UPDATE ON Craniofacial Pain and Dystonia

North American Neurotoxin Association

NORTH CENTRAL HEADACHE SOCIETY
Neuropathic Facial Pain

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Disclosures

• I have no disclosures.
• I will be discussing the off-label use of treatments in this talk.
Neuropathic Facial Pain

- Trigeminal neuralgia and neuropathy
- Glossopharyngeal neuralgia
- Nervus intermedius neuralgia
- Other less common facial pain disorders
- Excluded from this talk: Trigeminal autonomic cephalgias (TACs) and occipital neuralgia.
Trigeminal Neuralgia

- ICHD-3 criteria:
  - At least three attacks of unilateral facial pain fulfilling the following criteria:
  - In one or more division of the trigeminal nerve but not beyond that distribution
  - 3 of the follow 4 characteristics:
    - Paroxysmal attacks lasting fraction of a second to 2 minutes
    - Severe pain
    - Electric shock-like, stabbing, shooting, or sharp
    - Precipitated by innocuous stimuli to that side of the face
  - No clinically evident neurologic deficits
Trigeminal Neuralgia

- Source: ufhealth.org
Classic TN vs. Neuropathy

- Classic: idiopathic cause or later discovered to be caused by neurovascular compression
- Trigeminal neuropathy: other secondary causes, such as herpes zoster, multiple sclerosis, tumor, etc.

Source: ICHD-3
Clinical Features

- V3 or V2 > V1
- Older patients > 50, women > men
- Pain provoked by triggers with refractory period after repeated triggers
- Can have constant dull pain in between, rarely occurs during sleep
- No autonomic features although tearing has been described
- Remissions are possible

TN vs. TACs

- There can be some diagnostic confusion with TN vs. TACs, particularly SUNCT/SUNA.
- When TN is in V1 (rare), there can be diagnostic overlap.
- TN has refractory period, SUNCT/SUNA does not.
- TN should have no autonomic features.
- SUNCT/SUNA can have sawtooth pattern of attacks, although not always.
- Some treatment overlap between the disorders, I am working on a systematic review on this currently.

Diagnostic Imaging

- Obtain a brain MRI with and without contrast when assessing a patient with TN. Add FIESTA or MP-RAGE (depending on software at your institution) and 3D CISS (constructive interference in steady state) sequence.
- If there is neurovascular compression and the patient is refractory to medication, he can be assessed for gamma knife treatment or microvascular decompression at a center with neurosurgeons who perform these procedures.
- Imaging also will help identify other secondary lesions causing trigeminal neuropathy.
Treatments

• Systematic Review for AAN in 2008:
  – Effective: Carbamazepine
  – Probably effective: oxcarbazepine
  – Possibly effective: lamotrigine, pimozide, baclofen

• Screen for HLA allele B*1502 in patients of Asian decent before starting carbamazepine or oxcarbazepine

• Consider IV phenytoin or lidocaine for acute severe exacerbations, or SPG block


Other Treatments

• Clonazepam, gabapentin, pregabalin, topiramate, tizanidine, levetiracetam, lacosamide

• Ziconotide (intrathecal infusion), an N-type calcium channel blocker

OnabotulinumtoxinA

• Several open label and RCTs supporting onabotulinumtoxinA use in trigeminal neuralgia.
• Recent systematic review by Morra et. al shows that onabotulinumtoxinA is safe and efficacious in trigeminal neuralgia
  – Adverse events included facial edema and asymmetry
  – Different studies used different amounts of onabotulinumtoxinA and in different locations, further studies are needed to determine optimal amount and location
Overall, the main effect (patients with >50% reduction of pain score from baseline to endpoint favored botulinum toxin A: (RR = 2.87, 95 % CI [1.76, 4.69], P < 0.0001).

Mean VAS score was significantly lower for the botulinum toxin A group at the end of month 1, 2, and 3.

Mean paroxysm frequency per day was also significantly lower in the botulinum toxin A group.

Followup ranged 8-12 weeks.

Painful tic convulsif

- Not in ICHD-3, but described in literature as coexistent trigeminal neuralgia and hemifacial spasm.
- Usually caused by neurovascular compression in posterior fossa.
- Several case studies showing efficacy for botulinum toxin A in this disorder.

Glossopharyngeal Neuralgia

• ICHD-3 criteria:
• At least three attacks of unilateral pain fulfilling the following criteria:
  – Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw and/or in the ear
  – Pain has 3 of the following 4:
    • Recurring in paroxysmal attacks lasting from a few seconds to 2 minutes
    • Severe
    • Shooting, stabbing, or sharp
    • Triggered by swallowing, coughing, talking, or yawning
  – No neurologic deficit
Glossopharyngeal Neuralgia

- Must rule out secondary cause, including Eagle syndrome:
  - Elongation of the styloid process or calcification of the stylohyoid ligament.
  - Pain in neck with head position change, pain with palpation of styloid process, foreign body sensation in throat, hypersalivation, otalgia, dysphagia, voice change
  - Can even cause carotid dissection
  - Diagnosis: lateral views of skull base and cervical spine
  - Most cases acquired, trauma #1
  - Treatment usually surgical

Eagle Syndrome
Glossopharyngeal Neuralgia

• Glossopharyngeal neuralgia can even cause cardiac syncope in rare cases, due to nerve fibers connected to carotid sinus and dorsal motor nucleus of CN X affected.

• Treatment medically is similar to that of trigeminal neuralgia.

