Article published online: 2022-08-16

Case Report

# Trigeminal Neuralgia Secondary to Cerebellar Arteriovenous Malformation: A Report of Two Cases

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## Abstract

Trigeminal neuralgia (TN) secondary to cerebellar arteriovenous malformation (cAVM) is a rare condition with only few reports existing in the literatures. Given to its rarity, the treatment armamentarium is still controversial. We reported our experiences treated two cases of TN secondary to cAVM using different strategies. The first case was successfully treated by a combination of gamma knife radiosurgery and microvascular decompression (MVD) of the trigeminal nerve. The second case was successfully treated by one-step microsurgical AVM resection and MVD of the trigeminal nerve. Postoperative immediate pain relief was achieved in both patients. Microsurgical procedure is still playing an important role in treating TN secondary to cAVM.

**Keywords:** *Cerebellar arteriovenous malformation, microsurgery, multimodal treatment, trigeminal neuralgia*

## Introduction

Trigeminal neuralgia (TN) classically presents as a brief and severe paroxysmal facial pain involving one or more trigeminal nerve divisions. TN has an incidence of 4–5/100,000 and up to 25/100,000 in the geriatric population.[1] The majority of TN (80%–90%) is caused by a neurovascular compression at the Obersteiner–Redlich (OR) zone which is located approximately 1–3 mm distal to the trigeminal nerve origin at the brainstem. Secondary TN is due to any underlying disease resulting in compression or irritation at the OR zone such as an arteriovenous malformation (AVM), brain tumor, aneurysm, and multiple sclerosis.

Cerebellar AVMs (cAVMs) are rare, accounting for 7%–15% of all intracranial AVMs. The most common clinical presentation is brain hemorrhage.[2,3] A cAVM presenting with TN is extremely rare with only 17 published in the English literature over 60 years.[4,5] Endovascular embolization and/or stereotactic radiosurgery have been the most common method of treatment in the past 20 years. Surgery was only reported in five cases, and the total AVM resection was achieved in just two cases. A cAVM presenting with TN is a rare and complex clinical entity, and multimodal treatment must consider not only patient's debilitating facial pain but also the natural history of cAVM. To date, there is no consensus on the optimal treatment for these cases.

In this study, we reported our experiences in the management of two patients presenting with secondary TN due to cAVM. Our treatment strategies were tailored according to individual clinical status and our local treatment capabilities.

## Case Reports

### Case #1

A 39-year-old Jehovah witness female presented with progressive right-sided V2 and V3 TN over the past 3 years. Magnetic resonance imaging (MRI) and digital subtraction angiogram (DSA) showed a grade 5 right cAVM with feeding vessels from the right superior cerebellar artery (SCA), anterior inferior cerebellar artery (AICA), and posterior inferior cerebellar artery [Figure 1a and b]. The compression of the trigeminal nerve was suspected to be from the right SCA, AICA, and pontomesencephalic vein, which represented the main venous drainage



**How to cite this article:** Tjahjadi M, Wijaya V, Serrone J, Caropeboka MS. Trigeminal neuralgia secondary to cerebellar arteriovenous malformation: A report of two cases. Asian J Neurosurg 2020;15:745-9.

**Submitted:** 11-May-2020 **Revised:** 14-Jun-2020
**Accepted:** 04-Jul-2020 **Published:** 28-Aug-2020

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**Access this article online**

