Extracranial arteriovenous malformation and subsequent contralateral cavernous sinus dural arteriovenous fistula showing abducens nerve palsy: illustrative case

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BACKGROUND Extracranial arteriovenous malformations (AVMs) are rare clinical entities and on rare occasions cause neurological symptoms. The authors report a case of an extracranial pterygoid AVM and a subsequent contralateral cavernous sinus dural arteriovenous fistula (dAVF) presenting with abducens nerve palsy.

OBSERVATIONS An 80-year-old woman was referred to the authors' hospital with left abducens nerve palsy followed by right ophthalmalgia. Magnetic resonance imaging (MRI) showed abnormal vessel staining in the left pterygoid and the right inferior petrosal sinus (IPS). Cerebral angiography revealed a left pterygoid AVM draining into the right IPS via the cavernous sinus (CS). A dAVF in the right CS was also revealed. The right ophthalmalgia disappeared spontaneously, and, 4 months later, the left abducens nerve palsy also disappeared after conservative management. Follow-up MRI showed spontaneous regression of the AVM and dAVF. The disappearance of the dAVF was considered to be due to spontaneous regression of the left pterygoid AVM and the consequent decrease in venous pressure of the CS, and the symptoms eventually disappeared.

LESSONS The authors treated an extremely rare case of extracranial AVM with dramatic changes in vascular structure and symptoms. Understanding of the pathophysiology between symptoms and dynamic changes in the vascular structure is essential for providing the appropriate treatment.

<https://thejns.org/doi/abs/10.3171/CASE21265>

KEYWORDS abducens nerve palsy; arteriovenous malformation; cavernous sinus; dural arteriovenous fistula

Arteriovenous malformations (AVMs) can occur anywhere in the body, but extracranial AVMs are rare and, on very rare occasions, can lead to the development of neurological symptoms.¹ The etiology of AVMs and dural arteriovenous fistulas (dAVFs) remain unclear, and there are some reports of spontaneous regression.²⁻⁴ We report a case of a left pterygoid AVM draining into the cavernous sinus (CS), which could have contributed to the development of left abducens nerve palsy and the formation of a contralateral CS dAVF. With conservative treatment, the vascular lesions disappeared spontaneously, and the symptoms improved. This case report includes a review of related literature, and a reasonable mechanism is proposed. Cases of extracranial AVMs leading to the development of neurological symptoms are also reviewed.

Illustrative Case

An 80-year-old woman was referred to our hospital with a 1-week history of diplopia. On admission, the diplopia was found to be due mainly to left abducens nerve palsy, and the patient also developed pain in the right eye with the diplopia's appearance. Contrast-enhanced magnetic resonance imaging (MRI), including a 4-dimensional time-resolved magnetic resonance angiography using a keyhole method performed at the time of admission, showed early visualization of the right CS and inferior petrosal sinus (IPS) following the vessels in the left pterygopalatine fossa, suggesting the presence of a shunt at the same sites (Fig. 1). A few days after admission, cerebral angiography was performed. Right common carotid artery angiography showed a dAVF fed by the branch of the

ABBREVIATIONS AVM = arteriovenous malformation; CS = cavernous sinus; dAVF = dural arteriovenous fistula; IPS = inferior petrosal sinus; MRI = magnetic resonance imaging.

INCLUDE WHEN CITING Published August 9, 2021; DOI: 10.3171/CASE21265

SUBMITTED April 28, 2021. **ACCEPTED** May 24, 2021.

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FIG. 1. Contrast-enhanced 4-dimensional time-resolved magnetic resonance angiography using keyhole method performed on admission shows early visualization of the right cavernous sinus and inferior petrosal sinus (double arrows) following the vessels in the left pterygopalatine fossa (arrows), suggesting the presence of a shunt at the same sites.

external carotid artery in the right CS, but no regurgitation into the cortical veins or contralateral outflow was observed (Fig. 2). Left common carotid artery angiography showed a deeply stained vascular lesion in the pterygopalatine fossa, suggesting the presence

of a pterygoid AVM. The diameter of the nidus was about 3 cm. Superselective angiography of branches of the external carotid artery failed because the microcatheter could not be placed stably in these branches. The left maxillary artery and ascending pharyngeal artery were the main feeding arteries, and the drainer vein belonged to the left external jugular vein system. Regurgitation to the contralateral CS and IPS was observed via the pterygoid and clivus plexuses. No abnormal vascular image was found in the left CS, and the left IPS was normally perfused in the venous phase. Venous return stagnation from the left sigmoid sinus to the external jugular vein suggested venous hypertension around the external jugular vein system (Fig. 3).

