Venous Stroke with Intracranial Dural Arteriovenous Fistula

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Abstract- We present a 51-year-old man who developed a biphasic clinical course with an initial myelopathy followed by a lower brainstem syndrome caused by intracranial dural arteriovenous fistula. Brain magnetic resonance imaging (MRI) on T2-weighted imaging demonstrated an extended high signal intensity lesion in the upper cervical cord, medulla oblongata, and lower pons. Cerebral angiography disclosed a fistula between the left external carotid artery (ECA) and the left superior petrosal as well as sigmoid sinuses. There was retrograde venous drainage from the left superior petrosal sinus to the perimedullary cervical veins, leading to venous congestion and venous infarct. Transarterial and then transvenous embolisations were performed and successfully abolished the fistula.

Key Words: Intracranial dural arteriovenous fistula, Myelopathy, Venous stroke, Magnetic resonance imaging (MRI), Angiography, Embolisation

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INTRODUCTION

Intracranial dural arteriovenous fistula (DAVF) is formed by abnormal arteriovenous connections located within the dura matter of cerebral venous sinus. It accounts for 10-15% of all intracranial arteriovenous malformations[(1)], yet one third of the posterior fossa intracranial arteriovenous malformations are purely dural[(2-3)]. Intracranial DAVF consists of abnormal direct communications between dural arteries and dilated dural veins with no intervening capillary bed[(4)]. They can be either congenital or acquired. Three major types (dural sinus malformation with intracranial arteriovenous fistula, infantile intracranial DAVF, and adult-type intracranial DAVF)[(5)] are categorized according to the pattern of vascular communication. Clinically, adult-type intracranial DAVF is the most common dural vascular malformations, andoften becomes symptomatic in patients aged between 40 and 60 years[(3)]. The clinical manifestation varies a lot according to the location and venous drainage pattern of the intracranial DAVF, with the venous outlet pattern of the intracranial DAVF being the single most important determinant of the clinical presentation and long-term prognosis[(6-7)]. Intracranial DAVF with perimedullary venous drainage was initially described by Woimant et al.[(8)] in 1982. Since then, only

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rare sporadic or small series of cases have been reported. The clinical courses of these cases were variable, but the period from the clinical symptoms to the final diagnosis usually took several months. Based on the clinical and angiographic findings, Cognard et al.[9] further classified intracranial DAVFs into five types. Type V intracranial DAVF is rare, and is characterized by abnormal spinal venous drainage and progressive myelopathy which is most likely ascribable to venous hypertension. Here we present a case of type V DAVF who developed unstable venous infarct encompassing the upper cervical cord and brainstem.

CASE REPORT

A 51-year-old male engineer was admitted to a community hospital due to fullness of abdomen and difficulty in urine and stool passage. At first, abnormal urine test led to a clinical diagnosis of acute pyelonephritis. But sudden onset of paraparesis and hiccup happened to him during hospitalization, with full recovery three days later. Unfortunately, another episode of paraparesis and easy choking developed 2 weeks later. Besides, general weakness rapidly progressing to quadriplegic state was found the next day. Deterioration of sphincter function and labile blood pressure were also noted. He was soon transferred to our emergency unit where a brain computed tomogram revealed no gross abnormality.

Tracing back his medical history, he had moderate hypertension with regular treatment for 10 years. Physical examination revealed normal findings except for a blood pressure of 156/80 mmHg and a pulse rate of 134/min. Neurological examination showed a horizontal nystagmus, weakness of bilateral soft palates, asymmetric quadriparesis (the muscle power of the right limbs was grade 4/5 and left limbs grade 3/5), dysmetria (more marked on the left side), generalized hyporeflexia, and an extensor-type plantar response on the right side. Blood biochemistry tests revealed only mild hypercholesterolemia. Blood routine tests revealed moderate leukocytosis (white blood cell count: 11,300/cmm).

Unfortunately, acute respiratory failure developed one day after admission. He was then intubated and transferred to the neurological intensive care unit.

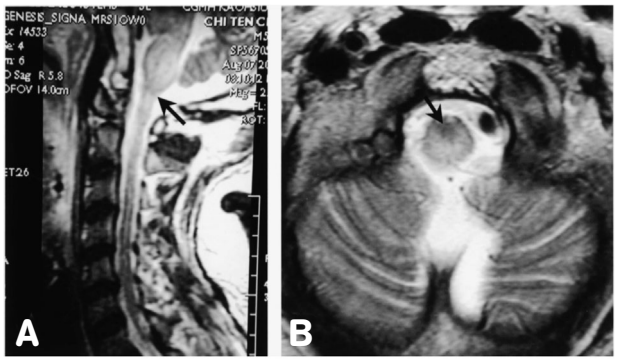

Figure 1. (A) Saggittal T2-weighted (4500/120[TR/TE]) MR image showed an extensive high signal lesion in the upper cervical cord, medulla oblongata and lower pons (arrow). (B) Axial T2-weighted (4000/120 [TR/TE]) MR image showed a high signal lesion in the medulla oblongata, whose ventral part was relatively spared (arrow).

