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Transverse-sigmoid Sinus Dural Arteriovenous Fistula Presenting with False Localizing Signs of Cavernous Sinus Dural Arteriovenous Fistula Due to Obstructed Cerebral Venous Outflow into the Bilateral Superior Ophthalmic Veins: A Case Report

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Abstract

Cavernous sinus (CS) dural arteriovenous fistula (dAVF) presents ocular symptoms and visual disorders due to retrograde drainage from the CS into the superior ophthalmic vein (SOV). Some papers reported non-CS dAVFs with those symptoms. We present a unique case of transverse-sigmoid sinus (TSS) dAVF with localizing signs of CS dAVFs resulting from congested cerebral venous outflow into the CSs and SOVs in an 86-year-old female patient. Right pulsatile tinnitus and chemosis appeared a few years ago. After experiencing progressive bilateral blurred vision and decreased visual acuity with papillary edema for a few months, she was admitted to our hospital. Cerebral angiography demonstrated right sigmoid sinus dAVF with retrograde venous reflux into the superior sagittal sinus (SSS) and contralateral TSS without cortical venous reflux. Under the influence of congestion of the SSS, cerebral venous outflow drained into the CSs subsequently into the SOVs. Trans-arterial embolization using ethylene-vinyl alcohol copolymer was conducted via the right middle meningeal artery. After treatment, right chemosis immediately improved. The 2-month follow-up examination revealed resolution of bilateral visual acuity and improvement of papillary edema. Our case demonstrates that retrograde drainage into the SOVs with not only directly arterialized shunted flow but also congested cerebral venous outflow can cause CS dAVFs-like symptoms.

Keywords: intracranial dural arteriovenous fistulas, cavernous sinus, transverse-sigmoid sinus

Introduction

Intracranial dural arteriovenous fistulas (dAVFs) are abnormal vascular connections between meningeal feeding arteries and the intradural venous system.[1)] Symptomatology of dAVF typically depends on the fistula location, venous drainage patterns, and available collaterals. Cavernous sinus (CS) dAVF classically presents with ocular symptoms and visual disorders due to abnormal retrograde venous drainage from the CS into the superior ophthalmic vein (SOV). Venous drainage into the SOV is a well-known phenomenon responsible for ocular symptoms, such as chemosis and exophthalmos.[2,3)] Contrarily, those symptoms are rare in intracranial dAVFs occurring at locations other than the CS.[1)]

Previous studies have reported non-CS dAVF cases with ocular signs and visual disorders.[4–7)] In most cases, detailed analyses of the pathophysiology show the fistulas located at the transverse-sigmoid sinus (TSS) or internal jugular vein (IJV). The ocular symptoms result from specific retrograde venous drainage routes via the inferior petrosal sinus toward the CS or the vein of Labbe to the superficial middle cerebral vein toward the CS, both subsequently passing into the SOVs.[4–7)] Moreover, the sphenoparietal sinus and uncal vein are also reported as intermediate routes of venous drainage leading to

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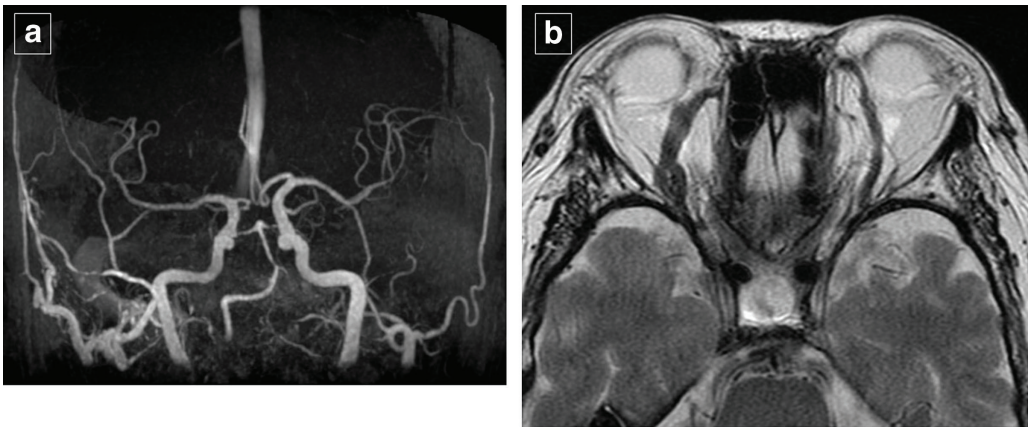

