JOURNAL OF APPLIED COGNITIVE NEUROSCIENCE
https://revistascientificas.cuc.edu.co/JACN

Short Survey

Reversible cognitive impairment and parkinsonism with leukoencephalopathy due to cranial dural arteriovenous fistula

Melanie Lara-Sanchez[1]

Daiana E. Dossi[2]

Sebastian Ameriso[3]

How to cite this article:
Lara-Sanchez, M., Dossi, D. & Ameriso, S. (2022). Reversible cognitive impairment and parkinsonism with leukoencephalopathy due to cranial dural arteriovenous fistula. *Journal of Applied Cognitive Neuroscience*, 3(1), e00244459.
https://doi.org/10.17981/JACN.3.1.2022.03

Manuscript received on 14th April 2022
Accepted on 22th June 2022

Abstract

Parkinsonism and rapid cognitive impairment may have many causes, but only a few have specific treatment. Cranial dural arteriovenous fistula (DAVF) represents 10%-15% of intracranial arteriovenous malformations. Clinical manifestations depend on location and venous drainage. It is common the presence of pulsatile tinnitus, bruits and headache or headache and papilledema. Progressive cognitive decline is an unusual presentation due to bilateral thalamic edema or cortical venous hypertension. Endovascular or surgical treatment can reverse disease symptoms. We present a 74-year old man with rapidly progressive cognitive impairment and parkinsonism and a subsequent diagnosis of dural arteriovenous fistula (DAVF). Brain MRI revealed diffuse leukoencephalopathy probably attributable to elevated venous pressure. He was treated with partial embolization with Onix and left transverse sinus angioplasty. Following the procedure, there was a substantial reduction of venous pressure signs and remarkable clinical and imaging improvement, persisting at two years follow-up. This case represents an unusual presentation of DAVF. It is important to suspect the diagnosis of DAVF in cases with rapidly evolving cognitive impairment, parkinsonism and leukoencephalopathy as it can be reversible with early treatment.

Keywords: Cognitive impairment; parkinsonism; leukoencephalopathy; cranial dural arteriovenous fistula

Resumen

El parkinsonismo y deterioro cognitivo rápidamente evolutivo puede tener múltiples causas, pero solo unas pocas cuentan con un tratamiento específico. Las fístulas arteriovenosas durales (FAVD) representan el 10%-15% de las malformaciones vasculares intracraneales. Las manifestaciones clínicas dependen de la locación de la misma y del drenaje venoso. La presentación más frecuente es tinnitus, soplos y cefalea o cefalea y papiledema. El deterioro cognitivo es una presentación inusual y puede ser debido a edema bitalámico o hipertensión venosa cortical. El tratamiento endovascular o neuroquirúrgico puede revertir los síntomas. Presentamos un caso de un paciente de sexo masculino de 74 años de edad, con un cuadro de deterioro cognitivo rápidamente evolutivo y parkinsonismo con un diagnóstico subsecuente de FAVD. La resonancia de cerebro mostró leucoencefalopatía probablemente atribuible a elevación de la presión venosa intracerebral. El paciente fue tratado con embolización parcial con Onix y angioplastía del seno venoso transverso izquierdo. Posterior al procedimiento se observó una marcada mejoría clínica e imagenológica, siendo persistente a los dos años del seguimiento. Este caso representa una presentación atípica de FAVD. Es importante sospechar este diagnóstico en casos de deterioro cognitivo rápidamente evolutivo con parkinsonismo y leucoencefalopatía ya que puede ser reversible con un tratamiento temprano.

