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A Case of Trigeminal Neuralgia Due to Cerebellopontine Epidermoid Cyst: Discrepancy between Intraoperative and Radiological Findings of Constructive Interference in Steady State (CISS)

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Abstract

Trigeminal neuralgia (TN) is characterized by lightning pain paroxysms in the somatosensory distribution of the trigeminal nerve accompanied by hypersensitivity to non-nociceptive stimuli. Epidermoid cysts sometimes cause TN. To plan the surgery, constructive interference in steady state (CISS) image is useful for understanding the tumor's location, extent, and relationship against the cranial nerves, and epidermoid cysts are shown as hypointense compared to cerebrospinal fluid (CSF). However, we herein describe a case with TN due to epidermoid cysts, whose intraoperative findings are different from the preoperative and postoperative CISS image. A 49-year-old woman has suffered from TN. CISS images revealed the prolonged trigeminal nerve and the hypointense tumor compared to the CSF at the right cerebellopontine angle. CISS image suggested that the tumor would surround the trigeminal nerve, reach into the Meckel cavity, and offend and compress the trigeminal nerve's root entry zone (REZ). However, contrary to our expectation, the trigeminal nerve was not surrounded by the tumor. Neuroendoscope revealed that the tumor compressed the REZ, but the tumor was not present in the Meckel cavity. We performed partial tumor removal around the trigeminal nerve, and her symptoms improved. However, the postoperative CISS image was similar to the preoperative one, and so we could not evaluate the remaining tumor. The pathological diagnosis was epidermoid cysts. Intraoperative findings are sometimes different from the pre- and postoperative CISS images, making it difficult to follow up the remaining part of the epidermoid cyst.

Keywords: constructive interference in steady state (CISS), diffusion-weighted image (DWI), epidermoid cyst, neuroendoscope, trigeminal neuralgia

Introduction

Trigeminal neuralgia (TN) is a clinical entity characterized by lightning pain paroxysms in the somatosensory distribution of the trigeminal nerve accompanied by hypersensitivity to non-nociceptive stimuli.[1)] The onset of pain can be triggered even by minimal stimulation such as talking, chewing, or light touch of the overlying skin. Although most often unilateral, the pain occurs sporadically and frequently repeats throughout the day. TN is rare, affecting an estimated 4–13 people per 100000 annually. Male-to-female prevalence ratios ranging around 1:1.6.[2)] Classically, it is generally agreed that TN's most common cause is compression and morphological changes of the trigeminal nerve. The "root entry zone (REZ)" is vulnerable. The surgical strategy aims to transpose the offending vessels from the REZ to other areas or resolve the trigeminal nerve's morphological changes, such as

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distortion, dislocation, and distension.[2] The surgery for TN is now performed for the young and the elderly,[3] and an exoscope is recently used as a new surgical method in addition to classical craniotomy surgery.[4] The incidence of TN by tumor is around 2%–10% of all TNs, and 38% of the tumors are epidermoid cysts.[5,6]

An epidermoid cyst is a slow-growing benign tumor from the epidermal origin. The cerebellopontine angle is one of the most common sites, threatening cranial nerves, cerebellum, and brain stem.[6] The Brain Tumor Registry of Japan (2001–2004) found that intracranial epidermoid cyst incidence accounted for 0.9% of all primary brain tumors. In the study, 6% of the cerebellopontine angle tumor are epidermoid cysts.[7] Its radiological findings are typically lobulated mass hypodense on CT, hypointense on T1-weighted image (T1WI), and hyperintense on T2-weighted image (T2WI). Especially, hyperintense in the diffusion-weighted image (DWI) is important to differentiate from an arachnoid cyst.[6,8–12]

It is important to know the tumor's size and positional relation to the cranial nerves, cerebellum, and brain stem to determine the surgical strategy for cerebellopontine angle tumors. Constructive interference in steady state (CISS) image is useful to understand the tumor's location, extent, and relationship against the cranial nerve because an epidermoid cyst is shown as a hypointense lesion compared to cerebrospinal fluid (CSF).[11,13,14] However, we herein describe a case with TN due to an epidermoid cyst, whose intraoperative findings are different from the preoperative and postoperative CISS image, making it difficult to follow up the remaining part of the epidermoid cyst.

