Introduction to Trigeminal Neuralgia

Thimayu Plangkoon JÖRNS
Objectives

- Historical background of the condition
- Epidemiology
- Clinical features
- Pathophysiology of TN
- Diagnostic criteria and differential diagnosis
- Clinical assessment of TN
Historical Background

11th century Arab physician Jurjani (1066 – 1136)’s book

“There is a type of pain which effects the teeth on one side and the whole of the jaw on the side which is painful. With the pain there is spasm of the face and severe anxiety..... The cause of spasm and anxiety is the proximity of the artery to the nerve.”

Nicolaus André (1756) described 5 cases tic douloureux (unbearably painful twitch)
Historical Background

John Fothergill (1773) – published a full account of *tic douloureux* and presented a paper to the medical society of London

*Fothergill’s disease*

(*Dolor Faciei Fothergillii*)

Pujol (1787) discussed the diagnostic confusion of *tic douloureux* and *pain of dental origin*
Historical Background

Charles Bell (1829) confirmed that trigeminal nerve (the V nerve) was identified as a distinct nerve from facial nerve.

Trigeminal Neuralgia (TN)
Epidemiology of TN

- TN is a rare condition
  (incidence = 4.5 / 100,000)
- The peak incidence is in 60-70 age group
- Women are more likely to get TN
- At increased risk of developing TN are patients with multiple sclerosis (MS is seen in 2-4 % of TN patients) and hypertension

Clinical features

- Character of pain
- Radiation
- Site
- Severity
- Onset of pain
- Provoking factors
- Relieving factors
## Character of pain

- **Sharp**, shooting, stabbing, unbearable, like lightning or an electric shock, cattle prod

- **McGill Pain Questionnaires (MPQ)**, 78 adjective words describing pain (Melzack, 1975)
Site and Radiation

- Distribution of trigeminal nerve
- Nearly always unilateral
- Total right 61%, Total left 36%, Total bilateral 3%
- V1 = 4%
- V2 = 17%
- V3 = 15%
- V1+V2 = 14%
- V2+V3 = 32%
- V1+V2+V3 = 17%
Likely sites of trigger TN pain
Severity

- TN pain can be suicidal
- The MPQ shows both sensory and affective elements are high
- VAS (Visual Analogue Scale)
Timing, duration and onset

- Paroxysmal attack and a pain-free (refractory) intervals between attacks, memorable first attack
- Pain of abrupt onset and equally abrupt termination
- Remission period
Differences between TN and neuropathic pain

- Paroxysmal with long pain free intervals
- No abnormal sensations
- No build up of pain, refractory periods
- Responds to surgical treatment
Provoking and relieving factors

**Provoking**
- Light touched provoked i.e. washing, shaving, smoking, talking, drinking, brushing teeth, making up
- Mechanical
- Thermal

**Relieving**
- Rest and relaxation
- Local anaesthetic agent
- Anticonvulsants (carbamazepine)
- Surgery
- Spontaneous remisssion
Why is the right diagnosis important?

- Proper managements and referrals
- Recruit patients into trials

“Words matter, because what you describe will make a different in treatment”
- Dr Kim Burchiel, chief of neurosurgery, Oregon Health Sciences University, Oregon, USA

“Before we start giving a lot of medicines, we ought to be sure of the diagnosis”
- Dr Joanna Zakrzewska, Oral Medicine, Barts and The London School of Medicine and Dentistry, London, UK
Trigeminal Neuralgia
IASP Classification (1994)

- Sudden, severe, brief, recurrent stabbing pain in the distribution of the trigeminal nerve (IASP, 1994)
  1. Trigeminal neuralgia (Tic Douloureux)
  2. Secondary trigeminal neuralgia from CNS lesions
  3. Secondary trigeminal neuralgia from facial trauma
Diagnostic Criteria of Trigeminal Neuralgia by the IHS (2004)

1. Classical trigeminal neuralgia
2. Symptomatic trigeminal neuralgia


**Diagnosis Criteria of TN (2004)**

**Classical trigeminal neuralgia**

The International Headache Society (IHS)

- Paroxysmal attacks of pain (fraction of a second to 2 minutes)
- Pain has at least one of the following characteristics
  - Intense, sharp, superficial or stabbing
  - Precipitated from trigger areas or by trigger factors
- Attacks are stereotyped
- No clinically evident neurological deficit
- Not attributed to another disorder

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**Diagnostic Criteria of TN (2004)**

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No clinically evident neurological deficit  
Not attributed to another disorder
Atypical Trigeminal Neuralgia (TN Type II)

- Sharp but also burning aching background pain
- Pain can persist over 2 minutes
- They tend to benefit less from interventional treatments

refractory pain free period
The Liverpool classification for TN

British Journal of Anaesthesia 87 (1) : 117-32 (2001)

The Liverpool criteria 2001 (T.J. Nurmikko and P.R. Eldridge, The Walton centre for neurology and neurosurgery, Liverpool, UK)

1. Typical TN
2. Atypical TN
3. Trigeminal neuropathy
A new classification for facial pain

Burchiel KJ. Neurosurgery 2003 Nov;53 (5): 1164-7

Burchiel classification

1. Trigeminal neuralgia type I
2. Trigeminal neuralgia type II
3. Trigeminal neuropathic pain
4. Trigeminal deafferentation pain
5. Symptomatic trigeminal neuralgia
6. Postherpetic neuralgia
7. Atypical facial pain
Burchiel’s classification

