Angiographically occult vascular malformation of the intracranial accessory nerve: case report

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Angiographically occult cerebral vascular malformations (AOVMs) are usually found in the supratentorial brain parenchyma. Uncommonly, AOVMs can be found within the cavernous sinus or basal cisterns and can be associated with cranial nerves. AOVMs involving the intracranial segment of the spinal accessory nerve have not been described. A 46-year-old female patient presented with a history of episodic frontal headaches and episodes of nausea and dizziness, as well as gait instability progressing over 6 months prior to evaluation. Imaging revealed a well-circumscribed 3-cm extraaxial T1-weighted isointense and T2-weighted hyperintense contrast-enhancing mass centered in the region of the right lateral cerebellomedullary cistern. The patient underwent resection of the lesion. Although the intraoperative appearance was suggestive of a cavernous malformation, some histological findings were atypical, leading to the final diagnosis of vascular malformation, not otherwise specified. The patient’s postoperative course was uneventful with complete resolution of symptoms. To the authors’ knowledge, this is the first report of an AOVM involving the intracranial portion of the accessory nerve. For any AOVM located within the cerebellomedullary cistern or one suspected of involving a cranial nerve, the authors recommend including immunohistochemistry with primary antibody to neurofilament in the histopathology workup.

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Angiographically occult cerebral vascular malformation (AOVM) is a clinicoradiographic term that describes a heterogeneous group of entities, including cavernous malformations (CMs), thrombosed arteriovenous malformations, venous malformations, capillary telangiectasias, and mixed vascular malformations.48 These account for 5%–15% of all cerebral vascular lesions, with a general population prevalence estimated at 0.3%–0.5%. Approximately half of AOVMs occur sporadically, while the other half are inherited in an autosomal-dominant pattern. Given that histologically most AOVMs are CMs, the current literature concerns patient populations with CMs. Indeed, only a minority of AOVMs are venous malformations, arteriovenous malformations, or mixed vascular malformations.52,56 CMs are found within the brain parenchyma in approximately 95% of cases, with spinal CMs accounting for 5%. Of the intracranial CMs, 80% are supratentorial and 20% are infratentorial.53 CMs usually have a characteristic appearance on MRI with a mixed T2 signal intensity reticulated core surrounded by a rim of decreased signal intensity. However, smaller lesions can appear as small areas of decreased signal intensity.24,57

Uncommonly, CMs (as the most common form of AOVMs) can be found within the cavernous sinus, basal cisterns, cauda equina, and cranial nerves.30,41,43,44 Although CMs have been previously reported to be associated with almost all cranial nerves, their involvement of the lower cranial nerves is extremely rare. We report a case of an angiographically occult vascular malformation (AOVM) arising from the intracranial portion of the spinal accessory nerve, with pathological confirmation of nerve fibers traversing the lesion.

Case Report

History and Presentation

A 46-year-old woman presented with a history of episodic frontal headaches and episodes of nausea and dizzy-
ziness, as well as gait instability progressing over the 6 months prior to evaluation. The patient denied difficulty with fine motor coordination, dysarthria, dysphonia, or dysphagia. Her medical history was unremarkable. Results of a detailed physical examination were completely normal at this time.

Brain MRI showed a well-circumscribed 29 mm (anteroposterior) × 23 mm (transverse) × 29 mm (craniocaudal) extraaxial mass centered in the region of the right lateral cerebellomedullary cistern. The mass appeared isointense on T1-weighted and hyperintense on T2-weighted imaging, and enhanced heterogeneously following gadolinium injection (Fig. 1). The lesion caused distortion and compression of the lower medulla and inferior aspect of the fourth ventricle. However, there was no focal signal abnormality within the medulla and no obstructive hydrocephalus. The initial working diagnosis included ependymoma or choroid plexus papilloma. A catheter angiogram showed no arterial feeders supplying the lesion.

Surgical Procedure

The patient underwent a midline suboccipital posterior fossa craniotomy and resection of the posterior arch of C-1. Intraoperative neuromonitoring included electroencephalography, somatosensory evoked potentials, brainstem auditory evoked potentials, facial electromyography, and lower cranial nerve monitoring. After dural opening and incision of the arachnoid, a reddish-purple lesion was seen elevating the right cerebellar hemisphere (Fig. 2A). The arachnoid was separated from the surface of the well-circumscribed mass. After stimulating the capsule with the nerve stimulator, the surface was coagulated and incised. The frozen pathology section was suggestive of AOVM. Once the central portion of the mass was resected, the capsule was folded inward and the arachnoid was separated circumferentially. There was at least one rootlet of the spinal component of the spinal accessory nerve that entered the central substance of the mass at its inferolateral pole. It could not be stripped from the lesion’s capsule (Fig. 2B). A component of the spinal accessory nerve was also seen exiting from the mass prior to coursing toward the jugular foramen (Fig. 2C). Again, this could not be separated from the tumor capsule. Therefore, a component of the spinal accessory nerve was included within the tumor and could not be salvaged. The medullary roots of the spinal accessory nerve, however, were well visualized and were preserved by dissecting the arachnoid away from the capsule of the tumor. The hypoglossal nerve fibers were also identified and preserved, as were the ninth and tenth cranial nerves. By dividing the involved rootlet of the spinal accessory nerve proximally and distally, the mass was freed and completely resected (Fig. 2D). The lateral brainstem and the inferior pial surface of the cerebellum were entirely intact, with no hemosiderin coloration.