**Website:** www.asianjns.org

**DOI:** 10.4103/ajns.AJNS_229_20

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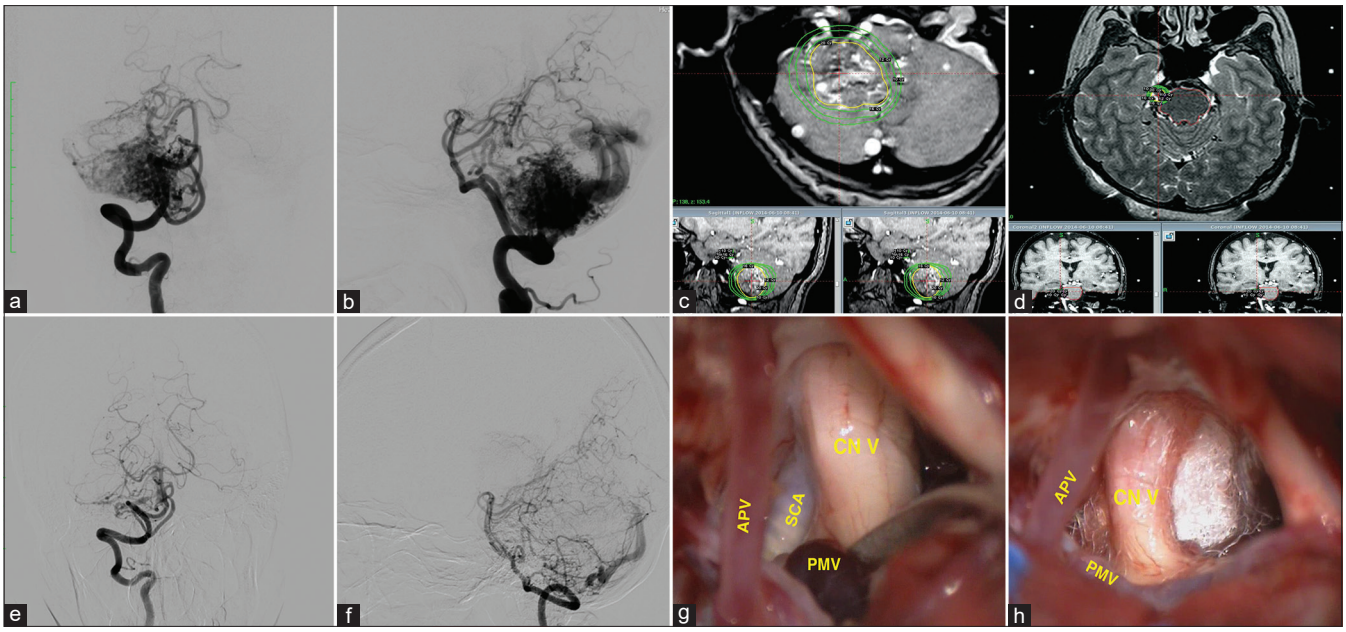


**Figure 1: (a and b) Anteroposterior and lateral projection of the pregamma knife radiosurgery cerebral angiogram shows a massive grade 5 cerebellar arteriovenous malformation, (c) gamma knife radiosurgery planning of 20 Gy at 45% isodose line for the arteriovenous malformation treatment, (d) gamma knife radiosurgery planning of 80 Gy at 100% isodose line for the trigeminal neuralgia treatment, (e) anteroposterior projection of postgamma knife radiosurgery cerebral angiogram taken 18 months after the treatment. These figures showed marked reduction of the arteriovenous malformation nidus, (f) Lateral projection of postgamma knife radiosurgery cerebral angiogram taken 18 months after the treatment, (g) compression sites of the root entry zone of the right trigeminal nerve by a dilated pontomesencephalic vein at its ventral side just after the nerve exits the brainstem and by the superior cerebellar artery at its dorsomedial side. (PMV – Pontomesencephalic vein; SCA – Superior cerebellar artery), (h) the trigeminal nerve after the nerve compression released**

from the AVM. The patient was referred to gamma knife radiosurgery (GKRS) service to treat both her AVM and TN. She received 80 Gy at the 100% isodose line for her trigeminal nerve and 20 Gy at the 45% isodose line for her AVM [Figure 1c and d]. After GKRS, the pain was relieved for several weeks but relapsed with gradual pain worsening in frequency, intensity, and duration. Gasserian ganglion glycerol injection and medical therapy with carbamazepine 900 mg, oxcarbazepine 600 mg, and pregabalin 300 mg were utilized without effective result. The patient then returned to our clinic at 18 months after GKRS in poor clinical condition with modified Rankin Scale (mRS) of 3, due to malnourishment, general weakness, and severe facial pain. A follow-up DSA was performed and showing significant obliteration of the AVM [Figure 1e and f]. Due to the improvement of the AVM status and her refusal to consent to blood transfusion, we decided to perform a microvascular decompression (MVD).

A standard right-sided retrosigmoid approach was performed. A 3 cm craniotomy bone flap was executed. Intraoperative finding showed that the OR zone of the fifth nerve was severely compressed by the SCA and dilated pontomesencephalic vein [Figure 1g and h]. The separation of these vessels was challenging due to the adherence of the arachnoid and neurovascular structures which we believed to be an effect of GKRS.

Postoperatively, the patient had immediate pain relief and was discharged at postoperative day 4 with mRS 1. Follow-up on 12, 24, and 60 months showed that the patient was still free of pain and medication and a mRS 0 at all-time points.

## Case #2

A 31-year-old female presented with right-sided facial pain in the V2 dermatome for 4 years. MRI of the brain showed a grade 3 cAVM at the right anterior lobe [Figure 2a and b]. A surgical attempt was performed at another hospital but was aborted due to excessive bleeding during dural opening. After the failure of the first surgery, she had neurological decline, and her TN was treated with maximum dose of carbamazepine, amitriptyline, and pregabalin. Gasserian ganglion radiofrequency ablation was also attempted to ease her pain. Despite all these efforts, her facial pain persisted and made her unable to eat solid food and to do other routine daily activities. She was recommended by the neurologist to get GKRS to treat her AVM, but due to the limited access to this service, she decided to visit our clinic.

