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The rare, unilateral headaches. Vågå study of headache epidemiology

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Abstract In the Vågå study of headache epidemiology, 1838 parishioners in the age group 18–65 years were included (88.6% of the relevant population). Each individual was questioned in a face-to-face situation. In this population, a search of rare unilateral headaches was also made, in spite of their presumed rarity. Trigeminal neuralgia was present in two cases. Two individuals with SUNCT traits were observed. Hemicrania continua may have been present in one individual. Also observed were: optic neuritis (n=1), herpes zoster (n=4); a case of unilateral headache upon neck rotation (chronic paroxysmal hemicrania variant? or "forme fruste" of the neck–tongue syndrome?); masseter muscle spasm (n=1); temporo-mandibular joint dislocation (n=1); and possible carotidynia (n=3). A particularly intriguing form of headache was a unilateral, neuralgiform (?) pain, associated with ipsilateral, regular jabs and allodynia, a combination observed in eight females. A couple of conditions that entirely defy rubrication are also reported.

Keywords Headache • Herpes zoster • Trigeminal neuralgia • CPH • SUNCT syndrome

Introduction

Unilateral headache (unilaterality without sideshift) is a special brand of headache [1]. Such headaches generally seem to have an organic origin [2–4]. Furthermore, they lend themselves to investigation, as the non-affected – or clinically relatively silent – side can be used as a control.

Cluster headache, the prototype of such a headache, has already been dealt with in the Vågå study; prevalence: 0.3%. Most headaches in this category are supposedly rare, with the exception of cervicogenic headache (CEH), which will be dealt with separately, and possibly with the exception of hemicrania continua (HC) [5], which has been claimed to be less rare than previously suspected [6]. Supraorbital neuralgia (prevalence Vågå 0.5% [7]) and neck-tongue syndrome (N-TS) [8, 9] (prevalence Vågå: 0.2%) have been treated separately, because of special problems attached to these headaches.

In the Vågå study of headache epidemiology, we also searched for the prevalence of rare, unilateral headaches.

Materials and methods

Details of the design of the Vågå study of headache epidemiology and of the demography of Vågå have been presented else-

where [10]. The material consisted of 1838 18–65-year-old citizens (F/M ratio: 1.05); mean age 35.9 years [11]. A questionnaire, also addressing rare, unilateral headaches, was in its entirety administered by the principal investigator (O.S.). A meticulous physical/neurological examination of the head, neck and face was included, and so was a short-version general neurological examination. If medically indicated, even neuroimaging could be performed.

This study was carried out several years prior to IHS II criteria of 2004. Therefore, the IHS I criteria [12] were adhered to whenever they covered a particular headache, e.g., headache in connection with optic neuritis and herpes zoster. For SUNCT [13], the description by IASP [14] was used. As for *carotidynia*, the diagnostic principles of Roseman [15, 16] and IHS were followed. According to Roseman [15], carotidynia is characterised by a unilateral, mostly moderate, throbbing head and neck pain, with self-limiting attacks. There is tenderness of a certain segment of the carotid artery – and a swelling. For the ultrashort headache episodes, the term *jabs* is preferred, as it indicates ≤1–3 second-long pain paroxysms [11], in contradistinction to "stabs", defined by the IHS as ≤1 second-long paroxysms [12]. *CEH* was diagnosed according to the CHISG criteria [17].

Hemicrania continua diagnosis

The diagnosis of hemicrania continua (HC) is mainly based on three features: (1) a pure hemicranial pain of mild to moderate intensity, without side-shift, (2) an absolute response to indomethacin, and (3) lack of effect of cluster headache drugs (ergotamine and triptans). Otherwise, HC is a symptom-poor disorder. There is ample evidence for a dual temporal pattern: intermittent and chronic. In this context, both jabs and female preponderance were free variables. The diagnostic principles laid down by the IASP [14] were adhered to.

Intensity of headache

Intensity of headache was graded according to a previously published scheme: 0–6.0+ [18]. "Features indicative of cervical abnormality" ("CF") were assessed on a scale 0–5.0+ [19].

Strikingly, many of the headaches herein were brought to the surface as a consequence of the routine questions: "Other headaches?" and "Headaches in other situations?"

Observations

Well known, or relatively well known, forms of headache

Trigeminal neuralgia

Trigeminal neuralgia was present in two citizens, one female, with a fairly typical picture, aged 42, and one male with a rather severe head trauma, aged 42 (prevalence: 0.16%). Both exhibited unilateral distribution of pain, along the branches I–III. Further details are given in Table 1.

