

# Straight sinus dural arteriovenous fistula presenting with reversible parkinsonism

## A case report and literature review

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### Abstract

**Rationale:** A dural arteriovenous fistula (DAVF) refers to an abnormal direct connection between an intracranial artery and a dural venous sinus. A DAVF presenting with parkinsonism is rare, and is therefore easily misdiagnosed. Therefore, early consideration of DAVF in the differential diagnosis of reversible parkinsonism is necessary.

**Patient concerns:** We present the case of a 51-year-old male with progressive parkinsonism.

**Diagnoses:** He was diagnosed as straight sinus occlusion. Imaging studies revealed a DAVF associated with cerebral hypoperfusion of the lenticular nuclei and frontal lobe white matter.

**Interventions:** Endovascular embolization was performed through his left occipital artery.

**Outcomes:** Treatment resulted in marked clinical improvement that a major improvement of parkinsonism was observed concomitant with no evidence of early venous drainage of this patient.

**Lessons:** DAVF should always be considered as a potential cause of progressive parkinsonism on account of its potential reversibility. Our case suggests a concomitant role of basal ganglia degeneration and frontal white matter hypoperfusion in the pathology of parkinsonism due to DAVF. However, the precise pathophysiology remains to be investigated.

**Abbreviations:** CSF = cerebrospinal fluid, DAVF = dural arteriovenous fistula, DSA = digital subtraction angiography, MELAS = mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes, MRA = magnetic resonance angiography, MRI = magnetic resonance imaging, PMA = posterior meningeal artery.

**Keywords:** dural arteriovenous fistula, endovascular closure, parkinsonism, straight sinus

### 1. Introduction

DAVF refers to an abnormal direct connection between an intracranial artery and a dural venous sinus. Such fistulas account for 10% to 15% of all intracranial vascular malformations.<sup>[1]</sup> Clinical features, natural history, and management options depend on the location and anatomy of the DAVF.<sup>[2]</sup> Common initial symptoms of DAVF include headache, tinnitus, proptosis, decreased cognitive function, and neurological deficits associated

with intracranial hemorrhage. However, DAVF with the onset of parkinsonism symptoms has been reported rarely.<sup>[3]</sup> We present a case of rapidly progressive parkinsonism caused by DAVF, associated with lenticular nuclei degeneration, with dramatic improved following endovascular embolization.

### 2. Case report

A 51-year-old man complained of a 1-month history of slowness of activities without apparent reason. He developed progressively reduced social interaction with slowness of speech and movement, as well as stiff facial expressions. Two weeks before admission, he became slow to eat and developed difficulty swallowing. He had a history of hypertension and hypercholesterolemia, but no history of head trauma, meningitis, toxic ingestion, or poultry exposure. Family history of Parkinson disease was absent. On admission, his vital signs were stable. He had no headache, and a machinery murmur characteristic of an arteriovenous fistula was not heard on auscultation of his head. Because this patient had no fluctuating cognitive defect or visual hallucination, we did not consider Lewy body dementia.

On neurological examination, he was fully alert and oriented with remarkable hypomimia, slow speech, hypophonia, and psychomotor slowness. He had generalized bradykinesia and brisk deep tendon reflexes. Rigidity was present in all 4 limbs, with some hypokinesia on rapid alternating movements. Mild weakness was observed in the lower limbs. No abnormal findings were detected on the remainder of the physical and neurological examination.

Serum chemistries, ceruloplasmin, lactic acid, thyroid hormone levels, inflammatory markers, and tumor markers are all

Editor: Peipeng Liang.

Jiali Pu and Xiaoli Si contributed equally to the study.

This work was supported by the National Natural Science Foundation of China (81520108010 and 81400933), and the Zhejiang Medical Science and Technology Plan Project (2016KYB119).

The authors report no conflicts of interest.