Fig. 1 Initial preoperative MRI and MRA. (a) MRA revealed the manifestation of the right TSS and SSS with signs of tiny feeding vessels assembling into the right sigmoid sinus. (b) MRI revealed the dilation of the bilateral SOVs with predominance on the right side. MRA: magnetic resonance angiography, MRI: magnetic resonance imaging, SOVs: superior ophthalmic veins, SSS: superior sagittal sinus, TSS: transverse-sigmoid sinus.

the CS.[6] In these cases, the symptoms are due to the specific flow pattern involving retrograde drainage into the SOVs, through the CS, directly from the fistulous points.

Herein, we present a unique case of TSS dAVF manifesting with localizing signs of CS dAVF resulting from obstructed cerebral venous outflow into the bilateral CSs and SOVs due to severe congestion of the superior sagittal sinus (SSS).

Case Report

An 86-year-old female patient was admitted to our hospital for progressive bilateral blurred vision and decreased visual acuity over the past few months. She had a history of right pulsatile tinnitus and ipsilateral chemosis for a few years, without any apparent cause.

The preoperative clinical signs and initial ophthalmological examination findings revealed bilateral normal intraocular pressure of 16 mmHg, bilateral papillary edema, and decreased visual acuity (20/133) on both eyes. There was no evidence of cranial nerve palsies.

The initial preoperative magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) findings demonstrated manifestation of the right TSS and SSS. Additionally, there were signs of tiny feeding vessels assembling into the right sigmoid sinus (Fig. 1a). Notably, bilateral SOV dilation was observed predominantly on the right side (Fig. 1b).

Diagnostic cerebral angiography showed extensive right sigmoid sinus dAVF with retrograde venous reflux into the SSS and contralateral TSS without cortical venous reflux (Fig. 2a–2d). Under the influence of heavy congestion of the SSS and the impairment of both superior and inferior petrosal sinus drainages, the cerebral venous outflow was severely hindered and could not flow into the SSS. Thus, outflow had concentrated into the bilateral CSs and subsequently into the bilateral SOVs and pterygoid plexus (Fig. 2e–2g). In this case, the right vein of Labbe was not fully developed, and the right IJV was occluded at the cervical level. This fistulous connection was mainly supplied by branches from the right middle meningeal (petrosquamous branch), posterior auricular, and occipital arteries. This dAVF is considered type 2a, according to the Cognard classification.[1] Left internal carotid angiography revealed that the fistulous point at the right sigmoid sinus was also supplied from the left ascending pharyngeal artery arising from the left internal carotid artery (Fig. 2h).

Transarterial embolization (TAE) using ethylene-vinyl alcohol copolymer (Onyx; ev3-Covidien, Irvine, CA, USA) was performed via the right petrosquamous branch of the middle meningeal artery (Fig. 3a). Under general anesthesia, a 7-French (Fr) Cook shuttle sheath (80 cm; Cook Medical, Bloomington, IN, USA) was advanced up to the external carotid artery via a femoral site. From the shuttle sheath, a 4 × 11-mm Scepter XC balloon catheter (MicroVention Inc., Aliso Viejo, CA, USA) with a microguidewire (ASAHI CHIKAI 14; Asahi Intecc Co Ltd., Aichi, Japan) and a Defrictor Nano Microcatheter (Medico's Hirata, Osaka, Japan) with a 4.2-Fr Fubuki guiding catheter (120 cm; Asahi Intecc Co Ltd.) with a microguidewire (ASAHI CHIKAI X 010; Asahi Intecc Co Ltd.), respectively, were navigated up to the portion just proximal to the fistula (Fig. 3b and 3c). Onyx 18 was injected from the Defrictor microcatheter with the Scepter XC for flow control. The total amount of Onyx 18 used was 10.2 ml over an injection time of 84 minutes.