Table 1 Trigeminal neuralgia (*n*=2)

Variable	Patient no.	
	I	II
Sex	F	M
Age, onset	39	36
Age, at present	42	42
Unilateral pain	+	+
Pain along branches I–III	+	+
Trigger points along branches I–III	+	+
Carbamazepin effect	+	(+)
Deficiency phenomena/neurological examination	–	+[a]

[a]Sensory deficits, whole trigeminal area, symptomatic side at present as well as previous neuropathological examinations. Also abducent n. palsy, symptomatic side, causing diplopia; and tinnitus. Head trauma at 36. Cerebral CT scan: negative

Herpes zoster and headache

The combination of unilateral herpes zoster and ipsilateral headache was present in four parishioners – two females and two males; age of onset: 30–53 years; prevalence 0.22%. The headache invariably was intense and circumscribed as regards time: the first weeks, chronic-fluctuating headache; later, an intermittent and relatively mild headache, before it stopped totally. In one, it was in the parieto-occipital area. In this parishioner, the headache was particularly intense ("as tooth ache": 5–6+ [18]) and throbbing, and there was exacerbation on coughing, anorexia and retching, and a weight loss of 7 kg. In the other ones, the pain corresponded to the first branch of the trigeminal nerve. In one case, an ophthalmic zoster with complications necessitated a corneal transplant some years afterwards.

Optic neuritis and retrobulbar neuritis

A 66-year-old female who had lost sight in her right eye at 36 was hospitalised for 6 weeks, and then gradually regained her vision during the first year. CSF was abnormal, with increased protein content. From the beginning of this episode, she developed an ipsilateral hemicrania and hemi-prosopalgia, at first chronic-fluctuating, and later episodic until it disappeared after 8–9 years. The peak headache was of 3–4+ intensity ("mild"/"moderate") [18]; it was symptom-poor, with no nausea and no pulsatile component of the pain. Otherwise she had been in good health. A "long-version", neurological examination

(at 66 years of age) showed minimal vision reduction and a whitish papilla, right side; otherwise neurological findings were unremarkable.

A 63-year-old female had had amblyopia of the left eye and pain in the ocular/periocular area and was diagnosed as retrobulbar neuritis at 20 years of age. Vision normalised within two years.

SUNCT-like headache

In two females, a state of low-frequency attacks and mild autonomic phenomena was present in the temporal area. Attacks could not be precipitated. In patient 1, a 40-year-old secretary, unilateral headache had been present for approximately 10 years. An initially recurrent pattern had recently become chronic, with 1–8 episodes/day of 1–2 min duration and of a mild degree (3+). Mild degree, unilateral lacrimation was present. Pain could spread to the symptomatic side shoulder area.

Patient 2 was 27 years old and had for the last 6 months had jabs (stabs) with jolts of 1–3 s duration in the left fronto-temporal area, appearing in volleys (with 5–6 jabs each), with intervals of weeks. Over the same period, several times per week, she also had pain episodes of 10–120 s duration in the same area as the jabs. During the latter attacks, the left eye was sore, and there was secretion from the left nostril "as clear as water", but no conjunctival injection or lacrimation. The jabs sometimes appeared in concert with this headache, but not always.

The female sex and low frequency of attacks count against a diagnosis of SUNCT (?). Jabs seem to be rare in SUNCT. A temporal location of pain is fully consistent with SUNCT [13, 14]. The closest one can come to classifying this case may be to term it: "SUNCT-like". No indomethacin test could be carried out.

Hemicrania continua

A total of 18 individuals (11 females and 7 males) exhibited a picture that *could* suggest HC: a hemicrania, mostly of a mild to moderate intensity. There were three cases of jabs. With one exception, the headache was intermittent. In the exceptional individual, a male aged 56, there was mild to moderate headache, without jabs. Indomethacin could, however, not be given to anyone, due to the compliance problems in Vågå: as alluded to previously in connection with the Vågå study, the inhabitants are generally sceptical to drugs. The highest attainable degree of diagnostic likelihood as regards a HC diagnosis, even in this particular patient, is "probable HC".

Temporo-mandibular joint (TMJ) dislocation

A 50-year-old female teacher had from 45 to 48 years of age had >40 episodes of unilateral TMJ dislocation. In connection with each episode, there was an ipsilateral, mostly moderate, decrescendo pain in the fronto-temporal area lasting hours. These attacks stopped entirely after she was taught how to reposition the jaw herself and employed dental appliances.

