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A case of trigeminal-vagal neuralgia relieved by peripheral self-stimulation

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Abstract

A case of simultaneous trigeminal and vagoglossopharyngeal neuralgia is described. No microvascular compression was seen at the trigeminal complex while the origin of vagoglossopharyngeal pain could have been due either to Chiari malformation or microvascular compression of the IX-X nerve complex. Decompressive surgery was ineffective. The patient could completely block his facial painful fits by strongly pinching the anterior axillary fold.

This case militates against peripheral theories of facial neuralgias, including microvascular compression and ganglion ignition focus theories, and supports a central origin thereof.

Key words : Trigeminal neuralgia ; vagoglossopharyngeal neuralgia ; microvascular compression ; trigeminal ganglion ignition focus theory.

Introduction

Idiopathic trigeminal neuralgia (ITN) (tic douloureux) is characterized by "sudden, usually unilateral, severe brief stabbing recurrent pains in the distribution of one or more branches of the Vth cranial nerve" (Merskey and Bogduk, 1994). Pain of ITN is extremely severe and it is usually described by patients as sharp, agonizing electric shock-like stabs felt superficially in the skin or oral mucosa. Attacks are generally of brief duration (few seconds, rarely up to 1-2 minutes) and are followed by a refractory period of up to a few minutes. Light tactile stimulation of more or less restricted sites within the area of trigeminal innervation (trigger points) may set off pain. ITN has a characteristic periodicity, with episodes occurring for weeks to months followed by spontaneous remission that may last months or years. In time, pain paroxysms may become more frequent and severe.

The genesis of ITN is unknown, but is generally believed to be due to demyelination of sensory fibers within the nerve root resulting from microvascular compression (MVC) at the trigeminal root entry zone (REZ) by both pulsating arteries or veins. This theory has gained almost universal acceptance in the neurosurgical literature. Microvascular decompression (MVD) is the standard operation for relief of ITN, generally following magnetic resonance (MR) angiography. MVD is performed trough a small retromastoid craniectomy, under general anesthesia. Any compressive vessel is isolated and separated from the neural elements. Pain relief is complete in about 80% of cases immediately after operation, but only about two thirds of the patients are still drawing benefit at 10 years (Barker, 1996).

The MVC hypothesis draws support from the efficacy of surgical decompression, but this hypothesis collapses on the basis of astute clinical observations (Adams, 1989 ; Canavero *et al*., 1997 ; Sweet, 1998 ; Bonicalzi and Canavero, 2000), including the disappearance of ITN for 8 years after MVD in a patient without MVC (Sweet, 1998).

We recently experienced a patient with long standing ITN and no evidence of MVC who could systematically and reproducibly block his attacks by intense mechanical stimulation in the C4-C8 territory.

Case report

This 70 year-old hypertensive hypoacusic man has complained of paroxysmal, shock-like painful fits involving the left trigeminal territory (branches II and III) plus a cutaneous area between the mandibular angle and the left hyoid horn, for about 30 years. Fits never involved the tonsillar sinus, the tongue or the acoustic meatus. The pain either awaked him from night sleep or prevented it. Over the past 20 years, cycles of carbamazepine (CBZ) up to 1200 mg reduced both frequency and intensity of the attacks. Gabapentin up to 1000 mg was ineffective. Instead, the patient found out that strong pinching of the left anterior axillary fold or – more rarely – left wrist between thumb and remaining fingers blocked the fits as they started. Three left percutaneous glycerolizations over three years (1996, 1997 and 1998) each afforded complete relief for 9-10 months. A fourth one in March 2000 was ineffective.

The neurological examination disclosed bilateral hypoacusia, slurred speech (due to pain), slight gait

ataxia, hyperreflexia of lower limbs, finger-to-nose and heel-to-knee tests abnormal on the left side and worsened by shutting the eyes. The patient almost continuously rubbed or pressed the left hemiface with his open left hand but could not explain why. This maneuver neither triggered nor relieved the pain. The right hand was constantly held against the left pectoral muscle with the thumb held outside and the other fingers in the armpit.

The corneal reflex was bilaterally normal ; there was a slight left trigeminal hypesthesia involving the II and III branches and static and dynamic asymmetry of the lips. The uvula was shifted to the left.

Speaking and swallowing triggered the pain, but there were no observable skin or intraoral trigger points. Fits lasted a few seconds each time. If the patient kept silent, a free interval of up to a few hours was possible.

During the fits the patient performed the above-described maneuver, which brought about an immediate block of the pain lasting for some minutes and allowing him to talk. This compression could not prevent the fits, though, but systematically blocked them.