Palabras clave: Deterioro cognitivo; parkinsonismo; leucoencefalopatía; fístula arteriovenosa dural intracraneal

[1] Fleni, Institute for Neurological Research. Contact mail: melaniels194@gmail.com.
ORCID: https://orcid.org/0000-0002-5405-9468

[2] Fleni, Institute for Neurological Research. ORCID: https://orcid.org/0000-0002-5823-880X

[3] Fleni, Institute for Neurological Research. ORCID: https://orcid.org/0000-0001-8933-0847

Introduction

Cranial Dural Arteriovenous Fistula (DAVF) represents 10%-15% of intracranial arteriovenous malformations, with an usual age of presentation between 50 and 60 (Brito et al., 2018). It is more frequent in men (Brito et al., 2018). Clinical manifestations depend on location and venous drainage. It is common the presence of pulsatile tinnitus, bruits and headache (DAVF with antegrade drainage) or headache and papilledema (DAVF with retrograde drainage) (Elhammady et al., 2017). Progressive cognitive decline is an unusual presentation with a few cases reported in literature. In these cases isolated rapidly progressive dementia or parkinsonism were described (Brito et al., 2018; Geraldes et al., 2012; Racine et al., 2008; Luo et al., 2014; Netravathi et al., 2010). Bilateral thalamic edema or cortical venous hypertension may be the main cause of acute cognitive impairment (Brito et al., 2018).

Endovascular or surgical treatment can reverse disease symptoms and the imaging changes in patients without hemorrhagic complications or irreversible ischemia (Brito et al., 2018; Yamakami et al., 2001).

We described a patient with rapidly progressive cognitive impairment and parkinsonism associated with leukoencephalopathy.

