Trigeminal Neuralgia
and
Endoscopic Microvascular Decompression Surgery

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Trigeminal Neuralgia

The Suicide Disease
Trigeminal Neuralgia

• A rare clinical entity characterized by severe, sudden, brief, mostly unilateral, stabbing pain in the distribution of the 5th cranial nerve
Trigeminal Neuralgia

- Condition of hyperexcitation of the trigeminal nerve causing:
  - Severe stabbing or lancinating pain
  - Triggered by light touch, chewing, brushing teeth, wind
  - Typically responsive to carbamazepine or other anti-epileptic agents
- Differential diagnosis includes:
  - Multiple Sclerosis
  - Cerebello-pontine angle tumor
  - Herpes Zoster
  - Dental caries
History

• Aretaeus of Cappadocia
  – Rome and Alexandria
  – 2\textsuperscript{nd} Century AD
  – Headache with “spasm and distortion of the countenance”
History

- Jurjani (Zayn al-Din Isma‘il ibn al-Husayn)
- 11th Century physician
- Persian province of Khvarazm
- “type of pain which affects the teeth on one side and the whole of the jaw on the side which is painful”
History

• Wells Cathedral
  – Somerset, England
  – The Tomb of Bishop Button, 1274
  – The toothache figure
History

• John Locke
• 17th Century Philosopher and Physician
• Described the facial pain of the Countess of Northumberland, wife of the English Ambassador to France in 1677
History

• “...I found in a fit of such violent and exquisite torment... it forced her to such cries and shrieks as you would expect from one upon the rack... her mouth was constantly drawn on the right side towards the right eare... These violent fits terminated on a suddaine, and then my Lady seemed to be perfectly well... there was not the least appearance of any alteration in her face... speaking was apt to put her into these fits... or touching her gums”
History

• Nicholas Andre
• Tic douloureux
• 1756
• Observations pratiques sur les maladies de l’urethre et sur faits convulsifs
History

• James Ewing Mears
• Study of the pathological changes occurring in Trifacial Neuralgia. *Medical News of Philadelphia* 1884
• First suggested Gasserian ganglionectomy as treatment
History

• Hartel
• 1914
• First detailed account of injecting (procaine) into Meckel’s cave percutaneously
History

• Walter Dandy
• 1934
• While performing partial sectioning of the trigeminal sensory root first described the presence of vascular compression of nerve root
• 50% of his cohort
History

- Gardner and Miklos
- First described decompressing vascular contact with sponge pledget
- 1959
History

- Peter Jannetta
- University of Pittsburgh
- Defined modern technique of MVD
- 1967
History

- Lars Leksell
- Stereotactic Radiosurgery
- 1971

"The tools used by the surgeons must be adapted to the tasks, and where the human brain is concerned they cannot be too refined."

-Lars Leksell, M.D., Ph.D.
Gamma Knife Developer, 1971
Epidemiology

• Annual Incidence 4-5 in 100,000
• Age: > 50 (avg 63)
• Female to male 1.8-1
• Laterality:
  – Right: 60%
  – Left: 39%
  – Both: 1%
Epidemiology

• Division:
  – V1 – 2%
  – V2 – 20%
  – V3 – 17%
  – V1 & V2 – 14%
  – V2 & V3 – 42%
  – all 3 – 5%
Pathophysiology

• Focal demyelination of root of trigeminal nerve

• At the level of the junction between central myelin (from oligodendroglial cells) and peripheral myelin (from Schwann cells)

• Called the Obersteiner-Redlich Zone (ORZ); or Root Entry Zone (REZ)
Pathophysiology

• Ephaptic transmission in trigeminal nerve from large diameter myelinated A fibers to poorly myelinated A-delta and C (nociceptive) fibers

• May explain why innocuous stimulation of trigger zones within distribution of same trigeminal branch causes attack
Pathogenesis