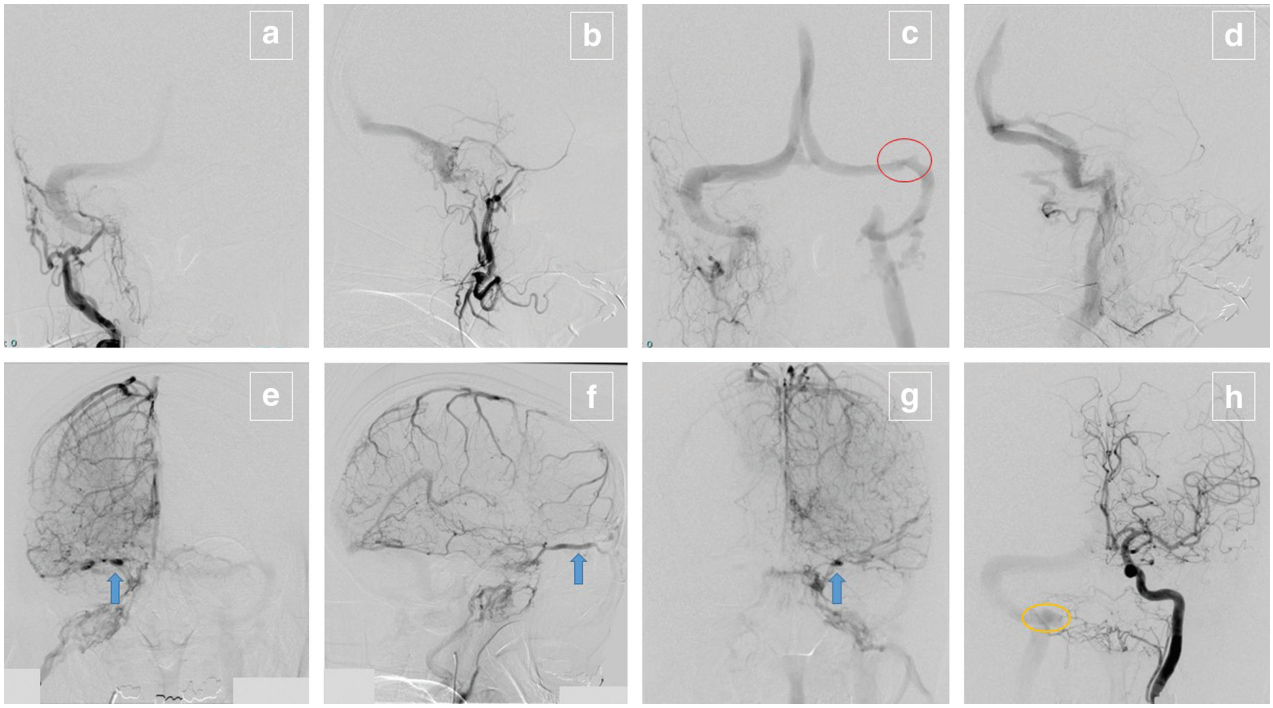

Fig. 2 Preoperative cerebral angiogram revealing a Cognard type 2a right sigmoid sinus dAVF. (a–d) Right external carotid angiography demonstrated an extensive dAVF at the right sigmoid sinus. This dAVF exhibited venous reflux into the SSS and left TSS without cortical venous reflux. (e and f) Right internal carotid angiography revealed a severely hindered cerebral venous outflow that mainly flowed out into the CS and subsequently into the right SOV and pterygoid plexus due to the congestion of the SSS. The right SOV was significantly dilated (blue arrows). In addition, there was mild stenosis in the left TSS (red circle). (g) Left internal carotid angiography revealed dilation of the left SOV (blue arrow) due to obliteration of cerebral venous outflow. (h) Left internal carotid angiography revealed that the fistulous point at the right sigmoid sinus (orange circle) was supplied from the left ascending pharyngeal artery arising from the left internal carotid artery. CS: cavernous sinus, dAVF: dural arteriovenous fistula, SSS: superior sagittal sinus, SOV: superior ophthalmic vein, TSS: transverse-sigmoid sinus.

