Trigeminal neuralgia caused by microarteriovenous malformations of the trigeminal nerve root entry zone: symptomatic relief following complete excision of the lesion with nerve root preservation


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Object. Within a series of 341 consecutive patients who underwent posterior fossa surgery for trigeminal neuralgia (TN), in five the cause was found to be a microarteriovenous malformation (micro-AVM) located in the region of the trigeminal nerve root entry zone (REZ). The surgical management and clinical outcomes of these cases are presented.

Methods. Patients were identified from a prospectively collected database of all cases of TN treated at one institution between 1980 and 2000. Presentation was clinically indistinguishable from TN caused by vascular compression. Preoperative imaging, including computerized tomography scanning (two cases) and magnetic resonance (MR) imaging and MR angiography (three cases), failed to demonstrate an AVM except for one case in which multiple abnormal vessels were identified in the trigeminal REZ on an MR image obtained using a 1.5-tesla magnet. All patients underwent a standard retromastoid craniotomy. In all cases a small AVM embedded in the trigeminal REZ was identified and completely excised, with preservation of the trigeminal nerve. All patients experienced immediate relief of pain following surgery. Postoperatively, in one patient a small pontine hematoma developed, resulting in permanent trigeminal nerve anesthesia in the V2 and V3 divisions. All patients were free from pain at a mean follow-up period of 30 months.

Conclusions. These rare lesions are usually angiographically occult, but may sometimes be identifiable on high-resolution MR images. Total microsurgical resection with nerve preservation is possible, although operative complications are relatively common, reflecting the intimate association between these lesions and the pons. Complete resection is advised not only for symptom relief, but also to eliminate the theoretical risk of pontine hemorrhage.

KEY WORDS • microarteriovenous malformation • trigeminal neuralgia • nerve preservation

Clinical Material and Methods

During a 20-year period (1980–2000), 341 patients underwent posterior fossa surgery for trigeminal neuralgia (performed by the senior author, H.B.C.) for TN (Table 1). A prospectively collected database of all patients with TN identified five in whom the TN was found at surgery to be caused by a micro-AVM. A review of medical records, operative notes, and radiological images in these patients was undertaken.

The mean age at onset of symptoms was 42 years (range 35–55 years). The male/female ratio was 1:4. The mean duration of symptoms of TN before surgery was 5.8 years (range 1–10 years). All patients presented with typical TN and displayed no other unusual neurological symptoms or signs. The pain was located on the right side in four patients and on the left side in one. Four patients experienced pain in the third division of the trigeminal nerve and one experienced pain in the first and second divisions. No patient had a history suggestive of either subarachnoid, brainstem, or cerebellar hemorrhage. One patient had a strong family history of TN.

Preoperative imaging was obtained in all cases. Two patients underwent contrast-enhanced CT scanning; in both cases the findings were normal. The remaining three pa-
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tients underwent MR imaging (with Gd enhancement) and MR angiography. In one patient, multiple vascular loops were identified on the symptomatic side; these impinged on and distorted the trigeminal REZ (Fig. 1). In another patient a single vascular loop was identified, causing possible compression in the region of the trigeminal REZ (Fig. 2). This loop was considered by the neuroradiologist to be the SCA; in fact, during the operation the vessel was identified as an arterialized vein (Fig. 3A). No patient underwent preoperative cerebrovascular angiography.

All five patients had initially been treated with carbamazepine. Four patients showed some initial response to this treatment; however, all experienced breakthrough pain and eventually complete relapse of their symptoms, which became refractory to the maximum tolerated dosage of carbamazepine. One patient failed to respond to carbamazepine and underwent emergency radiofrequency lesioning of the trigeminal ganglion; this resulted in complete relief of pain for 6 months prior to a relapse of the TN.