Because of the two separate acute exacerbating episodes in the clinical course, demyelinating disorder was considered first. Analysis of the cerebrospinal fluid (CSF) showed normal IgG index, normal cell count, normal protein level, and euglycorrhacia. Brain MRI on T2-weighted imaging demonstrated an extensive high signal intensity lesion involving the upper cervical cord, medulla oblongata and lower pons (Fig. 1A) with intriguing sparing of the ventral medulla oblongata (Fig. 1B). On T1-weighted imaging a low signal lesion was noted in the aforementioned areas of involvement. Because sparing of the ventral medulla could implicate occlusion of bilateral vertebral arteries except for short circumferential branches, a digital subtraction angiography was arranged. Cerebral angiography showed multiple DAVFs, arising from the meningeal branches of the left occipital artery and extending to the superior petrosal and sigmoid sinuses with dilated perimedullary vein (Fig. 2A). Abundant vascular markings extending from the left middle meningeal artery and ascending pharyngeal artery to the left superior petrosal sinus (Figs. 2B-C) were also noted. Most importantly, there was significant retrograde drainage from the left superior petrosal sinus to the perimedullary cervical vein, contributing to in situ congestion. Venous infarcts of the upper cervical cord and lower brainstem were therefore concluded.

Considering the difficulty in surgical access to the

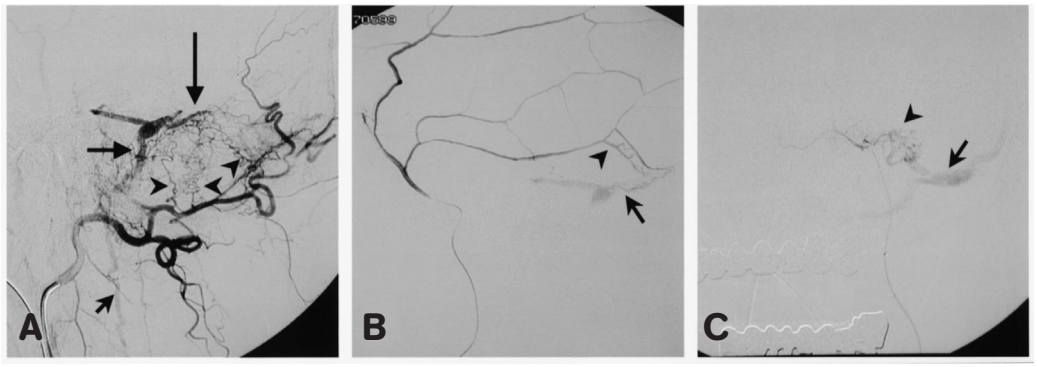

Figure 2. (A) Left ECA angiography (lateral view) showed multiple DAVFs extending from the meningeal branches of the left occipital artery (arrowheads) to the superior petrosal sinus (long arrow) and sigmoid sinus (medium arrow). Note also the dilated perimedullary vein (short arrow). (B) Left middle meningeal artery angiography (lateral view) showed DAVF extending from the left middle meningeal artery (arrowhead) to the superior petrosal sinus (arrow). (C) Left ascending pharyngeal artery angiography (AP view) showed DAVF extending from the left ascending pharyngeal artery (arrowhead) to the superior petrosal sinus (arrow)

