Trigeminal neuralgia related to arteriovenous malformation of the posterior fossa: Three case reports and a review of the literature

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Summary
Objective: To describe the rare association of trigeminal neuralgia (TGN) with a brain arteriovenous malformation (bAVM) of the posterior fossa.

Patients and methods: This is a report of three patients presenting with TGN due to vascular compression by a bAVM of the posterior fossa, with emphasis on clinical presentation, diagnostic imaging, management and follow-up. Magnetic resonance imaging (MRI) was performed with sequences in thin slices in the same section plane using a 3D time of flight (TOF) and axial T2-weighted driven equilibrium (DRIVE) of the posterior fossa.

Results: No bleeding episodes were documented in the three patients. MRI and digital subtraction angiography (DSA) showed a posterior fossa bAVM with a nidus surrounding the trigeminal nerve, fed by arteries from the carotid and vertebrobasilar systems. Within a few days, medical treatment effectively alleviated the symptoms, with no more pain during follow-ups at 6, 10 and 18 months. No invasive treatment was performed because the bAVMs were considered to have a low risk of bleeding.

Conclusion: TGN related to a bAVM can mimic classical TGN. MRI and DSA are the imaging methods of choice. Medical treatment remains the first line of therapy, but if that fails, multimodal invasive treatment may be an alternative for pain relief.

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Introduction

The most frequent clinical presentations of brain arteriovenous malformations (bAVMs) are hemorrhage, seizure, chronic headache and focal deficits (not related to cerebral...
hemorrhage) [1—3]. A bAVM is rarely revealed by secondary trigeminal neuralgia (TGN), an association that has been described in only a few case reports [1,4—13]. According to a recent study, 1.3% of all bAVMs are revealed by TGN, and 9.8% are posterior fossa bAVMs [5]. In published cases, different treatments have been performed for TGN related to bAVM compression, often using a multimodal approach. Analgesics were used for pain relief and, in persistent cases, more invasive treatments such as embolization with coils or liquid embolic agents, radiosurgery or surgery have been used [5—13].

This is a report of our experience with three patients presenting with a TGN due to vascular compression by a posterior fossa bAVM, with emphasis on clinical presentation, diagnostic imaging, management and follow-up.

Patients and methods

During a 7-year period from March 2003 to May 2010, 101 patients were diagnosed with bAVMs in our department. Among these patients were included those who presented with a bAVM in the posterior fossa with TGN as the primary presenting symptom. Each of these patients underwent magnetic resonance imaging (MRI), performed with a 3T whole-body MRI scanner (Achieva, Philips Healthcare, Best, The Netherlands), with digital subtraction angiography (DSA) imaging, using a biplane angiography unit (AXIOM Artis, Siemens, Erlangen, Germany), within 1 week of the MRI examination. MR imaging was performed with sequences in thin slices in the same section plane, using a 3D time of flight (TOF) with 1.1 mm thickness and 0.55 mm spacing, and axial T2-weighted driven equilibrium (DRIVE) with 1 mm thickness and 0.5 mm spacing of the posterior fossa. The sagittal and coronal views were reconstructed in the plane of the trigeminal nerve. DSA imaging was performed by selective contrast injection of all territories feeding the bAVM in standard orthogonal anteroposterior, lateral and oblique projections.

Also studied were each patient’s medical history, clinical presentation, diagnostic modalities, bAVM angioarchitecture, management and follow-up. A review of the literature regarding this special association was also carried out, along with a discussion of the clinical TGN presentation especially in relation to a posterior fossa bAVM, the pathophysiology and the main goal of treatment for such cases.

Results

Of the 101 patients with a cerebral AVM discovered during the study period, 11 (10.8%) patients had a posterior fossa bAVM and three (3.0%) patients presented with a typical TGN as their first symptom. No bleeding episodes were reported in these three patients. Thus, in three of 11 (27.3%) patients with posterior fossa bAVMs, the bAVM was associated with TGN.

For each patient, T2 DRIVE and 3D TOF imaging showed numerous high-flow blood vessels in contact with the cisternal portion of the fifth cranial nerve, including the root entry/exit zone (REZ) and central nervous system (CNS) segment. It was difficult to determine whether the feeding arteries or the nidi was responsible for the compression. Also, no signs of recent or former bleeding were observed.

