Dural Arteriovenous Fistula Presenting with Status Epilepticus Treated Successfully with Endovascular Intervention

Chun-Yu Lee, Shin-Joe Yeh, Li-Kai Tsai, Sung-Chun Tang, Jiann-Shing Jeng

Abstract-

Purpose: Status epilepticus was rarely reported as the initial manifestation of intracranial dural arteriovenous fistulas. Successful treatment with endovascular intervention has not been reported in the literature.

Case Report: We report three cases of dural arteriovenous fistulas initially presenting with various types of status epilepticus, including generalized tonic-clonic status epilepticus, complex partial status epilepticus, and nonconvulsive status epilepticus. The status epilepticus of these patients was successfully terminated through aggressive endovascular intervention in conjunction with anti-epileptic drugs.

Conclusion: These cases highlight the importance of intracranial dural arteriovenous fistulas in diagnosing patients with status epilepticus. Moreover, directly treating dural arteriovenous fistulas plays a crucial role in addition to anti-epileptic drugs therapy in controlling seizures in patients with dural arteriovenous fistulas related status epilepticus.

Key Words: Dural arteriovenous fistula; Electroencephalography; Endovascular procedures; Status epilepticus

Acta Neurol Taiwan 2015;24:52-56

INTRODUCTION

Dural arteriovenous fistulas (dAVFs) account for less than 10% of intracranial vascular malformations[1]. Pathologically, it is an abnormal direct connection between meningeal arteries and dural sinuses or meningeal veins, which can lead to venous hypertension, cerebral venous sinus thrombosis (CVST), increased intracranial pressure, intracerebral hemorrhage, intracranial calcification, or venous infarction[2]. There are also various clinical manifestations of dAVFs. Around 3-10% of dAVF patients suffer from seizures which usually co-occur with CVST[2,3]; however, status epilepticus (SE) has been rarely reported in dAVF patients before. Here, we report three cases of SE secondary to dAVFs even without concomitant CVST in one case. We focus on the crucial role of aggressive endovascular intervention for dAVFs to halt refractory seizures.

From the Stroke Center and Department of Neurology, National Taiwan University Hospital, Taipei, Taiwan.
Received September 19, 2014. Revised January 26, 2015.
Accepted February 10, 2015.
Correspondence to: Dr. Shin-Joe Yeh, Department of Neurology, National Taiwan University Hospital, No 7, Chung-Shan South Road, Taipei 100, Taiwan
E-mail address: shinjoeyeh@gmail.com

CASE REPORT

Case 1:

A 49-year-old man suffered his first episode of generalized convulsive SE, which failed to be controlled by phenytoin and continuous midazolam infusion. Electroencephalography (EEG) revealed bilateral pseudoperiodic lateralized epileptiform discharges (Fig. 1A). Computed tomography angiography (CTA) showed intracranial dAVFs of Cognard type IV with abnormal vascular connection in the sinus wall of superior sagittal sinus (SSS) and drainage into it, resulting in the narrowed lumen of SSS. Angiography confirmed luminal stenosis of the SSS (Fig. 1B). Besides, the absence of thrombus in the SSS was confirmed by the balloon angioplasty which yielded no thrombus and immediate recanalization after balloon angioplasty (Fig. 1C). Angioplasty by balloon dilatation of SSS was followed by subtotal trans-arterial embolization of the dAVFs. Seizures were controlled within 5 days by AEDs, and the patient recovered well. However, two months following hospital discharge, simple partial SE recurred on the left limbs accompanied with progressive quadriparesis. CTA demonstrated restenosis of the SSS and residual dAVFs. Stenting of the SSS and near total embolization of dAVFs were achieved without incident (Fig. 1D). Seizures were terminated, and muscle power rapidly recovered following the procedure. After hospital discharge, focal seizures lasting for a few seconds to minutes recurred about once every 1~2 months without progression to SE, and there was no dAVF recurrence on brain MRI in the following 10 months.

