Chapter 10
Pain Disorders

Trigeminal Neuralgia

This disease involves intermittent, sudden, brief, severe, sharp (electric shock or stabbing) pains in the face (in the trigeminal distribution), which are usually triggered by light touch and usually respond to carbamazepine or oxcarbazepine. The pains are often described as "shooting" or "radiating" in nature. There can also be a minor component of constant achy pains in the trigeminal distribution. Spontaneous remissions are common. The definition of trigeminal neuralgia as any idiopathic spontaneous facial pain, subcategorized as TN1 if such pain is predominantly episodic and TN2 if it is predominantly constant, is not advisable. Such a broad definition will, necessarily, include many patients who clearly do not have true trigeminal neuralgia and will not likely benefit from the standard trigeminal neuralgia procedures (microvascular decompression/MVD or trigeminal denervation). Of note, if a patient experiences clear dramatic pain relief after one of the standard trigeminal neuralgia procedures, that suggests that trigeminal neuralgia was probably the correct diagnosis.

Trigeminal neuralgia should specifically be distinguished from post-herpetic neuralgia (which follows a shingles rash outbreak, usually in a V1 distribution, and often involves numbness and a predominantly constant achy pain), nerve injury pain (which can follow from some nerve injury, such as might be caused by dental work, and often involves numbness and a predominantly constant achy pain), and chronic paroxysmal hemicrania (in which the pain usually centers in and around the eye, involves autonomic features, and is very responsive to Indomethacin). Trigeminal neuralgia should also be distinguished from other types of idiopathic neuropathic facial pains (sometimes called "atypical facial pain") that are usually more constant and achier or burning in nature and are usually best treated with medicine and conservative management (see Case 10.1).

M. H. Brisman, *Put Down the Knife*,
https://doi.org/10.1007/978-3-031-48499-5_10

Case 10.1
This is a 41-year-old man with 15 years of intermittent pains in the right side of his face. He was otherwise healthy. The pains would come on every day. They occurred in the right jaw and could radiate back toward the ear. At times, the pain would radiate toward the neck and toward the left side. Flexeril and Xanax did not help. When he got the pain, he felt that taking ibuprofen prevented the pains from getting worse. The pains were achy, dull pains, about 4/10 in severity, on a scale of 1–10. The pains would develop gradually over hours and usually occurred in the afternoons. Exam was normal and brain MRI was unremarkable. This was felt to be a neuropathic facial pain. He was started on gabapentin 200 mg TID. His pain was completely resolved by the medicine.

Trigeminal neuralgia is usually caused by a blood vessel, usually an artery, contacting or compressing the trigeminal nerve root. It can also be caused by multiple sclerosis, or a brain mass contacting the trigeminal nerve root. Sometimes, no cause can be found. The first line treatment is with the antiseizure medicines carbamazepine or oxcarbazepine. Gabapentin is usually used as the second line medicine. Other antiseizure medicines can also be used. Dilantin is useful for acute uncontrolled pain in the emergency room because it can be loaded quickly. Work-up for trigeminal neuralgia is with a brain MRI. A fine cut T2 weighted sequence (FIESTA or CISS) may demonstrate the offending blood vessel. That said, vascular contact against the trigeminal nerve (or the facial or glossopharyngeal nerves for that matter) is a common occurrence and in no way confirms the diagnosis of a cranial nerve hyperactivity syndrome such as trigeminal neuralgia. Furthermore, a significant neuro-vascular contact can be present that is not fully appreciated on FIESTA imaging, and lack of "definitive" vascular compression should not discourage the performance of an MVD in an otherwise appropriate operative candidate.

For those patients for whom medicines do not adequately control the pain, or for whom the side effects of the medicines are not tolerable, a procedure is appropriate (microvascular decompression, percutaneous rhizotomy, or radiosurgery). Of note, one of the trigeminal neuralgia procedures is not likely to help a patient who has a facial pain syndrome other than trigeminal neuralgia. The microvascular decompression (MVD) is a good choice for younger patients (under about 65–70), who are healthy and do not have multiple sclerosis [69]. MVD is more likely to be effective if the offending vessel is an artery (usually the superior cerebellar artery) and is more likely to be successful if the offending artery was distorting the nerve. Neuronavigation is helpful in identifying the transverse-sigmoid junction. Intradurally, the petrosal vein can be sacrificed, if needed. The use of brain retractors should be avoided as this increases the chance of an eighth nerve injury. Furthermore, neuromonitoring changes of either the eighth or seventh nerve during the micro-dissection suggest an imminent retraction injury to the eighth nerve and