• Vascular Compression
• MS
• Posterior fossa tumor
Pathophysiology

• Vascular compression of the trigeminal nerve at the REZ
  • Most commonly by SCA (80%)
  • Persistent primitive trigeminal artery
  • Note: vascular compression may be seen in up to 50% of autopsies in patients WITHOUT TGN
Pathophysiology

- Vascular Compression

Irritation of the trigeminal nerve root by neurovascular compression

Hyperactivity of trigeminal nerve nucleus
Pathophysiology

- MS: plaque within brainstem
  - poorly responsive to MVD
  - 2% of patients with MS have TGN
  - 18% of patients with bilateral TGN have MS
Pathophysiology

- Posterior fossa tumor
Diagnostic Criteria

• 1) Paroxysmal attacks (< 2 min)
• 2) Pain characteristics (at least 4)
  – Distribution along 1 or more divisions of CN V
  – Sudden, intense, sharp, superficial, stabbing, burning
  – Severe intensity
  – Trigger areas or activities (eating, talking, etc)
  – Asymptomatic between attacks
• 3) No neurologic deficit
• 4) Stereotyped in individual patient
• 5) Exclusive of other causes by history, physical

(adapted from International Headache Society)
Classification of Facial Pain

• The surgeon, however, is chiefly concerned with the question: “What cases of neuralgia are suited for operative treatment, and what are the best methods to employ?” The answer, obviously, should depend upon a scientific classification, based solely upon the causes of neuralgia; at present such a classification is impossible.

• J. Hutchinson (1905)
Classification

• Typical trigeminal neuralgia

• Atypical trigeminal neuralgia

• Atypical facial pain (AFP)
Classification

• TGN type I: episodic
• TGN type II: constant
• Trigeminal neuropathic pain: unintentional injury
• Trigeminal deafferentation pain: intentional injury (gangliolysis, or rhizotomy)
• Postherpetic neuralgia
• Symptomatic trigeminal neuralgia: MS
• Atypical facial pain: 2nd to somatoform disorder
  • Kim Burchiel, 2003
Progression of Trigeminal Neuralgia Over Time

Early in Disease Course
- Periods of Exacerbation
- Periods of Remission

Later in Disease Course
Differential Diagnosis

- Herpes Zoster – pain is not paroxysmal
- Dental disease
- Orbital disease
- Temporal arteritis – tenderness over STA
- Intracranial tumor – usually has sensory deficit
History and Physical

• Physical exam: should be normal in TGN – any deficit in previously un-operated patient should prompt search for tumor or other lesion
History and Physical

- Assess all divisions of CN V – including corneal reflexes
- Assess masseter function (bite) and pterygoid function (on opening mouth the chin deviates to the weak side)
Imaging

• Old teaching: imaging required only when atypical features are present

• Current wisdom: Advanced imaging may be able to identify, pre-operatively, vascular compression affecting the trigeminal nerve
Medical Management

- Carbamazepine (Tegretol): complete or acceptable relief in 69%
  - If 600-800 mg/d are tolerated and give no relief, the diagnosis of TGN should be questioned
- Lyrica
- Neurontin
Medical Management

- Dilantin
  - may use in IV form for patients in too much pain to open mouths for oral Tegretol
- Capsaicin (Zostrix)
  - Red Hot Chili Pepper
- Clonazepan (Klonopin)
  - Works in 25%
- Amytriptyline
Medical Management

• Status Trigeminus: a rare manifestation of TGN characterized by rapid succession of tic-like spasms triggered by seemingly any stimulus
  – IV Tegretol (if available) or IV Dilantin
Percutaneous Ablation

- Most employ the technique/landmarks initially described by Hartel

1) Beneath the medial aspect of the pupil

2) 3 cm anterior to the external auditory meatus

3) 2.5 cm lateral to the oral commissure
Percutaneous Ablation

• Finger intraorally and inferior to lateral pterygoid wing, 18 guage needle introduced at the point 2.5 cm lateral to oral commissure.