Case 2:

A 71-year-old man was admitted for acute onset of right hemiparesis with right facial twitching, followed by altered consciousness and sustained convulsions of the right upper limb. EEG revealed periodic lateralized epileptiform discharges in the left cerebral hemisphere (Fig. 2A). Convulsive seizures were controlled by AEDs, whereupon consciousness also improved. CTA, MRI, and angiography revealed intracranial dAVF of Cognard type IV near the left transverse-sigmoid sinus, with venous infarction, multiple intracerebral hematomas, as well as focal wall thickening and stenosis of the left transverse-sigmoid sinuses (Fig. 2B-D). The dAVF was completely occluded by trans-venous embolization, and no additional seizure was recorded either clinically or electroencephalographically thereafter in the following six months.

Case 3:

An 80-year-old woman presented with acute onset of left hemiparesis, and five days later she suffered from deterioration of muscle power and clouding of consciousness. Brain MRI and cerebral angiography showed dAVF of Cognard type IIa+b at the right transverse sinus, with sinus thrombosis of bilateral transverse-sigmoid sinuses, as well as diffusely engorged cortical veins (Fig. 3A and 3B). EEG revealed evolutional changes in periodic lateralized epileptiform discharges and episodic lateralized fast activities in the right hemisphere followed by background attenuation (Fig.

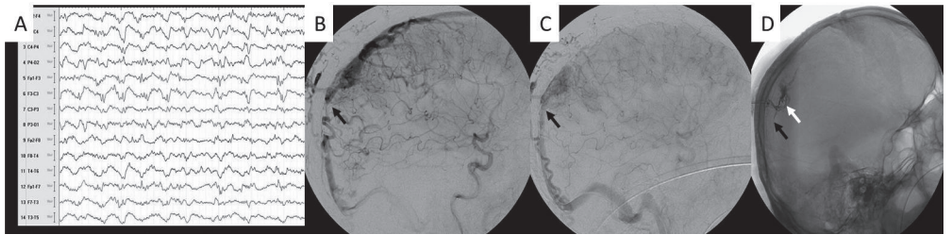

Figure 1. EEG and digital subtraction angiography from case 1. (A) The EEG shows bilateral pseudoperiodic lateralized epileptiform discharges at a recurrence rate of 1 Hz. (B) Delayed angiogram from right common carotid artery shows dAVFs around the stenotic posterior one-third of SSS (black arrow). (C) Delayed angiogram from right common carotid artery after balloon angioplasty shows revascularization of the original stenotic segment of SSS (black arrow). (D) Digital subtraction angiography shows stent placed in the SSS (black arrow) and Onyx liquid embolic agent (white arrow) used for TAE.

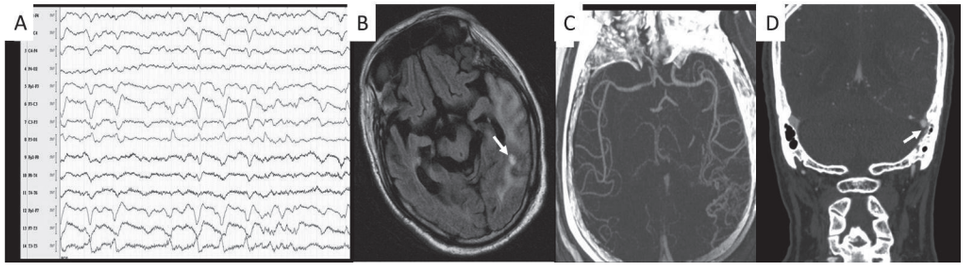

Figure 2. EEG, brain MRI, CT angiography, and digital subtraction angiography findings of case 2. (A) The EEG shows periodic lateralized epileptiform discharges in the left cerebral hemisphere. (B) T2-FLAIR MRI shows venous congestion in the left temporo-occipital lobes with a small hematoma (white arrow). (C) CT angiography shows dural arteriovenous fistulas near the left transverse-sigmoid sinus. (D) Delayed-phase CT angiogram shows focal wall thickening and stenosis (white arrow) in the left transverse sinus.