should prompt the surgeon to pause and then redirect the surgical activity. An endoscope can sometimes enhance the view, particularly distally toward the entrance of Meckel's cave. Veins contacting the trigeminal nerve can be cauterized and divided, but caution must be taken not to injure the nerve itself through a heating effect. Ideally at the end of the MVD procedure, there will be no blood vessels or implants (like Teflon felt) contacting the trigeminal nerve (see Figs. 10.1 and 10.2). Repeat MVDs should usually be avoided. The risk for repeat MVDs is much higher due to scarring, and postoperative pain relief that is obtained is frequently due to denervation, so these operations are generally no more than open rhizotomies.

Recent considerations for avoiding the use of Teflon felt during MVD are not unreasonable, as the felt itself often causes an aseptic meningitis (though this can usually be minimized by putting patients on a tapering 3-week course of dexamethasone after the surgery). If at the time of MVD no clear offending vessel is found, the nerve can be injured slightly by gently massaging the nerve or making two tiny grooves in the surface of the nerve with a micro-dissector (a modified combing technique). An "open rhizotomy" can be performed in the same way, in the rare case that the less invasive denervating techniques are not successful.

For older patients, patients with serious medical problems, patients with multiple sclerosis, patients who have already had an MVD, and patients who just prefer a less invasive alternative, a denervating procedure (such as percutaneous rhizotomy or Gamma Knife) is reasonable.

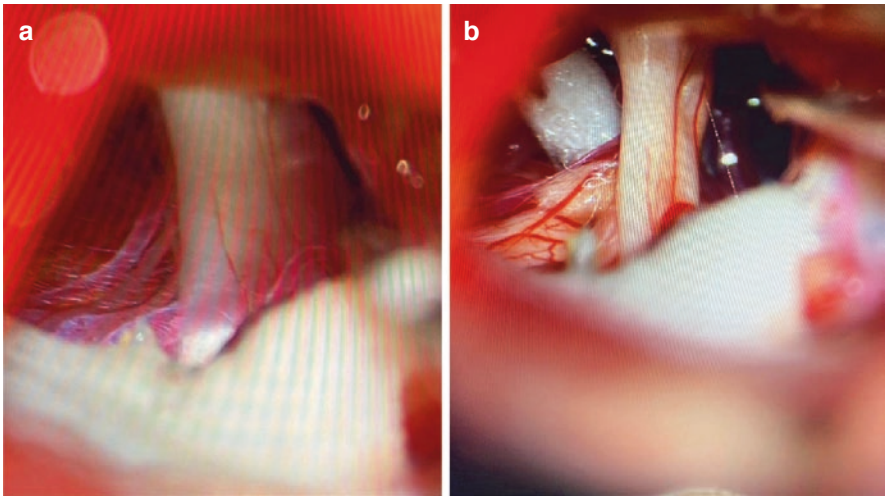

Fig. 10.1 This is a 62-year-old man with right-sided trigeminal neuralgia that was refractory to medical and conservative measures. Intra-operative view under the surgical microscope showing the loop of the superior cerebellar artery coursing along the pons and compressing and flattening the trigeminal nerve at the root entry zone (**a**). After the artery has been moved away toward the tentorium with Teflon felt, the trigeminal nerve is noted to be completely decompressed (**b**). The patient had immediate and lasting relief from his trigeminal neuralgia pain

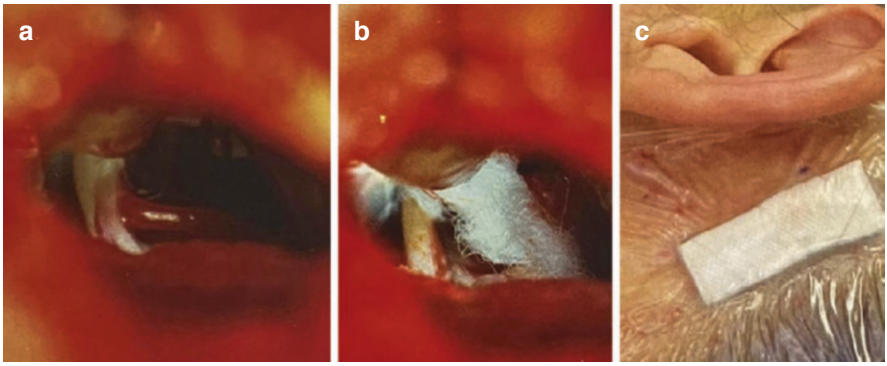

Fig. 10.2 This is a 59-year-old woman with left-sided trigeminal neuralgia who had failed medical and conservative measures. This was the view under microscope of her MVD. At surgery, a loop of the superior cerebellar artery was noted to be significantly compressing the trigeminal nerve root (**a**). The artery was separated from the nerve with micro-dissectors and pushed away with a piece of Teflon felt (**b**). The entire operation was done through an incision about 1.5 inches long (**c**). Postoperatively, the patient's pain was gone and has remained so