Carotidynia (IHS: 6.6.2.)

The principal investigator (O.S.) has been sceptical about this headache diagnosis throughout his professional life. There were three possible cases in the Vågå series: two females and one male. The following case is reported in detail to demonstrate the difficulties encountered in correct categorisation at the grassroots level. A 44-year-old mailman had, from the age of 30, had repetitive episodes of pain of a few days' duration in the right forehead area. Just prior to headache onset, he had had an automobile accident – and again at 38. The initially periodic headache eventually developed into a continuous, fluctuating headache. It was pressing, non-pulsating; at the peak of pain (=moderate), there could be photo- and phonophobia and nausea. There were no *local*, autonomic abnormalities. He was followed up for more than a year. At one time, there was, on physical examination, a soft nodule in the symptomatic side, mid-cervical area, approximately 0.75 cm in diameter, overlying the carotid artery and clearly tender on being pressed against the corresponding transverse process (the Fay sign [20]). The nodule disappeared within 2–3 weeks, and with it also the tenderness. It is doubtful whether the headache really was worse than usual during the "period of swelling". The right supraorbital nerve was occasionally tender. Local anaesthetic blockade of this nerve during an exacerbation led to a partial, transitory improvement. He also drank ca. 10 cups of coffee per day. He, nevertheless, had a *unilateral* headache. "CF" was at 1.0+ (mean, unassorted Vågå cohort: 0.79+; scale 0–5+) [19]. There was a slight reduction in movements in the neck in all directions. Neurological/ENT/ophthalmological examinations and CT scan of the brain were all negative. If one disregards the lump over the carotid artery, CEH could be a diagnostic alternative; also IHS migraine criteria were present to a weak degree. This case was categorised as carotidynia, with a big question mark attached. It might as well have been placed in a category: non-diagnosable headaches (see Discussion).

In two female migraineurs, 55 and 74 years old, respectively, a minor, bulging mass was present over one carotid

artery; their usual headache did not appear changed then. In the first one, palpation caused mild, brief nausea. Auscultation of the carotid area was invariably negative.

To put this situation into perspective, the principal author has seen two other cases (not during the Vågå study) with a similar nodule, combined with ipsilateral headache, of 2–4 weeks duration, in whom there conceivably could be a causal relationship between the two. One case has been summarily reported [21]. Such nodules were systematically searched for in the Vågå study.

Cerebrovascular disorders

In telegram style, it can be mentioned that there was one case each of chronic subdural haematoma (female, 54 years old) and intracerebral haematoma (male, 50 years old); the mainly unilateral headache in both was removed upon craniectomy.

Less well-known headaches

Unilateral headache upon head rotation (CPH variant?; "forme fruste" of neck-tongue syndrome? or: non-definable, unilateral headache?)
A 38-year-old female had, for approximately 6 years, had stereotyped attacks, appearing at intervals of two weeks to six months; they only appeared *immediately* upon neck rotation to the right, but not – by far – upon every rotation. The intense right-sided pain started in the neck and quickly spread to the ipsilateral temporal area. Due to the severity of the attack, she had to sit down quietly; over the course of 5–10 min the pain slowly decreased to zero. The pain was non-pulsating and was not accompanied by nausea or photo- or phonophobia, nor by any definite, local autonomic phenomena. Neck rotation was moderately reduced and particularly to the symptomatic side, i.e., by ca. 15°; otherwise, there were only moderately expressed neck signs ("CF": 1.0+, *vs*. a mean of 0.79+ [19]). Attacks could not be provoked iatrogenically. There had been no direct, or indirect, trauma to the neck.

This unilateral headache could be precipitated mechanically; it, in other words, fulfilled two cardinal criteria of CEH [17]. The precipitation mechanism, however, seemed to differ from that in "ordinary" CEH, where a *protracted* period of malpositioning (for instance neck rotation) usually is necessary to elicit an attack, the ensuing attack only gradually increasing in intensity. In the present case, an ultra-brief exposure sufficed, and the response was abrupt. The briefness and stereotypy of exposure/response are more reminiscent of a CPH variant with mechanical precipitation of attacks [21]. The exposure time is *usually* somewhat longer in CPH though, and the lack of macroscopical, autonomic phenomena is at variance with the typical CPH picture.