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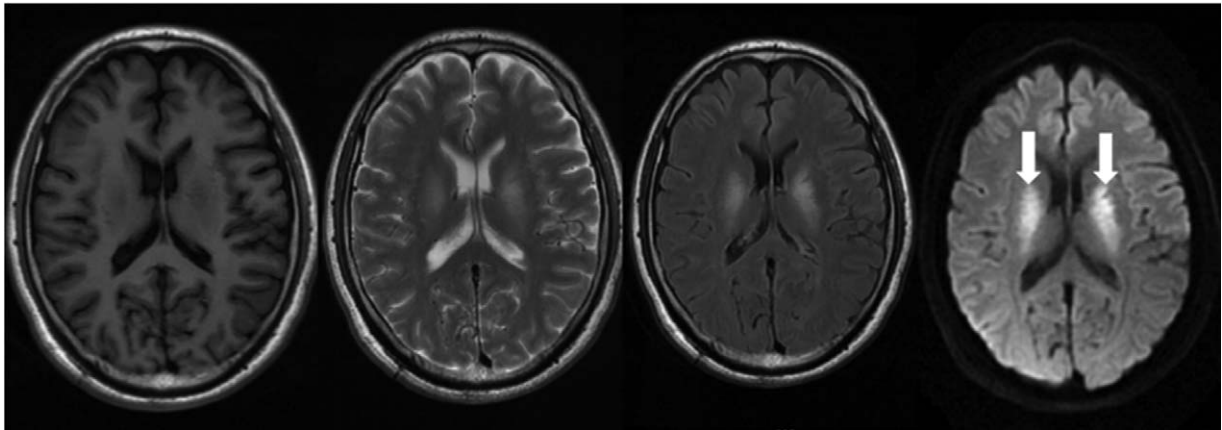
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Medicine (2017) 96:49(e9005)

Received: 17 July 2017 / Received in final form: 19 September 2017 / Accepted: 9 November 2017

