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Arachnoid cyst alone causes hemifacial spasm: illustrative case

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BACKGROUND Hemifacial spasm (HFS) due to an arachnoid cyst at the cerebellopontine angle is rare. Here, the authors reported such a case and analyzed the mechanism of facial nerve hyperactivity by reviewing the literature.

OBSERVATIONS A 40-year-old man presented with right HFS for the past 3 years. Preoperative magnetic resonance imaging revealed a right cerebellopontine angle cystic mass with high intensity on T2-weighted images, low intensity on T1-weighted and diffusion-weighted images, and no contrast effects. Cyst excision and decompression of the facial nerve using a lateral suboccipital approach to monitor abnormal muscle response (AMR) resulted in permanent relief. The cyst was histologically compatible with an arachnoid cyst.

LESSONS In the present case, when the cyst was dissected, the AMR disappeared and no offending arteries were detected around the root exit zone. Therefore, the cyst itself was responsible for HFS, for which AMR was useful. Limited cases of HFS due to arachnoid cysts without neurovascular compression have been previously reported. The authors suggested that pulsatile compression by the cyst results in facial nerve hyperactivity and secondary HFS.

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KEYWORDS cerebellopontine angle; arachnoid cyst; hemifacial spasm; pulsation

A neurovascular conflict in the root exit zone (REZ) of the facial nerve causes hemifacial spasm (HFS). A curable treatment option for patients with HFS is microvascular decompression surgery.[1] Intracranial lesions, such as tumors, trauma, and infections, may also cause HFS; however, the incidence is rare. Arachnoid cysts are cerebrospinal fluid (CSF)-filled malformations of arachnoid tissue. Congenital, genetic, and traumatic factors have been suggested as underlying mechanisms for the formation of arachnoid cysts.[2] The incidence of arachnoid cysts in adults is $<2\%$.[3,4] The temporal fossa is the most common location. However, in a population-based study, 5.2% of arachnoid cysts were located in the cerebellopontine angle (CPA).[5] There are a limited number of reports on arachnoid cysts in the CPA associated with HFS.[6–12] Most of the cases demonstrated vascular compression on the REZ due to vascular displacement by the cyst. Herein, we report a case of HFS due to an arachnoid cyst in the CPA without neurovascular involvement that was successfully treated with cyst excision. We review the literature regarding secondary HFS and discuss the mechanism of HFS without vascular involvement based on changes in intraoperative abnormal muscle response (AMR).

Illustrative Case

A 40-year-old man presented with right HFS for the past 3 years, with gradually worsening paroxysmal contractions of the right orbicularis oris and orbicularis oculi muscles. Other neurological examinations were normal. Magnetic resonance imaging (MRI) showed a cystic mass occupying the right CPA with low intensity on T1-weighted and diffusion-weighted images and high intensity on the T2-weighted image. Contrast-enhanced MRI revealed no enhanced lesions in the CPA (Fig. 1A–C). Fast imaging employing steady-state acquisition images revealed a cyst displacing the facial acoustic nerve complex rostrally (Fig. 1D and E).

The patient received surgery through a lateral suboccipital approach to monitor the AMR and auditory brainstem response. The AMR was

ABBREVIATIONS AMR = abnormal muscle response; CPA = cerebellopontine angle; CSF = cerebrospinal fluid; HFS = hemifacial spasm; MRI = magnetic resonance imaging; REZ = root exit zone; TN = trigeminal neuralgia.

