Bilateral thalamic lesions caused by a complex intracranial dural arteriovenous fistula: a rare case report

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To the Editor: Bilateral thalamic lesions are uncommon, but the different types of pathogenesis can be classified into metabolic processes, toxic exposures, infection, vascular lesions, and tumors.[1] Among the possible forms of pathogenesis, intracranial dural arteriovenous fistula (dAVF) is extremely rare.[1] Several articles have reported on cases of bilateral thalamic lesions associated with dAVFs. However, due to differences in the supply arteries and drainage veins that were involved, as well as the treatment methods that were used, dAVFs result in a range of clinical outcomes.[2,3] Here, we report a rare case of complex intracranial dAVF that presented with dementia-like symptoms and bilateral thalamic lesions. We described the patient’s symptoms, the findings from magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), and digital subtraction angiography (DSA), and the therapeutic process and clinical outcome of the endovascular treatment.

A 38-year-old man was admitted with a 5-day history of progressively worsening forgetfulness, headache, dizziness, and blurred vision. He was relatively healthy except being a former cigarette smoker for 10 years. The patient presented normal vital signs on physical examination. The neurologic examination revealed mild short-term memory impairment and sluggish reaction times. The remainder of the examination was normal. Brain MRI demonstrated a symmetric, bilateral thalamic hypointense on T1-weighted images which was hyperintense on fluid attenuated inversion recovery (FLAIR) and T2-weighted sequences with no free diffusivity changes [Figure 1A and 1B]. Despite 2 days of mannitol treatment to reduce intracranial pressure, the patient’s status continued to worsen. He exhibited increased confusion and speech abnormalities. Further MRI brain imaging revealed that the right thalamus was predisposed to multiple microhemorrhagic lesions in susceptibility weighted imaging [Figure 1C]. The brain MRA visualized the straight sinus, Galen vein, and the large, curved, and abnormal vessels that were connected to the branches of the left posterior cerebral artery [Figure 1D and 1E]. Magnetic resonance venography (MRV) showed malformed blood vessels located in the awning area, while the straight sinus presented with a thick, proximal end, an uneven thick distal end, and a confluens sinuum variation [Figure 1F]. The possibility of the presence of a dAVF was raised.

Then, DSA was performed, which demonstrated a circuitous and complex dAVF in the left posterior thalamus, with multiple arterial feeders and involvement of the deep venous system. The arterial supply arose from branches of the following bilateral arteries: the left occipital artery, left posterior cerebral artery, and right middle meningeal artery. Among these three arteries, the meningeal branch of the left occipital artery was the main artery that contributed to the dAVF. The meningeal branch of the left occipital artery was partially redirected into the straight sinus, Galen vein, intracerebral vein, and superior sagittal sinus by the malformed dilated meningeal vein. A portion of the artery was connected to the posterior superior sagittal sinus through cortical veins and the other was directly diverted into the straight sinus [Figure 1G and 1H; Supplementary Figure 1, http://links.lww.com/CMJ/A334]. The branch of the left posterior cerebral artery was the secondary artery involved in the dAVF and it drained into the straight sinus and the cortical veins [Figure 1I; Supplementary Figure 2, http://links.lww.com/CMJ/A335]. The right middle meningeal artery was the least involved artery, and it primarily drained into the posterior sagittal sinus. These findings were consistent with a Cognard type IIa+b dAVF.

Interventional endovascular treatment was then performed. The dAVF was successfully embolized at the
Galen vein and the straight sinus using a liquid blood suppository and a transarterial approach via the left posterior cerebral and left occipital artery. The patient was re-examined using angiography, which revealed that the treated fistula was no longer present [Supplementary Figure 3, http://links.lww.com/CM9/A336]. The patient’s health status began to improve during his hospitalization, and he was discharged to his home 7 days after surgery. Three months following the interventional therapy, the clinical evaluation revealed a remarkable recovery in this patient, with improvement in his forgetfulness and confusion. In addition, the previously observed bilateral thalamic signals had disappeared on the follow-up MRI.

Bilateral thalamic abnormalities can be caused by a wide range of etiologies. However, among the vascular etiologies, bilateral thalamic lesions that present dementia-like symptoms and are caused by dAVFs are rare. Here, we reported a case of a complex, comb-like dAVF, which caused bilateral thalamic lesions and progressively worsening forgetfulness and headache.

Bilateral thalamic abnormal signals on MRI may have different characteristics which indicate different diagnoses. For dAVFs, dilated draining veins are often suspected when viewed with MRA and/or MRV. Such observations can provide evidence for a diagnosis of dAVF. In general, DSA is considered the gold standard for the diagnosis of dAVF.

dAVFs represent 10% to 15% of all intracranial arteriovenous malformations. The symptoms of dAVFs can vary depending on their size, specific aspects of the venous drainage, and location of the fistula. As this case presented, memory deficits and confusion are the main symptoms, which are due to the retrograde flow of the fistula into the straight sinus and Galen vein, which resulted in thalamic edema and functional deficits. It is noteworthy that thalamic dementia associated with
intracranial dAVF is considered to be a reversible cognitive impairment as symptoms are reduced after interventional therapy. The classification system for dAVFs is well documented. The revised classification published by Cognard et al in 1995 consists of five main types. In our case, the arterial supply predominantly arose from the left branches of the bilateral arteries. The involved vessels included the left occipital artery, left posterior cerebral artery, and right middle meningeal artery. The venous drainage was retrograde into the straight sinus, Galen vein, and dilated cortical veins, which is consistent with a Cognard type IIa+b dAVF.

Bilateral thalamic lesions have a poorer neurobehavioral prognosis compared to unilateral lesions. The clinical course of dAVFs with retrograde flows of intracranial veins shows a high risk of progressive neurological deficits and ultimately death. Therefore, early intervention through endovascular treatment or surgical therapy is necessary. In the past, a single-hole dAVF was considered to be the most amenable to endovascular treatment, while other forms of dAVF were suitable for surgery. With the development of interventional medicine, more dAVF cases have been successfully treated using endovascular treatment in recent years. However, complex dAVFs often need repeated embolizations and still relapse easily. Our patient was treated using only endovascular embolization, which obliterated the dAVF with no complications.

dAVF should be considered as one of the vascular etiology for bilateral thalamic abnormal lesions. The use of conventional brain MRI imaging combined with DSA should be considered to confirm the diagnosis of dAVF. dAVF can be treated effectively by endovascular embolization but follow up will be needed since the fistulas may come back sometimes.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent form. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflicts of interest
None.

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