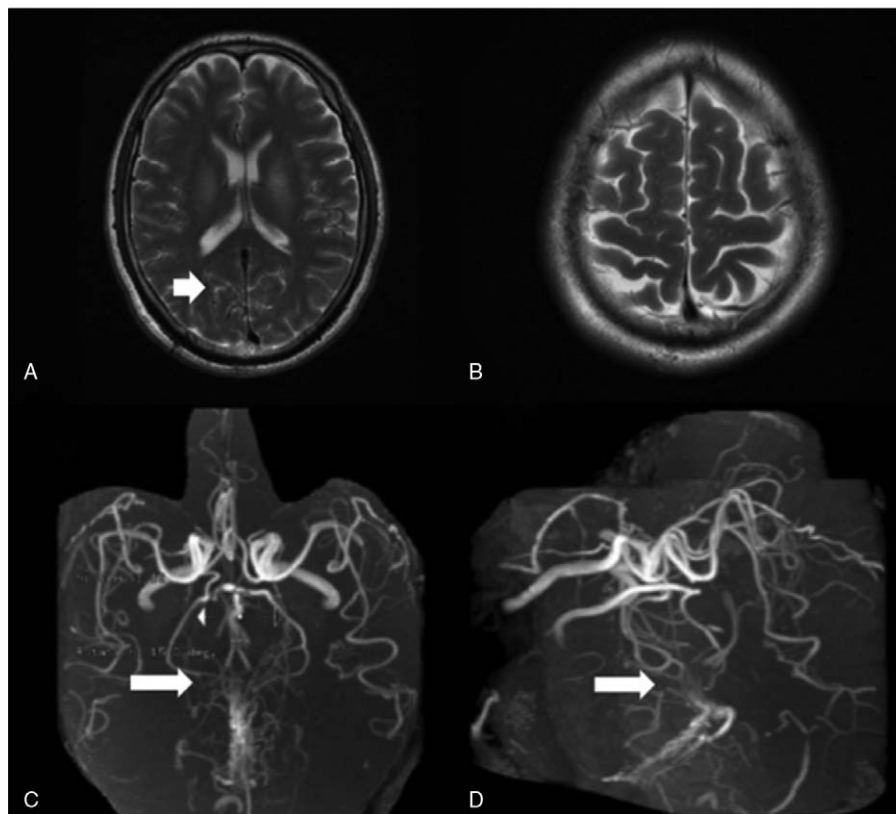
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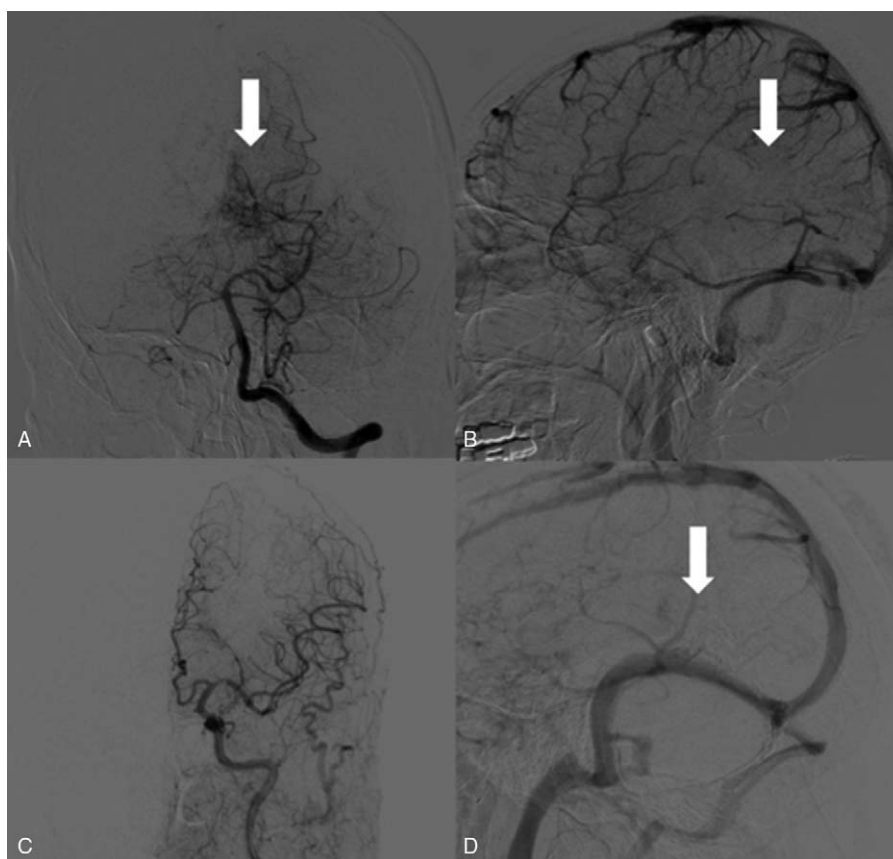
**Figure 1.** Preoperative cranial MRI scans revealed flow void clusters in the location of straight sinus and posterior fossa. Hyperintense signals were found in the medial part of lenticular nuclei bilaterally in T2, Flair, and DWI on sagittal view.

unremarkable. To rule-out autoimmune encephalitis, mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS), and Creutzfeldt-Jakob, we performed lumbar puncture. The cerebrospinal fluid (CSF) tests were normal. T2-weighted magnetic resonance imaging (MRI) revealed flow void clusters in the straight sinus and posterior fossa (Figs. 1 and 2A). In addition, T2-weighted MRI revealed hyperintense signals in the medial part of lenticular nuclei bilaterally and frontal lobe white matter (Figs. 1 and 2A and B). The patient was diagnosed

with Parkinson syndrome and treated with Madopar (62.5 mg TID). Symptoms gradually improved. Magnetic resonance angiography (MRA) revealed multiple vermiform enlarged vessels in the posterior fossa, particularly adjacent to the straight sinus and torcula (Fig. 2C and D). Digital subtraction angiography (DSA) revealed a DAVF adjacent to the tentorial notch, fed by the posterior meningeal artery (PMA), the meningo-hypophyseal trunk, and a branch of the left internal carotid artery (Fig. 3A and B). The cortical venous drainage



**Figure 2.** Preoperative cranial MRI and MRA scans. (A) T2-weighted MRI revealing flow void clusters in the straight sinus and posterior fossa. Hyperintense signals were found in the medial part of lenticular nuclei bilaterally. (B) T2-weighted MRI showing hyperintense signals in frontal lobe white matter. (C and D) MRA showing multiple vermiform enlarged vessels in the posterior fossa, especially adjacent to the straight sinus and torcula on sagittal view.



