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Focal status epilepticus in dural arteriovenous fistula detected after a two-step clinical course: a case report

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1. Case report

We describe a case of intracranial dural arteriovenous fistula (dAVFs) in left parieto-insulo-occipital regions and left cerebellum presenting with acute myelopathy followed by focal status epilepticus in a two-step clinical course.

An 82-year-old male was admitted to our Neurology Unit for sudden weakness in the legs and fall upon waking in the morning, low back and proximal leg pain, fever, diarrhea. He had history of arterial hypertension, renal failure, dyslipidemia without any previous neurological disease. Neurological examination in emergency room showed inability to walk, weakness in both legs especially in the left side, hyperreflexia without spasticity or sensory level. Absence of sensory deficits or urinary symptoms and the prevalence of weakness in the left lower limb made this clinical presentation more consistent with brain stroke than spinal stroke. Moreover on admission he had complaints of low back pain and fever suggesting a second diagnostic hypothesis of myelitis. Therefore, a computed tomography (CT) with angiogram was required; CT images were interpreted as normal thus a diagnostic lumbar puncture (LP) was performed. LP showed slight pleocytosis: 18 cells/mm^3 of which 68 % lymphocytes. Although no pathogen was detected by Film–Array, a transverse myelitis was initially assumed, therefore patient was treated with aciclovir 10 mg/Kg t.i.d. for a 2 week course. Spinal MRI did not show lesions at cervical, thoracic, lumbo-sacral sites. Instead, motor evoked potentials (MEPs) documented delayed conduction time in both lower limbs. In the following days neurological examination revealed significant improvement of limb weakness, the patient was afebrile, he had regained walking so he was discharged with diagnosis of possible transverse myelitis. Two weeks later, the patient was readmitted in Hospital with fever, vomiting, mild confusion, aphasia and myoclonic jerks of right limbs and face with oro-mandibular involvement. Electroencephalogram (EEG) showed slowing and continuous lateralized sharp waves in left parieto-temporo-occipital (p-t-o) regions, suggesting a focal status epilepticus (Fig. 1). Myoclonic jerks of right limbs and face were related to left p-t-o sharp waves.

Administration of IV lorazepam 4 mg induced the disappearance of sharp waves on EEG and of myoclonic jerks. IV valproic acid therapy (VPA) induced transitory recovery of focal status epilepticus. Subsequently the patient showed further seizures in the same regions, therefore VPA dosage was increased until complete recovery. In the following days, he presented progressive worsening of mental status: language, executive functions and attention were assessed at bedside because he was unable to undergo a systematic cognitive assessment. A further LP showed slight pleocytosis (18 cells/mm^3) with lymphocytes prevalence. Brain MRI showed T2 and FLAIR hyperintense lesions in left parieto-insulo-occipital regions and left cerebellum, with oedema and ventricular compression (Fig. 2a). Cerebral post-contrast T1-sequences showed multiple vascular components and a dilated middle left meningeal artery, suggesting intracranial dAVF Type III (Fig. 2b).

A second course of aciclovir was given and, after brain MRI, methylprednisolone was added in therapy. Cerebral angiography confirmed the diagnosis of dAVF, involving the left transverse sinus with afferents