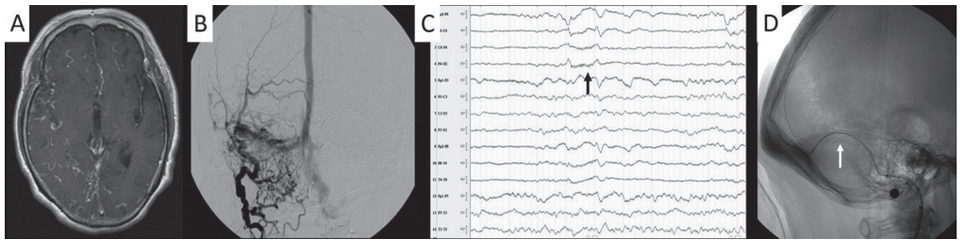

Figure 3. Brain MR angiography, cerebral angiography, and EEG results from case 3. (A) MR angiography shows dural arteriovenous fistulas (dAVFs) in the region from the right transverse sinus to the torcula. (B) Cerebral angiography shows that most of the feeders of the dAVFs are from the right occipital artery. (C) EEG shows periodic lateralized epileptiform discharges and episodic lateralized fast activities (black arrow) in the right hemisphere followed by background attenuation. (D) Digital subtraction angiography shows a stent (white arrow) placed in the right sigmoid sinus.

3C). Phenytoin was administered for nonconvulsive SE, which dramatically improved her consciousness within 24 hours (i.e. Glasgow Coma Scale improved from E3M5V4 to E3M6V5). Follow-up EEG revealed no epileptiform discharge. Further improvement in neurological status was achieved through trans-arterial embolization of the dAVF and stenting of the bilateral transverse-sigmoid sinuses (Fig. 3D). The patient recovered well without recurrence of seizure in the seven-month follow-up.

DISCUSSION

This report outlines the courses of treatment used in three cases of intracranial dAVFs presenting with various types of SE. The presentations were categorized as generalized convulsive SE (case 1), complex partial SE (case 2), and non-convulsive SE (case 3). All of the cases presented dAVFs with or without CVST, and SE was controlled successfully through a combination of AED therapy and endovascular intervention for dAVFs. Case 1 was characterized by the recurrence of SE in association with residual dAVFs and re-stenosis of the venous sinus following the first endovascular intervention. For this patient, SE was again brought under control following endovascular treatment. In cases 2 and 3, SE was effectively controlled with no signs of recurrence following the completion of dAVF treatment combined with AED therapy. These cases demonstrate

the importance of combining endovascular intervention with AED therapy to treat SE in patients with dAVFs. Our report further illustrates that SE can recur following incomplete dAVF treatment.

Seizure is not a common presentation of intracranial dAVF[2,3], and only 2 cases of dAVFs presenting as SE have been reported previously[4,5]. The first case was treated by embolization with n-butyl-2-cyanoacrylate; however, this approach failed to achieve angiographic occlusion of the dAVFs, and the patient subsequently died of intractable SE[4]. The SE of the second case was treated successfully with craniotomy and ligation of the arteries feeding dAVFs[5]. SE is considered as a neurological emergency requiring immediate administration of antiepileptic drugs and anesthetic agents, and is associated with high short-term mortality (16% to 39%) even under intensive medical treatment[6]. The cases outlined here demonstrate for the first time that endovascular intervention for dAVFs is an important step to control SE in dAVF patients.

Seizures among patients with dAVFs are primarily attributed to venous hypertension caused by either dAVFs or coexisting venous sinus outflow obstruction which can be achieved by CVST or venous sinus stenosis. Among patients with intracranial dAVFs, 39% had concomitant CVST[7]. There is a hypothesis about the mutual relationship of causality between dAVF and CVST. The chronic venous ischemia caused by CVST may enhance the secretion of angiogenic factors, leading to the genesis of dAVF. On the other hand, the high turbulent flow of dAVF may result in intimal damage and thrombosis in the venous sinus, promoting the formation of CVST. The patient of case 1 demonstrated that SE can occur in dAVF patients with narrowing of sinus and the SE was controlled after stenting of the stenotic sinus in addition to embolization of dAVF, suggesting the importance of recanalization of the narrowed venous outflow to the control of venous congestion-related regional epileptogenicity. Animal models have shown that serum proteins may induce epileptogenicity in the cortex with disrupted blood-brain barrier.[8] These findings indicate that the leakage of serum-derived components by intracranial venous hypertension may contribute to the epileptogenesis in dAVF patients. Furthermore, SE itself can lead to disruption of blood-brain barrier, resulting in a vicious cycle of self-sustained epileptogenicity in dAVF patients[9].

