Global Year Against HEADACHE
Oct 2011 - Oct 2012

Trigeminal Neuralgia and Persistent Idiopathic Facial Pain

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 International Headache Society (IHS) differentiates between classical trigeminal neuralgia, often caused by microvascular compression at the trigeminal root entry to the brainstem, and symptomatic trigeminal neuralgia, caused by a structural lesion other than vascular compression.

Persistent idiopathic facial pain (PIFP), previously termed atypical facial pain, is a persistent facial pain that does not have the characteristics of cranial neuralgias and cannot be attributed to a different disorder. The facial pain occurs daily and persists throughout the day. Generally, it is limited to one particular area on one side of the face at disease onset, is deep and poorly localized, and is not associated with sensory loss or other neurological deficits. Investigations including X-ray of the face and jaws or cranial computed tomography (CT) or magnetic resonance imaging (MRI) do not demonstrate any relevant abnormality. The pain may be initiated by surgery or injury to the face, teeth, or gums, but it persists without any demonstrable local cause.

Epidemiology

TN and PIFP are rare diseases, and studies on their prevalences 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%. 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 over 90% of cases. The peak age of onset is between the ages of 50 and 60 years. Anxiety and depression, as well as deterioration of quality of life, are common consequences of the disease. Other cranial neuralgias and PIFP are far less frequent than TN. Data on their prevalence in the general population are not available.

Pathophysiology

Current opinion is that TN is caused by a proximal compression of the trigeminal nerve root close to the brainstem (root entry zone) by a tortuous or ectasic 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 inappropriate ephaptic propagation toward adjacent fibers. Thus, tactile signals coming from the fast myelinated (A-beta) fibers can directly activate the slow nociceptive (A-delta) fibers, resulting in the high-frequency discharges characteristic of trigeminal neuralgia.

The pathophysiology of PIFP is unknown. The available literature suggests that abnormal sensitization of the trigeminal nociceptive system may play a crucial role in the development of PIFP.

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 very little evidence and includes add-on therapy with lamotrigine (400 mg/day) or a switch to lamotrigine, baclofen (40–80 mg/day), or pimozide (4–12 mg/day). Other antiepileptic drugs have been studied in small open-label studies. Treatment with phenytoin, clonazepam, gabapentin, pregabalin, topiramate, and valproate, as well as tocainide (12 mg/day), has also been suggested as beneficial.

The treatment of choice for PIFP is tricyclic antidepressants such as amitryptiline (50–100 mg/day). Selective serotonin and norepinephrine reuptake inhibitors (duloxetine, venlafaxine, and mirtazapine) are used as well.

Surgical Treatment

If medical treatment is not successful, 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 over 80% remaining pain free after 1 year, 75% after 3 years, and 73% after 5 years. It is, however, a major surgical procedure that entails 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, or hematomas. The most common long-term complications include aseptic meningitis (11%), 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%). Gasserian ganglion therapies require short-acting anesthetics and are primarily 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 improved 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 makes it a last-reserve option for patients who cannot undergo open surgery or who have blood coagulation problems (e.g., patients who are taking warfarin).

References

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