Int. j. adv. multidisc. res. stud. 2023; 3(4):806-808

Received: 21-06-2023
Accepted: 01-08-2023

International Journal of Advanced Multidisciplinary Research and Studies

ISSN: 2583-049X

Arteriovenous Malformation and Cognitive Impairment: A Diagnostic Challenge

[1] Brian Madariaga-Cortés, [2] Marlene Arbeu-Reyes, [3] Sabatha Schravesande-De la Macorra, [4] Paul David Uribe-Jaimes

[1, 2] Department of Neurology, Hospital Universitario de Puebla, BUAP Puebla, Mexico
[4] Department of Neurology, American British Cowdray Medical Center, Mexico City, Mexico
[3] Physical Rehabilitation Doctor, Universidad La Salle, México City, Mexico

Corresponding Author: **Brian Madariaga-Cortés**

Abstract
We present the case of a 62-year-old man with a history of chronic alcoholism, who attended medical evaluation for cognitive impairment and progressive behavioral changes for 5 years. The clinical presentation was initially attributed to a history of alcoholism, until a massive fronto-parietotemporal arteriovenous malformation was found. This case highlights the importance of carrying out an adequate approach to cognitive impairment, which includes neuroimaging support.

Keywords: Dementia, Major Neurocognitive Disorder, Arteriovenous Malformation

Background

Arteriovenous malformation (AVM) has a congenital origin and is composed of a tangle of arteries and veins connected by fistulae, where the central part is a nidus. Represent a possible source of intracranial hemorrhage, but these malformations can also manifest with neurologic disorders secondary to ischemic penumbra from vascular steal. In the latter case, the clinical manifestations are less obvious and characteristic, and may include a varied clinical spectrum ranging from focal deficits to generalized malfunction of the brain parenchyma resulting in dementia [1].

Dementia, or major neurocognitive disorder, is a syndrome of progressive decline in cognitive function and/or behavior that impacts upon daily life functioning and has multiple potential causes. The diagnosis rests primarily on clinical assessment, including screening tests, with neuropsychological, neuroimaging and biomarker support where appropriate. Although clinical judgment remains essential, etiologic confirmation is required, to the extent possible, before a diagnosis is taken for granted. Dementias secondary to CVMs constitute a probably underestimated subpopulation of patients of great interest because they present with devastating but potentially reversible cognitive impairment.

Case Report

A 62-year-old man was assessed for presenting a 5-year history of progressive difficulties in household chores and financial control, visual and auditory hallucinations, and temporal-spatial disorientation. There was no family history of dementia.

This patient, with completed primary school, worker occupation, with a history of right focal motor seizures with altered consciousness, generalized since age 41 years old. Severe alcoholism since the age of 20. He used drugs to seizure control (carbamazepine 600 mg/d and phenytoin 100 mg/day).

His wife reported that he had a five-year history of progressive difficulty instrumental activities of daily living, such as how to drive car and repairing furniture, changing light bulbs, shopping and finance control. At the same time, he presented difficulties in the acquisition of new information, and presented topographical and temporal disorientation. His functionality decreased progressively with partial dependence for basic activities of daily life (bath, toilet and dress); aggression was the predominant neuropsychiatric symptom (no apathy, irritability or insomnia). He was assessed three times during that time, attributing the neurological deficit to a history of alcohol consumption without performing auxiliary, laboratory, or imaging studies. In August 2022, he went to the ER for right hemiparesis without investigation. Finally, two months later, go to the neurology outpatient clinic. Neurological examination revealed the right hemiparesis, right tactile and painful hypoesthesia, poor fluidity, temporal and spatial disorientation, The neurological examination revealed hemihypoesthesia, decreased verbal fluency, as well as temporal and spatial disorientation, which when evaluated by means of the Montreal Cognitive Assessment (MoCA), found a score of 6, with significant deficits in executive function, attention, delayed recall and abstraction, in relation

to major cognitive impairment, as well as frontal release data. In MRI, was found a massive left frontal, temporal, and parietal AVM, who had a diffuse nest at the frontal and left temporal level with arterial supply at the expense of the right M2 and ipsilateral internal carotid artery, venous drainage towards the basal vein of Rosenthal, vein of Galen and superior cerebral veins (Spetzler Martin grade V). (Fig 1) Normal serum paraclinical findings including vitamin D, B12, thyroid profile, and serum electrolytes. Treatment with endovascular embolization was considered, but was not accepted. The patient continues with conservative treatment, rehabilitation and monitoring in the outpatient clinic.