Because no regurgitation into the cortical vein was observed, the patient was conservatively managed. The right ophthalmalgia disappeared spontaneously after cerebral angiography, and the left diplopia and abducens nerve palsy gradually improved. MRI performed 1 month after the onset showed a decrease in the shunting flow in the pterygoid AVM, and stagnation from the left sigmoid sinus to the external jugular vein had disappeared, although the condition of the dAVF in the right CS had not changed significantly (Figs. 3 and 4). The diplopia and left abducens nerve palsy disappeared completely 4 months after the onset. MRI at that time revealed spontaneous regression of the right CS dAVF and the left pterygoid AVM (Fig. 4C and F). Because the symptoms and the vascular structure had changed dynamically, we decided to diligently continue detailed imaging follow-up.

Discussion

Extracranial AVMs have been reported in the field of oral surgery, but they are generally rare.⁵ Many of the draining tracts involved with extracranial AVMs are, as one might suspect, extracranial and thus are not often encountered by neurosurgeons. Cases with drainer veins entering the skull and involving the development of neurological symptoms are extremely rare. To the best of our knowledge, there have been only 5 cases of extracranial AVMs with neurological symptoms, including our case, reported in the literature so far (Table 1).⁵⁻⁷ The average age of these 5 cases was 53.2 years. The locations included the foramen magnum, sphenoid bone, and pterygoid. Neurological symptoms such as pulsatile tinnitus

FIG. 2. **A and B:** Right common carotid angiograms show a dAVF with the branch of the external carotid artery as the feeding artery in the right cavernous sinus. No regurgitation into the cortical veins or contralateral outflow is observed. **A:** Anteroposterior (A-P) view. **B:** Lateral view. **C and D:** Left common carotid artery angiograms show a deeply stained vascular lesion in the pterygopalatine fossa fed by the left maxillary artery and ascending pharyngeal artery. Regurgitation to the contralateral cavernous sinus and inferior petrosal sinus is observed via the pterygoid and clivus plexus. **C:** A-P view. **D:** Lateral view.

FIG. 3. Left common carotid angiogram, late venous phase, shows venous return stagnation from the left sigmoid sinus to the external jugular vein (arrow), suggesting venous hypertension around the external jugular vein system.

and abducens nerve palsy varied, depending on the drainage location, and in case 3 in Table 1, a cerebral hemorrhage developed. Our case was especially interesting, not only because of the rarity of the extracranial AVM leading to the development of neurological symptoms but also because of the unusual radiological presentation with dAVF development. In this case, the following 2 points were characteristic. First, it was speculated that the left pterygoid AVM had drained into the skull, causing an increase in bilateral CS venous pressure, which could have contributed to the left abducens nerve palsy and development of right CS dAVF. Second, the dAVF disappeared spontaneously with the regression of the AVM.

In this case, right ophthalmalgia occurred after the left abducens nerve palsy. MRI and cerebral angiography showed a dAVF in the right CS, which could have been the cause of the right ophthalmalgia. There was no evidence in the radiological findings that could directly explain the left abducens nerve palsy. However, Yokokawa et al.⁸

reported a case in which dAVF led to an increase in pressure in the CS on one side, causing cranial nerve symptoms on the opposite side. Pressure increase in the left CS is necessary to explain the occurrence of the left abducens nerve palsy. Golub and Bordon⁹ also reported that the pterygoid plexus is a complex of veins with comprehensive connections to surrounding veins, including the CS, inferior ocular vein, and facial vein. Therefore, it was speculated that the drainage of the left pterygoid AVM first returned to the ipsilateral CS rather than the right CS, and the pressure in the right CS increased. By the time we had performed cerebral angiography, the AVM drainage to the ipsilateral CS was already occluded, and only the shunt to the right CS via the clivus plexus remained. There is also a report that intracranial AVM could be affected by venous thrombosis, altering the drainage route.¹⁰ In our case, there was also stagnation from the left sigmoid sinus to the external jugular vein. The most likely hypothesis is that the earlier thrombosis occurred below AVM levels, resulting in extracranial-to-intracranial drainage changes, which seemed to be the first trigger for a series of changes in vascular lesions.

The etiology of dAVF is generally thought to occur after events such as venous sinus thrombosis, head trauma, craniotomy, and tumor-induced sinus compression.¹¹ A previous report¹¹ stated that increased venous pressure may contribute to the development of dAVF. Also, there are case reports in which dAVF in the CS on one side caused increased bilateral intracavernous pressure, resulting in bilateral ocular symptoms.⁸ Therefore, in our case, the right CS dAVF appears to have resulted from increased intracavernous pressure due to drainage from the left pterygoid AVM. Spontaneous regression of the AVM reduced the intracavernous pressure and caused amelioration of the symptoms.