Surgical Technique

After arrival in the operating room, all patients were placed in the park-bench position, with the head immobilized in a Mayfield clamp. The surgical approach was made through a small superior retromastoid craniotomy and the exposed in the standard manner. Arterialized veins in the region of the trigeminal REZ were exposed in the standard manner. Arterialized veins in the region of the trigeminal REZ were identified in all cases (Fig. 3A); these vessels were followed into the nidus of the AVM. The nidus was composed of a small knot of arterioles and small arteries located within the trigeminal REZ and the adjacent pons. In all cases the nidus was less than 1 cm in diameter. The nidus was frequently found to be predominantly subpial in location (Fig. 3B) and was not always immediately apparent. The feeding vessels originated from intrinsic pontine arteries in all cases, with additional feeding vessels provided by the anterior inferior cerebellar artery in one case and the SCA in another. In this latter case, the SCA loops also impinged to a minor degree on the trigeminal nerve root and were, therefore, mobilized and separated from the nerve by using Teflon slings secured to the tentorium cerebelli with fibrin glue. No dural feeding vessels were identified. In all cases total extirpation of the nidus was achieved, with preservation of the trigeminal nerve root (Fig. 3C). In some cases abnormal vessels passed into the trigeminal fibers as they entered the pons. The fibers were separated in line with the long axis of the nerve and the vessels were coagulated. The draining veins—usually superior cerebellar, pontine, and petrosal bridging veins—were also coagulated. In all cases, no evidence of previous hemorrhage was noted at the operation.

Results

All patients experienced immediate relief of pain following surgery. Three patients underwent postoperative angiography, which demonstrated no evidence of residual AVM. Postoperatively, in one patient a small cerebellopontine hematoma developed, resulting in transient symptoms of facial weakness, ataxia, and vomiting (all of which completely resolved), permanent trigeminal nerve anesthesia in the V2 and V3 divisions, and hypesthesia with an absent corneal reflex in the V1 division. Three patients had facial hypesthesia, with an absent corneal reflex in one patient (Table 2). There were no other postoperative complications. One patient experienced recurrence of pain 4 years after surgery; in that case the symptoms resolved in response to a regimen of carbamazepine (400 mg/day) and have not recurred. The mean follow-up period was 44 months (range 9–109 months). During the follow-up period so far there have been no episodes of hemorrhage, and all patients remain free from pain.

Discussion

The term “cryptic AVM” was used by Tsubaki, et al., to describe the only other series of patients in which TN was caused by small AVMs involving the trigeminal nerve REZ. In that series, six of seven patients underwent angiography, which demonstrated positive findings in one case, and contrast-enhanced CT scanning, which was nondiagnostic in all cases. No patient underwent MR imaging. The term “cryptic AVM” is not clearly defined in the literature. Crawford and Russell originally coined the term in 1956 to describe vascular lesions that remained clinically silent before hemorrhage. A more common interpretation is that cryptic AVMs are angiographically occult. This does not necessarily relate solely to small AVMs, but may include larger lesions that are angiographically occult because of the presence of vessel thrombosis or hematoma.

Many lesions described as cryptic AVMs are, in fact, cavernous angiomas. The term “cryptic cerebrovascular malformation” is less specific and can be used to describe any of the four classic angiographically occult vascular anomalies: cavernomas, micro-AVMs, venous angiomas, and telangiectasias.

We propose that the term “microarteriovenous malformation” should be used to describe these lesions. Yaşargil defined micro-AVMs as a subclass of AVMs in which the diameter of the nidus is less than 1 cm. We believe that this term provides a better definition of these lesions, the imaging manifestations of which are notoriously variable, because the definition is based on the anatomical absolute of a nidus diameter. Furthermore, because cerebral angiography is not recommended for patients presenting with TN without atypical features, it was not performed preopera-

<table>
<thead>
<tr>
<th>Cause of TN</th>
<th>No. of Patients (%)</th>
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<tbody>
<tr>
<td>vascular compression</td>
<td>269 (78.9)</td>
</tr>
<tr>
<td>tumor</td>
<td>6 (1.8)</td>
</tr>
<tr>
<td>meningioma</td>
<td>2 (0.6)</td>
</tr>
<tr>
<td>trigeminal schwannoma</td>
<td>1 (0.3)</td>
</tr>
<tr>
<td>metastasis</td>
<td>5 (1.5)</td>
</tr>
<tr>
<td>micro-AVM</td>
<td>1 (0.3)</td>
</tr>
<tr>
<td>AVM</td>
<td>45 (13.2)</td>
</tr>
<tr>
<td>multiple sclerosis</td>
<td>6 (1.8)</td>
</tr>
<tr>
<td>w/ vessel</td>
<td>6 (1.8)</td>
</tr>
<tr>
<td>w/o vessel</td>
<td>6 (1.8)</td>
</tr>
<tr>
<td>idiopathic (rhizotomy performed)</td>
<td>341 (100)</td>
</tr>
</tbody>
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T A B L E 1
Summary of all causes of TN from a prospectively collected database (1980–2000)
tively in any patient in our series. Although the lesions we describe are clearly similar to those described by Tsubaki, et al., we would prefer to reserve the term “cryptic” (if it is to be used at all) for confirmed angiographically occult AVMs. Of note, one case reported by Tsubaki, et al., cannot be classified as a true AVM because a dural feeding vessel was present.