In contrast to SE in CVST without dAVFs, which is usually treated with anticoagulants and AEDs[10], seizures that occur concurrently with dAVFs should be treated aggressively through endovascular intervention or surgical ligation, which have been shown to rapidly reverse venous hypertension and to reduce potential epileptogenicity[11]. A previous report has demonstrated that the use of AEDs alone fails to control seizures in a dAVF patient with CVST; however, the seizures can be reduced dramatically after embolization of the dAVFs[12]. The findings reported in this current study highlight the importance of timely endovascular intervention, including embolization of the dAVFs as well as stenting to revascularize the narrowed or thrombosed sinus, in the control of SE in patients with dAVFs.

To summarize, the cases reported in this study demonstrate that a diagnosis of intracranial dAVFs should be considered when dealing with patients with SE regardless of whether there is concomitant CVST. Endovascular intervention for the dAVFs and narrowed or thrombosed sinus to treat venous hypertension plays a crucial role in the control of seizures and the prevention of seizure recurrence in dAVF patients presenting with SE.

REFERENCES

1. Hacein-Bey L, Konstas AA, Pile-Spellman J. Natural history, current concepts, classification, factors impacting endovascular therapy, and pathophysiology of cerebral and spinal dural arteriovenous fistulas. Clin Neurol Neurosurg 2014;121:64-75.
2. van Dijk JM, terBrugge KG, Willinsky RA, Wallace MC. Clinical course of cranial dural arteriovenous fistulas with long-term persistent cortical venous reflux. Stroke 2002;33:1233-1236.
3. Cognard C, Gobin YP, Pierot L, Bailly AL, Houdart E, Casasco A, Chiras J, Merland JJ. Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. Radiology 1995;194:671-680.
4. Rabinov JD, Yoo AJ, Ogilvy CS, Carter BS, Hirsch JA. ONYX versus n-BCA for embolization of cranial dural arteriovenous fistulas. J Neurointerv Surg 2013;5:306-310.

5. Ou SF, Chi CS, Shian WJ, Mak SC, Wong TT. Dural arteriovenous malformation with symmetrical calcification of the basal ganglia: a case report. Zhonghua Yi Xue Za Zhi (Taipei) 1994;54:204-208.
6. Rossetti AO, Lowenstein DH. Management of refractory status epilepticus in adults: still more questions than answers. Lancet Neurol 2011;10:922-930.
7. Tsai LK, Jeng JS, Liu HM, Wang HJ, Yip PK. Intracranial dural arteriovenous fistulas with or without cerebral sinus thrombosis: analysis of 69 patients. J Neurol Neurosurg Psychiatry 2004;75:1639-1641.
8. Seiffert E, Dreier JP, Ivens S, Bechmann I, Tomkins O, Heinemann U, Friedman A. Lasting blood-brain barrier disruption induces epileptic focus in the rat somatosensory cortex. J Neurosci 2004;24:7829-7836.
9. Correale J, Rabinowicz AL, Heck CN, Smith TD, Loskota WJ, DeGiorgio CM. Status epilepticus increases CSF levels of neuron-specific enolase and alters the blood-brain barrier. Neurology 1998;50:1388-1391.
10. Kalita J, Chandra S, Misra UK. Significance of seizure in cerebral venous sinus thrombosis. Seizure 2012;21:639-642.
11. Feyissa AM, Ponce LL, Patterson JT, Von Ritschl RH, Smith RG. Dural arteriovenous fistula presenting with exophthalmos and seizures. J Neurol Sci 2014; 338:229-231.
12. Berryman R, Imam I, Whitfield PC, Mukonoweshuro W, Salih I. Progressive seizures in a patient with congenital coagulopathies. BMJ Case Rep 2011:bcr 0220113842.