Dural arteriovenous fistula: a clinical model of thalamic dementia?

TO THE EDITOR: We greatly enjoyed reading the article published by Holekamp et al.2 that reports their experience in the diagnosis and treatment of thalamic dementia caused by a dural arteriovenous fistula (dAVF) in 4 new cases (Holekamp TF, Mollman ME, Murphy RKJ, et al: Dural arteriovenous fistula—induced thalamic dementia: report of 4 cases. J Neurosurg 124:1752–1765, June 2016). In this paper the authors also include a thorough review of the scarce number of similar previously described cases, with an excellent discussion of the results in light of the relevant scientific literature published on this subject to date. In 2014 we had the opportunity to share with the medical community our modest experience in the management of a single case of thalamic venous ischemia caused by a tentorial dAVF;1 and we would like to remark on some key concepts that may prove useful for the professionals involved in the management of this rare entity.

To begin, it should be noted that the clinical manifestations of thalamic dysfunction could be easily overlooked or misdiagnosed. Arterial ischemia of the thalamus can produce 4 well-defined syndromes (tubero-thalamic, inferolateral, paramedian, and/or lateral posterior choroid) depending on the feeding artery involved.5 Nevertheless, an impairment of the common venous drainage of deep structures, which occurs in some dAVFs, gives rise to hyperemic changes in the whole thalamus, with bilateral involvement and a variable implication of the caudate nuclei, depending on the individual venous pattern of drainage.1 Such widespread disturbance often results in a complex constellation of neuropsychological symptoms and signs—including deficits in arousal, attention, memory, cognition, behavior, ocular motility, coordination, and gait—that have been gathered by Holekemp et al. under the term “rapidly progressive dementia.” Although in our opinion this term entails an excessive simplification, at the same time it rightly highlights the characteristic temporal evolution of the disease in a clinical scenario in which a specific localizing diagnosis can seldom be established. In our patient, the predominance of negative neurological symptoms (hypersomnia, abulia, anergia) led initially to an erroneous diagnosis of major depression, supported by the absence of significant abnormalities on the brain CT scan (Fig. 1A). One month later, the patient was referred to a neurologist because the symptoms were gradually worsening. During the neurological examination, the patient exhibited a marked bradykinesia and an unsteady gait. His verbal language, perseverant and bradypsychic, denoted temporospatial disorientation and unawareness of his illness. He also presented with impairment of vertical gaze. Brain CT and MRI findings (Fig. 1B and C) led us to complete the diagnostic workup with angiography (Fig. 1D and E).

In the review performed by Holekamp et al., almost all cases of dAVF-induced thalamic dementia were caused by lesions involving the tentorial area.2 This represents a recognized complex subgroup of dAVFs, considering their structure (which frequently includes the presence of multiple arterial feeders), the technical difficulties that may arise during the endovascular navigation of the deep venous system, and the potential impairment of critical neuroanatomical structures involved.3 In the review performed by Holekamp et al., the endovascular treatment of the dAVF did not cause significant complications, yet the failure rate of this technique reached 25% of cases.2 Moreover, in the absence of long-term follow-up data, the recurrence rate of dAVFs after embolization cannot be confidently ascertained. In contrast, the cure rate of dAVFs reached 100% following surgery, with only 1 patient developing a postoperative complication (hydrocephalus).2 It can be concluded from the available data that surgery represents an effective, safe, and definitive modality of treatment, which often constitutes the unique option in this subset of patients.2 For the purpose of proper surgical planning it is absolutely essential that neurosurgeons become familiar with the excellent systematization of tentorial dAVFs performed by Lawton et al., in which 6 major types of lesions are recognized, each one requiring a specific procedure, from the interruption of a single vein (as occurred in our case) to high-skill, demanding approaches including skeletonization of the major venous sinuses.3

Although the neurological symptoms produced by thalamic venous congestion are potentially reversible, a complete recovery was recorded in only 33% of patients.2 As Holekamp et al. state, early diagnosis and rapid treatment of the dAVF leads to restoration of anterograde venous circulation and cessation of the venous congestion of the brain parenchyma. Nevertheless, sustained venous ischemia may lead to permanent structural changes in both thalami, hence limiting the potential of recovery from
the neurological deficits.\textsuperscript{1,4} These pathological changes, excellently illustrated by the authors in the pathological photomicrographs obtained from Case 3, include gliosis, petechial hemorrhages, and microinfarctions.\textsuperscript{2} From a radiological perspective, although the bithalamic FLAIR hyperintense signal was reported to have been resolved in all cases (as occurred in ours), these underlying structural changes may still become evident in other MRI sequences, such as gradient echo sequences (Fig. 1F–I).

The pathological changes associated with sustained thalamic venous ischemia were also reported in the unique fatal case of this series, as a result of the natural evolution of the dAVF.\textsuperscript{4} The authors argued that the microscopic findings were similar to those described in so-called subacute diencephalic angioencephalopathy (SDAE), a rare and cryptic entity whose diagnosis is based on pathological findings and only made after exclusion of any other concurrent disease.\textsuperscript{6} Interestingly, most of the rare cases of SDAE reported to date lacked an angiographic study, or, when performed, it demonstrated the presence of a dAVF that the authors did not correlate with the disease.\textsuperscript{6} All this evidence suggests that an impairment of the deep venous system—including the presence of a nondiagnosed tentorial dAVF—might play a role in the pathogenesis of some cases that were diagnosed with SDAE.

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