Challenging vascular compression as a cause of trigeminal neuralgia

To The Editor: As the treatment of patients suffering from trigeminal neuralgia (TN) is a significant portion of my practice, I have always looked forward to Dr. Burchiel’s contributions to the literature. In their recent paper, Lee et al.² (Lee A, McCartney S, Burbidge C, et al: Trigeminal neuralgia occurs and recurs in the absence of neurovascular compression. Clinical article. J Neurosurg 120:1048–1054, May 2014) state that “compression of the trigeminal nerve by a blood vessel . . . is still thought to be the most common cause for TN,” but they also state that “the hypothesis that TN is caused by neurovascular conflict must be challenged.” They note that 99.94% of individuals with trigeminal neurovascular compression (NVC) do not have TN.

It is clear that not all cases of TN are caused by vascular compression. However, anecdotally, it is also clear to me that sometimes trivial, unnamed, small vessels cause TN. I suspect that these vessels “fly under the radar” of many surgeons intraoperatively and are also undetected by even the best MRI. Just because vascular compression was not seen on preoperative imaging or was not visualized intraoperatively does not mean that it was not present. Neither the tools nor the surgeon is 100% accurate. Despite these imperfections, however, neurosurgeons can treat the majority of patients with TN (those who do not have multiple sclerosis or a posterior fossa or trigeminal tumor) using the Jannetta procedure. Most stop the medication they were taking for TN.

Without a doubt, vascular compression can lead to cranial nerve dysfunction. Is there still doubt about the cause of hemifacial spasm? Absolutely not.

Use the authors’ logic on our spine patients. Approximately 60% of MRI studies performed in patients older than 60 years who have never had low-back pain, sciatica, or neurogenic claudication are significantly abnormal.¹ Do we now challenge the notion that sometimes these abnormalities lead to painful conditions that are relieved by surgical intervention?

Rather than proposing a challenge to the vascular compression hypothesis, perhaps the challenge is to perform a better vascular decompression or to obtain better MRI studies. Perhaps the challenge is to find yet another cause of TN. Surely there are undiscovered factors in the TN story. Challenging accepted dogma may be healthy, but challenging Peter Jannetta’s work that has led to the successful treatment of so many grateful patients seems off-base. It would be a shame to unnecessarily reignite some of the past vitriolic discussion against Dr. Jannetta’s landmark contributions to neurosurgery and patients with TN.

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Disclosure
The author reports no conflict of interest.

References

Response: We thank Dr. Kalia for his analysis of our work and would like to comment on the underlying theme of his letter: the tenet that NVC is, with minor exceptions, the underlying cause of TN.

Using his standard, “just because vascular compression was not seen on preoperative imaging or was not visualized intraoperatively does not mean that it was not present,” approaches a religious conviction. We submit that if NVC cannot be imaged or visualized, it is not there. The fact that MRI or MR angiography (MRA) agrees so strikingly with the surgical findings validates the premise that nothing is being “missed” intraoperatively. As an aside, I agree with Dr. Kalia that hemifacial spasm (HFS) appears in many ways to be the exemplar of NVC producing a cranial nerve disorder. In practice, we have never seen a patient with HFS who did not have NVC at the root of the facial nerve. As imaging has improved over the past decade, we have also never seen an MRI or MRA study with 3D reconstruction that did not clearly demonstrate NVC. Clearly, NVC seems to play an essential, and universally detectable, role in this disorder.

Microvascular decompression is the single most important surgical advancement in the treatment of TN in the past 6 decades. It remains the most successful surgery, long term, for this disorder. Nevertheless, the success of this surgery should not be a pretense to stop thinking about this condition. If Gardner and Jannetta had not challenged conventional thinking, they would not have made their seminal contributions to the treatment of TN.