Spontaneous regression of AVM is also rare, and some studies suggest that it occurs at a rate of 0.8% to 1.3%.¹ Several factors have been implicated, including trauma, intracranial hemorrhage, tumors, hypercoagulation, and the use of contraceptives.^{3,4} Minakawa et al.² reported that older age and smaller arterial feeders and draining veins regressed more commonly. Vodoff et al.¹² suggested that steroid use may be associated with regression of AVMs. In their case, old age and the small size of the nidus were factors favoring spontaneous regression. The diagram presented in Supplemental Fig. 1 illustrates the presumed relationship between the timeline of symptom onset and resolution and changes in vascular lesions.

Although craniotomy and interventional radiology were performed in similar cases (summarized in Table 1), we elected to manage the lesions conservatively with success. However, conservative management requires careful follow-up because most patients do not show spontaneous regression. It is important to carefully observe changes in clinical symptoms; particular attention should be paid to the signs of newly developed intracranial reflux, such as ocular hyperemia and exophthalmos. In addition, it is necessary to perform imaging evaluations on a regular basis. If conservative treatment is not effective, it is necessary to carry out intravascular treatment promptly. In cases such as ours that present with neurological symptoms, radiological findings may depict only a part of a series of changes involved in the vascular lesions. Clinicians should be aware that extracranial AVMs can present with neurological symptoms. Understanding the pathophysiological condition is essential to provide appropriate treatment. This case provides interesting insight into the onset and resolution of a dAVF and an AVM, as well as the symptomatology.

FIG. 4. Time course of MRI findings. **A–C:** Contrast-enhanced 4-dimensional time-resolved magnetic resonance angiography (MRA) using keyhole (**A:** on admission; **B:** 1 month later; **C:** 4 months later) showing spontaneous regression of vascular lesions (double arrows). **D–F:** Source MRA images (**D:** on admission; **E:** 1 month later; **F:** 4 months later) clearly demonstrate spontaneous regression of pterygoid AVM (arrows).

Observations

We report a case of a pterygoid AVM and a contralateral dual AVF to the CS that spontaneously regressed. Extracranial AVMs can present with neurological symptoms, and understanding the pathophysiological condition is essential to providing appropriate treatment.

Lessons

We report our experience with a case of an AVM and a dAVF with dynamically changing symptoms and vascular structure. Although it is a very rare case, it provides interesting insight into the mechanisms of onset and the resolution of dAVF and AVM, as well as their symptomatology.

TABLE 1. Summary of reported cases with extracranial AVMs showing neurological symptoms

Case No.	Authors & Year	Sex	Age (yrs)	Symptoms	Treatment	Prognosis
1	Malik et al., 1994 ⁵	M	48	Pulsatile tinnitus Headache	Craniotomy	GR
2		F	49	Pulsatile tinnitus Syncope	Endovascular treatment Craniotomy	GR
3	Tateshima et al., 2008 ⁶	M	46	Pulsatile tinnitus ICH	Endovascular treatment	SD
4	Park et al., 2013 ⁷	F	43	Ophthalmalgia Abducens nerve palsy	Endovascular treatment	GR
5	Present case	F	80	Ophthalmalgia Abducens nerve palsy	Conservative	GR

GR = good recovery; ICH = intracerebral hemorrhage; SD = severe disability.

Acknowledgments

We thank Dr. Alexander Zaboronok, MD, PhD, Department of Neurosurgery, Faculty of Medicine of the University of Tsukuba, for professional and English revision and Thomas Mayers of the Medical English Communication Center, Faculty of Medicine, University of Tsukuba, for native English revision.

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Disclosures

Dr. Matsumaru reported receiving personal fees from Medtronic Japan Co., personal fees from Medicos Hirata Co., personal fees from Asahi Intecc Co., personal fees from Kaneka Medix Co., personal fees from B. Braun Medical, personal fees from Stryker Japan Co., and personal fees from Terumo Co. outside the submitted work. The remaining authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Nakamura. Acquisition of data: Nakamura. Analysis and interpretation of data: Yanaka, Nakamura, Takahashi. Drafting the article: Tsukada, Yanaka, Nakamura, Takada, Sato. Critically revising the article: Yanaka, Sato, Matsumaru. Reviewed submitted version of manuscript: Ishikawa, Tsukada, Onuma. Study supervision: Yanaka.

Supplemental Information

Online-Only Content

Supplemental material is available with the online version of the article. *Supplemental Fig. 1*. <https://thejns.org/doi/suppl/10.3171/CASE21265>.

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