Craniofacial neuropathic pain syndromes in neurosurgery
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Outline
• Trigeminal neuralgia
• Supraorbital neuralgia
• Glossopharyngeal neuralgia
• Geniculate neuralgia
• Postherpetic neuralgia
• Cluster headaches
• Trigeminal autonomic cephalgias

Trigeminal neuralgia
Epidemiology
• Peak incidence - ages of 50 and 60 years
• Annual incidence in the UK - 27 cases per 100,000
• Female/male ratio – 1.8:1
Aetiology
• Vascular compression of trigeminal nerve (most commonly superior cerebellar artery at root entry zone)
• Tumours – esp. posterior fossa tumors (vestibular schwannoma)
• Multiple sclerosis – plaque within brainstem

https://www.drugs.com/cg/images/en3067038.jpg
Presentation – pain characteristics

<table>
<thead>
<tr>
<th>SOCRATES</th>
<th>Description</th>
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<tbody>
<tr>
<td>S - site</td>
<td>in distribution of one or more branches of trigeminal nerve, unilateral</td>
</tr>
<tr>
<td>O - onset</td>
<td>Sudden, frequency of onset ranges from a few to hundreds of attacks a day</td>
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<tr>
<td>C - character</td>
<td>stabbing, electric-shock like</td>
</tr>
<tr>
<td>R - radiation</td>
<td>Nil</td>
</tr>
<tr>
<td>A - association</td>
<td>tingling, numbness</td>
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<tr>
<td>T - time course</td>
<td>Recurrent, occurs in paroxysms (from seconds to minutes), periods of remission (months to years) - tend to shorten over time</td>
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<tr>
<td>E - exacerbating/relieving factors</td>
<td>touch to the face, eating, cold winds, vibrations, shaving, brushing teeth</td>
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</table>

3-5% of patients will have bilateral pains (rarely simultaneous onset of pain)

Neurological exam most commonly normal

Differential diagnosis

- Dental pathology
- Temporomandibular joint dysfunction
- Migraine
- Giant cell arteritis (temporal arteritis)
- Cluster headaches
- Postherpetic neuralgia after shingles

Investigations

- Clinical diagnosis
- MRI scan - to rule out other diagnoses/underlying pathology
  - Demyelination plaques that might indicate multiple sclerosis.
  - Intrinsic brain lesions in the thalamus or trigeminal brain stem pathways such as lacunar infarctions.
  - Cerebellopontine angle mass lesions such as tumour, epidermoid, dermoid, or arachnoid cyst, aneurysm, or arteriovenous malformation.

Management

- Conservative - education, triggers avoidance
- Medical - Carbamazepine, Oxcarbazepine, Baclofen, Gabapentin, Lamotrigine, Botulinum toxin
- Surgical
  - Rhizotomy – glycerol, compression, radiofrequency
  - Stereotactic radiosurgery – radiation injury to trigeminal nerve root
- Microvascular decompression
  - Trigeminal nerve root entry zone exploration via posterior fossa craniectomy and displacement of vessel impinging the nerve
  - 90% of patients obtain pain relief, with over 80% still pain-free at one year
  - Patients with MS not considered due to poor response rate
  - Complications - cerebrovascular event, CSF leaks, meningitis, seizures, deafness, death, Diplopia due to IVth, Vth, VIIIth nerve palsy, numbness, recurrence

**Supraorbital neuralgia**
- Affects supraorbital branch of frontal nerve (branch of ophthalmic division of trigeminal nerve)
- Presentation
  - Unilateral pain in distribution of supraorbital nerve
  - Chronic continuous/remitting-intermittent
- Causes
  - Primary (idiopathic)
  - Secondary – trauma, chronic pressure
- Treatment
  - Medical – Gabapentin, pregabalin, topical capsaicin,
  - Surgical – rhizotomy (alcohol, radiofrequency ablation), nerve decompression
Glossopharyngeal neuralgia

- **Presentation**
  - Severe pain in distribution of glossopharyngeal and vagus nerves (throat, base of tongue)
  - Radiates to ear/neck
  - Associated with salivation, coughing, hypotension, syncope, cardiac arrest
  - Triggered by swallowing, talking, chewing

- **Treatment**
  - Poor response to medical therapy due to pain severity
  - Microvascular decompression