Case Report

A 49-year-old woman has suffered from right TN for 4 years. She presented with paroxysmal lightning pain at the right mandibular region, involving V2 and V3 divisions. We first treated with carbamazepine 200 mg/day and pregabalin 50 mg/day but stopped them due to rash, liver dysfunction, and drowsiness. The preoperative Barrow Neurological Institute Pain Intensity Score was IV. MRI revealed a mass at the right cerebellopontine angle with hypointense on T1WI, hyperintense on T2WI, non-enhanced on contrast-enhanced T1WI using gadolinium, and low-dense on CT, mimicking arachnoid cyst. The lesion was hyperintense on DWI, and so we diagnosed an epidermoid cyst. The size was 30 × 25 × 15 mm from the ambient cistern to the hypoglossal nerve. Basi-parallel anatomical scanning (BPAS) and CISS images revealed the prolonged and distorted trigeminal nerve and the hypointense tumor compared to the CSF. CISS image suggested that the tumor would wholly surround the trigeminal nerve, reach into the Meckel cavity, and offend and compress the REZ of the trigeminal nerve (red circle in Fig. 1A). There are no obvious offending vessels on CT angiography. The tumor seemed to contact the facial and auditory nerves, as well as the cerebellar tentorium near the trochlear nerve.

Considering that 1) the symptom was only related to the trigeminal nerve, 2) epidermoid cyst would have a slow-growing rate, and 3) we should avoid cranial nerve palsy, we planned partial removal via the retrosigmoid approach with the prone position. We also hypothesized that the CISS image would reveal the rest of the tumor postoperatively, enabling us to follow up at the outpatient and to plan the two-stage surgery in case.

After 10 cm linear skin incision above the right asterion, we performed a 4-cm craniotomy above the transverse and sigmoid sinuses. We incised dura to the knee of sinuses and reached the trigeminal nerve at the cerebellopontine cistern. Contrary to our expectation based on the preoperative CISS image, the trigeminal nerve was not surrounded by the tumor (Fig. 2A). Neuroendoscope revealed that the REZ was compressed by the tumor (Fig. 2B), but the tumor was not present in the Meckel cavity (Fig. 2C). Gently retracting the cerebellum, we removed the tumor and its capsule around the trigeminal nerve, avoiding spreading the contents (Fig. 2D). We confirmed that there was no tumor around the REZ (Fig. 2E), and the tumor removal rate was estimated as 30%. We transposed the petrosal vein after tumor removal (Fig. 2F), which had not hit the trigeminal nerve before tumor removal (Figs. 2A, 2B, 2D, and 2E). We finally sutured the dura and skin, adequately using fibrin glue to avoid CSF leakage. The intraoperative findings suggested an epidermoid cyst, same in the pathological examination.

Her symptom improved so that the Barrow Neurological Institute Pain Intensity Score just after the surgery was II, and that on a postoperative day 6 was I without any complications. The postoperative images on day 3 revealed the bit loose trigeminal nerve. However, the BPAS, CISS images, and DWI did not show the removed part of the tumor, and the images were almost similar to the preoperative ones (Fig. 1B). Those radiological findings on day 39 seemed similar (Fig. 1C). These unchanged findings made it difficult for us to follow up the patient's tumor at the outpatient using these MRI sequences.

