Trigeminal Neuralgia (2008)

Steven J. Scrivani, D.D.S, D.Med.Sc.

Associate Professor of Dentistry
Tufts School of Dental Medicine
The Craniofacial Pain and Headache Center
Tufts University
Boston, Massachusetts

Introduction

Trigeminal neuralgia (TN) is a well-recognized, relatively unique disorder characterized by sudden, unilateral lancinating attacks of severe, electric-like facial pain. Between attacks of intense momentary pain, most TN patients are paradoxically symptom-free and have a normal clinical examination. A review of history of TN illustrates the devastating impact that the disease has on afflicted individuals and upon the history and evolution of thoughts to its causes and medical and surgical treatments.

TN has an estimated incidence of 4/100,000, it is most common in patients over age 50, on the right side of the face, and women are affected slightly more often than men. Several lines of research support the view that trigeminal neuralgia results from a chronic partial nerve injury between the Gasserian ganglion and the proximal trigeminal nerve root. However, despite recent advances in our understanding of trigeminal disorders, the basic pathophysiology of TN remains largely unknown. Clinical Classification

Trigeminal neuralgia is a clinical syndrome; there's no laboratory test or radiographic study that can reliably confirm the diagnosis. Instead, the diagnosis of TN rests on a series of signs and symptoms that uniquely define the disorder, and the clinician's ability to recognize that specific diagnostic pattern.

Although the major features of trigeminal neuralgia were known for centuries, White and Sweet made a significant contribution by articulating diagnostic criteria for TN that were both precise and succinct. Their criteria rapidly gained popular clinical acceptance, and are currently used by clinicians worldwide. White and Sweet emphasized five major clinical features that, in their opinion, established the diagnosis of TN.

In trigeminal neuralgia:

The pain is paroxysmal.
The pain may be provoked by light touch to the face (trigger zones).
The pain is confined to the trigeminal distribution.
The pain is unilateral.
The clinical sensory examination is normal.

Under the current IHS criteria, the International Classification of Headache Disorders II (ICHD II), clinical cases of TN fall into two groups: "classical" (primary) and "symptomatic" (secondary) forms of TN. "Classical TN" refers to the "idiopathic" syndrome, meaning that the underlying cause of the disorder is not known. Over 90% of TN patients fall in this category. "Symptomatic TN" identifies a group of patients that exhibit the clinical syndrome of TN as a symptom of another disease process. The most common disorders associated with symptomatic TN are multiple sclerosis and benign tumors of the Gasserian ganglion, trigeminal root or cerebellopontine angle.

Diagnostic Evaluation

The diagnosis of TN is based primarily on a clinical history consistent with the diagnostic criteria. However, the general physical and neurological examinations are also very important. Imaging may prove valuable with a clinical presentation that is not consistent with the classic diagnostic criteria.

The physical examination entails a thorough evaluation of the head and neck with special emphasis on the neurological examination. Cranial nerve examination should be performed with special attention to hearing abnormalities and facial nerve abnormalities. In addition, clinical neurosensory testing of the trigeminal system should

include light touch, sharp touch, temperature, direction, and two-point discrimination. Note should be taken of any trigger areas and they should be appropriately mapped out. Aside from the trigger areas when present, and minimal hypoalgesia or hypoesthesia in some patients, the neurological examination is essentially normal.

Management

The current treatment of TN consists of medical and surgical therapies. Medical management consists of pharmacologic and non-pharmacologic approaches, while surgical management consists of numerous peripheral and intracranial procedures.

It is also important to emphasize that the current therapy suffers from a lack of rigorous clinical trials that might provide a scientific foundation for important clinical treatment decisions.

Mathews and Scrivani have developed an algorithm for the differential diagnosis and management of craniofacial pain disorders (Fig. 1). This algorithm incorporates historical data, physical examination data and some diagnostic testing to guide medical and surgical management strategies for patients with craniofacial pain.

Pharmacological Therapy

The current first line treatment is medical therapy with the anticonvulsant/antiepileptic drugs (AEDs) such as: phenytoin (Dilantin), carbamazepine (Tegretol), baclofen (Lioresal), clonazepam (Klonopin) gabapentin (Neurontin), lamotrigine (Lamictal), topiramate (Topamax), oxcarbazepine (Trileptal), tiagabine (Gabatril), levetiracetam (Keppra), or zonisamide (Zonegran), in single or combination regimens.

Some of the AEDs have been carefully evaluated specifically for TN while others have been evaluated for generalized neuropathic pain disorders. Specifically for TN, carbamazepine, clonazepam, baclofen, lamotirgine, tizanidine and torprimate have been studies in randomized, placebo-controlled trials, while phenytoin, clonazepam, valroic acid, gabapentin, mexilitine, lamotigine and oxcarbazepine have been evaluated in uncontrolled, open-label or case series trials.