For percutaneous rhizotomy, the radiofrequency, glycerol, or balloon techniques can all be used [70, 71], and it is good practice for the surgeon to be prepared and set up to do any of these, depending on the circumstances during surgery (see Fig. 10.3). When the rhizotomy is performed, placement of a small metal marker over the middle of either ear can help confirm that the fluoroscopy is shooting a true lateral skull image. For radiofrequency lesioning, one lesion can be performed at 65–75° centigrade for 50–90 s. The need for higher voltage during testing stimulation suggests the need for higher temperatures for lesion generation. If the pain is only in the V3 distribution, a down curved electrode may be used. Intraoperative assessments of the extent of denervation are often unreliable. Radiofrequency is usually not used to create V1 lesions as this technique has a higher likelihood of causing keratitis. For the glycerol injection, it is preferable to see good CSF flow from the cannula and a good outline of the trigeminal cistern during an omnipaque injection. About 0.25 cc of sterile glycerol is injected with the patient sitting upright, and the patient is kept upright for 1–2 h. For the balloon technique, the balloon is inflated for 60–90 s to a pressure of about 1.5 atm. The glycerol or balloon techniques are preferred if there is a large V1 component to the pain. Rhizotomy benefit is usually noted immediately but can sometimes take up to a few weeks to be fully appreciated.

The diagnosis of trigeminal neuralgia with autonomic features is not clearly different from the diagnosis of Short-lasting Unilateral Neuralgiform headache with Autonomic symptoms (SUNA) and the subcategory of Short-lasting, Unilateral, Neuralgiform headache attacks with Conjunctival injection and Tearing (SUNCT). These may just be variants of the same disease [72, 73]. That said, these must be distinguished from chronic paroxysmal hemicrania (another "trigeminal autonomic cephalgia"), which involves sharp attacks of pain in and around the eye, with autonomic features, and complete response to Indomethacin (with poor response to carbamazepine and other anticonvulsants; see Case 10.2).

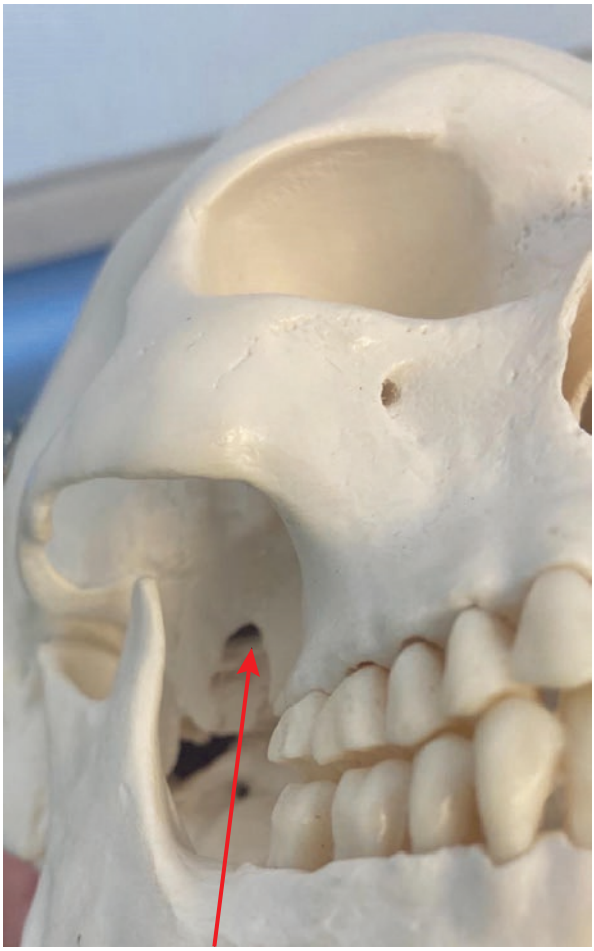

Fig. 10.3 A right-sided skull model with demonstration of access to the foramen ovale for purposes of a percutaneous trigeminal rhizotomy