Histopathology Findings

The resected lesion was examined using routine (H & E) stains, Masson trichrome, and immunohistochemistry using primary antibodies to smooth muscle actin (SMA), neurofilament, and factor VIII, together with appropriate positive and negative controls. Routine stains showed a vascular lesion composed of ectatic and dilated vascular

![Fig. 1. Sagittal T1-weighted images without (A) and with (B) gadolinium, coronal (C) and axial (D) T1-weighted gadolinium-enhanced images, and axial T2-weighted image (E) demonstrating a partially cystic, heterogeneously enhancing lesion in the cerebellomedullary cistern exerting significant mass effect on the brainstem. Lateral angiographic projection of the right vertebral artery injection with no evidence of vascular abnormality (F).](image)
channels. Walls of these vessels were markedly thickened and contained a basophilic amorphous material, within which were closely apposed spindle cells (Fig. 3A–C), most of which were strongly immunoreactive for SMA (Fig. 3D–F). Endothelium of the vascular channels was normal by both routine stains and factor VIII immunohistochemistry (not shown). Neurofilament immunohistochemistry confirmed the presence of immunoreactive nerve twigs at the edge of the lesion and occasional nerve twigs among the vascular channels themselves (Fig. 3D–F). Some of the nerve twigs showed degenerative change, including axonal “bulbs” (resembling neuroaxonal spheroids in the central nervous system) and fragmentation (Fig. 3E and F). Although histopathological findings were suggestive of a CM, the presence of a smooth muscle component was atypical for the classic diagnosis of CM. Also, there was a relative absence of altered blood pigment (hemosiderin) in or around the lesion, as seen typically in intraparenchymal CMs. The final pathological diagnosis was that of a vascular malformation, not otherwise specified.

Postoperative Course
The patient had an uncomplicated postoperative course. On postoperative Day 1, her brain MRI study demonstrated complete resection of the cerebellomedullary cistern lesion (Fig. 4). She was seen in the clinic 1 month after surgery and noted complete resolution of her preoperative symptoms. On physical examination, no weakness or atrophy of the right sternocleidomastoid and trapezius muscles was noted. The patient was recently seen 1 year after her intervention and remains asymptomatic, with no evidence of residual lesion on follow-up MRI.

Discussion
AOVMs and Cranial Nerves
AOVMs (most frequently CMs) frequently involve the brain parenchyma, but the involvement of the cavernous sinus, the basal cisterns, and various cranial nerves is well described.41,43,44 The most common cranial nerve associat-ed with an AOVM is the optic nerve, with at least 18 reported cases involving the optic pathways, typically presenting with sudden visual acuity deterioration.3,13,19–22,27,31,34,36,38,47,59 The oculomotor nerve was involved by an AOVM in 4 reported cases, most frequently presenting with ophthalmoplegia and diplopia.30,40,45,58 In 3 cases, the trochlear nerve was involved, presenting clinically with either progressive diplopia or hemisensory loss symptoms.11,14,17,39,47 Only one case of an AOVM involving the abducens nerve has been reported, which presented with recurrent episodes of diplopia.43 There are 8 reported cases of AOVMs involving the cranial nerve VII/VIII complex, with 5 described to originate from the facial nerve and 3 from the vestibular nerve.1,9,16,18,28,40,42 Approximately 40 cases of AOVMs within the internal auditory canal have been published, with no specification as to the cranial nerve origin of the lesion.5,7,15,46,49 The AOVMs in these locations most commonly present with sensorineural hearing loss, hemifacial spasm, or tinnitus.49 In addition, approximately 50 cases of cerebellopontine angle AOVMs have been reported in the literature.1,8,29,32 These lesions were not directly associated with the cranial

FIG. 2. Intraoperative photographs under 10× magnification. A: A reddish-purple lesion elevating the right cerebellar hemisphere. B: A fiber of the spinal component of the spinal accessory nerve entering the central substance of the mass at its inferolateral pole. C: A component of the spinal accessory nerve exiting from the mass (by the dissec-tor). D: Resection cavity after removal of the lesion. Figure is available in color online only.

