REVIEW ON THE CAUSES OF TRIGEMINAL NEURALGIA SYMPTOMATIC TO OTHER DISEASES

Florin Popovici[1], Athena Mergeani[2], Dan Popescu[2], Florina Antochi[2]

[1]Oral and Maxillofacial Surgery Hospital "Dan Teodorescu", Bucharest, Romania
[2]Neurology Department, Emergency University Hospital, Bucharest, Romania

ABSTRACT

Trigeminal neuralgia, also called tic douloureux, causes extreme, sporadic, sudden burning or shock-like face pain that lasts from few seconds to several minutes and can be physically and mentally incapacitating. Most cases are still referred to as idiopathic, although many are associated with vascular compression of the trigeminal nerve. A minority of cases are symptomatic to other diseases. The aim of this review is to show rare causes of trigeminal neuralgia described so far in the medical literature.

Key words: trigeminal neuralgia, trigeminal nerve, tic douloureux

Trigeminal nerve is the largest of all the cranial nerves. It transmits sensory information from the face, oral and nasal cavities and most of the scalp and caries motor supply to the muscles of mastication. Disease involving nerve or adjacent to it can cause trigeminal neuralgia or loss of sensory or motor function in the distribution of the nerve. The nerve can be divided into four segments: brainstem, cisternal, Meckel's cave and cavernous sinus and extracranial course. Trigeminal neuralgia, also called tic douloureux, causes extreme, sporadic, sudden burning or shock-like face pain that lasts from few seconds to several minutes and can be physically and mentally incapacitating. More than one nerve branch can be affected by the disorder. Most cases are still referred to as idiopathic, although many are associated with vascular compression of the trigeminal nerve. A minority of cases are symptomatic to other diseases. The condition is variable and patients may have just one episode. Most patients respond well to drugs; carbamazepine is usually the first line treatment. If drug treatment fails or is not tolerated, surgical treatments are available. Ablative surgical treatments are associated with facial sensory loss, almost no risk of severe complications or death, and a high rate of pain recurrence; microvascular decompression has a risk of severe complications or death and a lower relapse rate.

Although most cranial neuralgias are probably due to microvascular compression at the root entry zone, other etiologies need to be considered, especially in the young adult population in whom demyelinating disease, aneurysms, neoplasms, and infectious etiologies (post-herpetic, Lyme disease, etc) may be more common.

About 2-4% of patients with trigeminal neuralgia suffer from multiple sclerosis (Harris, 1950; Stookey and Ransohoff, 1959), while 2-2.5% of patients with multiple sclerosis have a form of trigeminal neuralgia (Parker, 1928; Garcin et al., 1960). Rushton and Olafson (1965) found 35 cases of combined trigeminal neuralgia and multiple sclerosis in a review of 1.735 cases of tic (2%) and 3.880 cases of multiple sclerosis (0.9%) seen at the Mayo Clinic. The neuralgia tended to occur early in the course of the disease and was usually bilateral.

Author for correspondence:
Florin Popovici, MD, Oral and Maxillofacial Surgery Hospital "Dan Teodorescu", 17-21 Calea Plevnei, District 1, Bucharest, Romania
e-mail: flrpop@yahoo.com

Trigeminal neuralgia may be associated with complex vascular lesions around the base of the brain and along the course of the trigeminal nerve.

Basilar artery ectasia may be associated with trigeminal neuralgia as a consequence of trigeminal nerve compression by the aberrant artery. In the literature there are reported cases of trigeminal neuralgia due to vertebrobasilar dolichoectasia as well as to anterior inferior cerebellar artery loop at the exit of the trigeminal nerve.

A variant type of the primitive trigeminal artery (PTA) is a rare anomalous vessel that originates from the internal carotid artery and directly supplies the territory of the anteroinferior cerebellar artery and/or the superior cerebellar artery. Although the PTA variant is frequently associated with intracranial aneurysms, it is extremely rare for the variant to lead to trigeminal neuralgia.

According to the National Institute of Neurological Disorders and Stroke, heredity may be a cause of trigeminal neuralgia. Some people inherit disorders of the blood vessels that affect the trigeminal nerve. People with a family history of trigeminal neuralgia or other blood vessel disorders may be more likely to develop this disorder themselves.