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Trigeminal neuralgia and the absence of neurovascular compression

To The Editor: I read with keen interest the article by Lee et al.7 (Lee A, McCartney S, Burbidge C, et al: Trigeminal neuralgia occurs and recurs in the absence of neurovascular compression. Clinical article. J Neurosurg 120:1048–1054, May 2014) and noted the provocative nature of their title and concluding statement regarding the etiology of trigeminal neuralgia (TN). In their retrospective review of 257 patients assessed over 18 years, they remarked the absence of neurovascular compression (NVC) in 29% and 18% of patients with TN Type 1 (TN1) and TN Type 2 (TN2), respectively, by utilizing MRI, intraoperative observations during microvascular decompression (MVD) surgery, and their own facial pain classification system.1,2 They concluded that “the hypothesis that TN is caused by neurovascular conflict must be challenged.” I respectfully suggest that another interpretation of their data is possible and that their conclusion should be challenged.

The authors have shown a strong correlation between MRI and operative findings, but they reported the absence of NVC in a significant proportion of their patients with TN1 and TN2. They have not, however, assessed the subset of patients with purely episodic pain commonly referred to as “classical TN.” This is an important distinction and should prompt a critical re-evaluation of their facial pain classification system. I suggest that their classifications of TN1 and TN2 facial pain lack specificity by including a significant proportion of patients who do not actually have TN and therefore are not expected to have a causative NVC. For example, their TN2 category is defined as an “idiopathic trigeminal facial pain that is aching, throbbing, or burning for more than 50% of the time and is constant in nature (constant background pain being the most significant attribute).”2 This definition is not compatible with the International Headache Society (IHS) International Classification of Headache Disorders (ICHD)-II 13.1.1 “classical trigeminal neuralgia,” in which pain is purely episodic with a caveat that “dull background pain may persist in some long-standing cases.” This has important clinical ramifications, as there is no substantive evidence that patients with predominantly constant facial pain—TN2 according to the authors classification—actually have TN or that they will benefit from the surgical interventions that are well established as effective for classical TN.

This line of reasoning also has important implications for their category of TN1, described as “sharp, shooting, electrical shock–like, episodic pain lasting several seconds, with pain-free intervals between attacks.”2 This definition is more in keeping with the IHS ICHD-II 13.1.1 classical TN, but there remains an important distinction. According to the authors’ definition, TN1 may also include patients suffering from additional, superimposed, or concurrent constant facial pain that may be prominent and severe for up to 49% of the day.1,2 The distinction between a level of constant pain for more than or less than 50% of the day has been set arbitrarily, and there is no clear clinical or etiological distinction between patients with TN1 and those with TN2. The TN1 and TN2 classifications also do not differentiate the important subset of patients with purely episodic, classical TN pain and other non-TN idiopathic craniofacial pain conditions.

The IHS ICHD-III (beta version) goes further to differentiate between 13.1.1.1 classical TN, purely paroxysmal, and 13.1.1.2 classical TN with concomitant persistent facial pain, the latter being defined by “recurrent attacks of unilateral facial pain fulfilling criteria for 13.1.1 classical TN and persistent facial pain of moderate intensity in the affected area.” Such a stringent differentiation between the purely episodic pain condition and one with up to a moderate associated constant pain element may be better suited to assessing the correlation between disease (that is, TN) and purported cause (that is, NVC).

The hypothesis that NVC is the root cause of classical TN is supported by two well-established principles: 1) vascular compression of the trigeminal nerve root is present in most patients and 2) atraumatic alleviation of such NVC with MVD surgery is usually successful in providing long-term relief from TN pain. The integrity of this NVC hypothesis is not diminished by the observation that not all patients with NVC develop TN or that technically successful MVD surgery does not always result in permanent disease cure. Clinical experience has indeed demonstrated a robust association between a diagnosis of classical TN and culprit NVC, as demonstrated in carefully reviewed MRI sequences, intraoperative observations, and long-term outcomes following MVD surgery. While MVD surgery pioneered by Jannetta6 does not guarantee a cure for TN, the success of this operation remains the strongest evidence that NVC causes TN. As suggested by Hutchinson in 1905,8 the selection of surgical treatment “should depend upon a scientific classification, based solely upon causes of neuralgia,” and this question appears to have been answered at least for classical TN.

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Disclosure

The author reports no conflict of interest.

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7. Lee A, McCartney S, Burbidge C, Raslan AM, Burchiel KJ: Trigeminal neuralgia occurs and recurs in the absence of neuro-
RESPONSE: We appreciate Dr. Kaufmann’s interest in our article concerning the etiology of TN. While the title may be “provocative,” we believe our interpretation of the data is fair. We further believe that in light of these data, “the hypothesis that TN is caused by neurovascular conflict must be challenged.”