The frontal and orbicular electromyography and somatosensory evoked potentials (median nerve) were normal. The electroencephalogram showed a few aspecific irregularities on the left (slow rhythms during somnolence) and short diffuse theta hypersynchronous sequences. Left brainstem auditory evoked potentials showed anomalies of the first portion of the acoustic pathway including the acoustic nerve. MR imaging showed Chiari I malformation with a downward displacement of the cerebellar tonsils 8 mm below the occipital foramen ; frontoparietal subcortical diffuse vascular encephalopathy of both hemispheres ; enlarged lateral ventricles and cisterns ; no MVC of the trigeminal nerve but an anomalous branch of the left posterior inferior cerebellar artery (PICA) coursing obliquely through the left cerebellopontine cistern.

On July 6 2000 surgery was performed. A fixed left cerebellar tonsil had to be debrided from arachnoditis. After displacing it, a large left PICA was seen elevating and stretching the XI and IX-X nerve complex, particularly the X, without a bona fide MVC. After mobilization, the vertebral artery left a groove on the bulbar surface at the anterior olivary margin. The X and XI cranial nerves were displaced downwards and kept so with fascia and muscle. No microvascular compression was seen at the trigeminal complex.

Postoperatively, the pain was improved, CBZ was discontinued but the above-reported maneuver was still reiterated.

In August 2000, the pain fully relapsed and did not improve over 6 months. CBZ was again partially effective while the pain-quenching maneuver produced 100% relief. The neurological examination was unchanged.

Discussion

Our patient suffered from typical ITN affecting the left II and III branches plus a left superior laryngeal neuralgia (a variant of vagoglossopharyngeal neuralgia) with simultaneous painful activation (an already reported combination, see in Loeser, 2001).

Nocturnal attacks in patients suffering from long-lasting ITN are not uncommon (Sjastaad *et al*., 1997). The pain referred to the angle of mandible could also be due to Chiari compression at olivary level. Chiari malformation is known to associate with painful facial syndromes (Kanpolat *et al*., 2001), but in our patient operation did not afford pain relief. While MVC of the IX and X cranial nerves rootlets was also found, MVD was ineffective.

This is the first reported case of ITN relieved by mechanical compression outside the trigeminal territory. Weber (quoted in Sweet, 1998) reported a patient suffering from a typical left second division neuralgia that could stop her pain by rubbing her left cheek with a wool cloth for about half a minute. Repetitive rubbing induced atrophy and pigmentation of her cheek. Sweet (1998) also reported a patient with MS-associated TN in whom pain spread from the trigeminal area to the ipsilateral radial forearm or to the shoulder and arm to elbow. There are sparse reports of ITN triggered by stimuli in remote locations (Loeser, 2001) but secondary activation of appropriate trigger areas cannot be ruled out. Our patient could unmistakably and reproducibly abolish his pain by mechanical stimulation of distant areas.

A recent peripheral theory of ITN, so-called trigeminal ganglion ignition focus (TGIF) theory, which also incorporates the MVC speculation, shifts the main generator of pain paroxysms in the gasserian ganglion (Rappaport and Devor, 1994). The TGIF theory states that trigger stimuli set off burst of activity in a small cluster of trigeminal ganglion neurons rendered hyperexcitable as a result of trigeminal ganglion or root damage, thus spreading from this TGIF to encompass wider portions of the ganglion. After a brief period of autonomous firing, refractoriness to further spontaneous activity ensues, due to an intrinsic hyperpolarizing process. According to this theory the primary abnormality resides in the trigeminal ganglion and root, rather than in the skin or the CNS, and, according to the originators, it would collapse if trigger areas of typical trigeminal pain would be found outside the trigeminal territory.

In the above-described case, the TGIF theory cannot explain the simultaneous trigeminovagoglossopharyngeal pain and its inhibition by mechanical stimulation of distant areas. A trigeminovagoglossopharyngeal discharge can be modulated by tactile/mechanical afferences travelling in high cervical roots only at central level.

An alternative theory, starting with Trousseau's suggestion that ITN should be termed epileptiform neuralgia, states that ITN has a central cause, with associated failure of intranuclear inhibition (Fromm *et al*., 1981 ; 1984). If we accept that ITN and glossopharyngeal neuralgia might be some sort of hyperfunctional disorder of the brain stem (as central theories do) the above-described pain-blocking effect from strong mechanical stimulation can be explained by active inhibition ("gate-like") of an anomalous generator : a strong volley of peripheral impulses may quench the trigeminal focus, just like surgical manipulation of the cranial nerves during MVD (Adams, 1989 ; Canavero *et al*., 1995 ; Canavero *et al*., 1997).

In conclusion, this report challenges both the MVC and ganglion ignition focus theories and supports a central origin of TN and VGPN.

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