Using TAE, near-complete packing of the entire right part of the sigmoid sinus was obtained (Fig. 3d–3e). Right carotid angiography performed immediately following TAE confirmed the obliteration of the fistula (Fig. 3f). Furthermore, right carotid angiography demonstrated cerebral venous blood flow normalization, which indicated that the SSS was restored as a functional component of normal cerebral venous return. Retrograde drainage of the SOV was still observed in the venous phase; however, drastic flow reductions were observed (Fig. 4a–4c). Left carotid angiography revealed that only a slight residual shunt at the extreme proximal segment of the right sigmoid sinus persisted, which was fed by the left ascending pharyngeal artery. However, further treatment was avoided because the distal segment of the right sigmoid sinus was completely obliterated by Onyx; consequently, the entire shunt drained only proximally and not into the intracranial side (Fig. 4d). Left carotid angiography also showed left cerebral venous drainage normalization, although retrograde drainage of the SOV was still observed in the venous phase (Fig. 4e). No complications occurred during and after the procedure. Following treatment, right chemosis resolved rapidly. The 2-month follow-up examination revealed gradual recovery of bilateral visual acuity (20/50 in the right eye and 20/67 in the left eye) and reduction of bilateral papillary edema. The intraocular pressure was slightly lower at 14 mmHg. MRI at 2 months following treatment revealed significant contraction of both SOVs (Fig. 4f).

The patient has consented to the submission of this case report to the journal.

Discussion

Our case demonstrates that retrograde drainage into the SOVs upon congested cerebral venous outflow concurrent with directly arterialized shunted flow can manifest with signs and symptoms consistent with CS dAVF. To our best knowledge, there are no reports of non-CS dAVF cases with ocular signs

