Actual management of essential trigeminal neuralgia

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ABSTRACT

Trigeminal neuralgia is a common cause of facial pain. Pathophysiology considerations, surgical armamentorium and results are mentioned. Because none of the currently available techniques is perfect, there is a continuing effort of neurosurgeons to develop new treatment methods. There are: microvascular decompression – which offer the best chance of permanent pain relief without sensory loss or dysesthesia, Gamma knife surgery, but also lesioning techniques.

It’s important for each neurosurgeon to be skilled not only in microvascular decompression, but also in at least one of the percutaneous treatment methods: radiofrequency rhizotomy, Gasserian ganglion glycerol injection, balloon compression. In this way treatment should be customized to the individual patient, informing properly of the risks, benefits of the main treatment options, also about the spectrum of technical and human competences locally available.

Key words: trigeminal neuralgia, dorsal root entry zone, surgical armamentorium

Trigeminal neuralgia, also known as “tic douloureux” still is a serious health problem, with a mean annual prevalence of 4 to 5 per 100,000 (1). However the incidence of trigeminal neuralgia increases with age, affecting the older age group, but also individuals in their second and third decades of life (2,3). There is a slight predominance of females to males 5.9 and 3.4 per 100,000 population, respectively; also the right side of the face is slightly more frequently affected than the left and rarely bilateral neuralgia can occur (1).

The most generally admitted pathophysiology suggests that trigeminal neuralgia is a multifactorial disease with a number of causes of which vascular compression of the trigeminal nerve root is the commonest (2-4), close to its entry into the pons, by an aberrant arterial or venous loop, most often at the central portion of the root (1-3). Through chronic pressure and pulsations, vascular compression causes focal demyelination, with groups of axons juxtaposed without interstitial glia. These focal alterations would be at the origin of neosynapses, called also ephapses, with direct
cross-talk between fibres. Ectopic spontaneous firing, chronically sustained, would generate central hyperactivity in the trigeminal system nuclei. The dorsal root entry zone is histologically defined as the zone where the peripheral myelination (schwannian) leave place to the central type of myelination (oligodendrocytic) – classically located 3 mm from the emergence. The location of this zone is highly variable and can spread more distally in some patients (4). Over the years, healthy criticism of the neurovascular compression hypothesis has arisen, the questions and answers that have been highlighted are:

- the incidence of vascular compression in cases of trigeminal neuralgia are found in 88.6% of cases, in about 5% of cases tumors (schwannomas, epidermoids, meningiomas) or arteriovenous malformation is found and in 7.5% of cases had no vascular compression or other lesions affecting the nerve; although biopsies taken at the site of a partial sensory rhizotomy and examined by electron microscopy can occasionally reveal unsuspected inflammatory demyelination (5,6)
- the incidence of trigeminal vascular compression in asymptomatic humans exists in 13% of the normal population (1)
- chronic vascular compression may cause the characteristic attacks of lancinating pain at light and unexpected stimuli by ischemic changes within the nerve, who is generating the segmental demyelination with ephaptic “crosstalk” between axons, but also by remyelination – which might explain the well-known phenomenon of spontaneous remissions and aberrant remyelination where several axons were enclosed by a single myelin sheath – explaining perhaps the occasional failure to improve or relapse after microvascular decompression surgery (1,7).

The diagnosis of trigeminal neuralgia must be done according to the International Headache Society definition; its definition of primary or secondary cases and typical versus atypical is also employed (3).

A typical primary, idiopathic, trigeminal neuralgia is clinical diagnosed on the basis of 6 criterion in the presentation (3):

- pain location on one side of the face
- no extension outside the trigeminal territory
- paroxysmal pain of the “electric shock” type, no pain between the attacks, no permanent pain at least at the beginning of the disease
- pain paroxysms that may occur spontaneously but more are triggered by stimuli
- no sensory deficit, no decrease in corneal reflex and no symptoms in other cranial nerve territories
- effectiveness of carbamazepine, at least initially

Atypical trigeminal neuralgia presentation (3) – not to be confused with atypical facial pain is characterized by spontaneous nonparoxysmal long lasting irritation in sensitive region of the trigeminal nerve: the pain becomes more frequent, pain free intervals shorten, an aching/burning background pain appears, sometimes with vasomotor phenomena.

Trigeminal neuralgia can be diagnosed as “primary” only after all specific causes have been eliminated by appropriate investigations. High resolution T2 like the CISS (Constructive Interference in Steady State) sequence with 3D Angio MR are the best MRI sequences for precise targeting of normal structures to demonstrate a neurovascular conflict (2, 3, 7).

Currently surgical armamentarium on patients suffering from essential trigeminal neuralgia has multiple alternatives because none of the currently available techniques is perfect, the form of treatment offered is influenced also by the spectrum of technical and human competences locally available thus explaining the continuing effort of neurosurgeons to develop new treatment methods (2, 3):

- microvascular decompression, first described in the 1950’s by Taarnhoj, treats the putative cause by separating the loop from the trigeminal nerve is established as the techniques of choice treating the aetiological cause of the disease as well as offering a very low risk of subsequent trigeminal nerve dysfunction. The procedure is performed under general anesthesia with a retrosigmoid craniotomy. This procedure in experienced hands has rare mortality, low morbidity consisting in facial palsy, deafness and should be the gold standard treatment for young
patients (2,3,8-10). On 1204 cases (8), results are excellent initial – analgesia in 82% of cases, 1 year after the first operation – analgesia is 75%, after the second operation 80%; 10 years after analgesia is in 64% of cases and 70% of cases after the second operation.