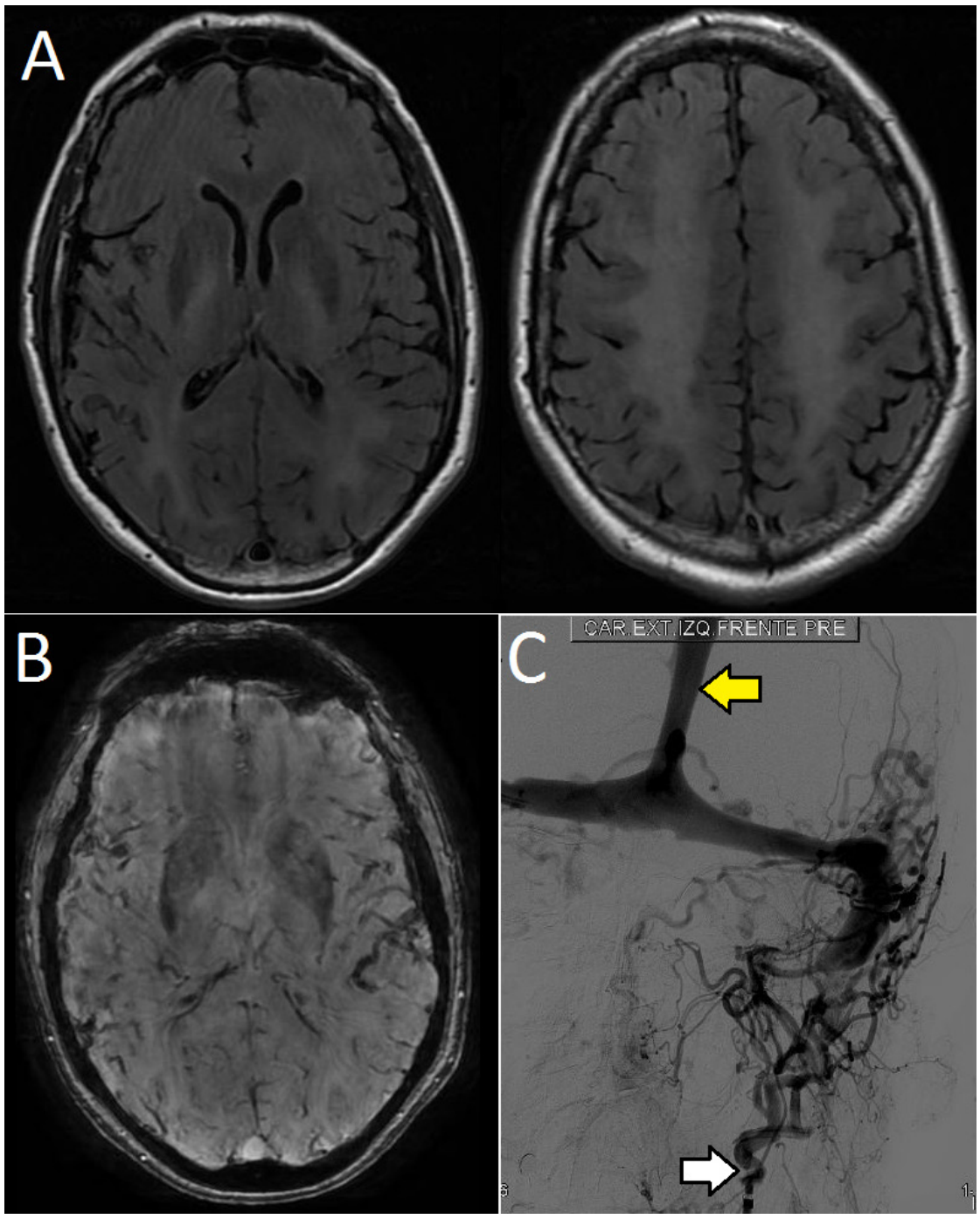
Case report

A 74-years-old man with a history of hypertension, was admitted to the emergency room with rapidly progressive cognitive and gait impairment. He had amnesic and executive dysfunction, complex visual hallucinations, tremor and gait impairment with frequent falls. The physical examination revealed disorientation in time and space and also, alteration in recall test, with a MoCA score (Brinjikji et al., 2020; Nasreddine et al., 2005) of 21 points out of 30, mild left hand weakness, gait disorder with short steps, symmetric action tremor and bradykinesia in four limbs. He was dependent on all activities of daily life. He has been previously treated with levodopa suspecting Parkinson disease, without improvement.

MRI showed extensive bilateral leukoencephalopathy in frontal, parietal and occipital lobes, with hyperintense signal in T2 and hypointense signal in T1 sequences. There was no gadolinium enhancement (Figure 1A, Figure 1B). Additionally, tortuous vessels around the left carotid internal artery near the jugular foramen were observed. It had increased signal in 3D-TOF in the homolateral rectus, transverse and sigmoid sinuses, suggesting a dural arteriovenous fistula with high venous pressure.

Diagnosis of left temporo-occipital DAVF was confirmed with digital subtraction angiography. It revealed occipital artery afference and drainage to the sigmoideus sinus with retrograde flow to the longitudinal superior sinus and deep veins (Figure 1C).