Case 10.2

This is a 51-year-old woman who presented with 3 years of worsening episodes of severe right facial pain. The pain was in and around the eye. The pain was initially more of a dull pain but was now characterized as primarily sharp intermittent pains. She also had some burning and throbbing pains that were not as bad or bothersome for her. Her pain was described as a stabbing pain that could be triggered by light touch or the wind. The pain could be severe and had brought the patient several times to the emergency room. She referred to her pains as a "raging" of her face. MRI of the brain was unremarkable. She felt she might have gotten slight relief from carbamazepine, but not significant relief of pain. Gabapentin had also not helped with the pain. She noted at times tearing of the right eye and a right nasal drip. She also noted at times

that there was a swelling under the eye itself. The sudden, sharp, severe pains in the trigeminal distribution, the focus of pain around the eye, the presence of autonomic features, the normal MRI, and the lack of response to carbamazepine and gabapentin suggested a diagnosis of chronic paroxysmal hemicrania. The patient was started on Indomethacin 25 mg orally twice a day. Her pain immediately and completely resolved, but the medicine bothered her stomach. The dosage was reduced to 15 mg BID, and she had excellent pain control with no side effects. Over time, she was able to taper off the indomethacin and restart it when the pain episodes would flare up.

For most patients with tumors as an etiology of the pain, it is usually easier to perform a denervating procedure, if needed, for pain control, and a radiosurgery treatment for the tumor. A denervating procedure is also preferred if the trigeminal nerve is compressed by a large ectatic basilar artery (see Fig. 10.4).

Gamma Knife can also be an effective treatment for trigeminal neuralgia [74]. For Gamma Knife initial treatments, 80 Gy to the 100% isodose line can be used for non-MS patients, and 85 Gy to the 100% isodose line for MS patients, with no more than the 20% isodose line touching the brainstem. For repeat treatments,

Fig. 10.4 This is a 40-year-old man with classic left-sided trigeminal neuralgia. His pain was controlled on medicines. Brain imaging showed compression of the trigeminal nerve and left anterior brainstem by a very large ectatic basilar artery (T2 axial MRI image). If medical management were to fail, an MVD would not be a good choice here

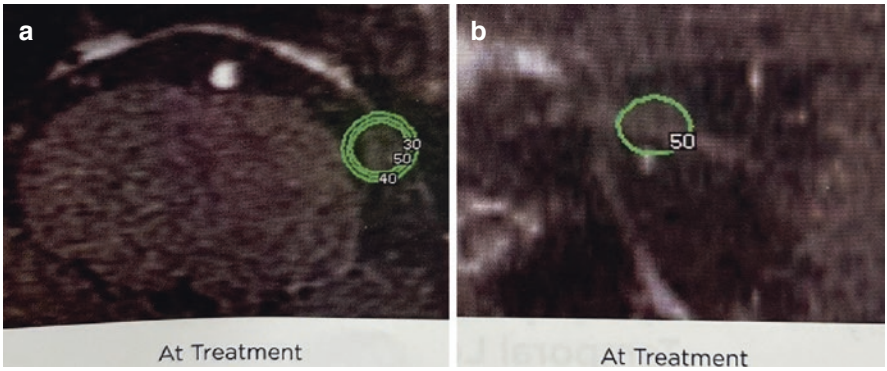

Fig. 10.5 This is a 60-year-old man with medically refractory trigeminal neuralgia in a left V2 distribution. He had an unsuccessful percutaneous rhizotomy elsewhere. He had a history of coronary artery disease and a triple bypass. Gamma Knife was performed (**a**: postcontrast T1 axial image during Gamma Knife treatment; **b**: postcontrast T1 sagittal reconstructed image during Gamma Knife treatment). Fourteen years later, he remains pain free, with no numbness, and requiring no medicines. Note, that my current treatments involve targeting the nerve a bit farther from the brainstem

appropriate doses range anywhere from 40 to 75 Gy to the 100% isodose line depending on the circumstances including time from the last Gamma Knife treatment and the degree of current facial numbness (see Fig. 10.5).

While either denervating procedure (rhizotomy or Gamma Knife) can be repeated, caution should be used in the timing of repeat Gamma Knife procedures as the full effect of these procedures can take quite a while to manifest itself. As such, it is ideal to wait at least 2 years between Gamma Knife trigeminal nerve treatments, including 2 years between treatment of the trigeminal nerve itself and an adjacent tumor (and vice versa).