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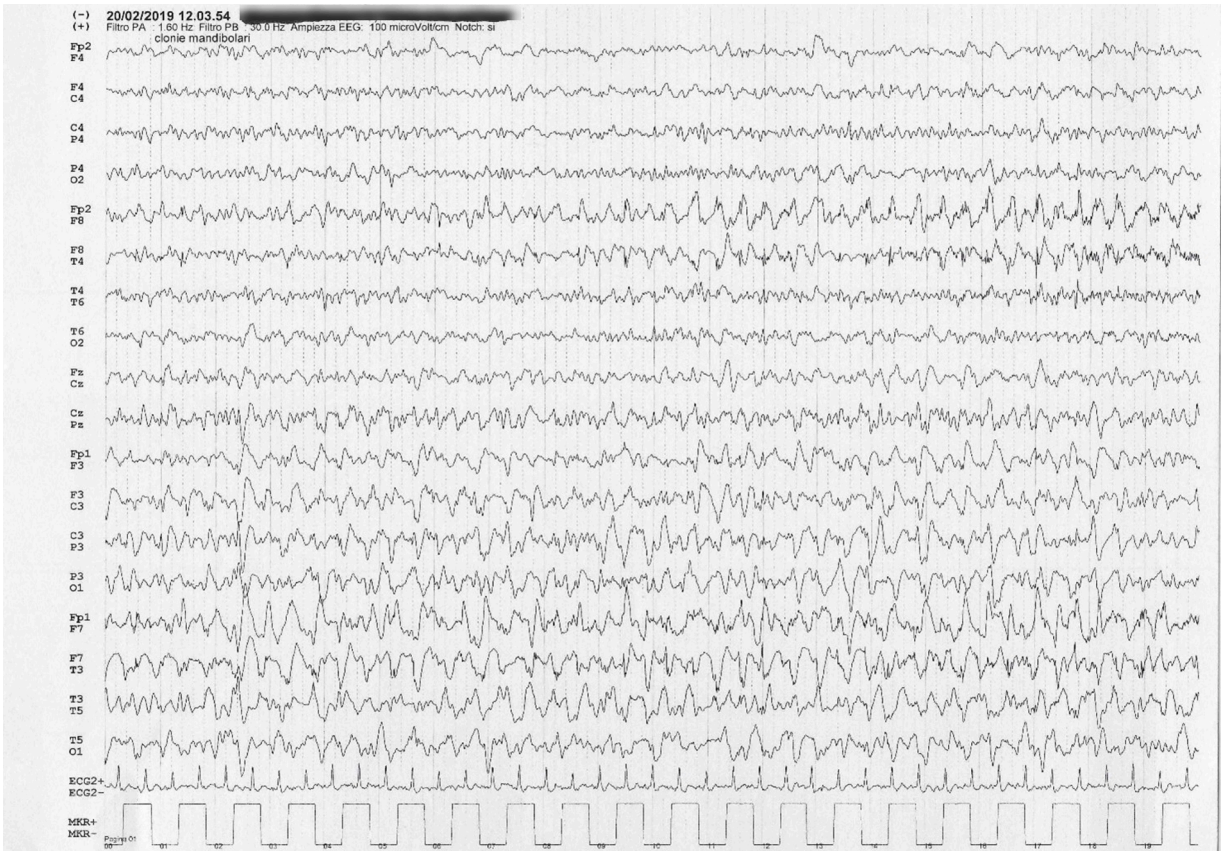

Fig. 1. EEG: 10-20 system, bipolar montage recording, viewed at a sensitivity of 100 uV/cm with the top 9 channels representing the right hemisphere, the bottom 8 channels representing the left hemisphere, the intermediate channels representing the vertex. Background is asymmetric with rhythmic irregular theta-delta activity on left hemisphere; repetitive high-voltage diphasic sharp waves are evident in left parieto-temporo-occipital (p-t-o) regions, with slight diffusion to right channels.

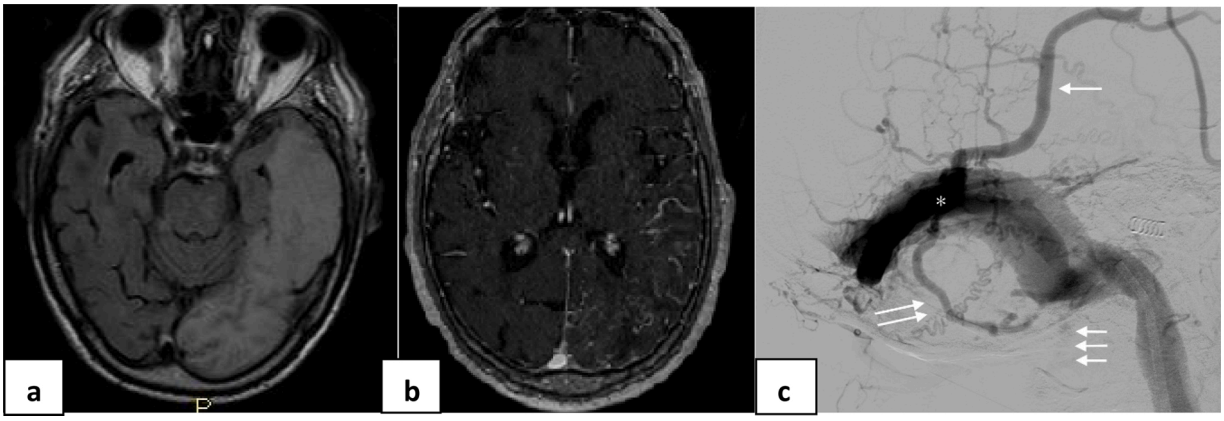

Fig. 2. Radiological findings. (a) MRI axial T2-FLAIR. Cortical-subcortical hyperintensity in left temporal, occipital lobe, ponto-mesencephalic regions due to congestive edema and venous hypertension without any arterial distribution. **(b)** MRI axial post-contrast T1 MPRAGE. Multiple dilated pial vessels in the subarachnoid space related to venous reflux with cortical or deep venous drainage. **(c)** Cerebral Angiography. Lateral view of the left external carotid artery. Asterisk: fistula point on left transverse sinus. Single arrow: vein reflux on homolateral Labbè Vein and in other supratentorial cortical veins. Double arrow: vein reflux in the infratentorial veins of the left cerebellar hemisphere. Triple arrow: absence of left jugular vein due to thrombosis.

from the left occipital artery, the left middle meningeal artery, the left posterior meningeal artery, the left medial tentorial artery of Bernasconi-Cassinari, with venous stasis and flow inversion in the left Labbè vein. Occlusion of the left jugular vein was also detected (Fig. 2c). Endovascular embolization was performed in the same day but was prematurely stopped because of sudden worsening of hemodynamic parameters, thrombocytopenia, anemia. We did not observe improvement in the following days. An extensive left cerebral and intraventricular hemorrhage occurred seven days later and caused death.