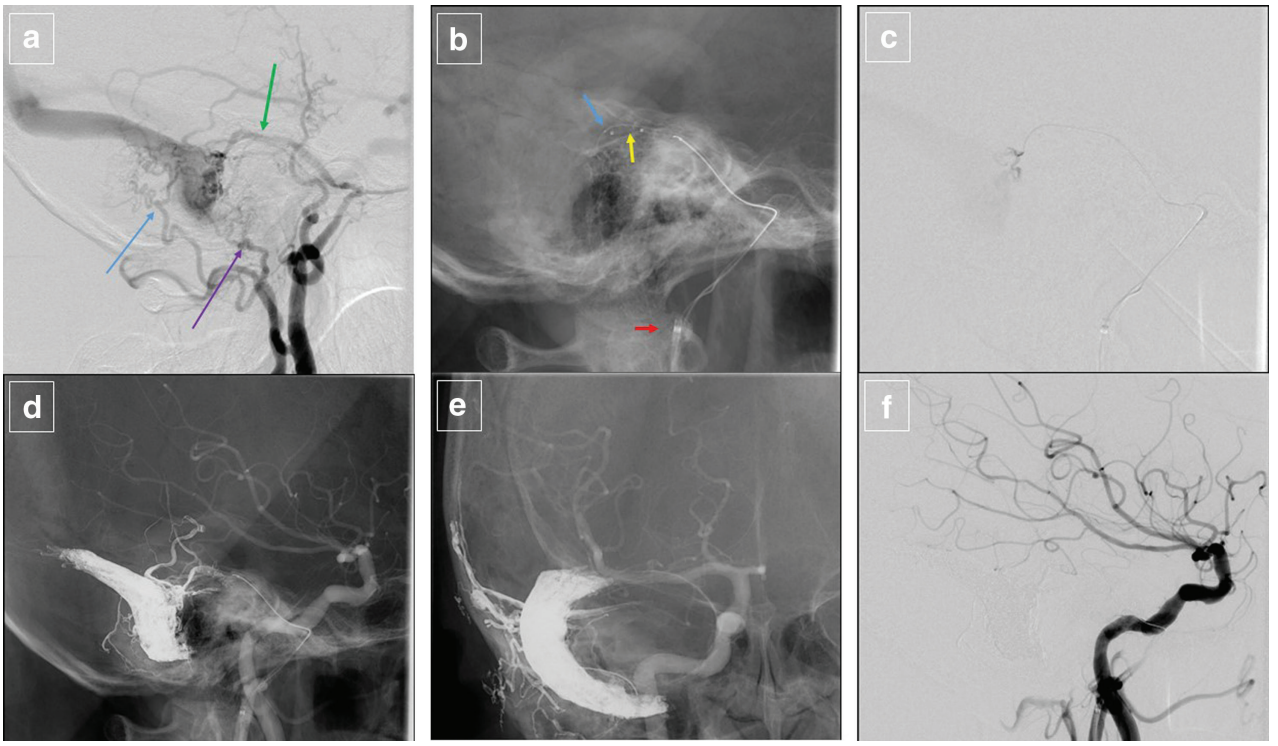

Fig. 3 Transarterial sinus occlusion using Onyx via the petrosquamous branch of the right MMA. (a) Right external carotid angiography demonstrated that this fistulous connection was mainly supplied by the branches from the right middle meningeal (petrosquamous branch) (green arrow), posterior auricular (purple arrow), and occipital (blue arrow) arteries. (b and c) A 4 × 11-mm Scepter XC balloon catheter (yellow arrow) and a Defrictor Nano Microcatheter (blue arrow) with a 4.2-French Fubuki guiding catheter (120 cm, red arrow) were, respectively, navigated up to the portion just proximal to the fistula. (d and e) Non-subtracted image of the skull demonstrated the Onyx cast of the sigmoid sinus and the petrosquamous branch of the MMA used for embolization. (f) Right common carotid angiography revealed the obliteration of the shunt immediately following treatment. MMA: middle meningeal artery.

and visual disorders with the same etiology in the existing literature.

Typically, TSS dAVFs present with diverse clinical symptoms, including pulsatile tinnitus, seizures, global cognitive decline, and intracranial hemorrhage.[8] Owing to retrograde drainage of the SOVs, CS dAVFs commonly manifest with ocular symptoms related to localized orbital venous congestion, such as chemosis, ptosis, exophthalmos, ophthalmoplegia, and arterialized conjunctival injection.[2] CS dAVFs can also cause visual disorders.[3] A prospective study involving 46 CS dAVF cases observed visual disorders in only three cases (6.5%), whereas ocular symptoms were observed in all cases.[9] The pathophysiology of visual disorders has not been thoroughly investigated. Kondo et al. demonstrated that visual disorders might be caused by ocular hypertension, retinal venous congestion, or both resulting from unsuccessful drainage of the SOVs.[3] Conversely, ocular symptoms related to papillary edema are not uncommon in non-CS intracranial dAVFs, which are often suggestive of increased intracranial pressure resulting from generalized intracranial venous hypertension.[7] However, progressive orbital venous congestion can also cause secondary papillary edema, together with glaucoma, choroidal effusions, retinal detachment, retinal vessel tortuosity, and sluggish flow with an associated branch or central retinal vein occlusion, macular edema, and ischemic optic neuropathy.[10,11] Except for pulsatile tinnitus, our patient's symptoms could be divided into three parts. The first two, chemosis and decreased visual acuity, can be categorized into ocular symptoms and visual disorders, respectively. Both symptoms were certainly due to orbital venous congestion with retrograde venous reflux into the SOVs. Furthermore, the mechanism of bilateral papillary edema could be associated with both generalized intracranial venous hypertension and progressive orbital venous congestion, although, in our patient, intracranial pressure was not measured. The relatively long period from onset of chemosis and tinnitus to

