

Trigeminal Disorders

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SCHWANNOMA OF CN5 → see p. Onc62 >>

GRADENIGO syndrome – *apical petrositis* (osteomyelitis) with localized meningitis involving CN5 & CN6:

- 1) facial sensory loss
 - 2) facial pain (e.g. in temporal region), headache.
 - 3) abducens paralysis
 - 4) may also involve CN7 (facial palsy), CN8 (deafness)
- in children, following suppurative otitis media or mastoiditis.
 - pain worse at night, aggravated by jaw or ear movement.
 - multiple approaches to infected petrous cells are possible:
 - a) if it is complication of otitis media: *simple mastoidectomy* → air cell track containing granulation tissue can be followed into petrous apex and adequate drainage can be obtained.
 - b) *middle cranial fossa approach*.

ONION-SKIN PATTERN FACE ANESTHESIA – caused by *damage to spinal tract of trigeminal nerve* in high cervical region.

RAEDER PARATRIGEMINAL syndrome:

- 1) intense pain in CN5₁ distribution
 - 2) lacrimation, conjunctival injection, rhinorrhea
 - 3) ipsilateral mydriasis (postganglionic Horner's syndrome).
- idiopathic or *pathology of carotid sympathetic plexus* (near Meckel cave).
 - may not actually represent distinct clinical entity.

TRIGEMINAL NEURALGIA (TIC DOULOUREUX)

- paroxysmal disorder of excruciating, lancinating painful spasms.

- most common neuralgia!!!
- one of the most excruciating pain syndromes!!! (may drive sufferers to suicide)
- *Aretaeus of Cappadocia* - first indication of trigeminal neuralgia.
- first adequate clinical description - *Fothergill in 1773*.

EPIDEMIOLOGY

- INCIDENCE 4-5 / 100.000 population; PREVALENCE 155 / 1 mln.
- slight **female** predominance (3:2).
- INCIDENCE peaks in middle age (> 50% cases onset in sixth or seventh decade), but occasionally may affect children.

N.B. appearance in young patient - suspicion of *demyelinating disease!*

ETIOLOGY

- a) **SECONDARY** (intrinsic and extrinsic *tumors* near gasserian ganglia, *multiple sclerosis* plaques*, syringomyelia, infarction, aneurysm, cholesteatoma, basilar impression);
 - *2-8% patients have MS; 4% MS patients have TN
- b) **IDIOPATHIC**
 - N.B. most idiopathic cases are due to *pulsations of aberrant vascular loop* compressing root at its entry zone! – **NVC (neurovascular contact)**
 - most commonly - superior or anterior inferior cerebellar artery; less commonly – vein.
 - with aging, blood vessels can become ectatic and atherosclerotic.

PATHOLOGY-PATHOPHYSIOLOGY

- **BIOPSY** - *focal demyelination* but no inflammatory cells.
- **ephaptic (nonsynaptic) neurotransmission** between demyelinated trigeminal axons - physiological substrate for paroxysmal pain (esp. if initiated by cutaneous stimuli).
- frequently, ectopic impulses are generated in trigeminal nerve secondary to *vascular compression*.

CLINICAL FEATURES

- brief lightning-like series of jabs (spasms); jab lasts fraction of second, episode lasts seconds ÷ few minutes.
- pain is reported as: lancinating, stabbing, searing, burning, electrical.
- intensity is such that patient winces or grimaces (hence the name *tic douloureux*).
- **unilateral** (in ≈ 5% **bilateral***, but simultaneous bilateral spasms are quite atypical).
 - *most often in MS patients!
- strictly affects divisions of CN V (in 15% all three divisions):

