Abstract

Trigeminal neuralgia is a very peculiar disease. The pain of trigeminal neuralgia, also known as “tic douloureux”, is characterized by episodic, paroxysmal, triggered pain in a distribution of 1 or more divisions of the trigeminal nerve. TN pain attacks may result from physiologic changes induced by a chronic partial injury to the brainstem trigeminal nerve root from a variety of causes. An early and accurate diagnosis of TN is important, because therapeutic interventions can reduce or eliminate pain attacks in the large majority of TN patients. (Gupta A, Singh SK, Sahu R. Trigeminal Neuralgia - A review. www.journalofdentofacialsciences.com 2012; 1(1): 27-31)

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Introduction

Trigeminal neuralgia (TN), also called tic douloureux, is the most common of the cranial neuralgias and chiefly affects individuals older than 50 years of age. It is a well recognized disorder characterized by lancinating attacks of severe facial pain. The diagnosis of TN is based primarily on a history of characteristic pain attacks that are consistent with specific, widely accepted research and clinical criteria for the diagnosis.2

TN is classified as either Classic TN when it is not associated with an underlying neurological disease or Symptomatic TN when no neurological disorder can be detected.3 The clinical examination, imaging studies, and laboratory tests are unremarkable in case of classic TN. While the signs and symptoms of TN are secondary to another disease process affecting the trigeminal system in case of symptomatic TN.2 Despite recent advances in our basic understanding of trigeminal pain disorders, the basic pathophysiology of TN remains largely unknown.

Many TN patients suffer pain attacks for months or years before the condition is finally diagnosed.3 This is unfortunate, because the severe pain attacks in TN can have a devastating impact on patients.4,5,6 A timely accurate diagnosis of TN is particularly important, because a variety of specific treatments can greatly reduce—or totally eliminate—TN pain symptoms in most patients.2,8 The dental medicine clinician is often the first to be consulted when patients experience these pain attacks and should be familiar with the syndrome in order to make an accurate diagnosis and initiate treatment.
History, Epidemiology and Demographics

Areataeus of Cappadocia is credited with the first clinical description of TN. At the end of the first century he described a condition (heterocrania) where "spasm and distortion of the countenance take place." Jujani, an 11th century Middle Eastern physician, also discussed a problem of unilateral facial pain with associated spasms and anxiety. He even suggested the pain resulted from "the proximity of the artery to the nerve."

In 1677 John Locke, a noted American physician and philosopher, accurately identified the major clinical features of TN in the Countess of Northumberland. He correctly recognized that her facial pain was not caused by dental pathology but rather by neuralgia of the trigeminal nerve.

In 1756 the French physician Nicolaus Andre recognized the unique nature of the syndrome, commenting that it was "exclusive and distinctive from all other diseases." He gave the name tic douloureux to the condition.

The English physician John Fothergill in 1773 also outlined the major clinical features of TN, clearly establishing the disorder as a discrete syndrome. TN has an incidence of approximately 4/100,000. Although familial examples of TN are reported, the large majority of cases occur spontaneously, and TN does not appear to be more common in any particular ethnic group, geographic region, or climate.

TN occurs in both genders (with a slight female predominance), and the diagnosis is most common over age 50. This age and sex distribution is notable, because most other common chronic headache and facial pain disorders develop at an earlier age and are more frequent in women. For example, migraine occurs predominantly in females (over 71%) and the mean age of onset of symptoms is 30.

The pain in TN typically consists of lancinating paroxysms. Attacks are most common in the second and third trigeminal divisions, and the right side of the face is more often involved than the left. In individual patients, the pain attacks are stereotyped, recurring with the same intensity and in the same distribution.

However, despite the intensity of the pain episodes, most TN patients are symptom free between attacks and most exhibit a normal clinical examination.

TN is a chronic disorder. Although temporary spontaneous remissions often occur over the clinical duration of the illness, most patients will experience episodic TN pain attacks for years unless appropriately treated.

ETIOLOGY AND PATHOGENESIS

The cause of TN pain attacks is not known. However, the fact that benign tumors and vascular anomalies that compress the trigeminal nerve root can produce symptoms clinically indistinguishable from classic TN strongly implies that injury to the nerve root is an important initiating factor in the disease.

Jannetta and colleagues expanded on this concept, showing that vascular compression is a common finding in patients with TN and that surgical decompression of the nerve root often effectively alleviates TN symptoms.

However, it is important to emphasize that TN pain attacks involve more than just an acute nerve injury, because cutting or compressing the trunk of a sensory nerve elicits, at most, only a brief discharge in damaged axons. The pathophysiology of TN probably evolves over days to weeks following injury. During this interval, the damaged sensory neurons go through a cascade of changes as part of the repair process that undoubtedly contributes to the signs and symptoms observed in clinical neurogenic pain disorders, including TN.

Following trauma, regenerating nerve fibers become relatively depolarized and physiologically more excitable.

Spontaneous action potentials originate from multiple sites and single action potentials may evoke sustained afterdischarges.

