Eur Neurol* 2020.
2. Rie Aoki and Kittipong Srivatanakul. Developmental venous anomaly: Benign or not benign. *Neurol Med Chir (Tokyo)* 56, 534-543, 2016.
3. Mullaguri N, Battineni A, Krishnaiah B, et al. (August 17, 2019) Developmental Venous Anomaly presenting with spontaneous intracerebral Hemorrhage, Acute ischemic stroke, and seizure, *Cureus* 11(8):e5412. DOI 10.7759/cureus.5412.
4. Meng et al. The association between cerebral developmental venous anomaly and concomitant cavernous malformation: an observational study using magnetic resonance imaging. *BMC Neurology* 2014, 14:50.
5. Cisneros O, Rehmani R, Garcia de Jesus K (April 03, 2019) Cerebellar Cavernous Malformation (cavernoma): A Case Report. *Cureus* 11(4):e4371. DOI 10.7759/cureus.4371.
6. Cortes Vela JJ, et al. Malformaciones cavernosas intracraneales: espectro de manifestaciones neurorradiológicas. *Radiologica*. 2012. Doi:10.1016/j.rx.2011.09.016.