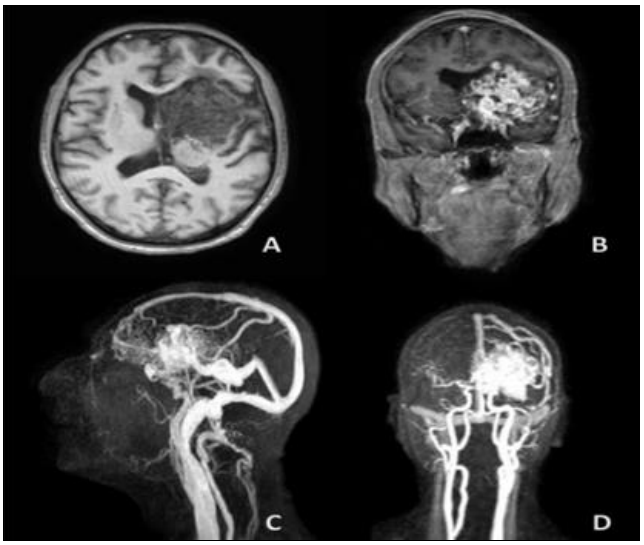

Fig 1: A, T1 with heterogeneous compact ovoid image. B. Blooming effect at base level lesion, denoting corresponding serpentine flow voids with venous drainage. C. Venous drainage with a tortuous course from the lesion towards the basal vein of Rosenthal, the vein of Galen and superior cerebral veins. D. Left internal carotid artery of longer size compared to its contralateral associated with aberrant branching at the of the M2 portion of the left middle cerebral artery

Discussion

In our patient, the functional decline was observed in recent years, evolving towards a manifest cognitive deterioration. Although her history reflects a degenerative condition with chronic and progressive impairment of cognition and function, we observed a cause of disability without hippocampal atrophy, beginning with no family or personal history of pathology. The patient presented a rare manifestation (cognitive impairment) of a massive AVM where there was involvement of multiple cognitive domains, behavioral involvement, and involvement of daily activities. The neurological deficit caused by AVMs is caused by several mechanisms, such as subarachnoid hemorrhage, intraventricular or intralobular hematoma, seizures and progressive neurological deficit, as in the case of our patient. There are very few data in the literature on the cognitive deficits observed in patients with AVM [1]. However, the observation made in 1948 is remarkable, when cognitive impairment was observed in 11 of 43 patients with AVM [2], manifested by "mental deficiency / deterioration" or "memory impairment." There are few data in the literature on cognitive deficits in geriatric patients with arteriovenous malformations.

Focal neurological deficit may be the first sign in 5% to 15% of AVMs in hospital studies, but they are rare [3]. In a larger sample, 7% of 735 untreated AVMs had focal neurologic deficits; with four possible outcomes: stable, progressive, fluctuating, or reversible. Among these focal deficits, cognition was compromised in 10 of 53 cases and progressive in four patients, and in another cohort, only two of 343 people had dementia [4]. Possible mechanisms are displacement of brain tissue (mass effect), the compressive effect of venous dilation, and neuronal loss from chronic hypoperfusion [4]. Perhaps the rarity of focal neurological signs could be explained by chronic massive lesions and chronic hypoperfusion in association with compensatory mechanisms, such as remote neuronal activation and reorganization of brain function [5]. The AVM, classified as massive, generated a greater involvement of the left cerebral hemisphere, compromising severely frontal medial and dorsal, temporal and parietal cortex but also thalamus, the hippocampus, the posterior cingulum, part of the limbic system, being severely affected in cognition and with neuropsychiatric alterations [6, 7] (Fig 2).