Geniculate neuralgia

- Affects nervus intermedius
- **Presentation**
  - Unilateral paroxysmal otalgia
  - Radiates to pinna
  - Associated with burning sensation around ipsilateral eye and cheek, prosopalgia, salivation, bitter taste, tinnitus, vertigo
  - Triggered by cold, noise, swallowing
- **Investigations**
  - Audiometry
  - Electronystagmogram
  - MRI
  - Angiography (to exclude aneurysm)
- **Treatment**
  - Medical – carbamazepine, phenytoin, valproate
  - Surgical – microvascular decompression

Postherpetic neuralgia
- Pain syndrome when pain persists for >1month after vesicular cutaneous eruptions caused by herpes varicella zoster virus (VZV) have healed

Aetiology
- VZV lies dormant in dorsal root ganglia of spine/trigeminal ganglion
- When weakened immune system VZV erupts causing inflammatory changes within the nerve leading to fibrosis

Presentation
- Post active infection
- Constant burning and aching pain - Spontaneous/triggered (touch)
- Dermatomal distribution (thorax > trigeminal nerve > limbs)
- Can be relieved by constant pressure
- Associated paraesthesia

![Nervus Intermedius](http://instruct.uwo.ca/anatomy/530/faclcomp.gif)
Treatment of postherpetic neuralgia

- Medical – Gabapentin, Oxcarbazepine, Amitriptyline, Capsaicin, Lidocaine patch, Intrathecal steroids
- Surgical
  - Cordotomy - selectively injuring pain-conducting nerve pathways
  - Rhizotomy - injuring nerve roots
  - Neurectomy - resecting a nerve
  - Sympathectomy – resecting a sympathetic nerve/ganglion
  - DREZ - dorsal root entry zone lesioning
  - TENS - transcutaneous electrical nerve stimulation

https://newsatjama.files.wordpress.com/2012/04/04-23-12-shinglesjpg011211f1-blog.jpg
Cluster headache

Presentation
- Severe unilateral recurrent episodic pain lasting between 15 minutes to 3 hours distributed between the outer canthus of eye and hair line
- Associated with nausea, restlessness and ipsilateral cranial autonomic symptoms (lacrimation, rhinorrhea, ptosis, miosis, eyelid swelling, facial sweating, conjunctival injection)

Pathophysiology
- Abnormal activity of posterior inferior hypothalamic grey matter

Treatment
- Medical – triptans, inhalation of 100% oxygen, topical lignocaine, ergotamine, preventative treatments (verapamil, lithium)
- Surgical
  - Neurosurgical ablation (glycerol injection, radiofrequency, radiosurgery)
  - Neuromodulation (deep brain stimulation, occipital nerve stimulation)

https://s-media-cache-ak0.pinimg.com/564x/42/99/90/429990b0757ee4eb294c8ec60c8ee029.jpg
**Trigeminal autonomic cephalgias**

- Group of primary headache disorders characterised by unilateral trigeminal distribution pain that is associated with prominent ipsilateral cranial autonomic features (conjunctival injection, lacrimation, nasal congestion, rhinorrhea, ptosis, eyelid oedema)

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<tr>
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<th>Description/characteristic features</th>
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<td>Cluster headache</td>
<td>As described before with prominent autonomic features</td>
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<tr>
<td>Paroxysmal hemicrania</td>
<td>Unilateral brief excruciating headaches with cranial autonomic features, responds well to indomethacin</td>
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SUNCT syndrome (Short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing)

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<td>SUNCT syndrome (Short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing)</td>
<td>Brief duration of pain attacks (5-250 sec) in ophthalmic distribution of trigeminal nerve, associated with prominent conjunctival injection and lacrimation, only partial response to carbamazepine, lamotrigine - most common treatment</td>
</tr>
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**Summary**

- Trigeminal, Supraorbital, Glossopharyngeal, Geniculate and Postherpetic neuralgia are the most common craniofacial pain syndromes managed by neurosurgeons
- There are new surgical treatment options emerging to treat cluster headaches and other trigeminal autonomic cephalgias
- Discussed conditions present with characteristic pain with specific distribution
- Diagnosis is clinical but patients are often investigated to exclude other diagnoses
- Management include conservative, medical and surgical options
References
2. Trigeminal Neuralgia; National Institute of Neurological Disorders and Stroke, February 2011
3. Trigeminal Neuralgia; Online Mendelian Inheritance in Man (OMIM)