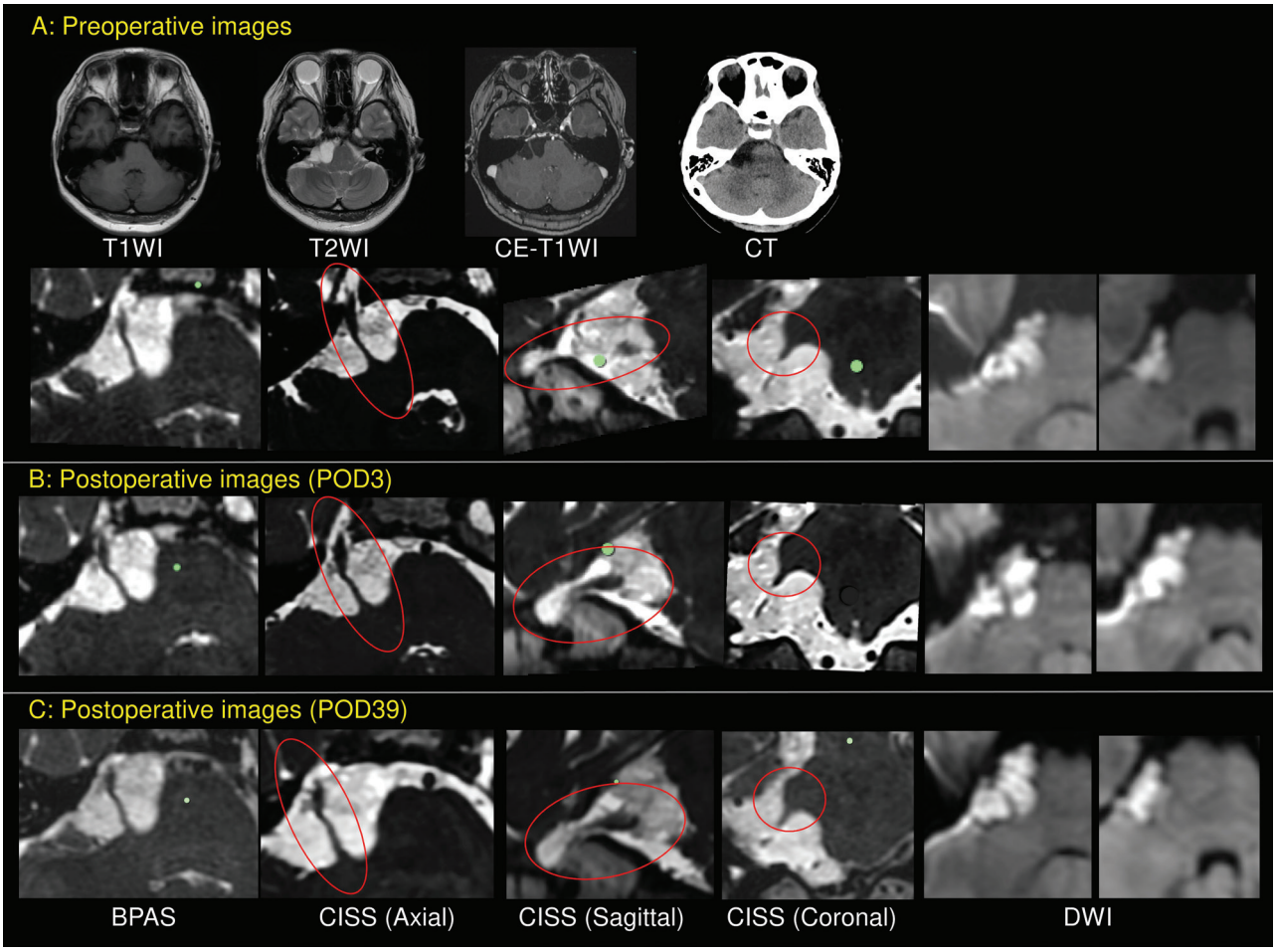

Fig. 1 Preoperative MRI revealed a mass at the right cerebellopontine angle with hypointense on T1WI, hyperintense on T2WI, non-enhanced on contrast-enhanced (CE-) T1WI using gadolinium, and low-dense on CT. The lesion was hyperintense on DWI, and so we diagnosed an epidermoid cyst. BPAS and CISS images revealed the prolonged and distorted trigeminal nerve and the hypointense tumor compared to the CSF. CISS image suggested that the tumor would wholly surround the trigeminal nerve, reach into the Meckel cavity, and offend and compress the REZ of the trigeminal nerve (red circle in A). The postoperative images on day (POD) 3 revealed the loose trigeminal nerve. However, the BPAS, CISS images, and DWI did not show the removed part of the tumor, and the images were almost similar to the preoperative ones (B). Those radiological findings on day 39 still seemed unchanged (C). BPAS: basi-parallel anatomical scanning, CISS: constructive interference in steady state, CSF: cerebrospinal fluid, DWI: diffusion-weighted image, REZ: root entry zone, T1WI: T1-weighted image, T2WI: T2-weighted image.