• Trajectory approximates intersection of coronal plane passing through the point 3cm anterior to tragus and sagittal plane passing through medial aspect of pupil.

• Lateral fluoroscopy used to direct needle into foramen ovale
Glycerol Rhizolysis

• Development of stereotactic technique to deliver gamma irradiation to the trigeminal ganglion

• Tantalum dust was injected into the trigeminal cistern using glycerol as a carrier

• Even without irradiation, patients had resolution of their pain
Glycerol Rhizolysis
Glycerol Rhizolysis

• Initial pain relief: 72-96%

• Recurrence rate: 10-92% (longer follow-up periods identify higher recurrence)

• The largest study of glycerol rhizolysis (N=522)
  – Recurrence at 2 years: 41%
  – Recurrence at 5 years: 83%
  – Recurrence at 6 years: 92%

• Median pain-free interval: 16-32 months
Glycerol Rhizolysis

• Complications:
  – Hypesthesia: 46-63%
  – Dysesthesia:
    • Minor: 10-15%
    • Major: 0-3%
  – Corneal anesthesia: 2-8%
  – Anesthesia dolorosa: 0-2.5%
Radiofrequency Thermocoagulation
Radiofrequency Thermocoagulation

• High rates of initial pain relief: 97-100%
• Recurrence rate: 13-37%
• Early studies showed a high rate of complications (dysesthesia and masseter weakness), and the procedure fell out of favor
• Subsequent modifications have reduced morbidity
Radiofrequency Thermocoagulation

- Using a curved-tip electrode in 500 patients treated with RT, only 9% of patients reported intermittent dysesthesia that did not require treatment.

- The largest series in the literature (N=1600) showed an overall recurrence rate of 25%:
  - 719 patients followed 5 years, recurrence: 43.3%
  - 365 patients followed 10 years, recurrence: 47.7%
  - Improved outcome with multiple procedures
  - After 5 yrs, 10 yrs, 92% and 94% reported absence of neuralgia with 1 or more RT procedures.
Radiofrequency Thermocoagulation

• Radiofrequency Thermocoagulation requires the cooperation of the patient

• This should be considered when deciding to proceed with this technique
Balloon Compression
Balloon Compression
Balloon Compression

• Initial relief: near 100%

• Recurrence rates: 10-33% (most recur in the 2nd year of follow up)

• Complications:
  – Dysesthesia: 4%
  – Bradycardia/hypotension: used by some as an indicator of penetration into the foramen ovale
    • Need for atropine administration rare
Stereotactic Radiosurgery

- Used to lesion CN V at the REZ
- Retrospective single institution reviews with little long term follow-up
- Patients described are typically older and have failed at least 1 other surgical intervention
Stereotactic Radiosurgery

• Largest study (N=220) from University of Pittsburgh
• Mean age 70 (29-92)
• >60% had undergone previous surgery
• >30% had a sensory disturbance
• Most treated with single 4mm isocenter with maximum dose of 70-80 Gy
• Median follow-up 2 years
Stereotactic Radiosurgery

- Median time to initial response: 2 months
- At 1 year complete or partial relief: 85%
- At 5 years complete or partial relief: 56%
- 10% experienced new facial numbness or paresthesias at 2 years (most complications observed within the first year)
Stereotactic Radiosurgery

• Another large cohort study (N=117)
• Similar patient profile
• Higher dose (4mm isocenter, 90 Gy)
• Complete pain free medication free:
  – 1 year: 57%
  – 3 years: 55%
• Slightly higher complication rate: 24%
Neurovascular Compression Syndromes

Irritation of the trigeminal nerve root by neurovascular compression

Hyperactivity of trigeminal nerve nucleus
Neurovascular Compression Syndromes

Pathophysiology

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Neurovascular Compression Syndromes

• **Trigeminal neuralgia**
  – “tic douloureux”
  – Superior cerebellar artery (SCA) typically culprit vessel
  – MVD 80-97% effective for typical TN

• **Hemifacial spasm**
  – Culprit vessels:
    • Anterior inferior cerebellar artery (AICA), Posterior inferior cerebellar artery (PICA), Verterbral artery, Venous structures
  – MVD 85-93% effective for classic HFS
Hemifacial spasm

• A syndrome of facial muscle spasms that typically begin with blepharospasm and progress to involve the lower facial muscles.