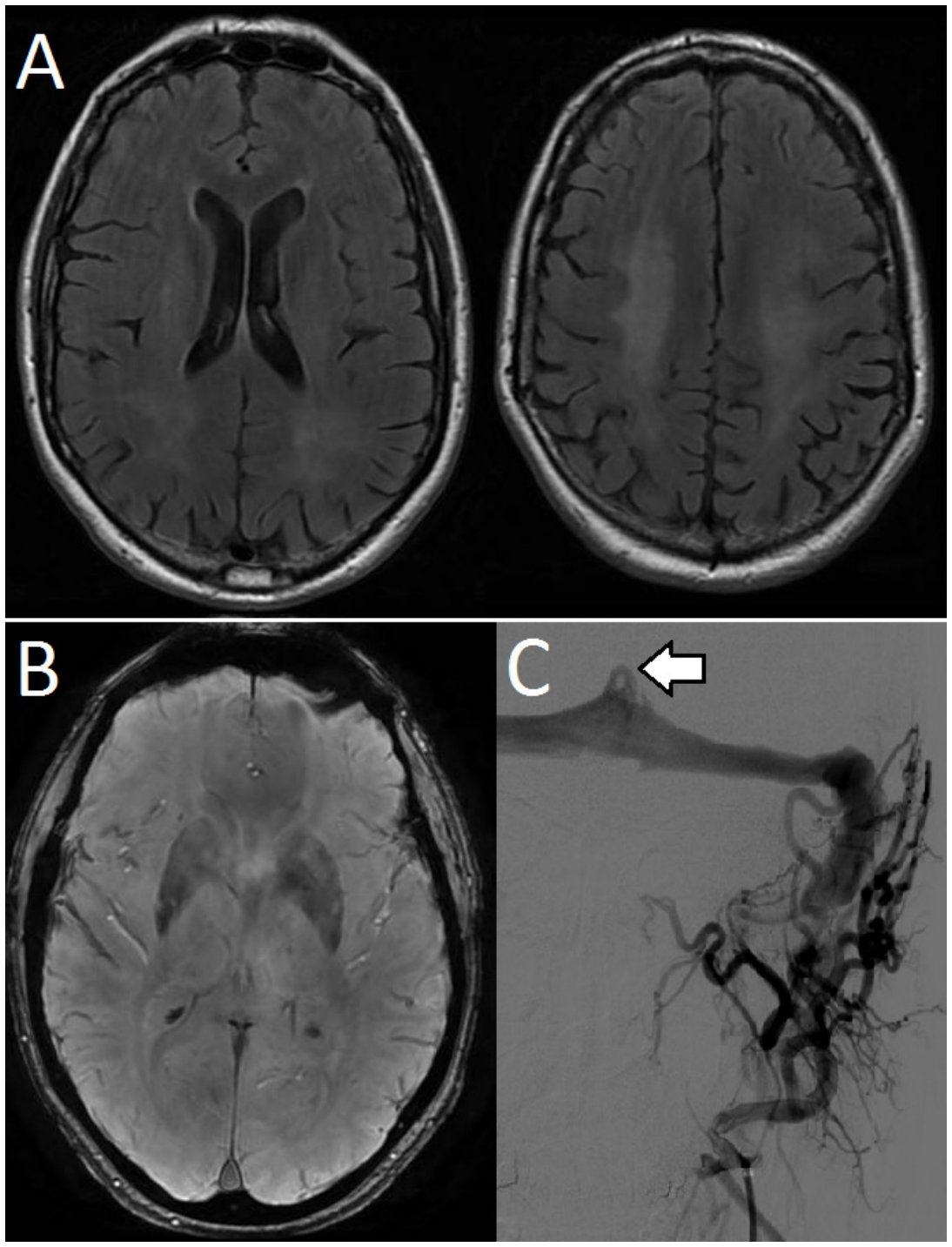
Figure 1.



Source: Authors.

He was treated with partial embolization with Onix and sinus angioplasty, obtaining a reduction of venous pressure and partial occlusion of the DAVF (Figure 2C). Two months later, the patient had achieved a substantial improvement of gait and cognitive functions with return to his premorbid status. A new MRI (six months after the diagnosis) showed reduction of the white matter hyperintensity in T2 sequence and reduction of dilatation of venous structures in SWAN (Figure 2A; Figure 2B). A subsequent embolization of the residual DAVF was completed five months after the first one, without complications.

Figure 2.



Source: Authors.

In a two-year follow-up, the patient evolved without fluctuations, with preservation of motor and cognitive functions, presenting a MoCA score of 26 out of 30.

Discusion

We reported a patient with cranial dural arteriovenous fistula with cognitive impairment and parkinsonism and diffuse leukoencephalopathy, being uncommon manifestations. These symptoms could be explained by a high venous pressure with retrograde flow, supratentorial edema and cortical dysfunction (Brito et al., 2018).

His improvement after endovascular treatment was remarkable, both clinically and radiologically as we can observe in control brain images, physical examination of gait and cognitive functions and recovery of independence for activities of daily living, persisting even at two-year follow-up, without neurological sequelae. The improvement after DAVF surgical treatment was previously reported in the literature, but mostly in patients without complications (Brito et al., 2018; Racine et al., 2008; Waragaia et al, 2006). Then, an early suspicion and diagnosis of this entity is necessary.

This report, although descriptive and lacking evidence to support a clear hypothesis, has the value of considering the coexistence of a cerebrovascular condition and the development of a complex potential neurodegenerative process.