Pharmacological therapy is effective for many patients, however for some; these medications do not relieve the pain and/or produce intolerable side effects with significant medical and functional morbidity. If medical therapy is unsuccessful or not tolerated, surgical treatment should be considered [see Figure 1 below].

Surgical Therapy

There are numerous surgical treatments for TN, yet currently several treatments are most commonly utilized. The surgical procedures currently commonly used for treatment of TN include:

1. Percutaneous stereotactic differential radiofrequency thermal rhizotomy (RTR)
2. Posterior fossa exploration and microvascular decompression of the trigeminal root (MVD)
3. Stereotactic radiosurgery – gamma knife radiosurgery or cyberknife radiosurgery

Treatment of Acute Attacks

Occasionally, patients may present in acute attacks, with frequent spontaneous, or easily triggered, high intensity jolts of pain. In this situation, some form of acute intervention is warranted because the patients functioning is generally severely affected by the pain attacks and if it continues, may alter their ability to be able to properly eat or drink. In such cases, there has been some reported success with local anesthetic trigeminal division nerve blocks typically with a long-acting local anesthetic (bupivicaine).

Occasionally, the application of a topical amide local anesthetic preparation (lidocaine 2-5%, EMLA) may be effective. However, there is little good data that shows that local anesthetic applications are consistently effective in stopping acute attacks and/or eliminating recurrence of pain after the duration of the applied anesthetic. Further, some patients may benefit from intravenous administration of fosphenytoin (Cerebyx), valproic acid (Depacon), or lidocaine. These infusions need to be conducted in a carefully monitored setting with appropriate medical attention and emergency equipment available (outpatient surgical center or in hospital).

Conclusion

While aggressive pharmacological therapy is generally considered “first-line” therapy for TN, the accumulated risk of

multiple pharmacological agents in the medically managed patients may well exceed the risk of complications with a well thought out management protocol, decision-making algorithm and a careful, skilled surgical approach.

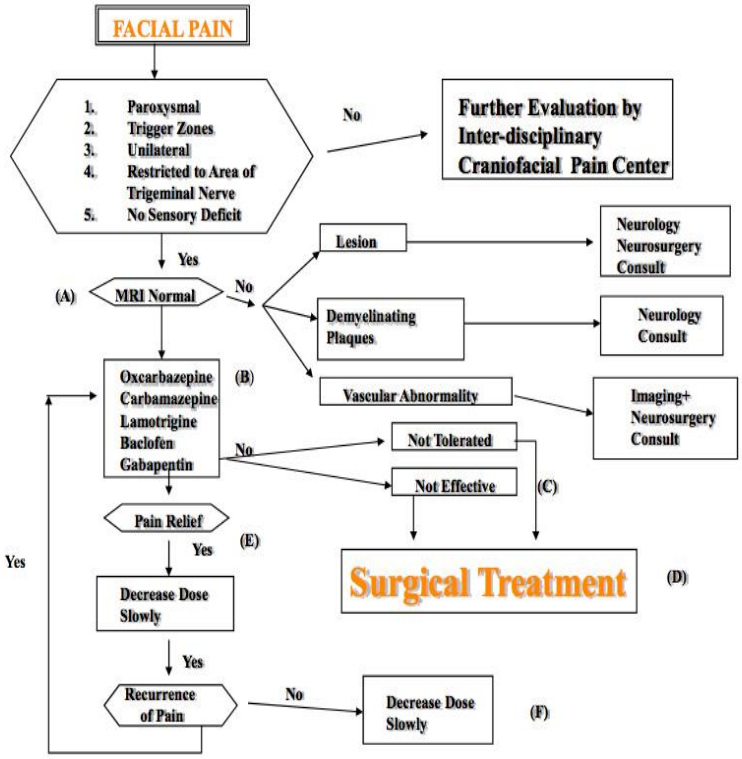

Figure 1

A. MRI of the brain, brainstem and base of skull. Further diagnostic studies are performed when physical examination or MRI findings are abnormal.
B. Medication is titrated progressively until pain is relieved or adverse effects occur. Often a second AED is added early, if single therapy is ineffective.
C. Drug allergy, idiosyncratic reaction, laboratory abnormalities, intolerable dangerous side effects or patient preference.
D. Local anesthetic trigeminal nerve blocks are performed in some patients as part of a further diagnostic evaluation prior to surgery.
E. Doses of medications can be tapered slowly if patient remains free after several weeks to months of pharmacotherapy.
F. Further decrease in dosage of medication is predicted on the patient's pain history. Many patients may need to be maintained on pharmacotherapy as preventive ther,apy.