- **percutaneous techniques** under a brief general anesthesia: **thermocoagulation, balloon microcompression, glycerol injection** produce a partial lesion of the trigeminal nerve – the induction of hypoesthesia particularly with thermocoagulation is essentially for prolonged pain cassation, are simple techniques, easy to repeat, readily suitable for the elderly (2,3; 11-13). Mean results: to 90% of cases analgesia instaled immediately or in the first 24-48 hours, if pain reappears – in 50% of cases 3 years after, procedure should be repeated (2, 3).

- **stereotactic radiosurgery** using Gamma Knife with 201 cobalt sources adopted in 1968 was based on Lars Leksell concept in 1951 who performed radiosurgery with a stereotactic X-ray beam on patients suffering from essential trigeminal neuralgia. Radiosurgery has the advantage of being the least invasive available procedure, is performed under local anesthesia, the rate of trigeminal dysfunction is remarkably low and comparable to that occurring with microvascular decompression (14-19). It’s efficacy is based on radiobiologic selective effect, to block ephaptic transmission without affecting normal axonal transmission, also by focal demielination, inflammation, ischemia, proven by MRI with contrast (17). After procedure, analgesia appear in 24 hours-8 month to 86-95% of patients, recurrency in 7% of cases and depends on the accuracy of the selected target (19).

On my own observations for idiopathic trigeminal neuralgia in a series of 578 patients treated between 1992-2008 (20) I can made the following conclusions:

- for 231 patients (39.96%) operated by microvascular decompression there was a neurovascular conflict in 207 patients with correlation between pain topography and site of the neurovascular conflict especially by megadolicho-vertebro-basilar artery; with good results even 1 year after procedure on 193 patients (complete pain relief, no hypoesthesia, no side effect). For those 24 patients without neurovascular conflict (4.13%) the root was not cut, pain relief was remarked on 18 patients, but on 13 patients pain recurred.

- for 347 patients (60.03%) operated by a destructive procedure: percutaneous selective thermo-rhizotomy I had good results on 239 patients even 1 year after procedure (68.87%), taking into account fibers somatotopy for selectivity. For good results (20) it is to be considered to perform such procedure on patients with pain installed less than 6 month, after X-ray control, lesion made at the triangular plexus on selected area, using neurophysiological testing (electrical stimulation 5c/sec, 0.2-0.4 V, checking evoked paresthesias in sensory territories and muscle twiches – clinically observable in masticatory muscles by motor root stimulation, also in facial muscles by sensory root stimulation using trigemino-facial reflex according to electrod location in V₁, V₂, V₃)

- I had recurrency in 68 patients (11.76%), pain recurred more frequently in patients with minimal trigeminal disfunction, compared with those who sustained facial numbness, paresthesias or dyesthesias.

To obtain best result in primary trigeminal neuralgia, avoiding harmful dyesthesia, keratitis, diplopia, anesthesia dolorosa masticatory deficit; in my opinion, a diagnosis and treatment algorithm should be imposed, based on several criteria (2,3).

If trigeminal pain persist postoperatively I suggest:

- **microvascular decompression** after percutaneous techniques failures
- **radiosurgical treatment**: especially to old patients after surgical procedures failures (microvascular decompression or percutaneous techniques)
- **trigeminal rhizotomy** if MRI, arteriography and even cerebellopontine angle exposure are normal
- **tractotomy – CT trigeminal nucleotomy guided or percutaneous/surgical Gasser stimulation** for iatrogen distesitia after
Medical history, neurologic examination, MRI, 3D Angio MR, cerebral CT with contrast, vertebral angiography

Surgical purpose: analgesia, without disesthesia, no mortality, less morbidity, to assure quality of life by optimum strategy for each case

Essential trigeminal neuralgia considering:
- patient age
- after medical treatment failure
- patient option
- surgeon preference
- treatment cost

Secondary trigeminal

Cerebellopontine Angle exposure
+ to remove all compressive factors
Alternatives: retrogasserian rhizotomy, radiofrequency rhizotomy, radiosurgical treatment

if age:
< 65 yrs, to patients without other illness, operation purpose:
analgesia without disesthesia,
with trigeminal neuralgia on ophthalmic branch or on all branches, with surgical risks
microvascular decompression
< 65 yrs, operation purpose:
analgesia with minor disesthesia,
with less surgical risks
percutaneous radiofrequency selective rhizotomy
> 65 yrs, noncooperative patients, anesthetic risk, who refuse cerebellopontine angle exposure:
1. peripheric neurectomy, alcholisation
2. for ophthalmic neuralgia –
glycerol percutaneous retrogasserian rhizotomy; CT guided trigeminal tractotomy-nucleotomy; balloon compression (this technique impose general anesthesia)
3. for maxilar and mandibular neuralgia – radiofrequency thermocoagulation, balloon compression, radiosurgical treatment
different operative techniques concerning trigeminal system

- nucleus caudalis drezotomy: for uni or bilateral ophthalmic pains, after failure of at least one trigeminal technique or for postoperative anesthesia dolorosa
- thalamic stimulation (ventral posteromedial nucleus) and motor cortex stimulation: for postherpetic trigeminal pains, postoperative anesthesia dolorosa.

Conclusion

For primary trigeminal neuralgia, treatment should be customized to the individual patient, informing properly of the risks, benefits of the main treatment options, also about the spectrum of technical and human competences locally available.

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