Most of all, the quick exposure and response resemble the pain provocation of N-TS attacks and in particular the putative, still not accepted [22], "forme fruste" of N-TS, where the tongue symptoms may not yet have developed. The pain part of N-TS attacks usually lasts only up to a minute or two. In the present case, pain intensity might be another distinguishing element, but not necessarily. The solitary episodes were too brief for attack intervention (indomethacin/anaesthetic blockades). The intervals between attacks were too long for continuous indomethacin medication to be justifiable. Although the clinical picture is distinct and clear-cut, diagnostic rubrication remains somewhat unclear. In theory, this could be a "missing link" between CEH and N-TS.

Mechanical (occipital) nerve irritation?
A 40-year-old man had at 32 and 39 years of age sustained indirect neck traumas. During the last one, there was probably an additional, mild, direct trauma against the occipital area; post-traumatically, there was a minimal, transitory headache. At 40 he had a 3-week period with a special symptomatology: an episodic, mild headache and a numbing feeling, starting in the neck and spreading to the vertex, probably more so towards the right side. These sensations only started when he was lying with the neck/occiput towards the pillow, and they disappeared almost immediately upon removing the head from the pillow. There was a continuous allodynia upon touch/combing in the vertex area during this period. "CF" was 0. A transitorily enlarged lymph node in the occipital nerve area could possibly explain this time-limited, weak, in all probability extracranial, headache.

Vague, unilateral neuralgiform headache with jabs
Not all unilateral headaches were easily interpretable. During the early phase of the study, a striking, non-definable, unilateral pain and jabs were united – *in space and time* in a couple of cases. Jabs were of particular interest during the Vågå study. Subsequently, a systematic search was made for similar cases. Additional traits would be free variables.

The basic, side-locked headache (n=8; prevalence: 0.4%) was mild to moderately intense; seemingly it most frequently occurred in the temporo-parietal and vertex areas, the duration varying largely, from a few weeks to 1–3 h (Tables 2 and 3). Jolts could be present with the jabs (Table 2). The headache started relatively early in life; mean age of onset: 29 years.

Table 2 Jabs "on top of" protracted, "neuralgiform" (n.) pain. Characteristics of the neuralgiform pain

Case	Sex	Age	Age at onset	Intensity*	Duration n. pain	Interval between bouts of n. pain
1	F	44	25	4–5	Few days	Months
2	F	27	Teens	2–3	1–3 h	Weeks–months
3	F	53	43	3	2–3 days	Weeks–months
4	F	37	20	3	1–3 weeks	2 weeks–3 months
5	F	33	31	4	1–2 days	Months–one year
6	F	37	30	3	1–2 days	Days
7	F	52	25	4	Few hours	Days
8	F	36	31	3	Hours–1 day	Days–weeks

Late ratio, continuous:remitting stage; 0:8 in present study. Previous review or hemicrania continua: 16:2 [24]
*On a 0–6+ scale [18]: 3="mild"; 4="moderate", 5="severe"

Table 3 Jabs "on top of" protracted, "neuralgiform" (?) pain

Case	Jabs duration (s)	Jabs per volley	Same localisation jabs and n. pain	Jolts	Vocalisation	CF[a]	Remarks
1	1–3	5–15	+	+	±	1	"Soreness" hair, feels "skin-less", symptomatic side
2	3–4	≤50	+	?	?	0	"Soreness" hair, painful area
3	1–3	≤4	+	+	–	1	"Soreness" hair, painful area
4	1–3	?	Probably	+	–	0	"Soreness" corresponding to I–III trigeminal branches[b]
5[c]	1–3	20–60	+	+	–	1	"Soreness" hair, symptomatic side, 4–5 cm wide area
6	1–3	20–30	+	+	–	1	"Soreness" hair (?)
7	1–3	–	+	+	±	1	"Soreness" hair, vertex
8	1–3	–	+	–	–	2.5	"Soreness" hair, vertex Ipsilateral, ear red, warm and more sore

n., neuralgiform
[a]*CF*, features indicating cervical abnormality; scale 0–5+; mean 0.79+ [19]; [b]Radiation of a feeling of "current and pins and needles" to the ipsilateral arm; [c]Jabs ~ weak

The invariable and striking symptomatic side "soreness of the hair" was or was not present outside headache episodes. There was thus a triad of symptoms in these cases: unilateral head pain, jabs and soreness of hair – in the painful area (Table 3). All were females, which was another free variable. Migrainous symptoms, like nausea and photophobia, apparently were non-existent. There was no particular tenderness over the supraorbital nerve and no forehead sensibility loss. In one patient (no. 8, Table 3) there was a red/hot ear.