We have shown that high-resolution MRI and MR angiography (MRA) can demonstrate the presence or absence of NVC with a high sensitivity and specificity. However, the diagnosis of TN is clearly based on a clinical history, not a neuroradiological interpretation. In patients with TN1, we found that 28.8% had no visible vascular compression on MRI or MRA. Our prior work has also shown that the rate of trigeminal NVC in an aged-matched asymptomatic population is 17%. These findings are further supported by a recent study1 of classical TN (ICHDI-II, 13.1.1), in which a meta-analysis showed that root entry zone NVC was detected in only 76% of asymptomatic and 17% of asymptomatic nerves—results strikingly similar to our own.

While an experienced clinician has little difficulty identifying a patient with classical TN (ICHDI-II, 13.1.1), in our experience very few patients with TN have purely episodic pain. Further, in our experience TN often “mutates” over time, such that pain that is initially episodic, lancinating, and electric shock-like can develop aspects of persistent pain after major attacks or between attacks. Unfortunately, patients with TN do not come in neat diagnostic categories, and categorization is a challenge. This makes the distinction between classical TN and concomitant persistent facial pain (ICHDI-II, 13.1.2) a subjective determination. For example, how does the ICHD-II definition distinguish a patient with pain that fingers for only a few minutes after an attack of classic pain from a patient in whom classic shock-like pain occurs on the background of a dominant, persistent aching or burning pain?

This subjectivity of diagnosis has confounded clinical research on TN, much as when research on traumatic brain injury was quiescent until the adoption of a common standard for the description of coma: the Glasgow Coma Score. “Stupor,” “semi-coma,” “obtundation,” and “lethargy” gave way to a numerical scale that allowed the study of traumatic brain injury. An explosion of research ensued. Trigeminal neuralgia is a condition that has been known for more than 300 years, and as yet there has never been a natural history study of the disorder. Terms such as “trigeminal neuralgia,” “atypical trigeminal neuralgia,” and “atypical facial pain” are simply not accurate enough to permit the longitudinal study of this disorder. With all due respect, the ICHD-II terminology is just the latest iteration of this subjectivity.

Almost a decade ago, we introduced a different terminology to describe TN that was based on the only true source of categorization we have for a pain syndrome, that is, the patient. This classification system is not dependent on the clinician’s interpretation, but rather on the patient’s experience. The distinction between TN1, as predominantly but not necessarily exclusively episodic pain, and TN2 has been shown to significantly influence outcome from MVD.6 In contrast to Dr. Kaufmann’s comments, patients with TN1 and TN2 do have about the same rate of NVC, and patients with TN2 do benefit from MVD, albeit not to the degree that TN1 patients do. He is correct, though, that there is “no clear clinical or etiological distinction between patients with TN1 and those with TN2.” In all likelihood, they are part of a continuum, with TN2 more the result of deafferentation producing a “central pain,” which is more resistant to the MVD procedure. Dr. Kaufmann should not interpret our findings as a disparagement of the association between vascular compression and TN. In fact, our data show that more than 70% of patients with TN1 or TN2 do have vascular compression of the nerve. This compares with an NVC base rate of 17% in the general population.3 Clearly, NVC is significantly related to TN. It simply cannot be the only factor, since TN occurs without any evidence of NVC in a substantial minority of cases. The additional observation that TN recurs after successful MVD at a surprisingly steady rate of about 4% per year also calls into question the theory that NVC is the root cause of TN.

The vascular compression theory of TN has provided patients with the most successful operation for this disorder, by far. However, the problem with powerful theories is that we often stop thinking about a disorder once we believe we have all the answers. In our opinion, there is still more to learn about this excruciating and fascinating condition.

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Timing of shunt placement after cranioplasty

To The Editor: We read with great interest the recent article by Heo et al. on the results of cranioplasty and ventriculoperitoneal (VP) shunt placement at the same time in patients with cranial defects and hydrocephalus (Heo J, Park SQ, Cho SJ, et al: Evaluation of simultaneous cranioplasty and ventriculoperitoneal shunt procedures. Clinical article. J Neurosurg 121:313–318, August 2014). Among the 51 patients who were studied, 32 underwent cranioplasty and VP shunt placement at the same time. In the remaining patients the average interval between cranioplasty and VP shunt placement was 2.4 months. The results of the study showed that simultaneous cranioplasty and VP shunt placement was significantly associated with complications and complications requiring reoperation.