Discussion

We report a case with TN due to the epidermoid cyst, which was partially removed. However, the pre- and postoperative MRI findings did not reflect the actual intraoperative findings. Our case raises the difficulty of radiological evaluation of epidermoid cysts.

CISS sequence was initially designed for MR myelography and MR cisternography.[14] CISS uses the gradient echo method, in which two scans with different phase control of the radio frequency pulse are performed. After the two-image acquisitions, the two images are combined to make the CISS images.[15] CISS is widely used for depicting structures around the brain stem. Ikushima first reported CISS's utility for depicting an epidermoid cyst because CISS can provide high-resolution images with good contrast between CSF and solid structures. This contrast may be obtained by the accentuation of T2 values between epidermoid cysts and CSF.[14] Murakami also reported CISS's utility, and the operative findings coincided well with CISS images, although the contrast between the epidermoid cyst and CSF was not sufficient. Therefore, they recommended that DWI should be combined to evaluate

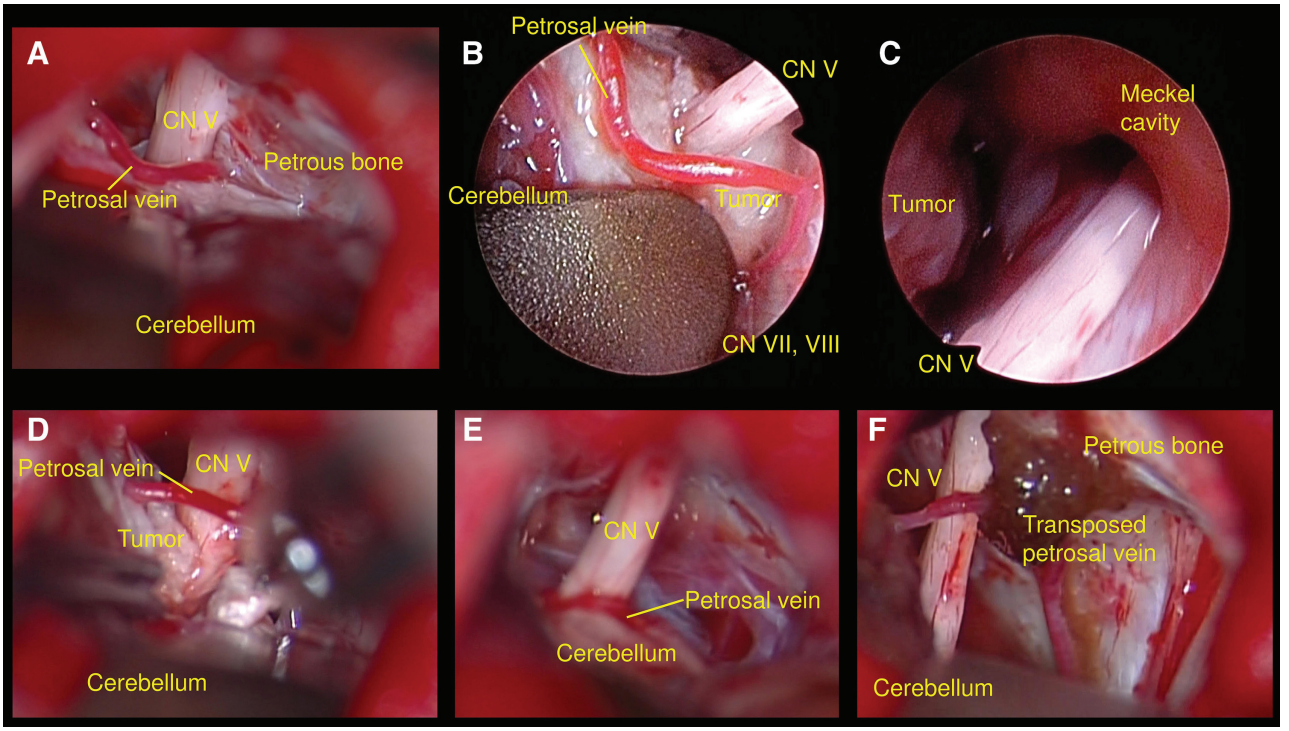

Fig. 2 Intraoperative findings: contrary to our expectation based on the preoperative CISS image, the trigeminal nerve (CN V) was not surrounded by the tumor (A). Neuroendoscope revealed that the petrosal vein did not hit the CN V. It also revealed that the tumor compressed the REZ (B), but the tumor was not present in the Meckel cavity (C). Gently retracting the cerebellum, we removed the tumor and its capsule around the trigeminal nerve, avoiding spreading the contents (D). We confirmed that there was no tumor around the REZ (E). We transposed the petrosal vein after tumor removal, which had not hit the trigeminal nerve before tumor removal (F). CISS: constructive interference in steady state, REZ: root entry zone.

the location.[13] Similar reports have been published that CISS is helpful to know the tumor location and extent,[8,10,11] but our case suggested CISS sometimes does not reflect the actual tumor location.

This may be because 1) the keratin in the tumor capsule might have been fluid-like as CSF, and it might have postoperatively come out to the removal cavity, or 2) physical effects by the tumor mass and chemical effects such as inflammation might have limited the CSF's fluidity and current. Further radiological studies like CSF current evaluation using four-dimensional velocity mapping[16] are needed. Another presumable reason is 3) that posterior fossa contents may