• Differential diagnosis for blepharospasm:
  ➢ Tardive dyskinesia
  ➢ Meige syndrome
  ➢ Facial Myokymia
  ➢ Bengin essential blepharospasm
  ➢ Dopamine agonists
Hemifacial spasm

• Etiology:
  ➢ Vascular compression of the facial nerve.
  ➢ Tumor compressing facial nerve
  ➢ Post-Bell’s palsy

• Treatment:
  ➢ Oral medications typically ineffective
  ➢ Can be managed temporarily with botulinum (BoTox) injections of facial muscles
  ➢ Definitive therapy: Microvascular decompression (MVD) of the facial nerve
    ▪ 85-96% efficacy
    ▪ Can take up to 3 years for full resolution
Hemifacial spasm

Neurovascular compression: primary culprit vessel, location, and severity*

<table>
<thead>
<tr>
<th>Vessel</th>
<th>Location</th>
<th>Severity</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>RExP</td>
<td>AS</td>
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<tr>
<td>AICA</td>
<td>4</td>
<td>26</td>
</tr>
<tr>
<td>PICA</td>
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<td>25</td>
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<tr>
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<tr>
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<td>3</td>
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<tr>
<td>total</td>
<td>12</td>
<td>74</td>
</tr>
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</table>
Glossopharyngeal neuralgia
- Culprit vessels: AICA or PICA
- MVD 90% effective

Disabling positional vertigo (DPV)
- Constant positional vertigo without hearing loss or vestibular dysfunction
- Associated with compression of the vestibulocochlear nerve
- Can respond to MVD
Neurovascular Compression Syndromes

- Geniculate neuralgia
  - “Hunt’s neuralgia”
  - Facial nerve neuralgia
    - paroxysmal otalgia
    - Prosopalgia (deep facial pain)
  - Can be caused by herpetic ganglionitis (Ramsay-Hunt Syndrome)
  - Can be associated with hemifacial spasm
    - “tic convulsif”
    - AICA typical culprit vessel
Intra-operative Considerations

• Positioning
• Anesthesia
• Neuromonitoring
• Microscopy
• Endoscopy
• Nerve decompression
• Closure
Beach Chair Position
Beach Chair Position
Teflon Back-table
Anesthesia

• Total IV Anesthesia (TIVA)
• Mannitol 25g
• Mild hyperventilation
• Dexamethasone – to prevent reaction to Teflon or glue
  – Continued for 12 days slow taper
• Antibiotics
• NO: Keppra necessary
Neuromonitoring

- Brainstem Auditory Evoked Response (BAER)
- Somatosensory Evoked Potentials (SSEP)
- Cranial nerve V and VII
1. Acoustic Nerve
2. Cochlear Nucleus
3. Superior Olivary Nucleus
4. Lateral Lemniscus
5. Inferior Colliculus
6. Medial Geniculate Body
7. Auditory Cortex (Heschl's Gyrus – Br: 41/42)
Keyhole Surgery

- Concept of accessing deep intracranial structures via small apertures
- Minimal bony and soft tissue disruption
- Minimal or no brain retraction
- Leads to fewer complications, shorter hospital length of stay and more direct lesion access
Microvascular Decompression
Endoscopic MVD
Post-operative Considerations

- Blood-pressure management
- Delayed Hydrocephalus Development
- Cerebrospinal Fluid Leakage
- Facial nerve weakness
- Hearing loss
- Failure to improve pain or spasms
Thank You

www.pacificneo.org
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