$3^{\text{rd}} (70\%) > 2^{\text{nd}} > 1^{\text{st}} (5\%)$

 - vs. postherpetic neuralgia most frequently affects CNV₁
- pain occurs (throughout day and night):
 - a) *spontaneously*
 - b) *precipitated* by stimuli (cutaneous, auditory, even draft of air); often *temporal summation* of stimuli is necessary to invoke response.
- > 90% have demonstrable **trigger point** - small area (on cheek, lip, nose) that can reproduce pain when stimulated (by facial movement, chewing, touch).
- *between attacks, there are no symptoms*, but patient is anxious about having another attack.
- some patients are unable to chew, eat, drink, shave, or brush their teeth for fear of triggering spasm (patients may appear emaciated, males disheveled).
- *no neurologic deficits!!!* (*subjective hyper- / hypo-esthesias* over face may be reported).
- after paroxysm, there is relatively **refractory phase** (2- min) during which is it difficult to trigger attack.
- *psychological problems* may occur secondary to chronic pain.
- disease lasts indefinite years (**severity steadily increases** – pain intervals shorten, pain becomes atypically constant, medically intractable).

DIAGNOSIS

- diagnosis can usually be made by history alone.
- **MRI** is only test always indicated (even if there is no loss of sensation or other abnormality on neurological examination!) - identifying etiologies of **SECONDARY CASES!** (in 15% cases tumor is found!)

coronal 3D time of flight MRA is centered on vertebral-basilar system; collapsed MRA is then superimposed on routine spin echo T1 images, which show cisternal portions of fifth nerve – vascular loop compression is accurately identified.
- **trigeminal reflex testing** can be screening to identify **SECONDARY CASES** (*trigeminal sensory deficits* identify **SECONDARY CASES**, but poor specificity - absence of these deficits cannot rule out **SECONDARY CASES**).
- laboratory studies are normal.

TREATMENT

Medical therapy (many require lifelong medication!):

- 1) **CARBAMAZEPINE!!!** – **first-line & most effective medication**;
 - started gradually; max daily dose 1200 mg;
 - follow serum levels, liver function tests, and white blood cell counts to avoid toxicity.
 - dose may be tapered once pain is controlled, since remission may occur.
- 2) **OXCARBAZEPINE!!** – alternative.
- 3) **GABAPENTIN** – efficacious as carbamazepine but with profoundly fewer side effects!
- 4) **BACLOFEN**
- 5) **LAMOTRIGINE**
- 6) **PHENYTOIN**; intravenous **fosphenytoin** (250 mg) is useful for acute severe attack.
- 7) valproic acid, clonazepam, pimozide

Surgical therapy

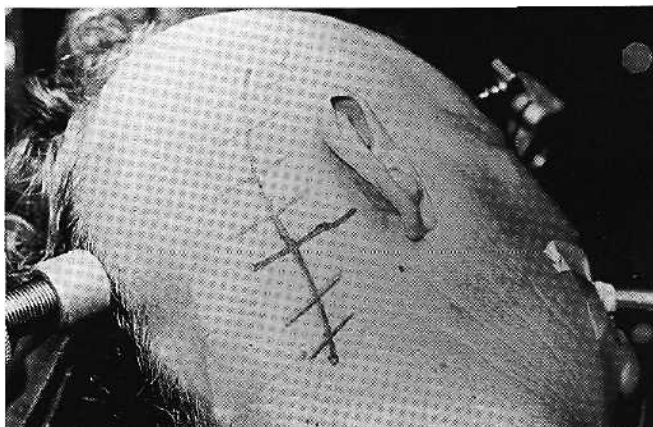
- 50% patients eventually have some kind of surgical procedure!
many experts believe that patients failing to respond to first-line therapy are unlikely to respond to alternative medications and suggest early surgical referral.

Distal procedures (neurectomies, nerve blocks) - generally not recommended - high incidence of early recurrence (indicated for patients with limited life span).

Proximal procedures:

Microvascular decompression (MVD), s. Jannetta procedure – classic, most effective procedure! (addresses etiology!)

