Cranial neuralgias

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13. Cranial neuralgias and central causes of facial pain

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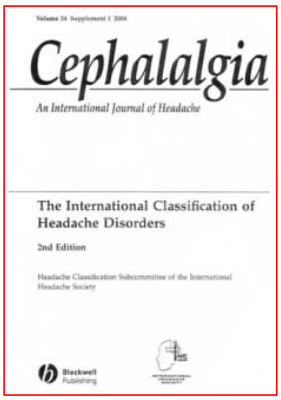
- 13.5 Nasociliary neuralgia
- 13.6 Supraorbital neuralgia
- 13.7 Other terminal branch neuralgias
- 13.8 Occipital neuralgia



13.1.1 Classical trigeminal neuralgia

- **Description:**
- **Trigeminal neuralgia is a unilateral disorder characterised by brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli including washing, shaving, smoking, talking and/or brushing the teeth (trigger factors) and frequently occurs spontaneously. Small areas in the nasolabial fold and/or chin may be particularly susceptible to the precipitation of pain (trigger areas). The pains usually remit for variable periods.**

13.1.1 Classical trigeminal neuralgia

- **Diagnostic criteria:**
- **A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C**
- **B. Pain has at least one of the following characteristics:**
  1. **intense, sharp, superficial or stabbing**
  2. **precipitated from trigger areas or by trigger factors**
- **C. Attacks are stereotyped in the individual patient**
- **D. There is no clinically evident neurological deficit**
- **E. Not attributed to another disorder**

13.1.2 Symptomatic trigeminal neuralgia

- **Description:**
- **Pain indistinguishable from 13.1.1 *Classical trigeminal neuralgia* but caused by a demonstrable structural lesion other than vascular compression.**

13.1.2 Symptomatic trigeminal neuralgia

- **Diagnostic criteria:**
- **A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, with or without persistence of aching between paroxysms, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C**

- **B. Pain has at least one of the following characteristics:**
  1. **intense, sharp, superficial or stabbing**
  2. **precipitated from trigger areas or by trigger factors**

- **C. Attacks are stereotyped in the individual patient**

- **D. A causative lesion, other than vascular compression, has been demonstrated by special investigations and/or posterior fossa exploration**

Trigeminal neuralgia - epidemiology

- not uncommon, with estimated incidence:
  4.5 cases per 100,000
- significantly more common with advancing age
- typical age of onset: 45 – 59 years
- nearly twice as common in women than men

Katusic S et al. 1990

Trigeminal neuralgia – pain localization

- Trigeminal neuralgia commonly afflicts the second or third divisions of trigeminal nerve
- Less commonly the combinations of V1/V2, V2/V3, or all 3 divisions
- Solitary involvement of the first division occurs in less than 5% of cases
- V1 and V3 involvement with sparing of V2 would be distinctly unusual and the diagnosis of trigeminal neuralgia should be in question

Barker FG II et al. 1996

Trigeminal neuralgia – pain lateralization

- The pain never crosses to the opposite side
- But, rarely it may occur bilaterally, in which case a central cause such as multiple sclerosis must be considered!

Trigeminal neuralgia – the pain attack

- sharp, stabbing, electric shock-like, lancinating
- can be so intense that the patient often winces in a tic-like fashion
- abrupt in onset and termination
- spontaneous or triggered (somatosensory stimuli within trigeminal area - specific "trigger zones" or somatosensory stimuli outside the trigeminal area, such as a limb or cross-modal sensory stimuli, such as a bright light or loud noise)
- lasting from a fraction of a second to 2 minutes
- usually is followed by refractory period during which pain cannot be triggered

Trigeminal neuralgia – between attacks

- patient is usually asymptomatic
- but, hypoesthesia and a dull background pain may persist
- there is often anxiety regarding the subsequent attack

