Idiopathic Trigeminal Neuralgia in the Healthy Patient

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1

Case Presentation

A 46-year-old woman presents to the clinic with a 10-year history of pain in the left side of the face. The pain is both in the forehead and the cheek. It is described by the patient as lancinating and like an electrical jolt. It occurs spontaneously, although it may also be triggered by wiping of the nose, touch, wind, and the brushing of teeth. The pain is episodic, and the patient is pain-free between episodes. The patient describes the initial onset of this pain 5 years prior as spontaneous and without inciting event. The initial occurrence of pain lasted only a week and was followed by a months' long remission. Since then, the patient has had several recurrences of her pain, each remitting several weeks later. She does state that the painful episodes have become longer and more intense with time, and that the remissions have not been completely pain-free more recently. After ineffective attempts to treat her condition with mild narcotic pain medications, she started taking Tegretol recently. This treated her pain effectively, but she has not been able to tolerate this medication due to granulocytopenia. Attempted treatment with baclofen and Neurontin has not been effective. Detailed neurological examination, including a full cranial nerve examination, is completely normal. Her eye and face appear normal, with a normal oral examination as well.

Questions

1. What is the likely diagnosis?
2. What diagnoses are important to exclude?
3. What is the most appropriate imaging modality?
4. What is the appropriate timing of further workup?

Assessment and Planning

The neurosurgeon suspects the diagnosis of trigeminal neuralgia. The lancinating nature of the pain, its trigeminal distribution, the presence of spontaneous remissions and recurrences, and the response to Tegretol are all hallmarks of this condition. It is important to note that trigeminal neuralgia is a rather uncommon cause of facial pain, accounting for approximately 10% of cases. It is therefore important to exclude other potential causes of trigeminal-distribution pain. These include such conditions as post-traumatic trigeminal neuropathy (such as post-dental procedure pain), atypical facial

pain (in which pain frequently goes outside the trigeminal distribution), and trigeminal autonomic cephalgia (a condition related to cluster headache that is accompanied by conjunctival injection, tearing, and rhinorrhea). Usually, the description of the pain or the history of its evolution is of significant value in excluding these trigeminal neuralgia look-alikes.

Since the diagnosis of trigeminal neuralgia is a clinical one, based upon the history and examination, the role of imaging is mainly in excluding conditions that cause so-called secondary or provoked trigeminal neuralgia. Whereas idiopathic trigeminal neuralgia has, by definition, no overt cause, the secondary trigeminal neuralgias are usually related either to mass lesions around the trigeminal nerve (such as tumors in the cerebellopontine angle) or demyelination of the central nervous system (such as multiple sclerosis). In addition to excluding the presence of inciting pathology, brain magnetic resonance imaging (MRI) may also allow the neurosurgeon to get detailed images of the root entry zone of the trigeminal nerve. These images are usually obtained in specialized sequences that accentuate the signal differences between cerebral spinal fluid (CSF), nerves, and blood vessels, allowing the neurosurgeon to appreciate the presence of a vascular loop causing compression of the trigeminal nerve at the root entry zone. The addition of time-of-flight source images of a magnetic resonance angiogram (MRA) can provide further anatomical information on the nature of the neurovascular contact.

Oral Boards Review—Diagnostic Pearls

1. Trigeminal neuralgia is a clinical diagnosis, based on the description of the pain, its distribution, and the history of its evolution. It is not a radiographic diagnosis, and the use of MRI is strictly adjunct in the assessment and planning of treatment.
2. The description of trigeminal neuralgia pain by patients is incredibly stereotyped. Deviation from description of lancinating, episodic pain should be regarded as a "red flag" that trigeminal neuralgia may not be the correct diagnosis.
3. The physical examination of the patient with untreated trigeminal neuralgia is invariably normal. Any trigeminal deficits (e.g., numbness, jaw weakness) should raise concern for lesional or secondary pain conditions.
4. Although the initial bouts of trigeminal neuralgia give way to pain-free remissions, with time, these remissions may not be strictly pain-free. Many patients develop a constant, frequently burning, component of facial pain that may persist after the lancinating pain has resolved. Such constant pain has been termed trigeminal neuralgia, type 2 (Type 2 TN) in counter-distinction to the lancinating pain, known as trigeminal neuralgia, type 1 (Type 1 TN). Many patients eventually come to have both Type 1 TN and Type 2 TN symptoms.