It is critical not to underestimate the potential bothersomeness of excessive trigeminal denervation. Facial numbness, dysesthesias, and achy pains can be very bothersome to patients (as can medicines, for that matter). Furthermore, numbness of the eye can lead to keratitis and even blindness and should be treated with frequent use of eye drops and regular visits with an ophthalmologist. Ultimately, the real issue is whether the denervation effects are perceived as a problem for the individual patient. While these effects often do lessen over time, it can often take a year or more for such an improvement to occur. As such, it is better to err on the side of creating too little denervation than too much, as further denervation can always be performed at a later time.

Glossopharyngeal Neuralgia

This rare condition has similar features to trigeminal neuralgia but involves the ninth cranial nerve. These patients may experience sudden, brief, severe, sharp pains in their deep ear or throat, triggered by light touch, and relieved with carbamazepine. Like trigeminal neuralgia, this disease is thought to usually be caused by compression of a blood vessel against the ninth cranial nerve. For younger patients who have failed medical management, an MVD is preferred [75], and for older patients, or patients with significant medical co-morbidities, a Gamma Knife procedure can be performed. If, at the time of MVD, no vessel is seen against the ninth nerve, consideration can be given to injuring the ninth nerve slightly with a massaging or combing technique. While cutting the ninth nerve (and possibly the upper one or two branches of the tenth nerve) is considered acceptable in these situations, such action should be taken only with great hesitancy as this can cause significant side effects that are bothersome to the patient, including dysesthesias and deafferentation pain (see Fig. 10.6).

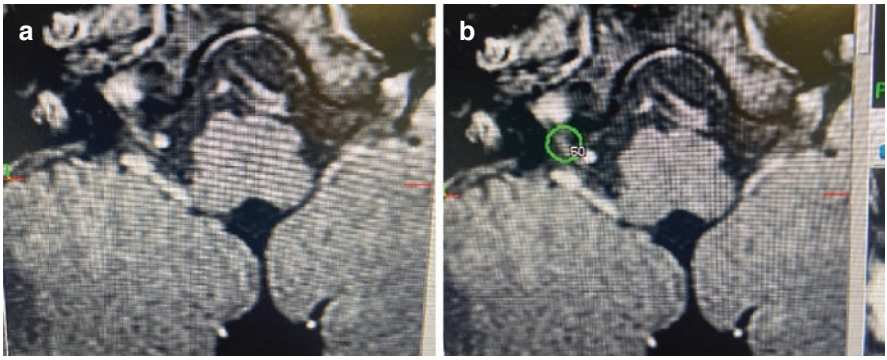

Fig. 10.6 This is a 22-year-old man with autism who is nonverbal. For the past year, he had experienced what seemed to be sudden severe episodes of pain in the right ear that lasted a few minutes and then would go away. Ear exam was normal. The pain was initially better on gabapentin, but he now had to increase his dose 3600 mg/day and was still having episodes of pain. He also still had pain episodes when carbamazepine was added. MRI showed a medium-sized artery contacting the right ninth cranial nerve. Because it was not possible for the patient to confirm the exact nature of his symptoms, it was decided to try a Gamma Knife treatment for presumed right glossopharyngeal neuralgia. Several weeks after Gamma Knife treatment, the patient had no more episodes of pain. (**a**) MRI axial postcontrast image at time of Gamma Knife treatment, showing a medium-sized artery contacting the right ninth cranial nerve; (**b**) MRI axial postcontrast image at the time of Gamma Knife treatment with targeting of the ninth nerve

Occipital Neuralgia

This is a condition in which people get sudden, brief, sharp, intermittent, severe pains on one side of the occipital region in the distribution of the occipital nerve. There can also be some constant and some achy component to these pains. The pains may seem to radiate and can be triggered by light touch in the occipital region. Occipital neuralgia can be caused by disease of the occipital nerve, including compressive masses or trauma. Often the cause is not known. Carbamazepine and gabapentin can help relieve these pains. Occipital nerve blocks can also be performed. For refractory cases, occipital nerve decompression in the posterior scalp can be considered. If other treatments are not successful, a trial of a peripheral occipital stimulator can be considered (with permanent internalization if the trial is successful).

Pain Procedures for Other Cranio-facial Pains

The procedures that currently seem to have the most potential are implantation of percutaneous peripheral nerve stimulators in the distribution of either the trigeminal nerve [76] or the occipital nerve [77]. These patients first undergo a trial implant and then permanent implant if the trial is successful. V3 distribution stimulators carry the extra challenge of potential lead migration due to movement of the mandible. These procedures are generally very low risk.