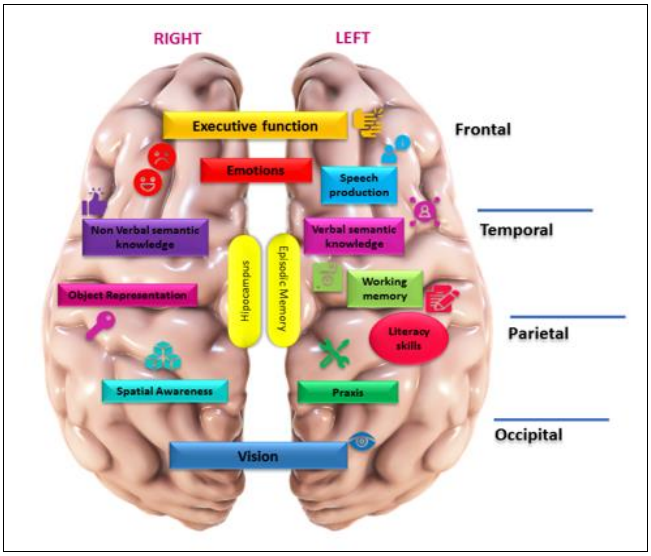

Fig 2: Representative scheme of mental functions predominantly mediated by different brain regions in each cerebral hemisphere

Table 1: The "ABCDEF" of clinical characteristics of common dementias

	Alzheimer disease	Frontotemporal dementia	Lewy body dementia	Vascular dementia
A Age of onset	> 65	< 65	< 65	< 65
B Brain pathology	Neurofibrillary tangles and amyloid plaques	Tau, transactive response DNA bindig protein (TDP-43), Pick bodies in cortex	α-synuclein Lewy bodies in cortex and midbrain	Arterioles with thickened vessel wall
C Clinical changes	Memory problems	Personality change	Fluctuating cognition, visual hallucinations, REM-sleep behavior disorder	Variable, focal neurologic symptoms
D Disability or motor symptoms	Apraxia	Frontal reléase signs	Parkinsonism	Focal weakness
E Evolution	Insidious onset	Insidious onset	Insidious onset, gradual with fluctuations	Abrupt or gradual stepwise
F Focal or diffuse changes by imaging	Hippocampal and generalized atrophy, temporal and parietal hypometabolism	Frontal /temporal atrophy and hypometabolism	Generalized atrophy, occipital hypometabolism	Strokes, lacunar infarcts

There are many etiologies of cognitive impairment, with the goal of the diagnostic process being to make a specific diagnosis, stage the disease, and identify any systemic diseases, psychiatric conditions, or delusions that might be contributing. The clinical profile of the patient's cognitive deterioration subtly fits with that of frontotemporal dementia (Table 1), which from the beginning would have encouraged the diagnostic process to rule out other pathological processes early [8]. The earlier the diagnosis, the greater the benefit in managing the disease. It is essential to have an structured approach to patients with cognitive impairment, which includes screening tests, laboratory studies to rule out reversible causes, even if clinically it meets the criterio for a specific type of pathology, Above all, it will always be essential to perform an imaging to study to rule out structural causes, as it happened in this case.

References

1. Brucki SMD, Gomes SMP. Arteriovenous malformation and dementia: A case report. Dement Neuropsychol. 2016; 10(3):244-246.
2. Olivecrona H, Riives J. Arteriovenous aneurysms of the brain: Their diagnosis and Treatment. Arch Neurol Psychiatry. 1948; 59:567-602.
3. Laakso A, Dashti R, Juvela S, Niemelä M, Hernesniemi J. Natural history of arteriovenous malformations: Presentation, risk of hemorrhage and mortality. Acta Neurochir. 2010; 107:65-69.
4. Ding D, Starke RM, Liu KC, Crowley RW. Cortical plasticity in patients with cerebral arteriovenous malformations. J Clin Neurosci. 2015; 22(12):1857-1861.
5. Choi JH, Mast H, Hartmann A. Clinical and morphological determinants of focal neurological deficits in patients with unruptured brain arteriovenous malformation. J Neurol Sci. 2009; 287:126-130.
6. Crawford PM, West CR, Chadwick DW, Shaw MDM. Arteriovenous malformations of the brain natural history in unoperated patients. J Neurol Neurosurg Psychiatr. 1986; 49:1-10.
7. Iwama T, Hayashida K, Takahashi JC, Nagata I, Hashimoto N. Cerebral hemodynamics and metabolism in patients with cerebral arteriovenous malformations an evaluation using positron emission tomography scanning. J Neurosurg. 2002; 97:1314-1321.
8. Johnson JCS, McWhirter L, Hardy CJD, *et al*. Suspecting dementia: Canaries, chameleons and zebras. Pract Neurol, 2021. practneurol-2021-003019. Doi:10.1136/practneurol-2021-003019