FIG. 3. Low-power view of the specimen showing abundant acute hemorrhage, and ectatic/dilated vascular channels separated by vari-ably thickened vessel walls. B: Magnified view from a representative portion of the AOVM showing lumens to be lined by normal endothelium; vessel walls are composed predominantly of spindle cells in a faintly basophilic matrix. C: Masson trichrome stain highlights thickened ves-sel walls in the AOVM. D–F: Immunohistochemical studies. D: Section immunostained with primary antibodies to SMA showing a significant element of smooth muscle cells in the vessel walls. E and F: Sections immunostained with primary antibodies to neurofilament. Arrow (in E) indicates several immunoreactive axons at the edge of the specimen, while arrowheads indicate nerve twigs among vascular channels. Many discontinuous nerve twigs are seen (F); the arrow indicates a dilated and swollen axon possibly representing degenerative change. Original magnification ×40 (A); ×95 (B–D and F); ×180 (E). Figure is available in color online only.
nerves; however, they were commonly exerting mass effect on them. Only one report exists of an AOVM involving the lower cranial nerves in a patient presenting with changes in the tone of voice. However, it is unclear if the lesion actually originated from one of the lower cranial nerves, and if so from which one. There is one reported case of a hypoglossal nerve AOVM presenting with tongue deviation and dysarthria.

**Imaging and Pathology Findings of Cranial Nerve–Associated AOVMs**

Imaging findings of AOVMs associated with cranial nerves may lack the classic reticulated appearance and resemble other benign lesions such as meningioma or schwannoma. To date, although multiple reports describe the finding of neuronal tissue within the AOVM, it remains challenging to ascertain whether the AOVM is originating from within the cranial nerve and splaying the nerve fibers at its periphery, or if the AOVM has originated outside of the nerve tissue and is encompassing some of its fibers. Intermingled cavernous and neural tissue has been described only in a minority of cases of cranial nerve–associated AOVMs. Although the described case was radiographically and clinically suggestive of a CM of the accessory nerve, some histopathological features were atypical of intracranial CMs and resulted in pathology diagnosing a vascular malformation, not otherwise specified. Finally, although less likely, it is possible that the lesion represented a mixed vascular malformation—a well-described entity containing elements of more than one angiographically occult type of vascular malformations.

**Cranial Nerve Associated AOVMs—Surgical Considerations**

Important surgical considerations and technique differences in the approach and resection of AOVM—most frequently CM—associated with cranial nerves merit discussion, as they differ from their more common intraparenchymal counterparts. In a parenchymal AOVM, the surgical strategy relies on development of a circumferential plane around the lesion and subsequent en bloc removal. In a cranial nerve–associated AOVM, this strategy must be modified to account for the intimate relationship between the lesion and the cranial nerve. The proposed surgical strategy is similar to that used in the resection of nerve sheath tumors, in which there is a focus on early identification of the involved nerve rootlet. In our case at least one rootlet of the spinal component of the spinal accessory nerve entered and exited the central substance of the mass and therefore could not be salvaged. The medullary roots of the spinal accessory nerve, however, were well visualized and were preserved by dissecting the arachnoid away from the capsule of the mass. Postoperatively, the trapezius and sternocleidomastoid muscle strength was intact, likely due to the meticulous identification and sacrifice only of the affected rootlet.

**Spinal Accessory Nerve and AOVMs**

To our knowledge, there is one reported case of an AOVM of the spinal accessory nerve in a patient who presented with progressive upper- and lower-extremity paresthesias and pain secondary to spinal cord compression. The lesion was located extracranially, at the level of the atlas. Histopathology was typical for a CM. Therefore, we report the first case in the literature of an intracranial accessory nerve AOVM located within the cerebellomedullary cistern confirmed by the histopathological documentation of nerve fibers coursing through the lesion. The differential diagnosis of a cerebellomedullary cistern mass should include meningioma, schwannoma, malignant peripheral nerve sheath tumors, ependymoma, and—although rare—AOVMs. For CMs located within the cerebellomedullary cistern as well as for any AOVM located within any cistern or suspected to be involving a cranial nerve, we recommend including immunohistochemistry with primary antibody to neurofilament in the histopathology workup.

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Disclosure
The 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: Martin, Terterov, McLaughlin. Acquisition of data: all authors. Analysis and interpretation of data: Terterov, McLaughlin, Vinters. Drafting the article: Terterov, McLaughlin. Critical revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Martin. Study supervision: Martin. Slide preparation and interpretation: Vinters.

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