have simply shifted, after craniotomy and CSF drainage, which might explain why there was no tumor near the porus trigeminus. Furthermore, 4) the CISS image can be affected by the magnetic susceptibility artifacts from the bone, vessel, and air in the mastoid bone, compared to other three-dimensional T2WIs like T2-SPACE (SIEMENS, Munich, Germany), T2-Cube (GE Healthcare, Illinois, USA), and T2-VISTA (Phillips, Amsterdam, The Netherlands). These three-dimensional T2WIs use the fast spin-echo method and can suppress the magnetic susceptibility artifacts by eliminating the magnetic field's inhomogeneity by 180° radio wave pulses every image acquisitions.[15] Therefore, it may be difficult to evaluate the tumor by the CISS image alone, and other three-dimensional T2WIs would be helpful. Further study on the difference between the CISS images and 3D-T2WIs is needed.

The surgery of epidermoid cyst aims to remove all tissue including the tumor capsule, with no damage to vital neurovascular structures. However, only 50%–80% can be completely removed, according to previous reports.[6,17,18] Few authors recommend the aggressive resection of every bit of the capsule to decrease recurrence rates.[19] Also, the lack of intraoperative monitoring limited our performance to manage adhesions of the capsule to the brain stem and cranial nerves.[20] Therefore, pre- and postoperative images to evaluate tumor mass are essential to follow up the remaining

tumor. However, our case suggested that the CISS images did not reflect the actual tumor volume and location. Therefore, when the diagnosis of epidermoid was made, the surgical strategy should have been well thought out because there is a possibility that we cannot follow up the rest of the tumor by CISS images.

We reported a case with TN due to an epidermoid cyst. However, intraoperative findings are different from the pre- and postoperative CISS image, making it difficult to follow up the remaining part of the epidermoid cyst. We should remember that the CISS images are sometimes not counted on for the epidermoid cyst evaluation.

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Ethical Approval

This study was approved by our hospital ethics committee. The patients in our report provided written consent forms.

Conflicts of Interest Disclosure

The authors report no conflicts of interest concerning the materials or methods used in this study, or the findings presented in this article. No sources of financial or material support were received. This article and content of this study were not published or presented previously.