Trigeminal neuralgia secondary to a posterior fossa arteriovenous malformations has been seldom reported in the literature. Trigeminal neuralgia has been rarely reported in association with dural arteriovenous malformations (DAVMs) such as dural arteriovenous fistula in Meckel's cave. There are described cases of trigeminal neuralgia resulting from compression of the trigeminal nerve at its root entry zone by a large tentorial DAVM. Surgical interruption of the draining vein at its exit from the tentorium resulted in complete angiographic obliteration of the fistula with immediate resolution of the facial pain. Dural arteriovenous fistula (DAVF) of the transverse-sigmoid sinus is described to produce TN and also does the cavernous angioma in the brachium pontis.

Chiari I malformation has been found to be associated with TN in 19 cases in the English-language literature. In patients refractory to medical treatment, suboccipital decompression leads to resolution of pain in about two thirds of patients. Is important to consider Chiari I malformation as a rare cause of TN that responds to surgical therapy.

Trigeminal neuralgia may be produced in some instances by intracranial tumors, usually in the posterior fossa (Dandy, 1936; O'Connell, 1978; Destee et al., 1980); the reports of Love and Woltman (1942) and Hamby (1943) suggest that such lesions, when present, are small.

Parapontine epidermoid cysts either encase fifth nerve but with intact arachnoid between the capsule and the nerve, or compress and distort the nerve with tumor capsule adherent or attached to the nerve surface. Resecting the tumor capsule's attachment to fifth nerve is critical in relieving pain, even though this method may damage the nerve.

Contralateral involvement of the trigeminal nerve due to a mass occupying the posterior fossa space (such as meningioma) has been reported. In these cases, however, the involvement of the fifth nerve was generally manifest by early impairment of facial sensation and decreased corneal reflex. Very rarely, a contralateral trigeminal neuralgia is the only symptom for a long time. Haddad and Taha summarised 21 such cases from the medical literature. Six of these patients had a typical neuralgia, 12 atypical, and in four cases the information available was not complete. There was a predominance of females, the meningioma being the most frequent tumour.

The most plausible mechanism to explain the pathogenesis of the contralateral trigeminal involvement is that the tumour, situated in the back portion of the posterior fossa, pushed the brainstem and caused compression of the nerve root at its point of entry into the tentorial foramen. In conclusion, an isolated trigeminal neuralgia, especially if carbamazepine resistant, may be due to a contralateral tumour of the posterior fossa, such as a meningioma, and requires CT of the head. Surgical treatment relieves pain in almost all cases.

Neurinomas of the trigeminal nerve are a rare entity: those located in the posterior fossa account for 20% of all cases. In the majority of cases, the clinical presentation begins with fifth cranial nerve involvement producing a constant pain associated with other cranial nerve palsy and cerebellar signs.

Trigeminal nerve tumors such as schwannoma and malignant lymphoma may present as trigeminal neuralgia. TN is also seen in patients with glioblastoma.

We encounted a case of TN secondary to leukemic infiltration of the trigeminal nerve.

Invasion of the cavernous sinus by pituitary adenoma may cause involvement of cranial nerves III, IV, V and VI, but trigeminal neuralgia as an isolated, initial symptom is very unusual.

Mandibular metastasis can cause TN, metastatic tumors to the oral cavity being relatively uncommon. 70% of cases are adenocarcinoma – most commonly from breast and lung, followed by adrenals, kidneys, prostate, thyroid and colon.

Spontaneous intracranial hypotension and trigeminal neuralgia are examples of pain syndromes arising from shifting anatomical relationships in the posterior fossa. Both conditions are reported occurring in the same patient and resolving following surgical closure of a cervical nerve root sleeve dural defect.

There are instances of hydrocephalus associated with paroxysmal facial pain where relief of the hydrocephalus by a CSF shunt led to remission of the pain. It seems reasonable to postulate that in these cases the basis for the paroxysmal facial pain and paresthesias lay in the sensory root, and was possibly due to stretching of the root by distal displacement of the brain stem due to the hydrocephalus.

Neuropathy can affect the nerve from its origin in brainstem to its peripheral branches.

The association of Charcot-Marie-Tooth (CMT) disease and TN is rare. CMT and bilateral TN is even rarer. A literature review has revealed only five cases with CMT and bilateral TN. The underlying neuropathy in CMT makes the trigeminal nerve more vulnerable to vascular compression than usual.