Focal demyelination at the site of compression may also allow electrical spread of excitation between adjacent sensory axons ("ephaptic" transmission). An ephaptic short-circuit of this type within the trigeminal nerve might explain the sudden "electric" jolts of pain that characterize the disorder.
Consistent with these views, Vos and colleagues demonstrated in rats that mild compression of the trigeminal infraorbital nerve results in behavioral evidence of a pain disorder associated with facial allodynia and hyperalgesia, but without actual sensory loss.

Histologic studies of the compressed nerve reveals focal inflammation, extensive demyelination-remyelination, and “neuroma in continuity.” Devor et al analyzed surgical biopsy specimens from TN patients who had presumed vascular compression. These trigeminal root specimens demonstrated evidence of inflammation, demyelination, and close apposition of axons; the reported changes are similar to those in the Vos rat model of trigeminal neuropathic pain. Love et al also examined trigeminal nerve specimens from 6 MS patients with TN. The specimens again revealed similar changes, even though the etiology of nerve injury in MS is quite different from traumatic compression.

Based on the morphologic and physiologic changes following partial nerve injury, Devor et al proposed an “ignition hypothesis” to explain the principal signs and symptoms in TN. In this model, a trigeminal injury induces physiologic changes that result in a population of hyperexcitable and functionally linked primary sensory neurons. The discharge of any individual neuron in this group can quickly spread to activate the entire population. Such a sudden synchronous discharge could underlie the sudden jolt of pain characteristic of a TN pain attack. This model is attractive, not only because it explains many of the key features of TN, but also because it encourages specific testable hypotheses that should stimulate advances in both basic science and clinical investigation.2

Medical Management and Treatment

(A) Non surgical

In most chronic neurogenic pain disorders, the therapeutic options are limited and therapeutic responses are only partial. TN is unique, because the large majority of TN patients respond to treatment and many have total elimination of pain attacks lasting months or years.2

Initial therapy for TN should consist of trials of drugs that are effective in eliminating the painful attacks. Anticonvulsant drugs are most frequently used and are most effective. Carbamazepine is the most commonly used drug and is an effective therapy for greater than 85% of newly diagnosed cases of TN. The drug is administered in slowly increasing doses until pain relief has been achieved. Skin reactions, including generalized erythema multiforme, are serious side effects. Patients receiving carbamazepine must have periodic hematologic laboratory evaluations because serious life-threatening blood dyscrasias occur in rare cases. Monitoring of hepatic and renal function is also recommended.

Patients who do not respond to carbamazepine alone may obtain relief from baclofen or by combining carbamazepine with baclofen.

Gabapentin, a newer anticonvulsant that has fewer serious side effects than carbamazepine, is effective in some patients but does not appear to be as reliable as carbamazepine.

Other drugs that are effective for some patients include phenytoin, lamotrigine, and pimozide.
Since TN may have temporary or permanent spontaneous remissions, drug therapy should be slowly withdrawn if a patient remains pain free for 3 months. Clinicians treating TN must be aware that drug therapy often becomes less effective over time and that progressively higher doses may be required for pain control.

(B) Surgical

In cases in which drug therapy is ineffective or in which the patient is unable to tolerate the side effects of drugs after trials of several agents, surgical therapy is indicated. A number of surgical procedures that result in temporary or permanent remission of the painful attacks have been described. These include procedures performed on the peripheral portion of the nerve, where it exits the jaw; at the gasserian ganglion; and on the brainstem, at the posterior cranial fossa.

Peripheral surgery includes cryosurgery on the trigeminal nerve branch that triggers the painful attacks. This procedure is most frequently performed at the mental nerve for cases involving the third division and at the infraorbital nerve for cases involving the second division.

The potential effectiveness of this procedure can be evaluated prior to surgery by determining whether a long-acting local anesthetic eliminates the pain during the duration of anesthesia. This procedure is usually effective for 12 to 18 months, at which time it must be repeated or another form of therapy must be instituted.

The most commonly performed procedure at the level of the gasserian ganglion is percutaneous radiofrequency thermocoagulation although some clinicians continue to advocate glycerol block at the ganglion or compression of the ganglion by balloon microcompression. An infrequent but severe surgical complication is anesthesia dolorosa, which is numbness combined with severe intractable pain.

The most extensively studied surgical procedure is microvascular decompression of the nerve root at the brainstem. In a report of 1,185 patients who were observed for 1 to 6 years, 70% of the patients experienced long-term relief of symptoms. It should be noted that 30% of the patients experienced a recurrence of symptoms and required a second procedure or alternative therapy. Complications were rare but included stroke, facial numbness, and facial weakness.

In summary, therapy for TN presently includes a variety of both medical and surgical approaches, each of which is effective for some patients. Drug therapy including trials of several drugs or combinations of drugs should be attempted before surgery is recommended. When surgery is necessary, the patient should be carefully counseled regarding the advantages and disadvantages of the available surgical procedures.

Clinicians should also remember that since spontaneous remissions are a feature of TN, procedures resulting in temporary relief might be all that is necessary for some patients.

References

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