In the present case, the patient had classic features of Type 1 TN, with a more recent minor contribution of Type 2 TN symptoms. Her examination was consistent with a diagnosis of trigeminal neuralgia. A prior routine brain MRI excluded any

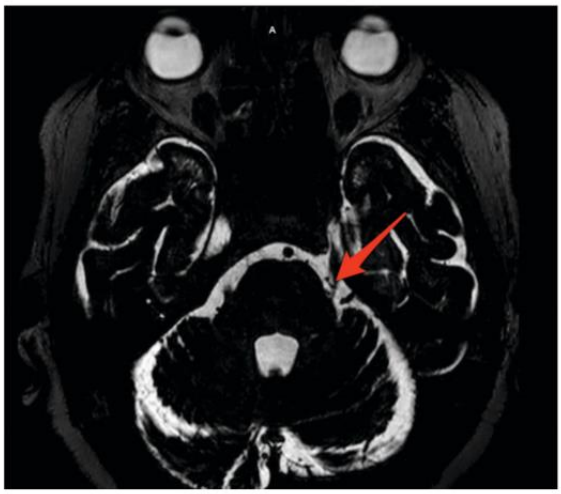

Figure 1.1. Thin-cut T2 MRI FIESTA sequence image demonstrating the typical appearance of neurovascular contact between a branch of the superior cerebellar artery and the trigeminal nerve entry zone on the left (arrow).

mass lesions around the trigeminal nerve, and the diagnosis of demyelinating disease was also discarded due to the lack of concordant history or any suggestive findings on her brain MRI. A high-definition MRI FIESTA (Fast Imaging Employing Steady-state Acquisition; termed CISS [Constructive Interference Steady State] on Siemens scanners) sequence showed a vascular loop making contact with the *left* trigeminal nerve at the root entry zone (Figure 1.1). This was confirmed by her MRA source images (Figure 1.2). Incidental note was made of a vascular loop making contact with the *right* trigeminal nerve as well.

Questions

1. How do these clinical and radiological findings influence surgical planning?
2. What is the significance of neurovascular contact contralateral to the patient's symptoms?
3. How would the absence of a neurovascular contact on the symptomatic side influence plans for surgery?

Decision Making

A history of unilateral, lancinating facial pain, with spontaneous remissions and recurrences and a response to Tegretol, is virtually diagnostic of trigeminal neuralgia. The absence of mass lesions along the trigeminal nerve means that the timing of surgical intervention would be strictly at the choice of the patient. If medical treatment is effective and well-tolerated, there is no reason to offer surgery to relieve symptoms, regardless of the presence of neurovascular compression. If the patient cannot tolerate

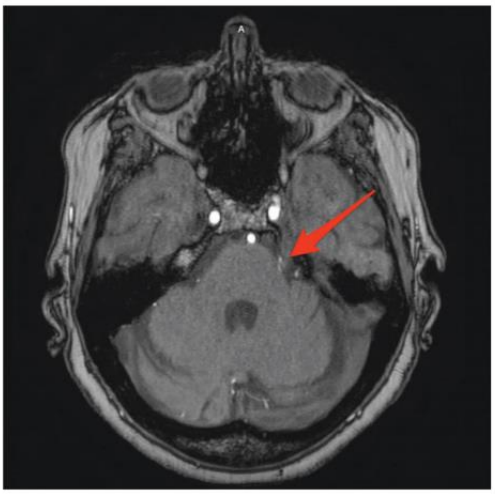

Figure 1.2. MR Angiography source image of the same region seen in Figure 1, demonstrating the vascular nature of the compressing structure at the root entry zone of the trigeminal nerve (arrow).

the medical treatment, or if medical treatment does not adequately control symptoms, however, surgery is indicated.

The choice of surgery for the treatment of idiopathic trigeminal neuralgia is complex and dependent on the surgeon's expertise and patient's preference. In general, however, patients who are young and healthy should be considered first for microvascular decompression (MVD) surgery, since it offers the most robust long-term results with the least risk of complications related to deafferentation. Other procedures for trigeminal neuralgia (outlined elsewhere in this book) all injure the trigeminal nerve to some extent and are subject to the risks related to such intentional nerve injury. In addition to age and health status, other factors that influence the choice of surgical procedure include the distribution of pain. Patients such as the one presented here, who have pain involving the ophthalmic (V1) division of the trigeminal nerve, are poor candidates for nerve-lesioning procedures that may put them at risk of corneal anesthesia, corneal exposure, and keratitis.

Questions

1. How does the absence of a convincing neurovascular contact on MRI influence MVD surgery?
2. What is the significance of neurovascular contact by a surface vein (rather than an artery)?

Surgical Procedure

MVD is a major procedure carried out under general anesthetic with a Foley catheter in place, IV access, and invasive arterial blood pressure monitoring. Brainstem auditory

evoked responses (BAERs) are routinely used, as excessive retraction on the cerebellum to visualize the trigeminal nerve may put hearing at risk. Facial nerve monitoring and trigeminal nerve monitoring may also be performed.