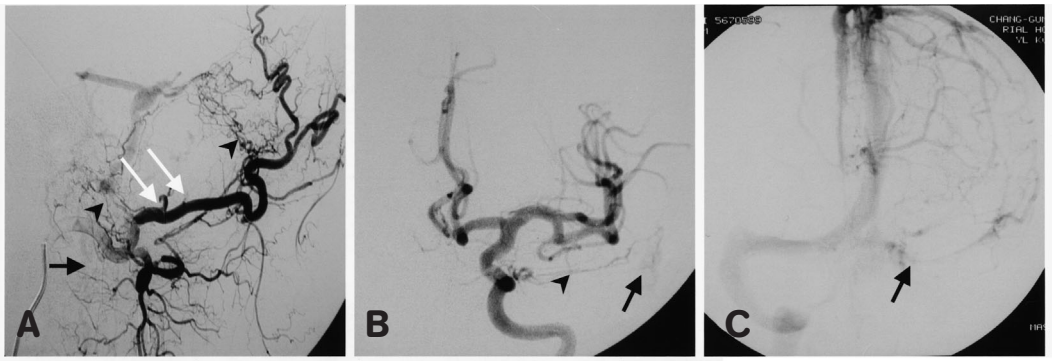

Figure 3. (A) Left ECA angiography (lateral view) showed post-transarterial embolisation of two supplying branches from the left occipital artery (white arrows). Multiple arterial feeders (arrowheads) grow after embolisation with opacification of the dilated perimedullary vein (arrow). (B) Left ICA angiography (AP view) showed a new DAVF extending from the left ICA siphon dural branch to the superior petrosal sinus. (C) Cerebral venous angiography (AP view) showed thrombosis in the left transverse and sigmoid sinuses (arrow).

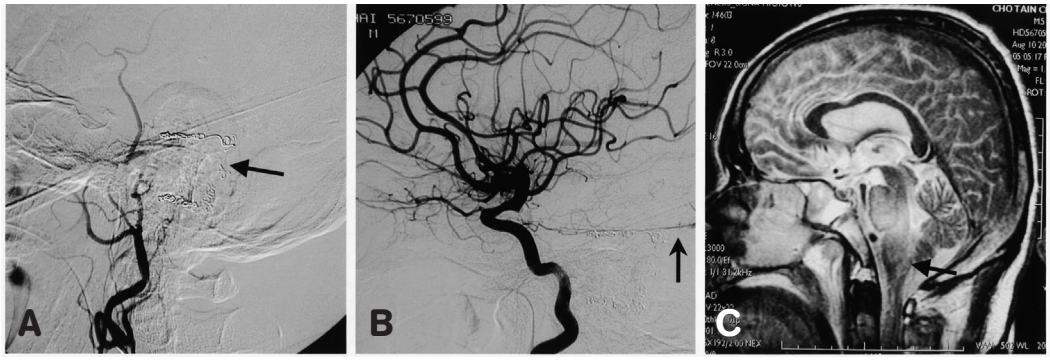

Figure 4. (A) Left ECA angiography (lateral view) showed successful transvenous embolisation of the left superior petrosal sinus and sigmoid sinus (arrow) with obliteration of dural AV fistula. (B) Left ICA angiography (AP view) showed obliteration of the dural AV fistula between left ICA siphon dural branch (arrow) and superior petrous sinus. (C) Saggittal T2-weighted (4500/120 [TR/TE]) MR image showed reduction of edematous changes in the medulla oblongata (arrow).

extensive vascular nidus, we chose endovascular intervention for therapeutic management at first. Transarterial embolisation was performed via the left ECA, and multiple feeding arteries including the occipital (Fig. 3A), middle meningeal and ascending pharyngeal arteries, were occluded with Ivalon powder (150-250μ) and NBCA glue. However, several newly formed feeders from the left occipital artery (Fig. 3A) and left ICA siphon dural branch (Fig. 3B) were noted at the same time and the fistulas remained. Venous approach was then exercised, but the passage of the catheter from the jugular vein to the left transverse and sigmoid sinuses was failed due to thrombosis of the left transverse and sigmoid sinuses (Fig. 3C). Thus, another transvenous embolisation procedure via the left sigmoid sinus, which was exposed by craniotomy, was undertaken. The left sigmoid and superior petrosal sinuses were successfully occluded with the procedure. A follow-up study of cerebral angiography showed complete obliteration of the fistulas (Figs. 4A-B) and a brain MRI revealed reduction of medullary edema (Fig. 4C) as well. Neurologically, improved muscle power on the four limbs (5/5 in bilateral upper limbs and 4+/5 in bilateral lower limbs) was found one week after embolisation. Nystagmus and dysmetria disappeared 2 months after embolisation. His autonomic and respiratory function recovered within 4 and 6 months following the therapeutic procedures, respectively.

DISCUSSION

Cognard et al. classified intracranial DAVFs into five types. Type I intracranial DAVF is located in the main sinus wall with normal antegrade flow typically a benign clinical course. Type II intracranial DAVF shows refluxes into the sinus (IIA) or cortical veins (IIB), with common complications of intracranial hypertension (20% in IIA) and cerebral hemorrhage (10% in IIB)[7]. Type III intracranial DAVF has direct cortical venous drainage without venous ectasia. The usual complication is cerebral hemorrhage in 40% of cases. Type IV intracranial DAVF has direct cortical venous drainage associated with venous ectasia, and cerebral hemorrhage happens in two thirds of cases. Type V intracranial DAVF is characterized by spinal venous drainage and therefore progressive myelopathy in half of the cases[9]. Hence, it is reasonable to choose conservative treatment in patients with type I or type IIA DAVF. On the other hand, types IIB, III, IV, and V DAVFs should be more aggressively treated because the lesions are vulnerable to cerebral hemorrhage or venous infarction.