Diabetes can produce almost any neuralgia nearly anywhere in the body, including trigeminal neuralgia. Diabetes damages the tiny arteries that supply circulation to the nerves, resulting in nerve fiber malfunction and sometimes nerve loss.

Infectious conditions may manifest as trigeminal neuralgia. There are cases of cerebellopontine angle cysticercosis, pons abscess and cases of bacterial infections (Shigella, Brucella, Leptospirosis, Lyme, Secondary syphilis, Mycobacterium leprae) manifested as trigeminal neuralgia.

Typical trigeminal neuralgia may follow an herpetic outbreak. This fact, taken in association with the other evidence that paroxysms of trigeminal pain may regularly precede the development of herpetic outbreaks occurring in the same area as the pain, a finding which is by no means unique, may indicate that the affected branches of the trigeminal nerve are the seat of an herpetic infection.

Granulomatous or inflammatory diseases, such as neurosarcoidosis or tuberculosis, may involve the nerve or ganglion at the site of Meckel's cave or cavernous sinus.

Trigeminal neuralgia has been reported with different rheumatic diseases such as rheumatoid arthritis, being an uncommon manifestation of relapsing polychondritis, systemic lupus erythematosus.

It is presumed that the TN in these cases was caused by compression of the trigeminal nerve from inflammation or ischemia secondary to vasculitis.

There is a report that describes a patient with mixed connective tissue disease (MCTD) and trigeminal neuralgia (TN). Features of TN, along with those of the 6 previously reported patients with TN and MCTD and response to steroid therapy, suggest that the TN in MCTD differs from the idiopathic form of the disease. Because the TN often precedes other features of the MCTD, it may be useful to test patients with atypical TN for the presence of anti-RNP antibodies in order to identify a subset with both systemic disease and a potential to respond to corticosteroid therapy.

Osteogenesis imperfecta may be added to Paget's disease as a potential cause of symptomatic trigeminal neuralgia in association with deformity of the skull base.

We encounted a rare case of a 31 years old female with Parry-Romberg facial hemiatrophy accompanied by trigeminal neuralgia exacerbated by a local infectious process-osteomyelitis of the mandibular body and angle, in whom the symptoms of pain were relieved after oral and maxillofacial surgical intervetion.

Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) is an autosomal-recessive disease associated with multiple deletions of mitochondrial DNA in skeletal muscle. MNGIE is a multisystem syndrome affecting muscle, peripheral, and central nervous systems and the gastrointestinal tract. There is a case in the literature of MNGIE with trigeminal neuralgia. An analogy is suggested between multiple sclerosis and MNGIE as a cause for trigeminal neuralgia in this patient.

Another rare associtión is familial oseodysplasia and trigeminal neuralgia.

Metabolic disorders such as gout, vitamin deficiencies (B1, B12, B3, B6, E), drug reaction (chloramphenicol, nitrofurantoin, sulfonamides, isoniazid) may be causes of TN.

According to the Beth Israel Medical Center Department of Pain Medicine and Palliative Care, stress may be a cause or trigger of trigeminal neuralgia. Stress can cause inflammation of blood vessels, which can compress or irritate the trigeminal nerve. In people who have trigeminal neuralgia due to another cause, stress and anxiety can trigger or worsen the severity of attacks.

CONCLUSIONS

The trigeminal nerve is the largest cranial nerve with both a sensory and motor function. The evaluation of patients with trigeminal neuralgia should include high-quality, thin-section, magnetic reso-

nance imaging scans, to exclude the possibility of vascular lesions and other structural lesions. In every examination for trigeminal symptoms, imaging of Meckel's cave, the cavernous sinus, the skull base foramina, the pterygopalatine fossa and the peripheral trigeminal nerve course should be included. On MRI, we always look for a normal gray and white matter and brainstem, a normal cisternal segment, a fluid filled Meckel's cave, a homogeneously enhancing cavernous sinus, a fat-filled superior orbital fissure and pterygopalatine fossa, a normal foramen rotondum and ovale, and a symmetrical appearance of the masticator muscles with normal fat planes of the skull base. In particular, patients who are being evaluated for surgical treatment of trigeminal neuralgia should undergo magnetic resonance imaging, with a focus on the course of the trigeminal nerve.

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