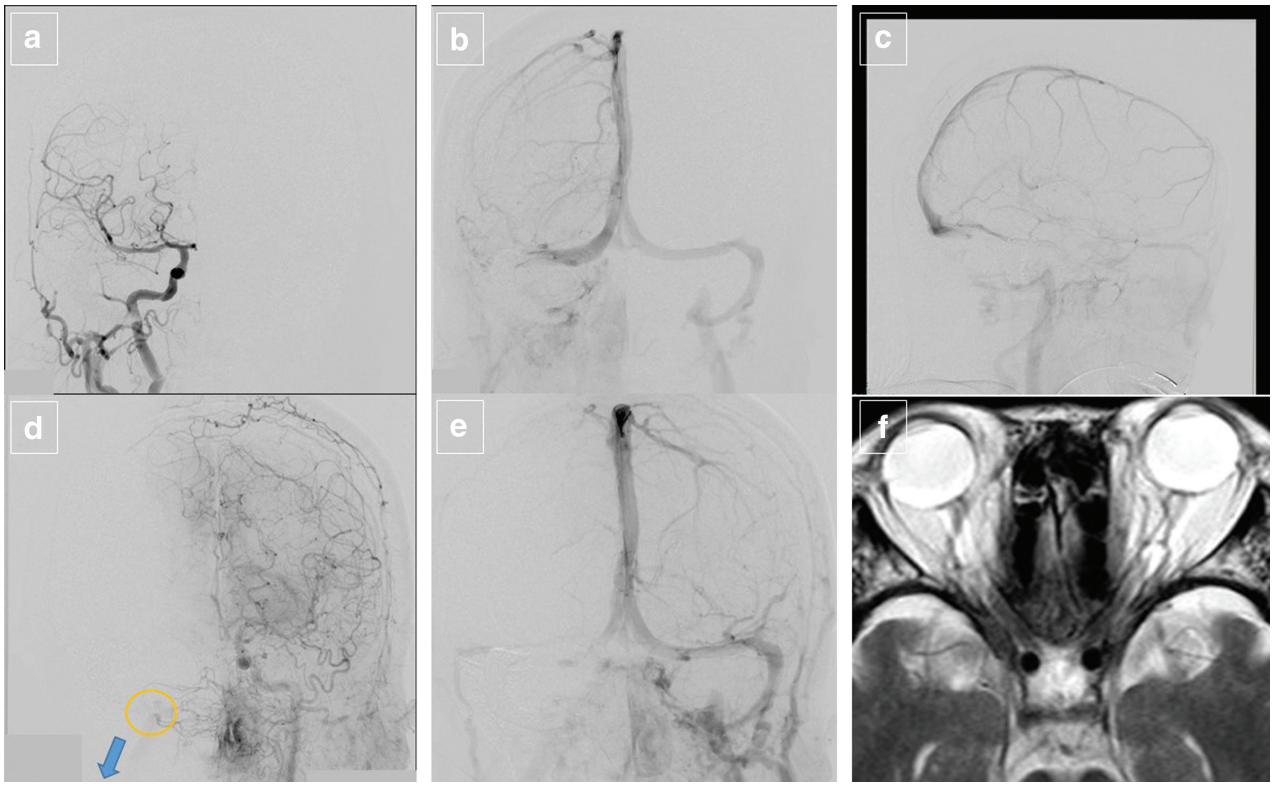

Fig. 4 Angiography immediately after treatment and MRI 2 months after treatment. (a–c) Right common carotid angiography revealed that the SSS was restored to be a part of normal cerebral venous return. Retrograde drainage of the SOVs was still confirmed in the venous phase; however, the flow drastically decreased. (d and e) Left common carotid angiography demonstrated only a residual shunt at the extreme proximal segment of the right sigmoid sinus (orange circle) fed by the left ascending pharyngeal artery. However, the shunt flow drained only proximally (blue arrow) and not into the intracranial side. It also showed left cerebral venous drainage normalization, although retrograde drainage of the SOV was still observed in the venous phase. (f) MRI 2 months following treatment revealed significant contraction of both SOVs. MRI: magnetic resonance imaging, SOVs: superior ophthalmic veins, SSS: superior sagittal sinus.

the manifestation of the decreased visual acuity suggests gradual progression of orbital venous congestion with non-arterialized, low venous retrograde outflow into the SOVs. Of note, there was no cortical venous reflux in our case. This is because the right vein of Labbe was not fully developed originally. The cause of the pathophysiology in this case was the SSS congestion. The congestion may be caused by the impairment of the right superior and inferior petrosal sinus drainages. In addition, the mild stenosis of the left transverse sinus (Fig. 2c) may influence the congestion. Robert et al. reported that non-CS dAVF presenting with ocular signs could be classified into four types owing to their pathologic mechanism: local venous reflux into the ophthalmic veins, massive venous engorgement of the cerebrum responsible for intracranial venous hypertension, compression of the oculomotor nerve by venous dilation, and intraorbital fistula with drainage into the SOV.[6] Our case study suggests that the precipitating mechanism for venous reflux into the ophthalmic veins should be divided into two subtypes: arterialized flow or congested cerebral venous outflow.