Clinical presentation of dAVFs may represent a challenging diagnostic issue, determined by both the pattern of venous drainage as well as the location of the fistula [1]. Mode of presentation and location of dAFVs are closely related. In our case, we observed an unusual clinical course: initial presentation with fever and sudden paraparesis suggesting transverse myelitis in differential diagnosis with stroke was followed

by recovery. After a few weeks, a focal status epilepticus required a second hospitalization, configuring a two-step clinical course: transient myelopathy followed by status epilepticus. Patient showed clinical symptoms suggesting initially spinal and afterwards brain involvement. First CT brain with angiogram, performed before the onset of status epilepticus was interpreted as normal. It did not allow early diagnosis; in retrospective examination, CT images showed slight prevalence of arterial vascularization on left cerebellar hemisphere and occipital lobe. Localization of dAVF in our patient can explain the unusual clinical course. Venous drainage pattern is the most important factor that influences clinical presentation [1,2]. In tentorial dAVFs, as in our case, neurological deficits may result from brainstem and cervical spine edema caused by perimesencephalic, pontine and spinal venous drainage [3]. dAFVs draining into perimedullary spinal veins may cause myelopathy and progressive tetraplegia. Thus, we can speculate that in our patient dAVFs drained into perimedullary spinal veins. Although spine MRI did not show any lesion, we can assume that the early symptoms, consistent with transient myelopathy, were caused by brainstem and cervical venous congestion related to infra-tentorial components of dAVF. This assumption may also explain the rapid recovery of strength in lower limbs. Status epilepticus and progressive neurological deficits were explained by venous congestion and subsequent thrombosis of supra-tentorial components of dAVF. Among intracranial dAVFs, tentorial dAVFs constitute less than 4 % [4]. Tentorial dAVFs are more prone to hemorrhage due to venous hypertension caused by retrograde leptomeningeal venous drainage [2]. In both the Borden and Cognard classification systems, the higher the fistula grade the worse the natural history [1,2]. Moreover, the higher the type, the more likely the dAVF is to be symptomatic as a result of increased venous congestion. dAVFs are rare but aggressive and potentially fatal vascular malformations. Atypical presentation can mimic other more common neurologic disorders delaying diagnosis. Therefore, our case characterized by clinical spinal and cerebral involvement, with focal status epilepticus as first cerebral symptom, is paradigmatic to suggest a diagnostic strategy that considers the hypothesis of a tentorial dAVF in the diagnostic work up of new onset epilepsy with apparently unrelated neurological symptoms.

Standard protocol approvals, registrations, and patient consents

Written informed consent has been obtained from patient's relative.

Data availability

Anonymized data will be shared by request of any qualified investigator.

Ethical publication statement

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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Disclosure statement

Dr. Quaranta, De Simone, Tavanti, Biraschi and Iani report no disclosures

Declaration of Competing Interest

The authors report no declarations of interest.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.seizure.2021.01.015.

References

[1] Borden JA, Wu JK, Shucart WA. A proposed classification for spinal and cranial dural arteriovenous fistulous malformations and implications for treatment. J Neurosurg 1995;82:166–79. https://doi.org/10.3171/jns.1995.82.2.0166.

[2] Cognard C, Gobin YP, Pierot L, Bailly AL, Houdart E, Casasco A, et al. Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. Radiology 1995;194:671–80. https://doi.org/10.1148/radiology.194.3.7862961.

[3] Khan S, Polston DW, Shields RW, Rasmussen P, Gupta R, et al. Tentorial dural arteriovenous fistula presenting with quadriparesis: case report and review of the literature. J Stroke Cerebrovasc Dis 2009;18:428–34. https://doi.org/10.1016/j.jstrokecerebrovasdis.2008.12.007.

[4] Tubbs RS, Nguyen HS, Shoja MM, Benninger B, Loukas M, Cohen-Gadol AA, et al. The medial tentorial artery of Bernasconi-Cassinari: a comprehensive review of its anatomy and neurosurgical importance. Acta Neurochir 2011;153:2485–90. https://doi.org/10.1007/s00701-011-1195-y.