Trigeminal neuralgia – dynamics

- attack frequency – from few to over a hundred daily
- pain attacks are characteristically absent during sleep
- symptoms often occur in bouts lasting weeks to months, and initially there are periods of spontaneous remission
- as the disease progresses, the pain-free intervals between attacks often grow shorter, and may eventually disappear

very brief headaches - differential diagnosis

	Paroxysmal hemicrania	SUNCT	Primary stabbing headache	Trigeminal neuralgia
M : F	1 : 3	1.3 : 1	1 : 2	1 : 2
Site of pain	V1	V1	V1	V1, V2, V3
Severity of pain	moderate to severe	severe	moderate to severe	severe
Attack duration	2 – 30 min	5 - 240 s	< 3 s	< 2 min
Daily frequency	> 5	3 - 200	sporadic	1 - > 100
Associated features	prominent	prominent	no	no except for V1
Trigger maneuvrs	no	yes / no	no	yes
Refractory period	no	no	no	yes
Indomethacin - responsive	yes	no	yes/no	no

Trigeminal neuralgia – more about it

- many, possibly most, patients with this condition have compression of the trigeminal root by tortuous or aberrant vessels
- classical trigeminal neuralgia is usually responsive, at least initially, to pharmacotherapy

Trigeminal neuralgia - treatment

EFNS GUIDELINES on the pharmacological treatment of neuropathic pain: 2010 revision AND *AAN-EFNS guidlines on trigeminal neuralgia management 2008*	Level of recommendation
Carbamazepine	A
Oxcarbazepine	B
Lamotrigine	C
Baclofen	C
Pimozide	C

A – effective, B – probably effective, C – possibly effective

Trigeminal neuralgia - treatment

- There is insufficient evidence to support or refute efficacy of:

- Gabapentin
- Clonazepam
- Phenytoin
- Tizanidine
- Topical capsaicin

Cruccu G et al. 2008 – AAN-EFNS guidlines on trigeminal neuralgia management

Case № 1

- ♂, 41, carpenter
- The first examination in Headache Center in December 2009
- The first headache – at the age of 31
- Past medical history – negative
- Social history – a heavy smoker, without alcohol or drug abuse, but taking alcohol provokes the pain
- Without relatives suffering from headache

Case № 1

- Attack frequency: 1 per 24h, during the day and night, without predomination
- Attacks occurs in periods – this is the 5th period in his life
- Pain localization: on the right, side-fixed, in the cheek area with radiation into the right temples
- Pain quality: stabbing
- Intensity: 9 (0-10)
- Lasting for 60 min
- Associated autonomic features: redness of the ipsilateral eye and ipsilateral rhinorrhoea

Case № 1

- Self-help with partial efficiency:
  - metamizole sodium, 2 x amp á 2,5 g/5 ml per os
  - cooling
- Examinations with normal findings:
  - neurological examination
  - blood testing
  - EEG

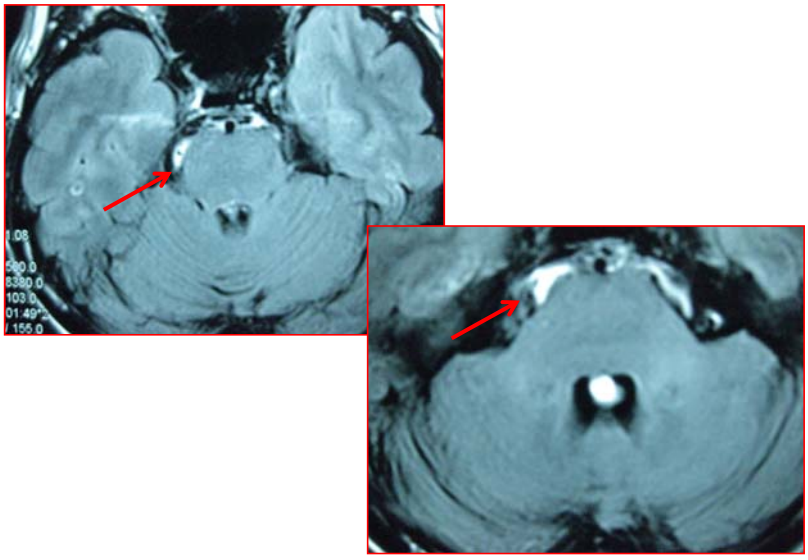
Case № 1

- Diagnosis?
- Sinus headache
- Cluster headache
- Trigeminal neuralgia

Case № 1

- **MRI :**
- **Neurovascular conflict - compression of the trigeminal root on the right**