**Figure 3.** Preoperative and postoperative DSA images. (A and B) Preoperative DSA revealing a DAVF adjacent to the tentorial notch, fed by the PMA, the meningohipophyseal trunk, and a branch of the left internal carotid artery. The cortical venous drainage associated with the varix drained into the vein of Galen, which flowed in a retrograde direction into the vein of Rosenthal, as well as into the internal cerebral veins. (C and D) Postoperative DSA images showing complete closure of the DAVF, with no evidence of early venous drainage. The straight sinus was still occluded, although there was normalization of blood flow in the deep cerebral veins.

associated with the varix drained into the vein of Galen, which flowed in a retrograde direction into the vein of Rosenthal, as well as into the internal cerebral veins (Cognard grade IIa + b). The straight sinus was occluded.

Endovascular embolization was performed through the left occipital artery. Complete closure of the DAVF was confirmed at the end of the procedure. Treatment resulted in marked clinical improvement, particularly of the patient's speech and movement. No obvious rigidity was found in all the 4 limbs after transarterial embolization. A follow-up angiogram showed complete closure of the DAVF, with no evidence of early venous drainage (Fig. 3C and D). The straight sinus was still occluded, although there was normalization of blood flow in the internal cerebral veins and vein of Rosenthal. There was no abnormal blush in the deep structures.

The patient returned for follow-up after 2 months. He had no slowness of movement or response, and his facial expressions were more varied than prior. Brain MRI revealed slight reduction in the hyperintense signal in the lenticular nuclei (Fig. 4A and B), compared with pretreatment MRI (Fig. 1). DSA revealed that the straight sinus was occluded and the tentorial DAVF was completely closed (Fig. 4C and D).