DSA showed a high-flow AVM with major feeders coming from the right SCA and right posterior cerebral artery, and major drainage was going through the superior petrosal vein to the transverse sinus [Figure 2c and d].

A simplified presigmoid approach modified by Juha Hernesniemi was performed to reach the AVM.[6] Intraoperative findings showed the superior and inferior divisions of the SCA feeding the AVM. A temporary clip

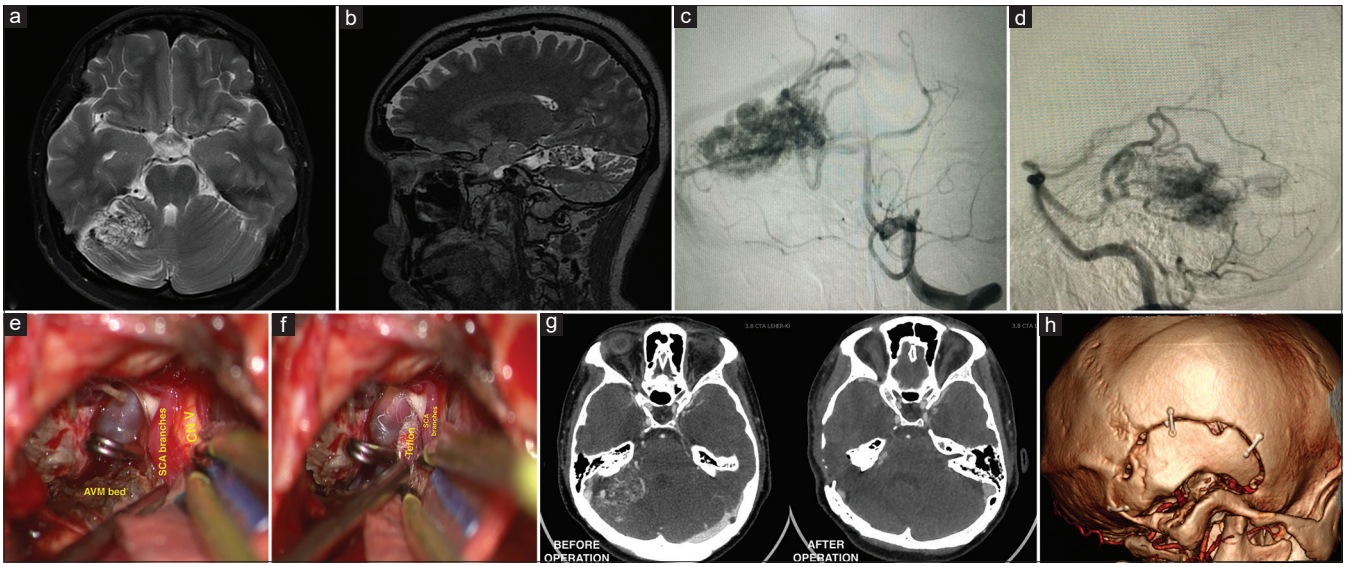


**Figure 2: (a) Magnetic resonance imaging study of case #2 showing a grade 3 arteriovenous malformation at the right anterior lobe of the cerebellum just below the tentorium (axial view), (b) Sagittal view of the magnetic resonance imaging (c) cerebral angiogram (anteroposterior view) showing a high-flow arteriovenous malformation with main feeders from the right posterior cerebral artery and right superior cerebellar artery. The arteriovenous malformation drained to the straight sinus through the tentorial bridging veins (d) Cerebral angiogram (lateral view) (e) Intraoperative finding of cerebellopontine angle cistern after arteriovenous malformation removal showed that the CN V was severely compressed by a dilated draining vein and branches of the superior cerebellar artery (CPA – Cerebellopontine angle; CN V – Trigeminal nerve; SCA – Superior cerebellar artery) (f) showed the separation of the nerve from the surrounding vascular structures and a Teflon patch inserted between these structures. (g) Pre- and postoperative computed tomography angiogram showed total removal of the arteriovenous malformation (h) Postoperative three-dimensional bone computed tomography showed the bony opening of simplified presigmoid approach**

was placed at the ambient segment of SCA. The AVM was circumferentially resected. The distal SCA branches feeding the AVM were coagulated and sectioned, and the SCA was mobilized from the fifth nerve. To ascertain the total decompression of the fifth nerve, we inserted a patch of Teflon surrounding the fifth nerve [Figure 2e and f].

Postoperatively, the patient experienced immediate pain relief with a mild-to-moderate degree of imbalance. The patient was discharged on postoperative day 7 with mRS 2 due to mild imbalance. Follow-up at 12 and 36 months showed that the patient was pain and medication free with mRS 1 due to her mild imbalance. Postoperative computed tomography angiography showed complete AVM removal [Figure 2g and h].