There are numerous reports documenting AVMs as a cause of TN; with these larger lesions, however, this symptom is seldom present in isolation, and symptoms and signs of other cranial nerve, brainstem, and cerebellar dysfunction are frequently identified, often in association with hemorrhage. In contrast, the patients harboring cryptic AVMs, who were described in the series reported by Tsubaki, et al., and those patients in the current series all had presentations indistinguishable from primary TN due to vascular compression. In our series symptoms were caused by direct compression of the nidus at the trigeminal REZ. In one case additional compression was caused by a pathologically enlarged SCA feeding vessel. A similar description of an SCA feeding vessel (to a large cerebellar AVM) presenting as TN has also been described.

Microarteriovenous malformations, such as those described in our series, are usually angiographically occult, although selective microcatheter views may increase the likelihood of detection. Contrast-enhanced CT scans and MR images or MR angiograms usually are also nondiagnostic. Our experience with high-resolution MR imaging has shown that a nidus may be identified in some cases. Furthermore, arterialized draining veins may also be mistaken for arterial compression of the trigeminal nerve, the nidus itself remaining occult on imaging studies. In our se-

![Fig. 1. Magnetic resonance angiograms (“un-MIPped,” Gd-enhanced, time-of-flight, axial sequence [fast field echo]), through the right cerebellopontine angle (CPA) revealing multiple abnormal vessels (thin arrows) in the region of the trigeminal REZ (thick arrow), which is consistent with a diagnosis of AVM. MIP = maximum intensity projection.](image1)

![Fig. 2. Magnetic resonance angiograms (“un-MIPped,” Gd-enhanced, time-of-flight, axial sequence [fast field echo], through the right CPA revealing a vessel (small arrow), thought to be the SCA, in contact with the right trigeminal REZ (large arrow). During the operation this vessel was found to be an arterialized vein; the associated micro-AVM, which involved the REZ and adjacent pons, was not exhibited on MR imaging.](image2)
ries the majority of feeding vessels originated from small intrinsic pontine arteries. These vessels would be extremely difficult to discern on cerebral angiography and would be too small to identify when other imaging modalities are used. Larger vessels that would be more readily identifiable were involved in only two cases.

The long-term risk of hemorrhage from AVMs is not clear. In the current series there was no evidence of previous hemorrhage in any AVM. Long-term follow-up studies have shown both ruptured and unruptured AVMs to have a similar risk of future bleeding, between 2 and 4% per annum, although the authors of a more recent study have suggested that previously ruptured AVMs carry a substantially higher risk of bleeding. The risk of hemorrhage is not higher for posterior fossa AVMs compared with supratentorial AVMs. Bleeding from small AVMs has been found to be more severe than bleeding from larger lesions, and patients with small AVMs have been found to be more likely to present with hemorrhage. Overall, however, smaller lesions are probably less likely to bleed. The natural history of untreated micro-AVMs may tend toward an increase in size, causing further neurological deficits from a direct mass effect or steal phenomenon. Taking these factors into account, there is a general consensus that, whenever technically possible, treatment of brainstem AVMs should be aimed at total microsurgical extirpation of the lesion.

