Botox injection in PALATAL MYOCلونUS

Lecture objectives

• By the end of this presentation you will have a clear idea about:
  – Palatal myoclonus.
  – Tinnitus associated with palatal myoclonus.
  – Botulinium toxin.
  – Technique of Botox injection.
Palatal myoclonus

• Palatal myoclonus is a rare condition, characterized by continuous rhythmic, or dysrhythmic jerks of the soft palate and pharyngeal muscles that are usually bilateral, at a rate varying between 40 and 240 per minute.

• Most patients present with annoying tinnitus.
History

• The condition was first reported by Kupper (1873) though Spencer (1886), coined the phrase “pharyngeal and laryngeal nystagmus”.
• Politzer (1878) was the first to describe this type of objective tinnitus. His impression was that movements of the tubal muscles caused the noises reported.

[Image of a timeline showing key events in the history of botulism and BTX-A use, including years and milestones such as 1822, 1895, 1940s, 1960s, 1970s, 1989, 1980s and 1990s, 2000, 2002, 2004. Details include identification of bacteria as cause of botulism, BTX-A isolated and purified, FDA approval for strabismus, FDA approval for cervical dystonia, and FDA approval for glabellar lines.]
Etiology of palatal myoclonus

• It is usually caused by a vascular ischemic lesion of the brainstem or cerebellum that disrupts the dentato-rubro-olivary pathway.

• The most common finding is hypertrophic degeneration of the inferior olivary nucleus involving the Guillain Mollaret triangle
  • dentate nucleus
  • red nucleus
  • inferior olivary nucleus
  • central tegmental tract
• Several other causes less frequently observed
  - multiple sclerosis
  - trauma
  - tumours
  - degenerative diseases
  - subarachnoid hemorrhage

• The time period between the occurrence of the lesion in the myoclonic triangle and the clinical manifestations of palatal myoclonus is not clear.
• It is stated that the symptoms most commonly occur within 10-11 months, although there might be a delay of several years.
• The movement can also spread to involve other branchial muscles, as well as the face, the eye, the neck, the diaphragm or the arm muscle groups.

Types

• Deuschl et al (1990). stated the existence of two types of palatal myoclonus:
  a. Symptomatic, in which a lesion in the brainstem or cerebellum is identified.
  b. Essential, in which no such lesion is documented.
Clinical presentation

• If restricted to the soft palate (typically essential PM), it may be asymptomatic.
• Often, however, an annoying clicking noise is heard by the patient, caused by opening and closing of the eustachian tube.

• The PM movement persists in the sleep state.
Diagnosis

- Diagnosis is usually based on clinical examination. However, MRI is essential for the definite diagnosis.
Palatal tremor in relation to brainstem tumour involvement

HISHAM EL BADAN, MD

Treatment of palatal myoclonus

• The treatment of PM is variable, with better results in those with PPM.
• In all patients it should be carried a trial with sumatriptan, even those with possible secondary PM, to differentiate clinically the PPM from SPM.
Palatal Myoclonus and Objective Tinnitus

- The production of objective tinnitus by palatal myoclonus has been reported in the literature as being related to palatal or middle-ear myoclonus.
- Tensor palatine muscle and the levator palatine muscles are involved in palatal myoclonus.
Tensor veli palatini muscle

- The Tensor veli palatini muscle originates from the edge of the greater wing of the sphenoid bone, adjoining petrosphenoidal fissure, and lateral wall of the eustachian tube.
- Its tendon joins the palatal aponeurosis within the soft palate.
- It is supplied by the fifth cranial nerve.

Levator veli palatini

- The levator veli palatini muscle arises from the inferior surface of the apex of the petrous bone medial to the eustachian tube.
- Its tendon merges with the upper and posterior surface of the palatal aponeurosis.
- Its nerve supply comes from the pharyngeal plexus and the eleventh cranial nerve.
The tensor tympani muscle and the stapedius muscle are involved in middle ear myoclonus.

Tensor tympani muscle

- The TTM arises from the cartilaginous part of the eustachian tube, the adjacent part of the greater wing of the sphenoid, and enters its 20- to 25-mm-long semicanal.
- Its tendon inserts in the neck of the malleus.
- Its nerve supply is the fifth cranial nerve.
Stapedius muscle

- The SM arises from the pyramidal process.
- Its tendon inserts in the neck of the stapes.
- Its nerve supply is the seventh cranial nerve.