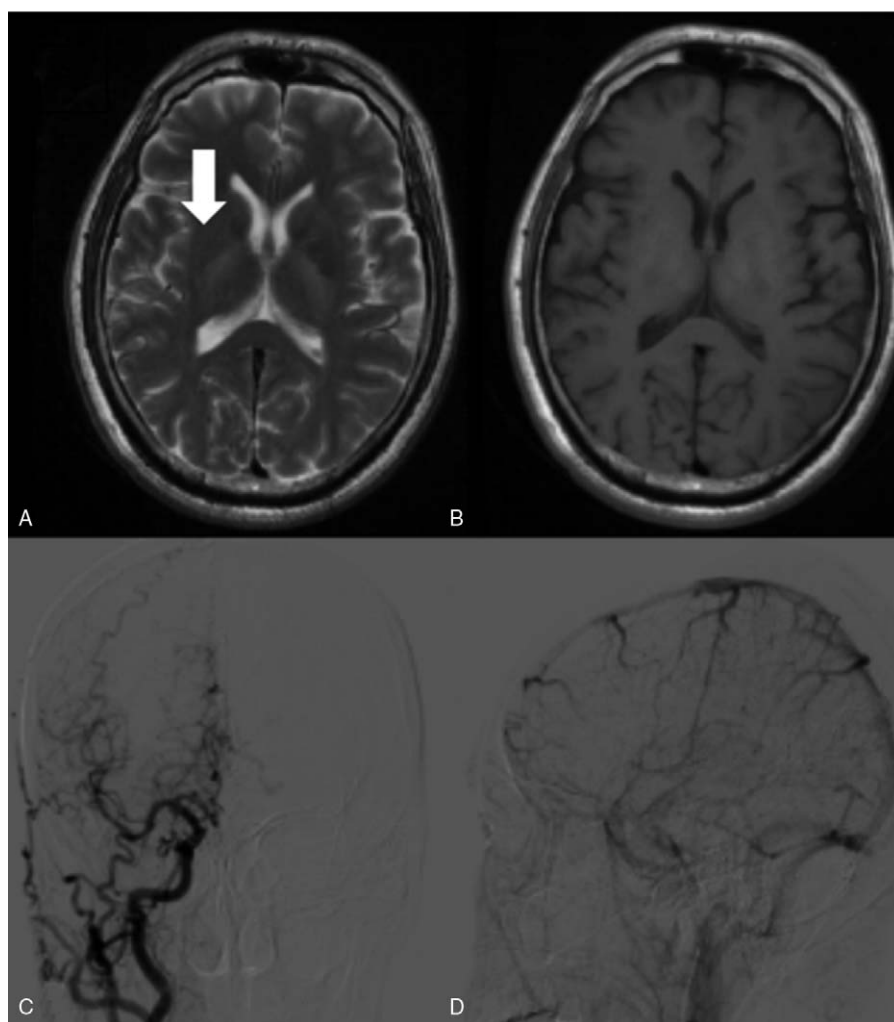
### 3. Discussion

Dural arteriovenous fistula (DAVF) is a rare type of acquired intracranial vascular malformation consisting of a pathological

shunt located within the dura mater.<sup>[4]</sup> DAVF is typically found in middle-aged adults with a median age of onset in the sixth decade.<sup>[5]</sup> DAVF presents with myriad clinical manifestations, depending on the anatomical location and the venous drainage pattern, but it rarely presents as parkinsonism.<sup>[6]</sup> Hence, DAVF is easily misdiagnosed. In this report, we present a rare case of reversible and treatable parkinsonism due to tentorial DAVF with acute thrombosis of the straight sinus, and lenticular nuclei congestion.

DAVF is an acquired and progressive arteriovenous shunt disease, consisting of >2 meningeal feeding arteries that drain into an intracranial vein or venous sinus. It can be separated into 2 types: type I sinus type—DAVF drains through an affected sinus; type II non-sinus type—DAVF with direct reflux to the cortical vein.<sup>[7]</sup> However, the etiology of DAVF remains controversial. Several lines of evidence suggest a 3-stage hypothesis for the formation of DAVF: Stage 1—venous sinus thrombosis is the initial event, possibly combined with other anatomic features that limit venous outflow; stage 2—nascent microscopic fistulas within the wall of the venous sinus, connecting the vaso vasorum to tiny venous tributaries, and enlargement; stage 3—recanalization of the thrombosed venous sinus.<sup>[8]</sup>

In this case, the pathophysiology of parkinsonism due to DAVF remains unclear. DAVF-associated parkinsonism has been attributed to hypoperfusion of the frontal white matter due to venous hypertension caused by either increased blood flow



**Figure 4.** Follow-up cranial MRI scans and DSA images. (A and B) MRI showed slight reduction in the hyperintense signal in lenticular nuclei bilaterally. (C and D) Postoperative DSA images revealing closure of DAVF and occlusion of the straight sinus.

through the draining veins or by obstruction to drainage.<sup>[9]</sup> Others suggest that parkinsonism in DAVF is associated with basal ganglia dysfunction secondary to impaired drainage of the deep internal veins, rather than of white matter lesions.<sup>[10]</sup> A recent study revealed that hemodynamic impairment could cause parkinsonism due to DAVF via an accentuation of the underlying dopamine deficiency in subjects with preclinical stage parkinsonism. This further supports the notion of basal ganglia dysfunction as the pathogenesis of the parkinsonism due to DAVF.<sup>[11]</sup> We observed cerebral hypoperfusion in both basal ganglia and frontal lobe white matter, suggesting a concomitant role of lenticular nuclei degeneration and frontal white matter hypoperfusion in the pathology of parkinsonism due to DAVF.

