GLOBAL YEAR AGAINST OROFACIAL PAIN
OCTOBER 2013 – OCTOBER 2014

Trigeminal Neuralgia

Definition

Trigeminal neuralgia (TN) is a unilateral painful disorder that is characterized by brief, electric-shock-like pains, is abrupt in onset and termination, and is limited to the distribution of one or more divisions of the trigeminal nerve. The revised International Classification of Headache Disorders -3 (ICHD-3) suggest three variants: (1) classical trigeminal neuralgia, often caused by microvascular compression at the trigeminal root entry to the brainstem; (2) trigeminal neuralgia with concomitant persistent facial pain; and (3) symptomatic trigeminal neuralgia, caused by a structural lesion other than vascular compression.

Epidemiology

TN is a rare disease, and studies on prevalence are scarce. Analyses of a few of the available studies suggest that the prevalence of TN in the general population might be between 0.01% and 0.3%, although studies carried out in primary care settings suggest that it may be much higher, around 12% per 100,000 persons per year. This higher percentage could be due to misdiagnosis, however. The gender ratio of women to men is approximately 2:1. TN can first appear at any age, but disease onset occurs after the age of 40 years in more than 90% of cases, and the peak age of onset is between the ages of 50 and 60 years. The incidence of TN in multiple sclerosis is higher than in the general population, and this disease is the only comorbidity that has been identified.

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Pathophysiology

Current opinion is that TN is caused by a proximal compression of the trigeminal nerve root close to the brainstem (dorsal root entry zone) by a tortuous blood vessel (an artery or vein), leading to mechanically twisted nerve fibers and secondary demyelination, probably mediated by microvascular ischemic damages. These changes lower the excitability threshold of affected fibers and promote cross-talk between adjacent fibers. Thus, tactile signals coming from the fast myelinated (A-beta) fibers can directly activate the slow nociceptive (A-delta) fibers, and sometimes C fibers, resulting in the high-frequency discharges characteristic of TN. Symptomatic TN can result from tumors (either benign or malignant), multiple sclerosis, or arteriovenous malformations.

Clinical Features

Location, radiation: The pain is unilateral, with only 3% of incidences being bilateral, and there is little radiation outside the trigeminal nerve area. The most commonly affected divisions are the second and third.

Character: Electric-shock-like, shooting, stabbing, or sharp in quality.

Severity: Moderate to severe, but it can be milder at times.

Duration, periodicity: Each attack of pain lasts between a few seconds and 2 minutes but can rapidly be followed by another attack. Between 10 and 70 attacks can occur in a day. There is often a refractory period between attacks. With disease progression, attacks tend to get longer. Spontaneous remission periods can occur, which initially can last for months or years, but over time the remission periods become shorter. In the condition termed “TN with concomitant persistent facial pain,” a prolonged period of burning and aching of lower intensity follows the sharp shooting pain and can last for hours.

Factors affecting it: Light innocuous stimuli to the affected side of the face provoke pain

Associated features: Very rarely are there any autonomic symptoms, and sometimes there may be sensory changes. Anxiety and depression, as well as deterioration of quality of life, are common consequences of the disease and resolve if there is no pain.

Symptomatic TN can present in an identical way to the features described above, and some patients will even have periods of remission.

Investigations

Magnetic resonance imaging (MRI) is the most useful imaging technique to determine the presence of lesions, such as cysts or tumors, vascular malformations, plaques of multiple sclerosis, as well as vascular compression of the trigeminal nerve.

Therapy

Medical treatment of TN is based on the use of antiepileptic drugs. First-line therapy should be carbamazepine (200–1200 mg/day) and oxcarbazepine (600–1800 mg/day), according to current evidence-based treatment guidelines. Second-line treatment is based on little evidence and includes add-on therapy with lamotrigine (400 mg/day) or a switch to lamotrigine or baclofen (40–80 mg/day). Other antiepileptic drugs have been studied in small open-label

studies. Treatment with phenytoin, gabapentin, pregabalin, and valproate is also been suggested to be beneficial. In an emergency, an intravenous infusion of fosphenytoin can be helpful, as well as local injections of lidocaine into trigger points (points from which pain arises).

Surgical Treatment

If medical treatment is not successful or results in marked deterioration in activities of daily living, surgical procedures should be considered. These include microvascular decompression of the nerve/vessel contact or destruction of the Gasserian ganglion. Microvascular decompression provides the most sustained pain relief, with 90% of patients reporting initial pain relief and more than 80% remaining pain free after 1 year, 75% after 3 years, and 73% after 5 years, with sustained improvements in activities of daily living. It is, however, a major surgical procedure that entails a craniotomy to reach the trigeminal nerve in the posterior fossa. The average mortality rate ranges from 0.2% to 0.5%, and up to 4% of patients suffer from major problems such as cerebrospinal fluid leakage, infarcts, hematomas, or aseptic meningitis. The most common long-term complications include mild sensory loss (7%) and hearing loss (10%).

Gasserian ganglion percutaneous techniques are destructive interventions that include radiofrequency thermocoagulation, balloon compression, and percutaneous glycerol rhizolysis. Ninety percent of patients report pain relief following these procedures. One year following radiofrequency thermocoagulation, 68–85% of patients are still pain free, but after 3 years the percentage goes down to 54–64%, and after 5 years only 50% of patients are still pain free. The most common side effects are sensory loss (50%), dysesthesias (6%), anesthesia dolorosa (4%), and corneal numbness with a risk of keratitis (4%). Surgery at the Gasserian ganglion level requires short-acting anesthetics and primarily consists of minor overnight procedures with an extremely low mortality rate.

In gamma knife surgery, a focused beam of radiation is aimed at the trigeminal root in the posterior fossa. One year after gamma knife surgery, 69% of patients were pain free without additional medication. After 3 years, 52% were still pain free. The development of pain relief can be delayed (for an average of 1 month). Side effects include sensory complications in 6% that may develop with a delay of up to 6 months, facial numbness in 9–37%, which improves over time, and paresthesias in 6–13%. Quality of life improves by 88%. The main disadvantage of gamma knife surgery is the cost, which limits its widespread use, and the delayed onset of both pain relief and sensory loss. It remains an option for patients who cannot undergo open surgery or who have blood coagulation problems (e.g., patients who are taking warfarin).

Patients need detailed information if they are to make the most appropriate decisions about the various options that are available. Such information may be obtained through patient support groups, which have websites and often hold national meetings attended by health care professionals.

References

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[2] Zakrzewska JM, Akram H. Neurosurgical interventions for the treatment of classical trigeminal neuralgia. Cochrane Database Syst Rev 2011;9:CD007312.

[3] Zakrzewska JM, Coakham HB. Microvascular decompression for trigeminal neuralgia: update. Curr Opin Neurol 2012 :3:296–301.

Online Resources

Guidelines for trigeminal neuralgia: http://www.aan.com

Patient support groups: http://www.tna.org.uk; http://www.endthepain.org/; http://www.tnaaustralia.org.au