References

1) Yamada K, Tanaka Y, Sumita K, Nemoto S, Maehara T: Computational fluid dynamics analysis of the offending artery at sites of neurovascular compression in trigeminal neuralgia using preoperative MRI data. *Neurol Med Chir (Tokyo)* 59: 415–422, 2019
2) Maarbjerg S, Di Stefano G, Bendtsen L, Cruccu G: Trigeminal neuralgia – diagnosis and treatment. *Cephalalgia* 37: 648–657, 2017
3) Amagasaki K, Uchida T, Hosono A, Nakaguchi H: Microvascular decompression surgery for elderly patients: a study based on proposals from the Joint Committee of the Japan Gerontological Society and the Japan Geriatrics Society. *Neurol Med Chir (Tokyo)* 60: 468–474, 2020
4) Shimizu T, Toyota S, Nakagawa K, et al.: Retrosigmoid approach in the supine position using ORBEYE: a consecutive series of 14 cases. *Neurol Med Chir (Tokyo)* 61: 55–61, 2021
5) Barker FG, Jannetta PJ, Babu RP, Pomonis S, Bissonette DJ, Jho HD: Long-term outcome after operation for trigeminal neuralgia in patients with posterior fossa tumors. *J Neurosurg* 84: 818–825, 1996
6) Kobata H, Kondo A, Iwasaki K: Cerebellopontine angle epidermoids presenting with cranial nerve hyperactive dysfunction: pathogenesis and long-term surgical results in 30 patients. *Neurosurgery* 50: 276–285; discussion 285-286, 2002
7) Committee of Brain Tumor Registry of Japan: Report of Brain Tumor Registry of Japan (2001-2004): 13th ed. *Neurol Med Chir (Tokyo)* 54: 9–102, 2014
8) Ben Hamouda M, Drissi C, Sebai R, et al.: Atypical CT and MRI aspects of an epidermoid cyst. *J Neuroradiol* 34: 129–132, 2007
9) Law EKC, Lee RKL, Ng AWH, Siu DYW, Ng H-K: Atypical intracranial epidermoid cysts: rare anomalies with unique radiological features. *Case Rep Radiol* 2015: 528632, 2015
10) Gizewski ER: [Epidermoid or arachnoid cyst: CISS, FLAIR and diffusion images as solution of the diagnostic dilemma]. *Rofo* 173: 77–78, 2001 (German)
11) Doll A, Abu Eid M, Kehrli P, et al.: [Aspects of FLAIR sequences, 3D-CISS and diffusion-weight MR imaging of intracranial epidermoid cysts]. *J Neuroradiol* 27: 101–106, 2000 (French)
12) Lopez Gomez P, Mato Mañas D, Marco de Lucas E: Epidermoid cyst extending along trigeminal nerve pathway with unusual imaging findings. *World Neurosurg* 146: 75–77, 2021
13) Murakami N, Matsushima T, Kuba H, et al.: Combining steady-state constructive interference and diffusion-weighted magnetic resonance imaging in the surgical treatment of epidermoid tumors. *Neurosurg Rev* 22: 159–162, 1999
14) Ikushima I, Korogi Y, Hirai T, et al.: MR of epidermoids with a variety of pulse sequences. *AJNR Am J Neuroradiol* 18: 1359–1363, 1997
15) Shimada K, Tanaka M, Taguchi M, Yamazaki I, Kasuga Y: [Comparison of magnetic resonance cisternography imaging techniques]. *J Jpn Assoc Rural Med* 68: 52–58, 2019 (Japanese)
16) Matsumae M, Kuroda K, Yatsushiro S, et al.: Changing the currently held concept of cerebrospinal fluid dynamics based on shared findings of cerebrospinal fluid motion in the cranial cavity using various types of magnetic resonance imaging techniques. *Neurol Med Chir (Tokyo)* 59: 133–146, 2019
17) Akar Z, Tanriover N, Tuzgen S, Kafadar AM, Kuday C: Surgical treatment of intracranial epidermoid tumors. *Neurol Med Chir (Tokyo)* 43: 275–280; discussion 281, 2003
18) Vernon V, Naik H, Guha A: Surgical management of cerebellopontine angle epidermoid cysts: an institutional experience of 10 years. *Br J Neurosurg* doi: 10.1080/02688697.2020.1867058 Epub 2021 Jan 7
19) Farhoud A, Khedr W, Aboul-Enein H: Surgical resection of cerebellopontine epidermoid cysts: limitations

and outcome. *J Neurol Surg B Skull Base* 79: 167–172, 2018
20) Aboud E, Abolfotoh M, Pravdenkova S, Gokoglu A, Gokden M, Al-Mefty O: Giant intracranial epidermoids: is total removal feasible? *J Neurosurg* 122: 743–756, 2015

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