Muscle actions

- The TVPM increases tension in the soft palate and opens the eustachian tube when it contracts.
- The LVPM elevates the soft palate and probably assists in the opening of the eustachian tube by the TVPM.
- The TTM pulls the malleus medially, increasing tension in the tympanic membrane.
- The SM pulls the stapes footplate away from the oval window.
• Anatomy and physiology provides some important relevant notes:
  – The TVPM and the TTM both are supplied by the fifth cranial nerve, and both have insertions in the cartilaginous portion of the eustachian tube.
  – In contrast, the SM and the LVPM are supplied by different cranial nerves and by the pharyngeal plexus, and neither has insertions in the cartilaginous portion of the eustachian tube.

• Very likely, palatal (TVPM) and middle-ear (TTM) myoclonus occur together. Both muscles are supplied by the fifth cranial nerve and belong to the same group of muscles.
• Middle ear myoclonus can be treated successfully by section of the TTM and SM tendons.

• SM contribution to the clicks is possible but unlikely because it is supplied by a different cranial nerve (nerve VII) and it is a small muscle (four times smaller than the TTM). However, it may be the cause of objective tinnitus after facial paralysis.
• The source of the actual clicking is postulated to arise from the breakdown of fluid surface tension as the walls of the eustachian tube open under the action of the peritubal muscles.

Treatment

• The tinnitus associated with palatal myoclonus has been traditionally managed:
  – with drugs such as anxiolytics, antidepressants and anticonvulsants.
  – with surgical procedures aimed at modifying palatal and eustachian tube function.
  – with adjunctive measures such as psychotherapy.
• The tinnitus however is often refractory to these measures though more recently white-noise masking has shown more favorable results in selected cases.
Botulinum toxin

- The drug is a noninfectious toxin haemagglutinin complex.
- whilst six toxin serotypes have been isolated only type A is in clinical use.

Mechanism of action

- The toxin binds to the motor end-plate and prevents acetylcholine release giving rise to pre-synaptic neuromuscular blockade resulting in flaccid muscular paralysis.
- Peak effect occurs within 7-14 days, and muscular function is reestablished within 3-6 months.
- Repeated use does not result in muscle atrophy or permanent degeneration.
Commercial forms

<table>
<thead>
<tr>
<th>Botox</th>
<th>Dysport</th>
<th>NeuroBloc</th>
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<tbody>
<tr>
<td><strong>Freeze-dried powder</strong></td>
<td><strong>Freeze-dried powder</strong></td>
<td><strong>A liquid protein complex</strong></td>
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<td>that clumps at the bottom</td>
<td>comes in a plastic box</td>
<td>vials</td>
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<td><strong>100 - 200 units of</strong></td>
<td><strong>Two 500 unit refrigerated</strong></td>
<td><strong>Three different sizes:</strong></td>
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<td>vials</td>
<td>2,500, 5,000 and 10,000</td>
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<td>units refrigerated vials</td>
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<td>kept at 2–8°C centigrade</td>
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</tbody>
</table>
Tools for Botox injection

RECONSTITUTION

Table 1: Dilution Instructions for BOTOX Vials (100 Units and 200 Units)*

<table>
<thead>
<tr>
<th>Diluent* Added to 100 Unit Vial</th>
<th>Resulting Dose Units per 0.1 mL</th>
<th>Diluent* Added to 200 Unit Vial</th>
<th>Resulting Dose Units per 0.1 mL</th>
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</thead>
<tbody>
<tr>
<td>1 mL</td>
<td>10 Units</td>
<td>1 mL</td>
<td>20 Units</td>
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<td>2 mL</td>
<td>5 Units</td>
<td>2 mL</td>
<td>10 Units</td>
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<td>4 mL</td>
<td>2.5 Units</td>
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<td>5 Units</td>
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<td>8 mL</td>
<td>1.25 Units</td>
<td>8 mL</td>
<td>2.5 Units</td>
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<tr>
<td>10 mL</td>
<td>1 Unit</td>
<td>10 mL</td>
<td>2 Units</td>
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</table>
• The product is vacuum-sealed.
• A thumb can be placed under the plunger of the syringe to control the rate of release of saline onto the powder or air can be injected into the vial to avoid too rapid reconstitution.

• The saline **must not gush** in and agitate the solution mechanically. Rotating the vial during injection also assists a gentle reconstitution.
INJECTION

• It is recommended to use one ml insulin syringes. These are essential for the dose to be gauged accurately during injection.

• Botulinum toxin is potent and very expensive, so each drop must be used to its maximum effect.
• Even 0.0125ml is effective in certain sites.
• Up to 0.3ml can be salvaged from the end of a bottle if the top is removed with a can opener.

Sites of injection
Palatal injections

Tubal injections
THANK YOU