Decompressive craniectomy has been associated with increased rates of complications. Among them, hydrocephalus is not an uncommon finding and requires treatment. The optimal timing for shunt placement in patients with hydrocephalus after decompressive craniectomy, however, has been a matter of debate. We have recently studied 63 cases involving patients who underwent decompressive craniectomy because of traumatic brain injury, middle cerebral artery infarct, or intracerebral hemorrhage. Among these patients, 23 developed hydrocephalus. In 11 cases, a VP shunt was placed and a cranioplasty was performed, either simultaneously or in a second stage. In the remaining cases, cranioplasty and placement of a ventriculostomy, with external CSF reservoir or monitor to measure the intracranial pressure, was performed simultaneously. After 3 to 5 days of serial pressure measurements and follow-up CT scans, a VP shunt was placed using the most appropriate opening pressure. Nine patients from the first group experienced treatment-related complications and 5 required reoperation. No patient in the second group required reoperation, and 2 patients did not require shunt placement.

Thus, we believe that the combination of cranioplasty and ventriculostomy followed by placement of a VP shunt later on, at the most appropriate opening pressure, is associated with fewer complications. Prospective studies with larger numbers of patients are obviously needed for clarification of the optimal timing for shunt placement in patients who develop hydrocephalus after decompressive craniectomy.

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Disclosure
The authors report no conflict of interest.

References

Response: We greatly appreciate Dr. Alexiou’s interest in our article. There is no consensus as to the timing and method of the cranioplasty and VP shunt placement in patients who require both operations after decompressive craniectomy. As he pointed out that cranioplasty and ventriculostomy followed by placement of a VP shunt later on, in the most appropriate opening pressure, is associated with fewer complications. Our study also showed that simultaneous cranioplasty and VP shunt placement was significantly associated with complications and complications requiring reoperation.

We think that when a patient needs both operations, especially with a bulging brain, CSF drainage via lumbar or ventricular puncture should be considered before the operation to control the intracranial pressure and avoid complications.

We again thank Dr. Alexiou for making his insightful remarks and hope that our report can help readers arrange their strategy for the cranioplasty and VP shunt placement in patients who require both operations after decompressive craniectomy.

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Pure arterial malformation of the posterior cerebral artery

To The Editor: We have read with great interest the article by McLaughlin and colleagues (McLaughlin N, Raychev R, Duckwiler G, et al: Pure arterial malformation of the posterior cerebral artery: importance of its recognition. Case report. J Neurosurg 119:655–660, September 2013). The authors reported for the first time the identification of a novel cerebrovascular pathological entity that, due to its particular anatomical features, has been called a “pure arterial malformation.” The authors suggested that this pathology differs from other similar vascular lesions such as dissecting aneurysms, dolichoectasia, or vascular malformations because of a presumably more benign prognosis. Since their report, another similar case has been published.

We have recently diagnosed a lesion whose angiographic characteristics were consistent with the mainstay diagnostic criteria for pure arterial malformations. In essence, this was a tortuous and redundant posterior communicating artery (PCoA) extending to the P 2 segment without signs of vessel dissection or arterialized veins.

This 1-year-old girl was admitted to our department because of a hemispheric stroke on the left side. She underwent an MR scan and, subsequently, a digital subtraction angiography study that showed moyamoya disease.
On the right side, apart from the stenosis of the distal internal carotid artery (ICA), we found a pure arterial malformation of the PCoA (Fig. 1). The patient underwent a combined revascularization procedure consisting of an extracranial-intracranial bypass and synangiosis on the right side. The arterial malformation was left untouched. The clinical course was uneventful.