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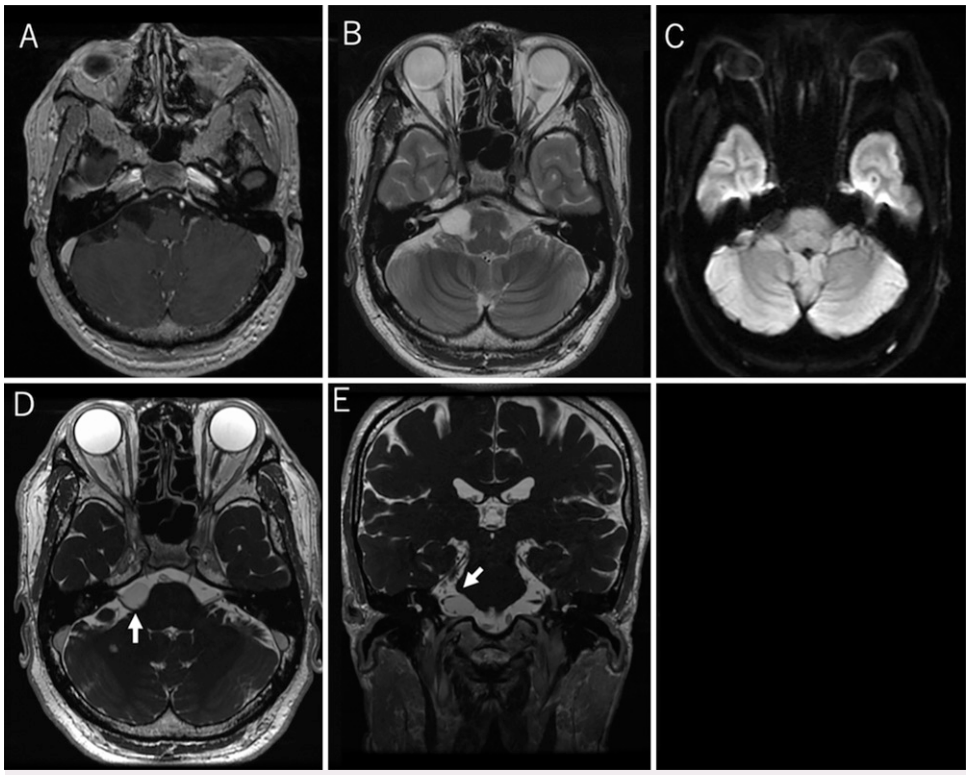

FIG. 1. Preoperative MRI showing low intensity on contrast-enhanced T1-weighted images (**A**) and high intensity on T2-weighted images (**B**). Diffusion-weighted imaging reveals no lesions (**C**). Axial (**D**) and coronal (**E**) fast imaging employing steady-state acquisition images showing the cyst compressing the facial nerve at the REZ (*arrows*).

monitored every 10 seconds during the intracranial procedures. AMR was detected before opening the dura mater (Fig. 2A). After draining the CSF from the cerebellomedullary fissure, we found a cystic lesion attached to the REZ (Fig. 2A). We assumed that there was no communication between the cyst content and the cistern of the CPA because the cystic lesion maintained its shape after CSF drainage (Fig. 2B). The cyst walls were dissected and resected (Fig. 2C). The content of the cyst was clear like CSF. No offending arteries were detected in the vicinity of the REZ. AMR disappeared after cyst perforation (Fig. 2C), and no recurrence of AMR was detected. Therefore, we confirmed that the cystic lesion caused the HFS as well as AMR. The pathological diagnosis demonstrated that the cyst wall was compatible with an arachnoid cyst (Fig. 2D). The spasm disappeared after surgery and the postoperative course was uneventful. The spasm was relieved 2 years after surgery.

Discussion

Observations

We encountered a case of an arachnoid cyst in the CPA causing ipsilateral HFS. After dissecting the arachnoid cyst, the AMR resolved and no vascular compression within the REZ was observed. Most cases of HFS are primary HFS, which are caused by vascular compression at the REZ of the facial nerve.[13] However, some cases of HFS are caused by tumors, trauma, and infections and are regarded as secondary HFS.[13] Zhang et al. reported that the incidence of secondary HFS was 0.3% in their study.[13] To our knowledge, eight cases of secondary HFS due to arachnoid cyst at the CPA have been reported.[6–12] In only two of these cases, the arachnoid cysts were responsible for HFS without vascular compression (Table 1).[6,7]

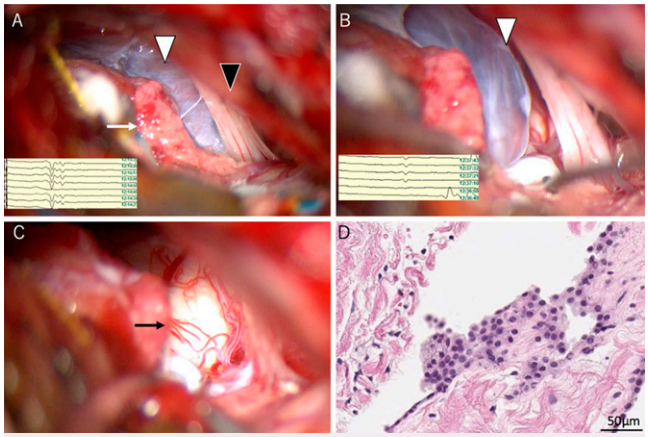

FIG. 2. A: Right cerebellomedullary cistern. The cyst wall (*white arrowhead*) occupies the medial CPA cistern. The content resembles CSF. The white arrow indicates the choroid plexus; the black arrowhead indicates the lower cranial nerves. An AMR was detected. **B:** Dissection of the cyst wall. The AMR disappeared (*white arrowhead*) after cyst perforation. **C:** After resection of the cyst wall, the REZ of the facial nerve (*black arrow*) is exposed. No offending arteries are observed. **D:** Pathological photomicrograph (hematoxylin & eosin staining, original magnification ×20). The cyst wall has simple or stratified squamous epithelia compatible with arachnoid cysts.