- indicated for *younger, healthier patients* (without MS).
- **general endotracheal anesthesia**.
- semisitting position with neck flexed and face rotated away from side of surgery (ipsilateral tentorium parallel to floor).
- **lazy-S incision** (two finger breadths behind hairline, central incision third behind ear).
- retromastoid 3 x 3 cm craniectomy.
- dura is opened in cruciate fashion - lateral and superior surfaces of cerebellar hemisphere are exposed.
- cerebellum is retracted inferiorly and medially; petrosal vein is coagulated and divided.
- visual examination reveals loop of *superior or anterior inferior cerebellar artery* impinging on nerve at root entry zone; vessel is dissected away from nerve.
- pieces of shredded **Teflon sheeting** (or **polyvinyl alcohol foam**) are wrapped circumferentially around nerve at entry zone.
- anesthetist performs *Valsalva maneuver* - surgeon watches nerve and implant under microscope to be sure that there is no implant movement.
- **RESULTS**: no sensory deficits!*; pain relief is longer than after rhizotomy (even may be cured!); pain recurrence rate - 3.5% per year.
 - ***otovestibular testing** is performed in all patients prior to surgery because hearing loss is occasional complication of MVD.



Source of picture: Marshall B. Allen, Ross H. Miller "Essentials of Neurosurgery: a guide to clinical practice", 1995; McGraw-Hill, Inc.; ISBN-13: 978-0070011168 >>

Percutaneous trigeminal (retrogasserian) rhizotomy - lesioning trigeminal rootlets with:

- a) sterile anhydrous **GLYCEROL**
 - may give relief for many months without any significant neurological deficit;
 - onset may be delayed up to several weeks;
 - reserved for CNV₁ cases (because of corneal denervation risk with other methods).
 - b) **RADIOFREQUENCY (RF) CURRENT**
 - must produce **HYPOESTHESIA*** in pain distribution (if complete anesthesia - risk of postoperative anesthesia dolorosa);
 - electrical current supposedly **ablates small pain fibers** while preserving heavily myelinated touch and proprioception fibers!
 - lowest recurrence rates of all percutaneous procedures!
 - *in case of cancer pain **ANESTHESIA** must be attained to achieve adequate pain relief.
 - c) **BALLOON MICROCOMPRESSION** (6 minutes of compression)
 - highest recurrence rates;
 - lowest risk of corneal anesthesia; highest risk of hearing loss
- limited **neuroleptic analgesia** (patient is easily arousable).
 - guide needle entry point - 2 cm lateral to mouth corner.
 - needle is aimed at inner aspect of ipsilateral pupil + at point 3 cm anterior to external ear canal at level of zygoma.
 - surgeon inserts index finger in patient's mouth (to palpate guide needle tip through mucosa as it is advanced, making certain it does not breach mucosa); thumbs and other index finger are used to advance needle.
 - when patient shows evidence of pain, anesthetists gives brief general anesthesia.
 - needle is advanced through *foramen ovale*; slowly advanced until slight release is felt (penetration through Gasserian ganglion into trigeminal cistern – there must be CSF flow).
 - **fluoroscopy** is used to check needle position; patient is recovered from anesthesia.
 - electrode position should be manipulated until *paresthesias (upon stimulation) are confined to distribution in which pain is located*.
 - **lesioning** is carried out:
 - a) **HEATING**; *continuous sensory testing is ideal* (but some patients need general anesthesia again due to strong pain produced).
 - b) **GLYCEROL** injection into trigeminal cistern with tuberculin syringe (maximum of 0.4 cm³); patient is sitting; before glycerol injection, needle position is confirmed by injection of contrast material.
 - **RESULTS**: after 5 years in ≈ 65% pain recurs.

Radiosurgery (gamma-knife) - least invasive procedure.

- single dose of 70-90 Gy to trigeminal root.

Bibliography for ch. "Cranial Neuropathies" → follow this [LINK >>](#)

Viktor's NotesSM for the Neurosurgery Resident
Please visit website at www.NeurosurgeryResident.net