Trigeminal and Glossopharyngeal Neuralgia

Gaddum Duemani Reddy, MD, PhD, Ashwin Viswanathan, MD*

KEYWORDS

• Trigeminal neuralgia • Glossopharyngeal neuralgia • Medical treatment
• Surgical treatment

KEY POINTS

- Trigeminal neuralgia and glossopharyngeal neuralgia are debilitating forms of paroxysmal facial pain and are diagnosed based on history.
- First-line therapy for both pathologic conditions is medication. Carbamazepine is the drug of choice; however, there are other medical options for patients unable to tolerate the side effects of carbamazepine.
- Surgical therapy with either microvascular decompression and/or an ablative procedure is often successful for medically refractory cases and can be considered early in such cases.
- Radiosurgery is emerging as a potential treatment modality for medically refractory cases and it should be considered in patients who cannot undergo more invasive treatment modalities.

INTRODUCTION

Trigeminal neuralgia (TN) is a clinical condition characterized by agonizing paroxysmal pain occurring in one or more divisions of the trigeminal nerve. The characterization of the disease process we are familiar with today, with afflicted individuals complaining of lancinating pain when chewing, speaking, or swallowing, began in the seventeenth century with the work of John Locke.[1] Since that time, our understanding of the disease process has advanced significantly. Today, TN is a condition for which there are several treatment options. However, it cannot always be cured and a subset of patients remains refractory to multiple forms of treatment.

Glossopharyngeal neuralgia (GPN) is another paroxysmal pain condition that is characterized by pain in the throat, pharynx, and ears. Affected individuals can lose consciousness because of bradycardia or asystole. The treatment strategy parallels

Disclosures: None.
Department of Neurosurgery, Baylor College of Medicine, 1709 Dryden Street, Houston, TX 77030, USA
* Corresponding author.
E-mail address: ashwinv@bcm.edu

Neurol Clin 32 (2014) 539–552
http://dx.doi.org/10.1016/j.ncl.2013.11.008 **neurologic.theclinics.com**
0733-8619/14/$ – see front matter Published by Elsevier Inc.

that for TN; however, given its rarity, less is known about its pathogenesis and the efficacy of various treatment modalities.

TRIGEMINAL NEURALGIA

Epidemiology

The only estimate on the prevalence of TN is from Penman[2] in his 1968 contribution to the *Handbook of Clinical Neurology*, in which he approximated 107.5 per million men and 200.2 per million women are afflicted by this condition. The incidence, however, was more extensively studied with early studies reporting approximately 4.3 new cases per 100,000 people annually.[3] The female to male ratio was estimated in these studies to be roughly 1.5 to 1[4–6] and there is known age dependence, with an annual incidence of 17.5 per 100,000 in individuals aged 60 to 69 years and 25.6 per 100,000 for those older than 70 years.[7]

More recently, European studies have found significantly higher incidence rates for TN, ranging from 12.6 to 27 per 100,000.[8–10] Similar to the older studies, these rates vary significantly with age, with incidences of less than 0.5 per 100,000 in subjects younger than 18 years compared with upwards of 80 per 100,000 subjects in older age groups. The female to male ratios in these newer studies are also significantly higher, at approximately 2.3 to 1.

Recent definitions of TN have separated it into two categories: classic, which is idiopathic in nature, and symptomatic, which is associated with an identifiable structural lesion excluding vascular compression. In cases of symptomatic TN, the condition can be associated with a more generalized demyelinating disease process such as multiple sclerosis.[3] In studies evaluating subjects with multiple sclerosis, TN occurs in approximately 2% of subjects,[11,12] with an increased risk of bilateral symptoms. TN has also been associated with other cranial nerve neuralgias, most notably GPN,[13] in which approximately 11% of subjects have associated TN.[14]

Familial cases of TN are rare but have been reported with estimates of approximately 4% to 5%[3,15] in patients with unilateral TN. Bilateral TN, however, has a higher familial association of approximately 17%,[15] suggesting a stronger hereditary component in this subpopulation. Although the data are sparse, case reports on families with a strong history of TN suggest an autosomal dominant inheritance pattern.[16–19]

Pathogenesis

The cause of TN is unknown. It is suspected, however, that both central and peripheral nerve dysfunction play a role. In the case of the peripheral nerve, it is hypothesized that vascular compression of the trigeminal nerve root at the root entry zone leads to chronic focal demyelination and afferent hyperexcitability. This can lead to hyperexcitability in the trigeminal brainstem complex, which subsequently responds to both nonnoxious and noxious stimuli in the same manner leading to the symptoms seen in TN.[20] However, in what is termed the ignition hypothesis, the injured sensory root itself can become a site of ectopic firing.[21] This in turn leads to some neurons continuously firing, which could be the source of baseline burning pain found in some TN; however, other neurons are silent but respond to momentary stimulation for a prolonged period after discharges.

Notably, several aspects of TN suggest a central role for the central action of effective medications. In addition, gray matter in the anterior cingulated cortex, parahippocampus, and temporal lobe was diminished in correspondence with the duration of the disease in patients afflicted with TN[22] suggesting a mechanism by which the disease process can alter the central nervous system, which, in turn, could prolong the disease.