The brain MRI of our patient revealed straight sinus thrombosis, which might be one potential cause of DAVF. It has been postulated that thrombosis of the dural venous sinuses may be the initial triggering event in the development of DAVFs.<sup>[12]</sup> Occlusion of a dural sinus or a cerebral vein may cause alterations in hemodynamics, which may contribute to arteriovenous shunts within the dura matter.<sup>[13]</sup> Another proposed mechanism suggests that venous thrombosis may result in abnormalities in local vascular growth factors, which

subsequently incite the development of the fistula by neoangiogenesis.<sup>[5]</sup>

To our knowledge, only 3 reports from China have described patients with DAVF who presented with parkinsonism with or without cognitive dysfunction. There have been 15 cases reported outside of China (Table 1). Most cases were reported in Japan. Fifteen of these patients (including the present case we reported) were male with a mean age of onset of 61.5 years (range 40–81 years). Most patients have no history of hypertension, diabetes, hyperlipidemia, or head trauma. The neurological presentations are various, depending on the anatomical location and on the venous drainage pattern. The time from onset to diagnosis of DAVF ranges from 1 week to 5 years.

Previous studies suggested that most DAVFs were found in the region of the transverse, sigmoid, or cavernous sinuses.<sup>[4]</sup> A recent review of parkinsonism due to DAVF indicated that its typical angiographical features include the location of the DAVF at the transverse-sigmoid sinus, reflux into the straight sinus, and probable venous congestion of the basal ganglia.<sup>[21]</sup> We find the most lesions of DAVF are superior sagittal sinus, transverse sinus and sigmoid sinus, and only one DAVF in the straight sinus has been reported (Table 1).

**Table 1**

**Neurological presentation and imaging findings of DAVF presented with progressive dementia and Parkinsonism.**

Case	Country	Age/sex	Underlying condition	Tremor	Rigidity	Brady-kinesia	Gait disturbance	Dementia	Time from onset to treatment	Hyperintense lesion on T2	The region of DAVF	Treatment	Outcome
1 <sup>[14]</sup>	China	50/M	Hypertension diabetes	-	+	+	+	-	1 y	Bilateral frontal lobe	Superior sagittal sinus	TAE	Improved
2 <sup>[15]</sup>	China	>60/M	NR	+	+	+	+	+	NR	NR	NR	L-dopa	Unimproved
3 <sup>[15]</sup>	China	>60/M	Head trauma	-	+	+	+	+	NR	Bilateral cerebral	Left transverse sinus	L-dopa	Unimproved
4 <sup>[16]</sup>	Japan	81/M	NR	-	+	-	+	+	6 mo	Bilateral cerebral white matter	Right transverse sinus	TAE	Improved
5 <sup>[16]</sup>	Japan	55/M	-	-	-	+	+	+	8 mo	Bilateral deep and subcortical white	Right sigmoid sinus	TAE	Improved
6 <sup>[16]</sup>	Japan	78/M	NR	-	+	+	+	+	9 mo	NL	Right sigmoid sinus	TAE	Improved
7 <sup>[12]</sup>	Japan	69/F	-	-	+	+	-	+	Several years	Bilateral cerebral white matter	Left sigmoid sinus	TAE	Unimproved
8 <sup>[17]</sup>	Japan	44/M	NR	-	-	+	+	-	1 wk	NL	Anterior cranial fossa	TAE	Improved
9 <sup>[10]</sup>	Europe	60/F	-	Bilateral action tremor	+	+	+	+	18 mo	Bilateral subcortical white matter	Superior sagittal sinus	TAE	Improved
10 <sup>[6]</sup>	Japan	75/M	-	Bilateral postural tremor	+	+	+	+	14 wk	NL	Superior sagittal sinus and right transverse-sigmoid sinus	TAE	Improved
11 <sup>[18]</sup>	Japan	65/M	NR	-	+	+	+	+	11 mo	Basal ganglia and deep white matter	Left transverse- sigmoid sinus	TAE	Improved
12 <sup>[19]</sup>	US	79/M	Tongue- base cancer	Bilateral postural tremor	+	+	+	-	2 y	NL	Left transverse sinus	TAE	Normalized
13 <sup>[20]</sup>	India	54/M	Diffuse headache	Bilateral postural tremor	+	+	+	+	3 y	Bilateral thalamus and globus pallidus	Bilateral transverse sinuses and straight sinus	TAE	Only minimal change
14 <sup>[20]</sup>	India	40/M	NR	-	+	-	-	+	3 mo	NE	Superior sagittal sinus	TAE	Unchanged
15 <sup>[21]</sup>	Japan	52/F	NR	-	-	+	+	+	3 mo	Basal ganglia and deep white matter	Right transverse- sigmoid sinus	TAE	Improved
16 <sup>[12]</sup>	France	69/M	Hypertension and diabetes	Right hand postural tremor	+	+	+	+	2 y	NL	Superior sagittal sinus	TAE	Improved
17 <sup>[22]</sup>	Canada	62/M	On warfarin for a remote history	Bilateral rest, movement, posture tremor	+	+	+	+	2 mo	Bilateral cerebral hemispheric white matter	Superior sagittal sinus, both transverse sinuses, torcula, and right sigmoid sinus	TAE	Improved
18 <sup>[22]</sup>	Canada	65/F	NR	Bilateral rest, movement, posture tremor	+	+	+	+	5 y	Bilateral cerebral hemispheric white matter	Transverse-sigmoid	TAE	Improved
19 (present case)	China	51/M	Hypertension hyperlipemia	-	-	+	-	+	1 mo	Bilateral lenticular nuclei	Straight sinus	TAE	Improved