Case № 1



Case № 1

- Prednisolone, 1 mg / kg /day, per os, during 5 days and than tapering of the dose
- During the prednisolone therapy:
- Gastroprotective therapy
- KCl, pulvis á 1g/day, per os
- On control examination - without headache

Case № 1

- Diagnosis?
- Sinus headache
- Cluster headache
- Trigeminal neuralgia

Case № 1

	Cluster headache	Trigeminal neuralgia
Attack frequency 1/day	+ (1 - 8)	+ (1 - >100)
Nocturnal attacks	+	-
Localization, cheek area, temporal	-/+	+
Pain quality - stabbing	-/+	+
Intensity -9 / 10	+	+
Attack duration – 60 min	+ (15 – 180 min)	- (< 2min)
Associated autonomic features	+	< 5%
No trigger maneuvers	+	-
Alcohol provokes attacks	+	-
Prednisolone responsive	+	-
MRI: neurovascular conflict	-	+

Case № 1 - Cluster-tic syndrome ?

- Cluster headache with coexistent trigeminal neuralgia
- Some patients have been described who have both:

  3.1 Cluster headache and 13.1 Trigeminal neuralgia
- they should receive both diagnoses
- The Trigeminal neuralgia elements of the attack occur in paroxysms of many seconds or minutes, always affect the maxillary or mandibular divisions of the trigeminal nerve, with spread into the ophthalmic division in some cases. These features may occur independent of Cluster headache elements but more often the two blend together.

- blood vessels were found to cross compress the trigeminal nerve

Solomon et al. 1985

Case № 2

- ♂, 68, retired engineer, married, father of two adult children
- Directed to the Headache Center by a dentist
- Past medical history – hypertension, regular therapy with metoprolol
- Without relatives suffering from headache

Zidverc-Trajković et al. Cephalalgia 2005

Case № 2

- Onset of symptoms – 6 months ago
- Since than, he experiences pain every day

- Attack frequency: 1-3/ 24 hours, exclusively during the day
- Pain localization: mandibular region on the right, right half of the tongue
- Pain quality: electric-shock like
- Intensity: 10/10
- Duration: 1 second
- Between attacks: sensation of "burned" skin in that area

Case № 2

- Examinations with normal findings:
  - neurological examination
  - maxillofacial examination
  - dental examination

Case № 2

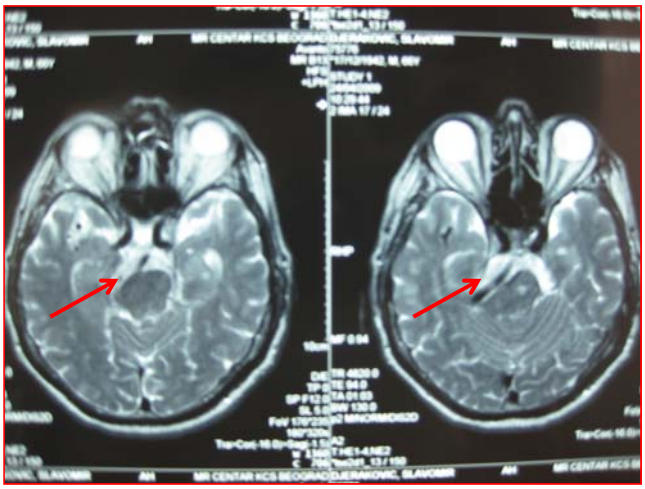
- Diagnosis?
- Burning mouth syndrome
- Tongue base tumor
- Trigeminal neuralgia V3