Concerning endovascular surgery for TSS dAVF, transvenous embolization (TVE) is recognized as the most effective treatment, including coil direct packing TSS, Onyx TVE, and balloon-assisted Onyx TVE.[12] Currently, TAE is extensively used, especially after the introduction of Onyx.[13] Particularly, in cases with venous sinus irregularities, such as focal occlusion or an isolated segment, TAE without reconstruction of the TSS is highly effective.[14] In our case, the right IJV was occluded, and the right sigmoid sinus was entirely affected and did not allow for significant normal venous drainage. Therefore, we assumed that the affected sinus might be permanently occluded without neurological sequelae.

In conclusion, in addition to the mechanism of retrograde drainage into the SOVs with arterialized

shunted flow, a non-CS dAVF with retrograde drainage into the SOVs of continually non-arterialized cerebral venous outflow can also show localizing signs consistent with CS dAVF. Severe congestion of the SSS can be an inciting factor to elicit this unique pathophysiology.

Conflict of Interest Disclosure

All authors have no conflict of interest.

References

1) Cognard C, Gobin YP, Pierot L, et al.: Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. *Radiology* 194: 671–680, 1995
2) Miller NR: Diagnosis and management of dural carotid-cavernous sinus fistulas. *Neurosurg Focus* 23: E13, 2007
3) Kondo R, Kumabe T, Yamamoto D, Koizumi H, Kuroda H, Miyasaka K: Visual disorders caused by cranial arteriovenous fistula with venous drainage into the superior ophthalmic vein. *Interv Neuroradiol* 25: 460–468, 2019
4) Ghorbani M, Asaadi S, Wipplinger C, Griessenauer CJ, Zangi-Abadi F, Mortazavi A: Dural arteriovenous fistulas with venous drainage patterns inducing ocular manifestations mimicking a carotid cavernous fistula: report of 2 cases. *World Neurosurg* 127: 216–219, 2019
5) Lv X, Zhang J, Li Y, Jiang C, Wu Z: Dural arteriovenous fistula involving the transverse sigmoid sinus presenting as chemosis. A case report. *Neuroradiol J* 21: 428–432, 2008
6) Robert T, Botta D, Blanc R, et al.: Ocular signs caused by dural arteriovenous fistula without involvement of the cavernous sinus: a case series with review of the literature. *AJNR Am J Neuroradiol* 37: 1870–1875, 2016
7) Spitze A, Gersztenkorn D, Al-Zubidi N, Yalamanchili S, Diaz O, Lee AG: Transverse and sigmoid sinus dural arteriovenous fistula mimicking idiopathic intracranial hypertension and carotid cavernous fistula. *Neuroophthalmology* 38: 29–35, 2014
8) Oh JT, Chung SY, Lanzino G, et al.: Intracranial dural arteriovenous fistulas: clinical characteristics and management based on location and hemodynamics. *J Cerebrovasc Endovasc Neurosurg* 14: 192–202, 2012
9) Pashapour A, Mohammadian R, Salehpour F, et al.: Long-term endovascular treatment outcome of 46 patients with cavernous sinus dural arteriovenous fistulas presenting with ophthalmic symptoms. A non-controlled trial with clinical and angiographic follow-up. *Neuroradiol J* 27: 461–470, 2014
10) Stiebel-Kalish H, Setton A, Nimii Y, et al.: Cavernous sinus dural arteriovenous malformations: patterns of venous drainage are related to clinical signs and symptoms. *Ophthalmology* 109: 1685–1691, 2002
11) Grumann AJ, Boivin-Faure L, Chapot R, Adenis JP, Robert PY: Ophthalmologic outcome of direct and indirect carotid cavernous fistulas. *Int Ophthalmol* 32: 153–159, 2012
12) Xu K, Yang X, Li C, Yu J: Current status of endovascular treatment for dural arteriovenous fistula of the transverse-sigmoid sinus: a literature review. *Int J Med Sci* 15: 1600–1610, 2018
13) Abud TG, Houdart E, Saint-Maurice JP, et al.: Safety of onyx transarterial embolization of skull base dural arteriovenous fistulas from meningeal branches of the external carotids also fed by meningeal branches of internal carotid or vertebral arteries. *Clin Neuroradiol* 28: 579–584, 2018
14) Eftekhar B, Morgan MK: Surgical management of dural arteriovenous fistulas of the transverse-sigmoid sinus in 42 patients. *J Clin Neurosci* 20: 532–535, 2013

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