In the three TGN patients, DSA revealed that the bAVM, fed principally by arteries from the vertebrobasilar system, had abnormal early venous drainage into the superior petrosal sinus. The nidi was compact, plexiform, small (< 3 cm), and located in the cerebellopontine angle (CPA) on the free edge of the tentorium cerebelli and surrounding the cisternal segment of the trigeminal nerve. No aneurysms were seen within the nidi, on the pedicle or in the circle of Willis. No stenosis was observed in the draining veins.

In all three patients, medical treatment with carbamazepine effectively alleviated symptoms within just a few days. In addition, there were no bleeding episodes recorded in these three patients; thus, no invasive treatment (endovascular treatment, radiosurgery or microsurgery) was necessary.

Observation 1

A 61-year-old man suffered from a right-sided TGN for 2 years; it involved the mandibular division with 10 days of pain every month, and was triggered by eating, tooth brushing and talking. On examination, there was no evidence of a trigger zone or neurological deficit. The pain decreased significantly with carbamazepine treatment (200 mg × 2/day). MRI and DSA revealed a bAVM fed by the right superior cerebellar artery and the meningohypophyseal trunk of the carotid siphon. The nidi was small, located in the upper cerebellopontine cistern, and drained into the right superior petrosal sinus and then into the right lateral sinus. Drug treatment allowed recovery with no more pain. Due to the side effects of carbamazepine (drowsiness, instability), the patient switched to oxcarbazepine (450 mg × 3/day), and was pain-free during the 10-month follow-up.

Observation 2

A 64-year-old woman had a history of right TGN of several years’ duration involving the maxillary and mandibular divisions, with pain interspersed by years of pain-free intervals. On examination, there was no evidence of neurological deficit, but a trigger zone on the right zygoma was detected. MRI and DSA revealed a bAVM (Figs. 1—4) fed by the middle cerebellar artery, the meningohypophyseal trunk of the carotid siphon and the external carotid artery (accessory meningeal artery). The small nidi surrounded the right trigeminal nerve REZ on the free edge of the cerebellar tentorium, and was drained by the deep venous system. Drug treatment with carbamazepine (400 mg × 2/day) allowed the patient’s recovery, with no more pain during the 18-month follow-up.

Observation 3

A 50-year-old man had an approximately 1-year history of lancinating episodic left TGN of the mandibular division, triggered by chewing and tooth brushing. On examination,
there was no evidence of neurological deficit, but a trigger zone at the left corner of the mouth was detected. MRI and DSA showed a bAVM with the small nidus surrounding the left trigeminal nerve REZ, fed by the left middle cerebellar artery, the left superior cerebellar artery and the inferolateral trunk of the carotid siphon. Initially, drug treatment with carbamazepine (200 mg × 2/day) brought about partial pain relief; however, the patient had to stop treatment due to a skin allergy. Because of the large number of pain episodes, microcompression of the gasserian ganglion was performed, which resulted in partial disappearance of the pain over the 6-month follow-up.

Discussion

Clinical presentation

Based on the various published cases and our above three cases, the clinical presentation of posterior fossa bAVM-related TGN is similar to that of classical TGN: facial pain, with no other neurological deficit, occurring intermittently in the affected dermatome [4]. No bleeding episodes were reported in either our three cases or, as far as we can tell, in the 59 patients described over the past 75 years with TGN related to bAVM [10]. Thus, TGN is rarely related to hemorrhage from a posterior fossa bAVM, and bAVMs revealed by
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Figure 3  Internal carotid angiogram of the patient in Observation 2: (A) anteroposterior view; and (B) lateral view. The meningohypophyseal trunk of the carotid siphon (arrow) is feeding the nidus.

Figure 4  External carotid angiogram of the patient in Observation 2: (A) anteroposterior view; and (B) lateral view. The accessory meningeal artery (arrow) is feeding the nidus.

TGN show no evidence of bleeding after diagnosis. However, Drake et al. [14] reported three cases of CPA bAVMs with TGN and subarachnoid hemorrhage on diagnosis, and the facial pain description was the same as in classical TGN.