F=female, M= male, NE=not examined, NL=normal, NR=not report, TAE=transarterial embolization therapy.

Here, we describe a case of reversible parkinsonism due to DAVF adjacent to the straight sinus and torcula. This can be characterized as a tentorial medial type of tentorial DAVF, which comprise about only 5% of all intracranial DAVFs.<sup>[8]</sup> Cerebral angiography revealed that the DAVF was fed by the PMA, the meningohypophyseal trunk and the left internal carotid artery, with retrograde drainage into deep cerebral veins and cortical veins. To our knowledge, there was one other case of tentorial DAVF associated with parkinsonism in our hospital several years ago. That patient, a 55-year-old woman, presenting with memory deterioration, decreased vision, gait disturbances, and bradykinesia. Her symptoms progressed rapidly over 3 months. She had a history of hypertension and hypercholesterolemia without regular drug treatment. Analysis of blood and CSF showed no abnormalities. Results of MRI showed abnormal signal on both sides of thalamus. DSA confirmed the diagnosis of DAVF in straight sinus and revealed a DAVF fed by the branches of internal carotid artery, external carotid artery, and the left vertebral artery. After endovascular embolization and surgical interruption of arteries feeding the DAVF, the symptoms gradually reversed. These 2 cases provide evidence that parkinsonism due to DAVF may related to localization adjacent to the straight sinus and torcula.

Open surgery and endovascular embolization are potential treatment options for DAVF (Table 1). With the invention of novel materials and devices, endovascular therapy has become the primary method for DAVF treatment. In our case, parkinsonism due to DAVF significantly improved after transarterial embolization therapy. Previous studies showed that the majority of patients with DAVF following endovascular embolization were relieved of their parkinsonian symptoms.<sup>[21]</sup> Unsuccessful treatment was reported in one case of delayed diagnosis with diffuse cerebral atrophy resulting from a thrombosed straight sinus.<sup>[20]</sup> This suggests that it is important to recognize parkinsonism due to DAVF in a timely fashion. However, maybe we have not collected all the literatures and the long-term efficacy of endovascular embolization needs be pursued.

#### 4. Conclusion

It is important to differentiate of parkinsonism due to DAVF from cases associated with neurodegenerative disease, as DAVF-associated deficits may be reversed by endovascular therapy. This reversal of the clinical course, correlated with changes in imaging studies, suggests that the pathogenesis of the parkinsonism due to DAVF is attributed to both basal ganglia dysfunction and frontal white matter hypoperfusion, while the exact pathophysiology of parkinsonism due to DAVF warrants further investigation.

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