- **TN type 1**
  - Spontaneous onset, >50% episodic pain

- **TN type 2**
  - Spontaneous onset, <50% episodic pain

- **Trigeminal neuropathic pain**
  - Unintentional trigeminal injury / trauma

- **Trigeminal deafferentation pain**
  - Intentional deafferentation

- **Symptomatic TN**
  - Multiple sclerosis

- **Postherpetic neuralgia**
  - Trigeminal *Herpes Zoster* outbreak

- **Atypical facial pain**
  - Somatoform pain disorder
The Delphi study
(Jörns TP and Zakrzewska JM, 2005)

Classical trigeminal neuralgia
Atypical trigeminal neuralgia

- Sharp shooting
- Pain free period
- Sharp, shooting
- Dull
- Refractory pain free period
- Background dull, aching pain
The Delphi study: TN
The problem of terminology
(Jörns TP and Zakrzewska JM, 2005)

<table>
<thead>
<tr>
<th>Participants</th>
<th>Classical TN</th>
<th>Atypical TN</th>
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<tbody>
<tr>
<td>Neurosurgeon 1</td>
<td>TN type 1</td>
<td>TN type 2</td>
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<tr>
<td>Neurosurgeon 2</td>
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<tr>
<td>Neurosurgeon 3</td>
<td>Typical TN</td>
<td>Atypical TN</td>
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<td>Neurosurgeon 4</td>
<td>Idiopathic TN</td>
<td>Atypical TN</td>
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<td>Neurosurgeon 5</td>
<td>TN</td>
<td>TN with atypical features</td>
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<td>Neurosurgeon 6</td>
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<td>Atypical TN</td>
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<tr>
<td>Neurosurgeon 7</td>
<td>Typical TN</td>
<td>NA</td>
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<td>Neurosurgeon 8</td>
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<td>Atypical TN / Facial pain Syn.</td>
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What would the diagnosis be if

- pain is more nagging aching but there are episodes of sharp pain
- intermittent but can sometimes be constant
- pain same area but tends to go down the neck
- provoked by stress, fatigue
- pain in other parts of the body
Chronic idiopathic facial pain (Atypical facial pain)

- **Character**
  - Nagging, dull, throbbing, sharp, aching

- **Severity**
  - Varies, mild to severe

- **Site, radiation**
  - Unilateral, bilateral, no anatomical area

- **Duration, periodicity**
  - Intermittent/constant
Chronic idiopathic facial pain (Atypical facial pain)

- **Provoking factors**
  - Chewing, stress
- **Relieving factors**
  - Rest, relaxation
- **Associated factors**
  - Pain in other areas,
  - personality changes
  - life events

Associated conditions:
- headache
- neck pain
- tinnitus
- back pain
- itchy skin
- irritable bowel
- pelvic pain
Pathophysiology of TN

- **Symptomatic** - tumours, AVM (arteriovenous malformations), multiple sclerosis

- **Idiopathic** - this could be due to a variety of mechanisms and result in different presentations
The Ignition Theory


Pathophysiology of Trigeminal Neuralgia: The Ignition Hypothesis

*Marshall Devor, Ph.D.  #Ron Amir, Ph.D., and ?Z. Harry Rappaport, M.D.
*Department of Cell and Animal Biology, Institute of Life Sciences, Hebrew University of Jerusalem, Jerusalem, Israel;
#Department of Neurosurgery, Rabin Medical Center, Peah Tikva, Israel
The Ignition theory

Pain fibre

Ectopic signal

Light touch fibre

**REVIEW ARTICLE**

Trigeminal neuralgia
Pathology and pathogenesis

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**Trigeminal Neuralgia**

Normally myelinated fibres

Remyelinated fibres

Demyelinated fibres
Investigations

- Haematology (Hb, folate and serum B12)
- U & E, LFT
- Dental radiographs
- MRI posterior fossa – compression, MS
- CT to exclude tumours

[Image of MRI posterior fossa]
## Psychosocial and Quality of Life

- McGill Pain Questionnaire (MPQ)
- Brief Pain Inventory (BPI)
- Hospital and Anxiety Scale (HAD)

### Psychosocial and Quality of Life Indicators

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### Psychosocial and Quality of Life Questions

**McGill Pain Questionnaire (MPQ)**

Please circle your response or ask for help if you are having problems.

1. Please rate your pain by circling the one number that best describes your pain at its WORST in the past week.

2. Please rate your pain by circling the number that best describes your pain at its BEST in the past week.

3. Please rate your pain by circling the one number that best describes your pain on the average.

### Brief Pain Inventory

**Brief Pain Inventory**

1. Please circle the one number that best describes how much pain you have RIGHT NOW.

2. Please circle the one number that best describes how much pain you have HAD IN A WHOLE DAY.

3. Please circle the one number that best describes how much pain you have HAD IN A WHOLE WEEK.

4. Please circle the one number that best describes how much pain you have HAD IN A WHOLE MONTH.

### Hospital and Anxiety Scale (HAD)

**Hospital and Anxiety Scale (HAD)**

1. Please circle the one number that describes how much you feel VII.

2. Please circle the one number that describes how much you feel VIII.

3. Please circle the one number that describes how much you feel IX.

4. Please circle the one number that describes how much you feel X.
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- Mrs Barbara Kenway
- All TN. patients

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- ผศ. ทพญ. นุษยา ศิริเทพ
- คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น
- ภาควิชาวิทยาชีววิทยา คณะทันตแพทยศาสตร์ มหาวิทยาลัยขอนแก่น
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Thank you for your attentions