Pathophysiology

Location of compression
The cisternal portion of the fifth cranial nerve is composed of a CNS segment, with a length of about 2.6 mm, and a peripheral nervous system (PNS) segment, separated by the REZ [15]. Several hypotheses have been proposed, describing mechanisms of TGN associated with vascular anomalies.

Jannetta [16] was the first to explain that the mechanical effect of the pulsating vessel must occur at the REZ of the cranial nerve to cause symptoms. De Ridder et al. [15] added that vascular compression can occur both at the REZ and along the CNS segment to cause symptoms, mainly because histological studies have demonstrated that the CNS segment is more sensitive to vascular compression than the PNS segment.

Type of vessel involved
Another feature found in TGN related to a posterior fossa AVM is contact between the fifth cranial nerve and a high-flow blood vessel, as observed in our three cases and in most of the cases reported in the literature [5,6,9].
Arterial vessels (main feeding arteries or nidus angioma) are often involved, but high-flow arterialized draining veins may also be involved — albeit rarely — as described by Sato et al. [12], who reported that a high-flow arterialized draining vein of a contralateral parenchymal AVM was responsible for compression. In such cases, a cisternal bAVM is more likely to be responsible for the TGN than a parenchymal bAVM because of the location of the nidus or main feeder arteries in the preoptic cistern around the cisternal portion of the fifth cranial nerve. Arterial vessel involvement corresponds to the vascularization of the fifth cranial nerve and the gasserian ganglion, which are highly variable from one patient to another, perhaps involving the branch of the meningoephrineal trunk from the internal carotid siphon and middle or accessory meningeal artery from the external carotid in the gasserian ganglion, or the middle cerebellar artery from the vertebrobasilar system and/or pontine artery from the basilar artery in the fifth cranial nerve.

**Imaging**

MR imaging is essential, and should be performed routinely in all patients with TGN to identify the cause and to exclude a structural mass lesion, such as a bAVM (2%), prior to any surgery [4,5]. Axial DRIVE and 3D TOF images are the most important sequences for analyzing the posterior fossa and trigeminal nerve, and for viewing bAVMs in the CPA. These need to be performed in thin slices and in the same section plane, and the analysis must be based on the native images to precisely identify the relationship between the fifth cranial nerve and the bAVM [17,18]. The presence of hemosiderin indicates previous hemorrhage. However, it remains difficult to identify all of the feeding arteries and drainage veins in the complex, and to reveal some associated aneurysms [3,19].

DSA remains the gold-standard imaging technique for bAVMs. DSA shows in detail the angioarchitecture (feeding arteries, nidus, venous drainage and associated aneurysm) of bAVMs, and is useful when planning the potential operative treatment of the bAVM [1,3,19].

**Treatment**

For a posterior fossa bAVM revealed by TGN, but with no evidence of bleeding, the main goal of treatment is to relieve the patient of pain. The decision to either completely obliterate an intact bAVM or to only intervene enough to relieve the TGN depends on the balance between the surgical risk for a lesion located in an eloquent area [3,20,21,23] and the risk of bleeding, which is lower for unruptured bAVMs [3,10,22]. Thus, medical treatment with anticonvulsants, such as carbamazepine, plays a major role and remains the first line of therapy. If medical treatment fails or if intolerable side effects occur, then invasive multimodal treatment becomes an alternative for pain relief [3,20,21,23]. Nevertheless, it is still important to consider the risk of bleeding when assessing any invasive treatment.

In the three cases reported here, the bAVMs were considered to have low risk of rupture, and medical treatment relieved the pain; for these reasons, no invasive treatment was necessary.

**Conclusion**

A posterior fossa bAVM is a rare cause of TGN. Although the pathophysiology remains vague, contact with the REZ or CNS segment of the fifth cranial nerve is a necessary finding, and vascular compression occurs with a high-flow blood vessel. Symptomatic medical treatment should be initially proposed to relieve pain in this rare etiology of TGN before considering multimodal invasive treatment. Of course, assessing the risk of bleeding is always important when considering invasive treatment.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

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