For larger AVMs, preoperative embolization may facilitate safe removal; however, there is little evidence to support its use in the management of AVMs, in which the feeding vessels may not be identifiable on angiograms, even when recent advances in superselective microcatheterization techniques have been implemented. Radiosurgery has also been advocated as both a primary and adjuvant treatment of brainstem AVMs; however, early edema may result in neurological deficits, the risk of hemorrhage is not immediately eliminated, the procedure may render subsequent surgical resections of lesions more hazardous, and there is a risk of late radiation-induced necrosis. It has been proposed that radiosurgery should not be used in the treatment of angiographically occult AVMs because there is no definite evidence showing that the treatment prevents these lesions from rebleeding. In a recent series, five of 30 patients suffered fatal rebleeding from small (mean diameter 1.26 cm) brainstem AVMs that had been treated with radiosurgery, within a mean follow-up period of 52 months. Furthermore, in the subgroup of patients with TN, radiosurgery would not offer a chance of immediate symptomatic relief, although symptomatic improvement has been reported in patients with TN who have been treated with radiosurgery, and the lesions themselves usually are only identified at surgery.

The micro-AVMs causing TN are not usually readily apparent during surgery and their identification requires a high index of suspicion together with careful inspection of the trigeminal REZ. In our series micro-AVMs were identified to be the cause of TN in 1.5% of all cases. This is substantially higher than the 0.6% incidence reported by Tsubaki, et al. It is a little surprising that similar cases have not been identified in other large series of patients undergoing surgical exploration of the posterior fossa, and it is possible that this entity is underrecognized. All these micro-AVMs were encountered during surgical exploration and were not identified on preoperative images. We therefore judged that resection of the lesions would be appropriate without performing intraoperative angiography, selective or otherwise. This decision was made in light of the previous publication by Tsubaki, et al., who had demonstrated that these micro-AVMs are not visualized by preoperative angiography. Recent advances in superselective microcatheter angiography may now facilitate identification.

Fig. 3. A: Operative view following initial exposure of the right CPA showing arterialized pontine veins (long arrows) crossing the trigeminal sensory root (thick arrow) and draining into the superior cerebellar vein (arrowhead). Note that the AVM nidus, located deep with respect to the pial surface, is not visible. B: The arterialized veins have been followed deep with respect to the pial surface and coagulated. The pial surface has been dissected to expose the nidus (arrows). C: Operative view of the right CPA following extirpation of the nidus. The trigeminal root (arrow) remains intact.
of these lesions, providing valuable additional information before resection is attempted.

The surgical technique demands extensive experience in posterior fossa microsurgery. As with microvascular decompression, there is little margin for error, making this an unforgiving procedure. All micro-AVMs in this series were subpial or parenchymal (within the brainstem), with feeding arteries supplying the micro-AVMs through the pons. Meticulous hemostasis of these perforating vessels is critical to avoid potentially life-threatening brainstem hemorrhage. If the surgeon is less experienced, it may be prudent to abandon the procedure, await further angiographic studies, and consider referral to an expert subspecialist.

The initial step in the surgical resection of these lesions is the identification of an arterialized draining vein on the cortical surface in the region of the trigeminal REZ. The bulk of the nidus frequently lies deep with respect to the pial surface and may not be immediately apparent. The draining vein is followed proximally, directly to the nidus. In the only series previously published, five of seven patients underwent resection of the trigeminal nerve root because the nidus was diffusely present within the root. We have shown that these lesions can be totally removed without sacrificing the nerve root. If the nidus of the micro-AVM involves the root we would advocate splitting the nerve along its long axis to access the nidus, which can then be directly coagulated and excised. Using this technique we achieved total removal of the lesion and no patient was left with complete facial hemianesthesia. Function of the motor root of the trigeminal nerve was preserved in all cases.

Postoperative angiography is recommended in all patients with micro-AVMs, even if initial angiography (if performed) is nondiagnostic. Superselective microcatheter views may help to identify residual small feeding vessels. Postoperative complications occur more frequently than those reported following straightforward microvascular decompression procedures, reflecting the intimate association of these lesions with both the trigeminal REZ and the adjacent pons.

**Conclusions**

These micro-AVMs are a rare cause of TN. High-resolution MR imaging may identify some, but not all, cases in which they occur preoperatively. Total microsurgical resection with nerve preservation is possible. Operative complications are relatively common, reflecting the intimate association of these lesions and the pons. Complete resection is advised not only for symptom relief, but to eliminate the risk of pontine hemorrhage.

**References**

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56. Tsubaki S, Fukushima T, Tamagawa T, et al: Parapontine trigem-

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