Patients are positioned either semilateral or lateral, with the head in Mayfield pins turned to expose the retroauricular region. The head is usually tilted downward in order to improve the surgical angle to the trigeminal nerve. The ipsilateral shoulder is frequently taped down to improve the angle of visualization. The incision used may be "C" shaped, linear, or "S" shaped, but it should always span the asterion so that the angle of the transverse sinus and sigmoid sinus can be defined in the bony opening. This angle must be developed to allow the surgeon access to the upper portion of the cerebellopontine angle. Once the scalp is opened, the surgeon can perform either a craniotomy or a craniectomy, based on the sinus angle. The edges of the transverse and sigmoid sinuses should be seen after the bony removal is complete. The size of the cranial opening is variable, but it is usually around 3 cm in diameter. The mastoid air cells should be waxed. The dura is opened and CSF is allowed to drain, until the cerebellum relaxes and CSF spontaneously drains from the opening. At this point, the surgeon gently retracts the upper portion of the lateral cerebellar hemisphere, proceeding under high magnification along the junction of the tentorium and petrous face, until the petrosal vein comes into view. This vein, emanating from the superior petrosal sinus, is frequently sectioned, as it commonly obscures the trigeminal nerve deep to it. If it is possible to preserve all or part of this venous complex, the surgeon may choose to do so, but never at the expense of access to the trigeminal nerve. The trigeminal nerve is usually seen as a thick white band, surrounded by arachnoid membrane. It runs close to, but significantly deeper than, the facial/vestibular/cochlear complex that is frequently encountered first during the surgical approach. After freeing up the arachnoid adhesions around the trigeminal nerve, special attention is paid to the vessel abutting the nerve. If this vessel is an artery, it can be manipulated and mobilized away from the nerve, and a piece of Teflon felt can be interposed between them. If the compressive vessel proves to be a vein, it may not be possible to mobilize it. In such cases, the vein is coagulated and divided, in order to release any compression from the nerve.

Following the completion of the decompression, the dura is closed. The bone flap may be replaced or a bone substitute used, but this is not a necessity. The incision is closed tightly in layers to reduce the risk of CSF leak.

Oral Boards Review—Management Pearls

1. It is important to position the head with adequate turning so as to expose the retroauricular region and with a downward tilt of the head to improve the surgical angle.
2. BAER monitoring is useful to reduce the risk of hearing loss with surgery, as cerebellar retraction and manipulation in the vicinity of the trigeminal nerve may cause unrecognized stretching of the nearby cochlear fibers.
3. Sacrifice of the petrosal vein may be necessary in order to provide adequate access to the trigeminal nerve.
4. The trigeminal nerve is deeper and larger than the VII/VIII complex.

5. Infrequently, there is a tubercle of the petrous face directly over the trigeminal nerve that may partially obscure it from view. It is important to work around this visual obstruction and to resist temptation to drill it down, as it is usually the representation of an air cell in the temporal bone.
6. It is important to thoroughly inspect the trigeminal nerve before placing any interposing Teflon.
7. Arterial compression may occur anywhere along the trigeminal nerve, although it is most commonly seen in its rostral/ventral aspect.

Pivot Points

1. If a healthy patient presents with classic features of idiopathic Type 1 TN, that person should be considered first for a MVD.
2. Imaging does not play a role in the diagnosis of trigeminal neuralgia, except to exclude the possibility of secondary, or provoked, trigeminal neuralgia.
3. FIESTA (or CISS) MRA sequences on MRI inform the surgeon about the anatomy of the neurovascular compression, helping direct the surgeon's attention at the time of surgery.
4. BAER monitoring during surgery is an important safeguard against excessive retraction.

Aftercare

Patients are usually observed in an intensive care unit setting on the first night after surgery. They are mobilized as soon as possible, and are usually allowed to leave the hospital in short order. A short course of steroids is often prescribed to lessen the incidence of aseptic meningitis after surgery. Routine imaging is not necessary. Small pseudomeningoceles are not uncommon, and they usually resolve spontaneously.

Complications and Management

The most frequent complication of surgery is aseptic meningitis, probably related to blood in the CSF or the presence of Teflon. Symptoms consist of headache, neck stiffness, and low-grade fever. The usual treatment of these symptoms consists of a short burst of steroids. Of course, on the differential for such symptoms is septic meningitis. If the patient presents with such symptoms but appears more ill than expected, or does not respond to steroids, a lumbar puncture is indicated to rule out infection.