Conclusion

It is possible that the sole presence of rapidly evolving cognitive impairment and parkinsonism, makes the diagnosis of DAVF unlikely. However, It is important to suspect this entity in those patients because it may be reversible with early treatment.

References

Brito, A.; Tsang, A. C. O.; Hilditch, C.; Nicholson, P.; Krings, T. & Brinjikji, W. (2018). Intracranial Dural Arteriovenous Fistula as a Reversible Cause of Dementia: Case Series and Literature Review. *World Neurosurg*, 121, e543–e553. https://doi.org/10.1016/j.wneu.2018.09.161

Elhammady, M. S.; Ambekar, S. & Heros, R. C. (2017). Epidemiology, clinical presentation, diagnostic evaluation, and prognosis of cerebral dural arteriovenous fistulas. *Handbook of Clinical Neurology*, 143, 99–105. https://doi.org/10.1016/b978-0-444-63640-9.00009-6

Geraldes, R.; Albuquerque, L.; Ferro, J. M.; Sousa, R.; Sequeira, P. & Campos, J. (2012). Rapidly Progressive Cognitive Impairment, Ataxia,and Myoclonus: An Unusual Presentation of a Dural Arteriovenous Fistula. *Journal of Stroke and Cerebrovascular Diseases*, 21(7), 619.e3–619.e5. https://doi.org/10.1016/j.jstrokecerebrovasdis.2011.01.002

Racine, C. A.; Lawton, M. T.; Hetts, S. W. & Josephson, S. A. (2008). Neuropsychological Profile of Reversible Cognitive Impairment in a Patient with a Dural Arteriovenous Fistula. *Neurocase: The Neural Basis of Cognition*, 14(3), 231–238. https://doi.org/10.1080/13554790802232677

Luo, Y.; Qi, J.; Cen, Z.; Hu, H.; Jiang, B. & W. Luo (2014). Show footnotes, Two cases of dural arteriovenous fistula presenting with parkinsonism and progressive cognitive dysfunction. *Journal of the Neurological Sciences*, 343(1-2), P211–P214. http://dx.doi.org/10.1016/j.jns.2014.05.059

Netravathi, M.; Pal, P. K.; Bharath, R. D. & Ravishankar, S. (2010). Intracranial dural arteriovenous fistula presenting as parkinsonism and cognitive dysfunction. *Journal of Clinical Neuroscience*, 18(1), 138–140. http://doi.org/10.1016/j.jocn.2010.04.047

Waragaia, M.; Takeuchib, H.; Fukushimaa, T.; Haisac, T. & Yonemitsuc, T. (2006). MRI and SPECT studies of dural arteriovenous fistulas presenting as pure rogressive dementia with leukoencephalopathy: a cause of treatable dementia. *EFNS European Journal of Neurology*, 13, 754–759. https://doi.org/10.1111/j.1468-1331.2006.01318.x

Yamakami, I.; Kobayashi, E. & Yamaura, A. (2001). Diffuse white matter changes with dementia caused by dural arteriovenous fistula:report of two cases. *Journal of Clinical Neuroscience*, 8(5), 471–475. http://doi.org/10.1054/jocn.2000.0796

Brinjikji, W.; Cloft, H. J. & Lanzino, G. (2020). Clinical Presentation and Imaging Findings of Patients with Dural Arteriovenous Fistulas with an Angiographic Pseudophlebitic Pattern. *American Journal of Neuroradiology*, 41(12), 2285–2291. http://dx.doi.org/10.3174/ajnr.A6811

Nasreddine, Z. S.; Phillips, N. A.; Bédirian, V.; Charbonneau, S., Whitehead, V.; Collin, I.; Cummings, J. L. & Chertkow, H. (2005). The Montreal Cognitive Assessment, MoCA: A Brief Screening Tool For Mild Cognitive Impairment. *Journal of American Geriatrics Society*, 53(4), 695–699. https://doi.org/10.1111/j.1532-5415.2005.53221.x