Facial pain: imaging perspectives

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Objectives
- Review the spectrum of facial pain
- Highlight the imaging features of facial pain syndromes

Overview
- Neuralgia
- Cranial neuropathy
- Trigeminal autonomic cephalgias
- Painful ophthalmoplegia
- Pure facial pain
  - Sinusitis
  - TMJ
  - Odontogenic
  - Oral/salivary
  - Persistent idiopathic facial pain (atypical facial pain)

Trigeminal neuralgia
- Excruciating, sudden, stabbing, lancinating, burning, or aching pain involving the distribution of one or more trigeminal nerve divisions
- Paroxysmal, 2-30 seconds, pain free intervals of hours to months
- Trigger zones, possibly around mouth or nostril
- Can be stimulated by touching, washing, brushing teeth, shaving, chewing, talking, wind, or can be spontaneous
  - Cross-modal stimuli: bright light or loud noise
- Brief refractory periods of insensitivity to triggers
  - Can give the patient an opportunity to brush his teeth
- Does NOT refer to just any trigeminal distribution pain!!!
  - Often used so anyway
  - Radiologists must be on guard for this to not miss an oncologic neuropathy

Trigeminal neuralgia
- Prevalence 3-6 / 100,000
- 70% of patients >60 y.o. at onset
  - 10% <40 y.o.
- Absence of physical findings or slight sensory impairment
- Usually V2 or V3
  - V1 in ~5%, uncommon isolated V1
- Bilateral 3%
Trigeminal neuralgia

- Reconsider if
  - Pain involves ear, occiput, neck, chest
  - Background pain
    - Unless chronic TN or ‘pretrigeminal neuralgia’
      - preTN
        - Atypical pain, dull, continuous toothache
        - Longer lasting, several hours
        - Triggered by jaw movements or drinking
        - Evolves into typical neuralgia
  - Sensory impairment or motor weakness
    - Suggests trigeminal neuropathy
    - Bilaterality
    - Young age

Trigeminal neuralgia - etiology

- Compression
  - Vascular loop in 80-90% of trigeminal neuralgia cases
  - Aneurysm, AVM
  - Schwannoma, meningioma, epidermoid, osteoma, other
  - Even contralateral mass resulting in distortion and stretching
  - Can be due to an interposed vessel compressing the nerve
  - Schwannoma of CN VIII, other CN, or even CN V
  - Compression in the root entry zone
    - First “few” millimeters
    - CNS myelin

- Vascular loop compressing the nerve
  - Asymptomatic NVC is common in the population
  - At most, 1 in 400 will have TN
  - Superior cerebellar artery is the most common
  - Compression of the proximal nerve
    - OR 10.4 (CI 2.9-37.4, p<0.001)
  - Indentation or displacement by the artery
    - OR 4.3 (CI 1.6-11.9, p<0.05)

Trigeminal neuralgia - imaging

- 44 y.o. woman with 8 years right V3 trigeminal neuralgia, chose gamma knife
- 53 y.o. F, L V2>V3 TN, outside imaging, no NVC demonstrated, effectively treated with balloon compression
**Trigeminal neuralgia- etiology**
- Demyelinating disease
  - 1.9% of MS patients have TN
  - (vs 3-6 / 100,000 in general population)
  - 4% of TN patients have MS
    - (vs 30-40 / 100,000 in northern US)
  - Foci of nerve root demyelination and axonal juxtaposition in both
  - TN was the first MS symptom in 5 of 35 MS patients with TN
  - Vascular compression may still exist
    - “Excellent” MVD outcome in 7 of 15 MS patients
  - Young patients
    - 90% of trigeminal neuralgia patients present after 40 y.o.
    - Bilateral

**Trigeminal neuralgia- imaging**
- Vascular loop compressing the nerve
- Mass compressing, stretching, or distorting the nerve
  - Schwannoma, meningioma, epidermoid, other mass
- Demyelinating disease
  - Multiple sclerosis
  - Peripheral demyelination (Charcot-Marie-Tooth)
  - Leptomeningeal metastasis, amyloid
  - Pontine infarct or “angioma”
42 y.o. with "trigeminal neuralgia of the V3 mandibular distribution." Began with tongue pain; Augmentin and an antifungal didn’t help. Moved into face, shooting pain caused by touch. Skull base neurosurgeon also called it "TN," with very mild hearing loss. CN8 at surgery. Teflon felt placed between CNS and SCA. Path schwannoma.

57 y.o. F with severe, lancinating, electric-like pain, V3 distribution, some V2/V3 numbness. Meningioma, resected, improved facial pain.

65 y.o. woman with R V3 "classical" TN, epidermoid, doing well on oxcarbazepine.

63 y.o. with 4 yr hx left V1 and V2 trigeminal neuralgia, shooting pain triggered by eating, brushing teeth, shaving, touch, wind. Presumptive arachnoid cyst.

55 y.o. left V2 and V3 lancinating pain with multiple triggers. Proven epidermoid at surgery.