• Surgical treatment includes nerve sectioning and microvascular decompression. Check MRI brain with and without contrast with FIESTA or MP-RAGE (depending on software at your institution) and 3D CISS sequence like with trigeminal neuralgia.

Nervus intermedius Neuralgia

- Small sensory branch of the facial nerve (cranial nerve VII) carrying general visceral efferent, special visceral afferent (taste), and general somatic afferent fibers
- ICHD-3 criteria:
- Three attacks of unilateral pain fulfilling the following criteria:
  - Located in auditory canal, can radiate to parieto-occipital area
  - 3 of the following 4 criteria:
    - Recurring in paroxysmal attacks lasting from a few seconds to minutes
    - Severe
    - Shooting, stabbing, or sharp
    - Precipitated by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region
  - No neurologic deficit
Nervus intermedius Neuralgia
Ramsay-Hunt

• Nervus intermedius neuralgia can be secondary to mass, vascular compression, or reactivation of herpes zoster virus (Ramsay-Hunt syndrome).
• Check MRI IAC and MRA.
• Triad of ipsilateral facial paralysis, ear pain, and vesicles in the auditory canal and auricle. Taste, hearing, lacrimation, vestibular function can also be affected.
• Reactivation of virus in geniculate ganglion affecting multiple CNs.
• Treated with steroids like Bell’s palsy, no evidence for efficacy of antivirals despite widespread use.

Nervus intermedius Neuralgia

- Again, treated similarly to trigeminal neuralgia when no major secondary cause is found.
- Surgical treatment includes nerve sectioning (with complications including partial facial palsy, ipsilateral xerophthalmia) or microvascular decompression.

Tolosa-Hunt

• Unilateral orbital pain associated with paresis of one or more of the IIIrd, IVth and/or VIth cranial nerves caused by a granulomatous inflammation in the cavernous sinus, superior orbital fissure or orbit.
• Unknown etiology
• Pain usually precedes ophthalmoparesis
• CN V1 and pericarotid sympathetic fibers can be affected as well, causing Horner’s

Tolosa-Hunt

- Investigation:
  - MRI with and without contrast, MRV
  - CBC, CMP, inflammatory markers, autoimmune workup, lyme ab, SPEP, ACE.
  - CSF studies: glucose, cell count with diff, protein, lyme, VDRL, cultures, ACE, cytology
- Prompt response to corticosteroids confirms diagnosis
- High dose IV solumedrol or high dose prednisone followed by taper of weeks to months.
- Radiographic improvement lags behind clinical improvement.

Raeder’s syndrome

• Also known as Paratrigeminal oculosympathetic syndrome
• Constant, unilateral pain in the distribution of the ophthalmic division of the trigeminal nerve, sometimes extending to the maxillary division, accompanied by Horner’s syndrome and caused by a disorder in the middle cranial fossa or of the carotid artery.
• Recommend immediate MRI /MRV w/wo contrast and MRA of brain and neck, or CTA/CTV
Raeder’s syndrome

- Source: aao.org
For help distinguishing 1st/2nd from 3rd order sympathetic pathology, use 4-hydroxyamphetamine drops

If third order is intact, 4-hydroxyamphetamine will cause neurotransmitter release and pupil dilation

If injured, there will be no dilation because the third order neuron releasing norepinephrine is damaged.
Burning mouth syndrome

- An intraoral burning or dysaesthetic sensation, recurring daily for more than 2 hours per day over more than 3 months, without clinically evident causative lesions.
- Rule out nutritional deficiency, xerostomia, HSV, allergic contact stomatitis.
- Otherwise considered idiopathic. Small fiber trigeminal neuropathy
- Usually start with TCAs, gabapentin. Systematic review also found alpha-lipoic acid (3 trials), klonopin (1 trial), CBT (1 trial) helped.

Atypical Facial Pain or Persistent Idiopathic Facial Pain

- Often used as a catch-all for facial pain not falling into previous criteria. Often lumped in with central-post stroke pain and central neuropathic pain attributed to MS.
- ICHD-3:
  - A. Facial and/or oral pain fulfilling criteria B and C
  - B. Recurring daily for >2 hours per day for >3 months
  - C. Pain has both of the following characteristics:
    - 1. poorly localized, and not following the distribution of a peripheral nerve
    - 2. dull, aching or nagging quality
  - D. Clinical neurological examination is normal
  - E. A dental cause has been excluded by appropriate investigations
Atypical Facial Pain or Persistent Idiopathic Facial Pain

- Very little quality evidence for any particularly interventions.
- Four RCTs have been done in persistent idiopathic facial pain: venlafaxine, fluoxetine, sumatriptan subQ, dothiepin (not available in U.S.)
- Harrison et. al found additional benefit with CBT in study with fluoxetine.

Atypical odontalgia

- Subform of persistent idiopathic facial pain
- Continuous toothache without any dental pathology
- Recent study cited 4 cases of efficacy of different periods, at different sites, of botulinum toxin A for atypical odontalgia.

Central post-stroke pain

- Depressingly, a systematic review found no conclusive benefit for any interventions in 8 eligible RCTs.
- Anticonvulsants, TCAs, naloxone, acupuncture
- While often these therapies and also SNRIs and mexilitene, among others, are used in various types of atypical facial pain, there is very little evidence supporting their efficacy as of yet.

Others

- Optic neuritis
- Headache attributed to ischemic ocular motor nerve palsy
- Recurrent painful ophthalmoplegic neuropathy (formerly ophthalmoplegic migraine) – treat with steroids. MRI enhancement.
Thank You!