Case № 2

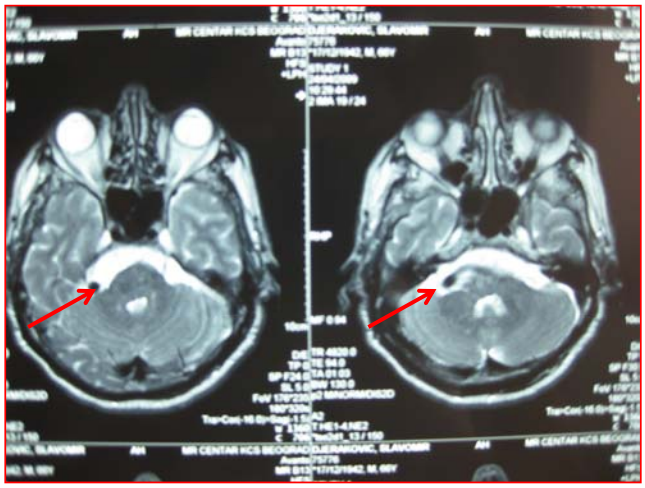
- MRI :
- Vertebrobasilar dolichoectasia with:

- compression of the neurovascular structures in the right pontocerebellar cistern
- compression of the right ventrolateral part of the pons
- neurovascular conflict with the right trigeminal nerve

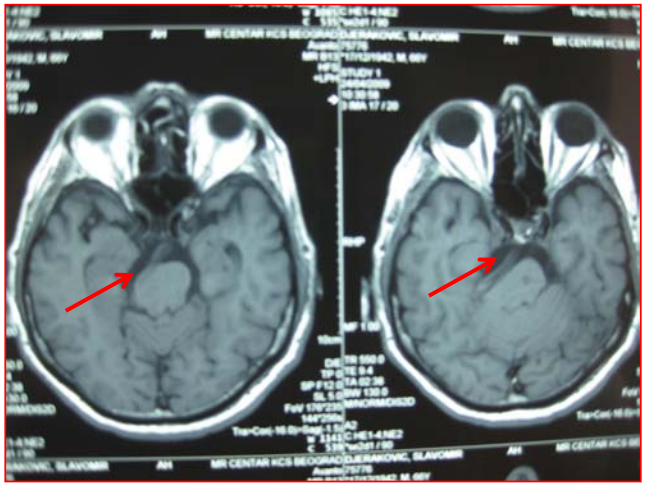
Case № 2



Case № 2



Case № 2



Case № 2

- Response on therapy
- Carbamazepine, 600 mg /day

- On control examination – pain-free

Case № 3

- ♂, 58, sales manager
- The first examination in Headache Center in November 2007

- Pain in the jaw started 6 months ago

- Past medical history – negative
- Social history – without tobacco, alcohol or drug abuse
- Without relatives suffering from headache

Case № 3

- Attack frequency: 10-15/ 24 hours, commonly during the night, always between 3 and 4h, awakening him from the sleep, but occasionally during the day

- Pain localization: maxillary region on the left
- Pain quality: stabbing
- Intensity: 9/(0 – 10)
- Duration: 4 - 5 seconds
- Between attacks his pain-free, but for only 10 minutes, and after that he experiences the next pain attack, and so on for dozen times

Case № 3

- trigger maneuvers and trigger zones: chewing, shaving, talking, touching in the left maxillary region

- without refractory period

- associated autonomic features: redness and lacrimation of the ipsilateral eye

Case № 3

- Diagnosis?
- SUNCT syndrome
- Paroxysmal hemicrania
- Trigeminal neuralgia

Case № 3

- Aspirin
- Diclofenac
- Paracetamol

- Carbamazepine 1000mg
- Gabapentin, 1200mg – partially effective

Case № 3

- ?
- Discordance between pain localization in V2 division of trigeminal nerve and occurrence of associated autonomic features, typical for the affection of the V1 division of trigeminal nerve

Case № 3

- MRI:
- Neurovascular conflict of the a. cerebelli superior and the root of trigeminal nerve on the left

Case № 3

- microvascular decompression (MVD) – performed in March 2008.
- First month after the MVD
  - the same pain is occurring only after the stimulation, on trigger maneuvers - chewing, shaving, talking, touching
  - without associated autonomic features
- Next 11 months – pain-free
- A year after the MVD - recurrent trigeminal neuralgia
  - The same pain is occurring on every movement of the jaw
  - Carbamazepine, 400mg, is efficient enough

Case № 3

- 14 months after the MVD
  - The same pain, irresponsive on Carbamazepine, 1800mg
- Gabapentin, 2100 mg, efficient enough only for 3 weeks
- Lamotrigin, 150mg, efficient enough since than till now

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