It should be mentioned that patient 7, who through the years has tried "most drugs" without any effect at all, recently has tried the new drug, pregabalin, 300 mg/day, and that has apparently removed her pain entirely, both the neuralgiform pain and the jabs.

A 24-year-old male with a soreness in the right side of the face at 21 years of age was ultimately not included, not because of the sex, but because of the following: conjunctival injection could be present, but no lacrimation or nasal secretion. The facial pain was of 4–5+ grade intensity and made shaving problematic for 4–5 days in a row. It could spread to the back of the head. There were no signs of migraine or CEH. This headache/facial pain may not be classifiable.

It should be mentioned that in several cases there was neuralgiform head pain with allodynia in a female, but no jabs.

Cephalic jabs, with spreading of pain to shoulder/arm
In two cases there were unilateral volleys of "non-precipitable" jabs of regular duration and intensity in the ear-

temporo-parietal area. In both, there was a spreading of the jabs to the ipsilateral shoulder, and in one of them also diffusely into the peripheral part of the arm.

Masseter muscle spasm

A 52-year-old male had had paroxysmal, spasmodic episodes in the chewing musculature on the left side, from the age of 32. The episodes appeared 2–3 times per week and lasted ≥1–3 min. The first author just by chance had the opportunity to witness an apparently typical episode, with marked left masseter musculature contraction. There was no apparent concomitant contraction of facial musculature/platysma. The teeth were clenched together. There was no observable simultaneous contraction on the opposite side. There were no visible, accompanying, autonomic phenomena. He described it as being like "hard leg cramps", phenomenologically. The level of the pain was 4–5+. The pain was mainly in the masseter area, but it also spread to the temporal area, where there was mild (3+) pain. After an episode, he was tender in the masseter area, and, to a lesser degree, in the temporal area. There was no local atrophy/sensory loss.

Some attacks appeared to start spontaneously. However, most attacks started when chewing, in particular in the cold, e.g., during a small snack on a ski outing. He felt that mental, as well as physical, stress played a predisposing role. Occasionally, he could provoke an attack by clenching his teeth together. CF was 1.5+. This disorder seemed to be distinct from hemifacial spasm.

At follow-up, 8–9 years later, the situation had improved: month-long interval between attacks; the temporal area pain was presently just barely noticeable. A less stressful life situation might have contributed to the improvement.

Masseter spasm (masticatory spasm) is a rather well known, but rare disorder [23–25]. It is mostly bilateral; combined with hemifacial atrophy, if unilateral. The spasms are described as being shorter (few seconds; twitches) than in our case. Head pain has (for this reason?) not been remarkable in previous cases.

Lifting and occipital pain

In connection with heavy lifting at a younger age, a 65-year-old male had an abrupt and intense neck ache that spread to the occipital area and one shoulder. It reached a maximum within a minute or so and from there on a slowly decreasing curve started, reaching zero in approximately a day, never to return. Many years later, and with good health at that age, no other exploration should be carried out.

A particular case with marked allodynia

A 37-year-old female was remarkable because of the large number of regular jabs that she had experienced, i.e., probably >60 000 in the course of 10 years [26]. She also had a particular unilateral headache; the jabs always appeared in the stigmatised area and only during headache. The pain started just anterior to the right ear and thence spread to the face, ocular area, neck, ear, occipital and parietal areas (where the most pain was felt), throat, gum and probably the posterior part of the tongue. During symptomatic periods, there was "soreness" on combing the hair (allodynia), on brushing the teeth and on swallowing – all on the symptomatic side. Chewing and swallowing could not precipitate the pain. There were 10–20 pain episodes per year, each of 3–5 days duration; the intensity was 3–4+ (mild-moderate). At pain maximum, there was a pounding sensation in the right parietal area. Migrainous symptoms, like nausea and phonophobia, were lacking, and there was no familial migraine. Localised autonomic features, like lacrimation, were lacking.

This picture by far exceeds the borders of HC [5, 27] in its recurrent form. Pain distribution, lack of appropriate precipitation mechanisms, and jabs and allodynia outside the cranial nerve IX area all count against glossopharyngeal neuralgia. The exterior, cephalic distribution of pain may fit with that recently observed in cases of combined sensitivity to indomethacin/sumatriptan [28], but the oral/pharyngeal symptoms do not. The "external" and pharyngeal symptoms seem to belong to the same complex, as they coexisted temporally (the lack of forehead pain must also be fitted in!).