MVD may also be complicated by CSF rhinorrhea, usually related to CSF egress from the dural opening through a mastoid air cell into the middle ear and through the Eustachian tubes into the nasopharynx. When present, CSF rhinorrhea is relatively straightforward to manage, given the superficial nature of the CSF egress pathway. A lumbar drain may be placed temporarily to allow the hole in the mastoid air cell to seal, but if this does not resolve the issue, a superficial re-exploration, with resealing of the air cells, will stop the leak.

Less frequent complications include trigeminal sensory loss, hearing loss, facial paralysis, or cerebellar infarction. Despite the manipulation of the trigeminal nerve inherent in this surgery, appreciable sensory loss does not typically occur after MVD surgery unless the manipulation is significant. Hearing loss, on the other hand, may occur with far less manipulation or retraction, especially in older patients. BAER monitoring is helpful to reduce the risk of this complication. If waveforms diminish during the surgical approach, it is advisable to temporarily cease manipulation and retraction and allow waveforms to recover before continuing. Facial paralysis is very unusual, and it is usually a sign of excessive retraction. It is frequently accompanied by significant hearing loss. Cerebellar stroke is usually a sign of significant cerebellar retraction, or suboptimal positioning at the start of surgery. Some authors claim that sacrifice of the petrosal vein predisposes to cerebellar infarction, but this claim is controversial, with a recent study that specifically examined this issue that reported no increase in risk of cerebellar stroke.

Oral Boards Review—Complications Pearls

1. Careful patient positioning is essential to improve visualization and reduce the need for cerebellar retraction during surgery, thus reducing the risk of the most serious complications of the operation.
2. The most common complications of MVD surgery, aseptic meningitis and CSF rhinorrhea, are relatively straightforward to manage.
3. BAER monitoring allows the surgeon to reduce tension on the facial and cochlear nerves, reducing the risk of cranial neuropathies as a result of surgery.

Evidence and Outcomes

Numerous retrospective studies have documented the results of MVD surgery in the treatment of trigeminal neuralgia, although there are no prospective studies comparing its efficacy to other modalities of treatment. MVD is expected to provide relief of trigeminal neuralgia pain in 95–99% of patients immediately after surgery, but like other surgical approaches to this disease, longer-term outcomes demonstrate a time-dependent recurrence rate that is not insignificant. In the largest published trial of MVD surgery, over a long follow-up period, Dr. Jannetta's group published a 70% pain-free outcome at 10 years. This is a significantly better long-term result than other surgical approaches to trigeminal neuralgia, such as percutaneous lesioning procedures and stereotactic radiosurgery. These favorable long-term outcomes come at a price, however, as this operation does have a higher major morbidity and mortality rate than the other, less invasive options.

Further Reading

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Idiopathic Trigeminal Neuralgia in the Elderly

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2

Case Presentation

An 86-year-old man with a history of a cardiac pacemaker placed for atrial fibrillation presents to clinic with a 5-year history of right-sided, episodic, lancinating facial pain in the maxillary (V2) distribution, consistent with trigeminal neuralgia (TN). He has used carbamazepine with good pain relief for years; however, his pain broke through and he was started on oxcarbazepine and then lamotrigine. His partner has noticed worsening memory since starting the additional medications, and his labs have demonstrated hyponatremia. In addition to his arrhythmia, his medical history is significant for hypertension, hyperlipidemia, prostate adenocarcinoma without evidence of metastases, and poorly compensated congestive heart failure. On detailed neurologic examination, he has trouble remembering objects, but cranial nerve and strength exams are normal. MRI demonstrates a segment of the right superior cerebellar artery causing compression of the trigeminal nerve at the root-entry zone.

Questions

1. What medications are commonly used to treat type 1 TN?
2. What are the common side effects of those medications?

Treatment Options—Medical

Treatment of TN —both in the young and the elderly—includes medical and surgical approaches. Type 1 TN is typically treated with anticonvulsant medications, including carbamazepine (CBZ), oxcarbazepine (OXC), lamotrigine, gabapentin, and baclofen, with varying success. The American Academy of Neurology (AAN) and European Federation of Neurological Societies (EFNS) recommend both CBZ and OXC as first-line drugs for treatment of TN.[1] Patients unresponsive to these first-line medications can try an additional medication—including lamotrigine, gabapentin, and baclofen, to name a few—or should be referred for possible surgical intervention. Although the common side effects—memory loss, leucopenia, hyponatremia, and dermatologic reactions—can occur in all patients, they are especially concerning in the elderly, given cross-reactivity with other medications via the liver's cytochrome pathway. In particular, the elderly can be sensitive to CBZ because older patients have lower levels of non-glycated albumin, a major ligand