TABLE 1. Patients with secondary HFS due to arachnoid cyst in the CPA

Authors & Year	Age (yrs)	Sex	Side	NVC	CN VIII Involvement	Treatment
Altinörs et al., 1991[6]	33	M	Lt	No	No	Cyst excision
Mastronardi et al., 2009[7]	42	F	Lt	No	No	Cyst excision & MVD
Higashi et al., 1992[8]	25	M	Rt	PICA	Tinnitus	Cyst excision & MVD
Takano et al., 1998[9]	59	F	Lt	AICA	Hearing loss	Cyst excision
Bonde et al., 2008[10]	26	F	Rt	AICA	No	Cyst excision & MVD
Ruiz-Juretschke et al., 2015[11]	71	F	Rt	AICA	No	Cyst excision & MVD
Ogawa et al., 2015[12]	66	F	Rt	AICA	No	Cyst excision & MVD
Present case	40	M	Rt	No	No	Cyst excision

AICA = anterior inferior cerebellar artery; CN = cranial nerve; MVD = microvascular decompression; NVC = neurovascular conflict; PICA = posterior inferior cerebellar artery.

Secondary HFS and Differential Diagnosis of the Cystic Lesion

For cystic tumors in the CPA, differential diagnoses include schwannoma, endodermal sinus tumor (neurenteric cyst), hemangioblastoma, cavernous malformation, meningioma, teratoma, epidermoid cyst, cysticercus, and ependymal cyst.[14–24] In the present case, MRI revealed a cystic tumor without any enhancement by the gadolinium-contrast agent. Epidermoid cysts, endodermal sinus tumors, and ependymal cysts are typically nonenhanced lesions found on the CPA. T1-, T2-, and diffusion-weighted images are also useful for differentiating cyst contents; however, pathological diagnosis by surgical excision is essential for diagnosing rare cystic lesions.

Potential Mechanisms of HFS Due to an Arachnoid Cyst

To clarify the cause of HFS, we continuously monitored the AMR, which confirmed the completeness of decompression during surgery.[25] Meta-analysis of the use of AMR demonstrated a high positive predictive value and low negative predictive value.[26] In the present case, the AMR disappeared after perforation of the cyst, and no vessels were conflicting the REZ of the facial nerve. We concluded that the cyst itself was responsible for the HFS as well as AMR in our case.

We collected 120 previously reported cases to analyze the relationship between tumor pathology and the presence of an offending artery in secondary HFS (Supplementary Table).[6–13,22,27–30] The most prevalent tumor pathology was epidermoid cysts in 121 patients, including the present case (Fig. 3). Despite the limited number of patients, including the present case, the incidence of no neurovascular conflict was 37.5% in eight patients with HFS with arachnoid cysts. The arachnoid cyst was in the cerebellopontine cistern and did not anchor the skull base or brainstem.

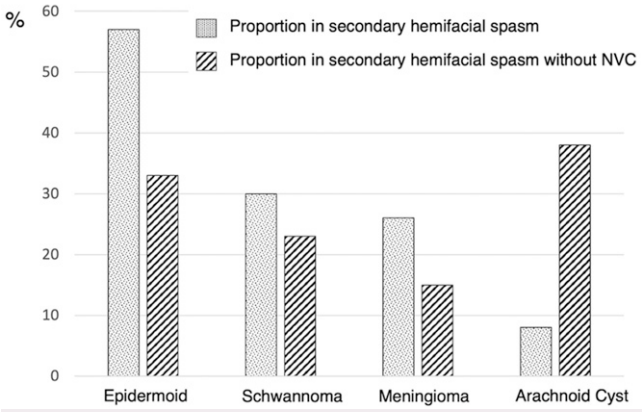

FIG. 3. Incidence of secondary HFS and neurovascular conflict in previously reported cases, including the present case.