Although reporting cases of pure arterial malformation may be important by itself to evaluate the clinical and epidemiological importance of this novel nosographic entity, our observation is not merely confirmatory because it differs from the previous ones for at least two reasons. First, our patient is a very young child, whereas all the cases reported in the literature were diagnosed in adulthood. The anatomical consistency of the lesion across different ages further supports the hypothesis of a benign malformation. Second, the association with moyamoya disease represents an absolutely rare occurrence that, on the one hand, may be the sign of a common genetic variant or, on the other hand, may be the corollary of increased hemodynamic stress in a predisposed patient.

**Disclosure**

The authors report no conflict of interest.

**References**


**Response:** We read with interest the case illustration presented by Lanterna and colleagues in their Letter to the Editor. Since the publication of our manuscript, in which we described in detail this vascular anomaly, 2 other examples of this entity have been reported. In addition, other colleagues have written to the senior author (N.A.M.), sharing well-documented cases of pure arterial malformations. In the cases reported by our group as well as the case reported by Lanzino's group, a dilated, tortuous, and redundant artery was found in the absence of other existing vasculopathy. In the case presented by Lanterna, the infant suffered from a left-sided stroke and right-sided probable moyamoya disease was diagnosed, which makes careful analysis of the angiogram so important, because many different vascular patterns may be encountered given the redistributed hemodynamic stress, as appropriately noted by Lanterna and colleagues. Two imaging sequences have been found to be critical in establishing the diagnosis of pure arterial malformations: 1) catheter angiogram with a 6-second frame, and 2) 3D angiogram. Only when reviewing the vascular anomaly on these imaging sequences could we reliably follow the dilated vessel through all its loops and turns.

We acknowledge that Lanterna and colleagues may have encountered another pure arterial malformation. However, in the setting of moyamoya disease, the enlarged vessel could also represent an enlarged perforator filling the posterior cerebral artery via perforator-to-perforator collateral vessels, which could explain the appearance of sluggish flow on the lateral views. Furthermore, on the images that are presented, it is difficult to assess if this represents a single redundant and tortuous vessel that can be followed or if there are many vessels. We would recommend completing the imaging assessment with the 2 described sequences when the patient presents for her follow-up catheter angiogram. This will also be informative regarding the potential impact of hemodynamic changes following the revascularization that was performed.

We encourage physicians to add their potential cases of pure arterial malformations to the growing body of literature and to share key imaging sequences confirming the diagnosis.

**Reference**

Cytochemical CSF analysis and antibiotic-impregnated external ventricular drains

To The Editor: We read with great interest the article by Stevens et al. (Stevens EA, Palavecino E, Sherertz RJ, et al: Effects of antibiotic-impregnated external ventricular drains on bacterial culture results: an in vitro analysis. Laboratory investigation. J Neurosurg 113:86–92, July 2010). Antibiotic-impregnated (AI) external ventricular drains (EVD) have their place in the prevention of CSF infections in patients with an external drain. The study by Stevens et al. has important clinical implications. We think that the question about the reliability of CSF cultures is not restricted to the use of AI drains, but it is a crucial issue in CSF infection management; in fact, therapy for CSF infection relies on endovenous and, in some cases, intrathecal antibiotic administration, and both endovenous and intrathecal antibiotic infusions can give false-negative cultures. In several years of experience we have observed that CSF cytochemical examination is a good marker of infection: concentration of glucose, proteins, and cell count can be altered in cases of mild shunt contamination or can persist at abnormal levels when microbiological cultures have already become negative. Reference values in our laboratory are 40–70 mg/dl for glucose, 20–40 mg/dl for proteins, and a cell count less than 5/mm³. It is our current clinical practice to treat patients with shunt infection by removing the infected shunt, placing a non–AI-EVD, and initiating endovenous antibiotic therapy that is continued also for a period of 24–48 hours after the new shunt has been implanted. In our practice the new shunt is implanted when we have obtained CSF samples on 3 consecutive days that have yielded negative microbiological cultures as well as normal glucose and protein levels and cell count. We also take into account cytochemical CSF examination findings when an AI-EVD has been used and the patient requires a permanent shunt. Unfortunately, we have not reviewed a series of patients, but in our experience, when the CSF cultures were negative and the chemical values were normal, shunt infections or reinfections were extremely rare. Therefore, we think that the CSF cytochemical examination is a safe and reliable marker of infection cure during antibiotic therapy and we encourage its use in clinical practice.

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