Ricolfi et al.[10] reported a 75 year-old woman with an intracranial DAVF between the left middle meningeal artery and left superior petrosal sinus showing similar MRI finding of brain to the patient reported here. In Ricolfi's report the patient developed a subacute quadriparesis, followed by swallowing difficulties, sphincter dysfunction and severe dysautonomia a few days later, and was finally cured by the transarterial embolisation. Our patient had sphincter dysfunction at first, followed by paraparesis and hiccup a few days later. Because hiccup may be related to drug[11], gastrointestinal reflex[12], or brain stem lesion[13], a possible localization of the lesion should include supraspinal structures, most likely in the brain stem. But no imaging of spinal cord or brain stem was performed in the community hospital. Two weeks later, the patient suffered from another episode of possible involvement of the pyramidal tract (paraparesis), the bulbar structures (easy choking), the spinocerebellar tract (dysmetria), and the autonomic nerves (dysautonomia such as labile blood pressure), leading to quadriplegia and respiratory failure. In summary, the patient showed rapid progressive myelopathy which included possibilities of both spinal cord lesions (sphincter dysfunction, paraparesis) and brain stem lesions (bulbar signs, pyramidal tract signs, spinocerebellar tract signs, dysautonomia, and respiratory failure). Therefore, we localized the lesions between the thoracic cord and brain stem. Because his clinical course is characterized by two separate episodes and two different anatomical locations, demyelinating disorder was considered first. Differential diagnosis included multiple sclerosis, vascular stroke, tumor, intervertebral disc herniation, syringomyelia, vascular anomaly and post-infectious demyelination. On brain MRI, an extensive high signal intensity lesion in the cervical cord and brain stem was found on T2-weighted imaging. In the meanwhile, a normal CSF result was obtained. Multiple sclerosis, tumor, interverte-

bral disc herniation, syringomyelia and post-infectious demyelination thus were excluded. A cerebral angiography revealed intracranial DAVFs with dilated perimedullary veins which could well be responsible for the myelopathy and venous stroke in this patient.

Intracranial DAVFs are located in the dural wall of the cerebral venous sinus, rather than within the sinus itself. The transverse and sigmoid sinuses are the most common sites. Except for carotid-cavernous fistulas, most intracranial DAVFs are solitary lesions. Multiple lesions are uncommon and account for only 7% of intracranial DAVFs[3]. However, most of the lesions have multiple feeding arteries, with the occipital artery or meningeal branches of the ECA being the most common ones. The tentorial or dural branches of the internal carotid artery and vertebral artery are also frequent feeders[14]. In this case report, we have also noted multiple feeders that were consistent with previous observations.

The pathogenesis of myelopathy in patients with intracranial DAVF is obscure. The most acceptable mechanism is underlied by venous hypertension which was initially proposed by Aminoff et al.[15] and expanded further by Merland[16]. The anterior spinal vein connects superiorly with the anterior medullary vein, which in turn communicates with the anterior pontomesencephalic vein and thence the petrosal vein. Pathological study shows mural thickening with hyalinization of both pial and intramedullary vessels which is consistent with chronic phase of venous hypertension[17]. Accordingly, venous hypertension built in the perimedullary cervical veins may be transmitted to the intrinsic vein of the spinal cord, resulted in reduced arteriovenous pressure gradient within the cord. Stagnation of blood flow and chronic hypoxemia might happen subsequently and lead to progressive necrosis of the spinal cord. We would speculate that the myelopathy in this reported case was caused by venous hypertension of the perimedullary veins.

Practically, the goal for the treatment of intracranial DAVF with perimedullary venous drainage is to occlude the drainage vein proximally (as it exits the arteriovenous shunt) by embolisation[18], surgery, or both. If there is only few feeders in the DAVF, transarterial embolisation usually is the first choice. In those DAVF with multiple arterial feeders, transvenous approach for shunt occlusion should be pursued either before or after transarterial embolisation. The embolisation treatment was considered successful in the occlusion of the fistula in our patient, although he was left with some neurological sequelae.

In conclusion, we would emphasize that intracranial DAVF may present with atypical myelopathy and low brain stem syndrome with an evolutionary clinical course. Proper decision in the management of such patients depend mainly on careful assessment of the clinical presentations and angiographic findings.

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