We have summarized the mechanism of HFS due to intracranial lesions with respect to tumor pathology (Fig. 4). Arachnoid cysts containing CSF and pressure from tumor compression do not seem

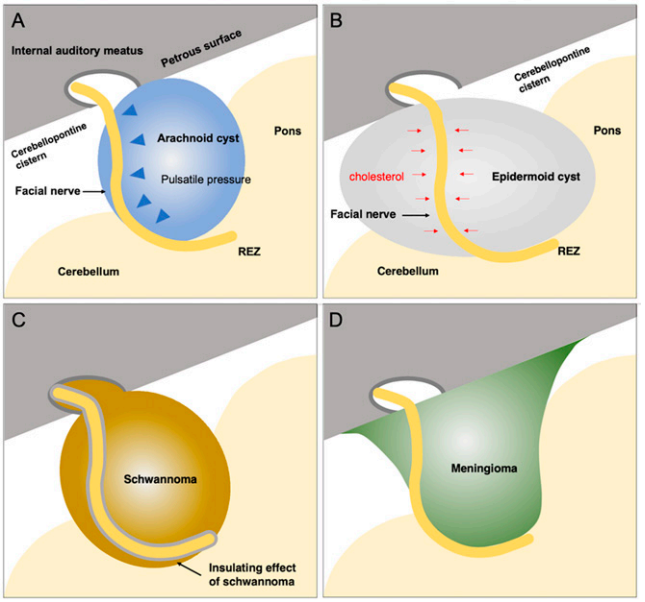

FIG. 4. Possible neurophysiological mechanisms of secondary HFS according to tumor pathology. **A:** Cardiac-related pulsatile CSF flow changes lead to pulsatile movement of the arachnoid cyst leading to facial nerve compression (*blue arrowheads*). Pulsatile stretching of the facial nerve causes demyelination and induces activation of voltage-gated and mechanosensitive ion channels (*blue arrowheads*).[28] **B:** An epidermoid cyst encasing or attaching to the facial nerve. Cholesterol in the tumor stimulates the facial nerve and induces HFS (*red arrows*).[33,34] **C:** The vestibular schwannoma is attached to the facial nerve without an arachnoid membrane; however, the schwannoma insulates the facial nerve.[32] The tumor anchors to the internal auditory meatus. Less pulsatile stretching of the facial nerve occurs with cardiac-related pulsatile CSF flow changes. **D:** Meningiomas attach to the petrous surface. Less pulsatile stretching of the facial nerve occurs with cardiac-related pulsatile CSF flow changes.

to be high. CSF flow dynamics have been extensively studied,[31] and MR technologies have revealed noninvasive CSF flow dynamics, such as cardiac-related pulsation. Flow changes would cause deviation of the arachnoid cyst following the cardiac cycle and stretching of the facial nerve (Fig. 4A). Liu and Zhong hypothesized that an ectopic action potential generated from the compressed nerve root was responsible for cranial nerve hyperactivity in the pathogenesis of HFS and trigeminal neuralgia (TN).[32] The compression of the arteries or tumor at the REZ results in demyelination of the nerves and induces activation of the voltage-gated and mechanosensitive ion channels after generation of the ectopic action potentials.[32] We suggest that repetitive pulsatile compression may cause the generation of spasms without vascular compression.

Epidermoid cysts also demonstrated a high incidence of no neurovascular conflict (33.3%, Fig. 3). Several authors have reported that the chemical effects of cholesterol on the nerve could cause the development of TN;[33,34] however, direct evidence of this mechanism has not been verified (Fig. 4B). The incidence of no neurovascular conflict in vestibular schwannomas and meningiomas was 23.3% and 15.4%, respectively. The low rate of vascular involvement in patients with vestibular schwannomas presenting with HFS may be related to the insulated root.[32] These tumors generally anchor the skull base or internal auditory meatus and hardly move after cardiac-related CSF flow changes (Fig. 4C and D). The arachnoid cyst without anchoring the skull base might cause HFS by cardiac-related CSF pulsation.

Lessons

Arachnoid cysts in the CPA are rare. Pulsatile compression of the REZ of the facial nerve may cause hyperactivity of the facial nerve and HFS. AMR monitoring is essential for clarifying the etiology of this rare phenomenon. Surgical treatment is the only curative treatment option for HFS due to a CPA arachnoid cyst.

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Disclosures

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: Higuchi, Ozaki, Iwadate. Acquisition of data: Ozaki, Nakano. Analysis and interpretation of data: Higuchi, Ozaki, Yamakami. Drafting the article: Higuchi, Ozaki. Critically revising the article: Higuchi, Nakano, Iwadate. Reviewed submitted version of manuscript: Higuchi, Ozaki, Nakano, Horiguchi, Yamakami. Approved the final version of the manuscript on behalf of all authors: Higuchi. Study supervision: Higuchi, Horiguchi.

Supplemental Information

Online-Only Content

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Supplementary Table. https://thejns.org/doi/suppl/10.3171/CASE2275.

Previous Presentations

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