## Discussion

The treatment for TN secondary cAVM still poses a great challenge. The difficulties arise from the need to address the neurovascular compression causing facial pain and concurrently address the formidable nature of a cAVM with a high risk of bleeding. The strong relation between the nerve compression and AVM makes it very rational to treat both conditions simultaneously. Due to its rarity and heterogeneity, no treatment consensus has been made to solve this problem, and one may not be practical either. Microsurgery, if feasible, offers a good option to completely remove the AVM and decompress the trigeminal nerve. However, in more complex cases, the treatment may need multimodality approaches to achieve a better clinical outcome. Of all treatment combinations, a combination of endovascular embolization and stereotactic radiosurgery seems to be the most favored one reported in the literature. Li *et al*. report only 15 cases of TN secondary to cAVM reported in the literature within the past 56 years including theirs.[4] Ten of these 15 cases were treated using embolization adjuvant therapy with satisfying pain outcome. Microsurgery by MVD was performed in five patients. Microsurgery for both MVD and AVM resection was only performed in three cases, and two of them were done in one surgery. Most of these surgeries resulted in an incomplete resection of the AVM, which exposes the patient to continued risk of hemorrhage from the AVM.

In addition, the AVM obliteration rate may not necessarily translate to TN relief. Intraoperative findings of our first case showed that even though most of AVM vessels were occluded [Figure 1d], the trigeminal nerve was still significantly compressed by dilated lateral pontomesencephalic vein and the SCA [Figure 1e]. This situation explained the patient's persistent facial pain even though follow-up DSA showed significant obliteration on the AVM. A similar experience on GKRS outcome was also reported by Sato in 2003. In their case, the AVM nidus was still prominent 1 year after the GKRS, and the trigeminal nerve was still compressed by the arterialized veins.[7] Above all, the GKRS efficacy on cAVM treatment is time dependent and has been reported to have total obliteration rates of 40%–50% at 3-year follow-up and

more than 70% at 5 years.[8-10] Therefore, in TN secondary to cAVM, GKRS alone for AVM treatment may not be adequate to cure the AVM. GKRS for classic TN has been well reported with poorer results compared to MVD. One systematic review of five prospective studies for the primary TN treatment showed that complete pain relief was achieved in 96% and 72% with MVD and GKRS, respectively.[11] In another meta-analyses comparing GKRS and MVD, short-term complete pain relief was achieved in 66% and 91%, respectively, and long-term complete pain relief was achieved in 44% and 69%, respectively.[12] The only downside of MVD was its postoperative complications that reached 11.6%.[12] Furthermore, in the case of TN secondary to cAVM, the risk of postoperative complications is presumably higher due to dilated vessels and arterialized veins which may obstruct the surgical corridor, compress the nerve, and are prone to rupture to potentially catastrophic hemorrhage. In 2017, a group from Beijing reported TN secondary to cAVM which was unrecognized before their first surgery. They had to terminate the MVD and eventually perform a successful second surgery 5 days later by combining AVM resection and MVD.[4] Our second case resembles their experience where the previous surgeons did not diagnose the AVM before the surgery and had to terminate the surgery after massive bleeding during dural opening. However, with adequate preparation, this case was successfully treated as has already been described above.

In our opinion, if the location and grading of the AVM can be safely accessed surgically, then surgery can be considered as one of the most effective ways to treat both the AVM and TN. Evidence shows that GKRS is a reasonable option to cure the AVM, but there are some concerns regarding the procedure. Concerns include the latency time to occlude the AVM, limited availability in developing countries, the possibility of post-GKRS extensive brain edema due to radiation necrosis or venous thrombosis, and finally, the efficacy of GKRS for TN reduces substantially over time.[8,12,13] Studies reported initial pain relief >90%, but it gradually decreasing to about 40%–70% after 5 years. AVM treatment through an endovascular approach is mostly performed as an adjuvant therapy to either surgery or GKRS. Curative embolization is only successful in 10%–20% of cases, and complication rates are over 30%.[14-16] Moreover, several factors such as higher cost, lack of pain relief, and lower rate of post-GKRS AVM obliteration after embolization made the treatment unappealing in most circumstances. As the case is relatively rare and AVMs are distinctly heterogeneous lesions, no adequate evidence will support a standard treatment algorithm. A tailored approach using a multimodality strategy based on clinical and social status should be embraced to treat this complex disease.

## Conclusion

We have reported two cases of TN related to cAVMs that were successfully treated using a microsurgical approach. Microsurgical procedures offer an important treatment modality in managing TN secondary to cAVMs.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot bechrological order guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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