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**Trigeminal neuralgia- imaging**

- Diffusion tensor imaging
  - Reduced fractional anisotropy of symptomatic nerve in TN
  - Reduced fractional anisotropy of symptomatic nerve, as well as increased radial diffusivity, correlation between FA reduction and visual analog scale, and trend toward mean diffusivity

- But why do it?
  - This is what you expect to see in idiopathic TN/NVC
  - NVC still remains somewhat a diagnosis of exclusion
  - I believe we image to exclude demyelinating disease, cisternal and skull base masses, and HN cancer
  - Requires 3T that will degrade imaging of the lower face due to slower T1 relaxation, and increased susceptibility and phase artifacts, reducing sensitivity for primary HN malignancy

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Liu EJR 2013

Lutz Radiology 2011

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Trigeminal neuralgia - treatment
• Spontaneous remissions are typical
• Carbamazepine (Tegretol)
  • A first line treatment
  • Effective in nearly 90% of cases
• Gabapentin (Neurontin)
  • Second line treatment
  • Off-label use
  • Often used in combination

Trigeminal neuralgia - treatment
• Microvascular decompression
  • Immediate complete relief in 82%, partial in 16%
  • 70% pain-free without medication at 10 years

Trigeminal neuralgia - treatment
• Glycerol injection
• Balloon compression
• RF ablation
• Gamma knife
  • Initial pain relief in 94%, generally < 1 month
  • 83% pain free at last visit (> 12 months)
  • Only 28.6% pain-free at 10 years

Other neuralgias
• Glossopharyngeal neuralgia
  • Paroxysmal oropharyngeal throat pain, triggered by swallowing, chewing, talking, yawning, laughing, or coughing
  • Due to vascular compression of the glossopharyngeal nerve in most cases; also compression by masses, demyelinating disease, or other pontine lesions
• Geniculate neuralgia
  • Paroxysmal pain involving ear, EAC, soft palate; trigger zone posterior wall of EAC
  • Nervus intermedius
  • Parasympathetic innervation to the lacrimal and nasopalatine glands, SMG, SLG
  • Sensory innervation to tongue, nose, ear
  • Herpes zoster? Vascular compression entiology?
  • MVD, nervus intermedius sectioning
• Supraorbital neuralgia
  • Paroxysmal pain along the supraorbital notch and on the midline forehead
  • Tenderness of the nerve at the notch
  • Charlin’s and Sluder’s neuralgia
  • Pain in medial angle of eye, radiating to eyebrow and orbit, nose, or jaw
  • Lacrimation, conjunctival injection, nasal congestion, sneezing, skin redness of the forehead. Differential diagnosis: cluster headache

Trigeminal neuropathy
• Trigeminal neuralgia
  • No sensory impairment or motor weakness
  • Exception: Anesthesia dolorosa
  • Preserved axons, “gain of function”
  • Background pain uncommon
  • Unless chronic TN or “pretrigeminal neuralgia”
• Trigeminal neuropathy
  • Pain followed by numbness and weakness
  • Facial or introral numbness increases the likelihood of underlying pathology

Trigeminal neuropathy
• Trauma
• Vascular
• Connective tissue disease (neuropathy or neuralgiform), radiation
• Neoplastic
  • Meningoencephalitis, schwannoma
  • Leptomeningeal metastasis
  • Perineural metastasis
• Infectious
  • Leprosy, VZV, HSV, Lyme, syphilis, fungal
  • Bacterial neuralgia – history of otitis media, ear: burning, constant pain
• Degenerative
• Toxic-metabolic
• Congenital
• Idiopathic trigeminal neuralgia
  • Spinal cord or autoimmune
• Amyloidosis
• Idiopathic intracranial hypertension

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68 y.o. man with right tongue and mandibular numbness, "V3 dull aching pain"; hx RA, longstanding TMJ pain

26 y.o. F presenting with gradual onset of left pharyngeal pain progressing to left SM and neck pain if touched and granulation tissue with microabscess formation and chronic inflammation.

Chronic inflammatory pseudotumor responded well to prednisone.

B-cell lymphoma

62 y.o. woman with Type 1 DM and L facial pain, Aspergillus
60 y.o. male with “perhaps a one- to two-year history of intermittent sensory symptoms affecting the lower half of his face on the left side.”

**Trigeminal neuropathy**
- Trigeminal trophic syndrome
- Result of “compulsive self-manipulation following trigeminal neuropathy”
- Anesthesia, facial paresthesias, secondary facial ulcerations
- Nasal ala > forehead, palate, ear
- Latency of years possible
- Treatment
  - Conservative: counseling, gloves
  - Pharmacologic: carbamazepine, amitriptyline, gabapentin
  - Transcutaneous electric stimulation, cervical sympathectomy

**Facial pain**
- Trigeminal neuralgia is narrowly defined.
  - The radiologist needs to think about what is really causing the facial pain: know what you’re looking for and don’t get blindsided by cancer!
- Trigeminal MR protocol
  - Complete face protocol (including DWI—epidermoid)
  - Coronal fiesta/ciss of cisternal trigeminal nerves and Meckel’s caves
  - Ax FLAIR and postGd T1 of brain
  - (Ax postGd T1 IAC with fatsat)
  - Facial weakness