This case can hardly be categorised among long-lasting, neuralgiform attacks with superimposed regular jabs (see previously). We have probably not seen an identical case. This case may be rare, even unique. Unfortunately, we lack information about drug response.

Other cases that to some extent defy classification

Jabs and dizziness

A 21-year-old car mechanic had frequent migraine without aura (M-A) attacks. In a period of two years (from 17), he had occasional, 5–10-s long, jab-like paroxysms, starting in the neck and spreading to the ear/posterior temporal area, apparently with a left-sided predominance. A peculiar feature was that during these short-lasting paroxysms, dizziness overshadowed the pain part. There were, however, no visual disturbances/disturbances of depth vision, ocular pain or nausea, as described elsewhere, and the jabs were never protracted [29]. Usually, these episodes were non-synchronous with M-A. They could not be precipitated. These jabs differ from nuchal jabs, which do not spread upwards from the neck. EEG and clinical neurological examination showed no abnormali-

ties, and CF was 0 [19]. During the examination (asymptomatic period), there were no signs of overventilation. The present classification does not seem to present any obvious rubric where such a disorder can be fitted in.

Non-classifiable cases
In 21 cases (14 females and 7 males), among them the 17 HC cases that were relegated, the unilateral headache seemed non-classifiable with the available time and examination equipment. Other solitary non-classifiable cases are mentioned in some of the subsections herein and elsewhere [e.g., 29].

Discussion

Headaches like migraine with aura and well defined and frequent, but not that important, ones like jabs [11, 30–32] are relatively easily categorised. In a circumscribed population, like in Vågå, there will – after categorisation of the major, frequent and recognisable headaches – be a remainder of headaches that are not easily classifiable. How should this category be handled? Time is limited during examination; one can only try to penetrate in the exceptional cases.

In order to extract solitary cases or groups of patients from the remaining hard-to-diagnose, relatively contourless mass, we primarily divided such cases into bilateral and unilateral ones, which is probably a meaningful subgrouping. The bilateral ones contained relatively unknown headaches like "headache after moderate H_2S exposure", which has been dealt with elsewhere [33]. Herein, unilateral headaches, including relatively unknown ones, have been dealt with.

A note on the classification of SUNCT

As indicated by the two last letters in the acronym, conjunctival injection and tearing have a cardinal position in this picture. However, the autonomic phenomena in SUNCT are by no means limited to these two variables [34]: rhinorrhoea/sweating (on a subclinical scale) and dilated vessels, both just below the lower lid anterior limbus and between the upper lid ciliae and the superciliae were all mentioned in the first communication [34]. Tachypnoea, relative bradycardia, blood pressure increment and probably also eyelid oedema can be part of the picture.

Eyelid oedema needs a special account. Such oedema is difficult to assess during short-lasting pain paroxysms, as in SUNCT. We have even tried to quantify the oedema in periods with particularly frequent attacks by a method suggested by F. Lembeck (personal communication to O.S.) [see also 35]; vibramycin apparently has the right molecular size to penetrate vessel pores in oedematous states and is fluorescent. However, we did not succeed in our wholehearted attempts.

It has long been clear that not every SUNCT patient exhibits the whole panorama of autonomic disturbances [e.g., 36]. It would be strange if conjunctival injection and lacrimation were *sine qua nons*. Under SUNCT (3.3.; [37]), it is stated that "[attacks are] very often accompanied by prominent lacrimation and redness of the ipsilateral eye". It is, however, a long way between "very often" (an expression that we can totally agree with) and what is stated under A 3.3: "all patients must have both conjunctival injection and tearing". This is *not* a "must". We do not use such terms in such connections. And that a *must* should be implied in the term SUNCT is a tendentious interpretation.

Also, to base the SUNCT diagnosis on one solitary sign, and even a relatively rare one at that, e.g., oedema, means moving into an unknown, hazardous terrain. Oedema is a highly unspecific sign, also in headache; we have seen it even in CEH. If, on top of that, one allows a low number of attacks (e.g., 1/day), one is bound to err, and more than just occasionally. A term – and its contents – should not be tampered with.

Hemicrania continua. A note on its diagnosis

One has to be a purist when trying to purify a clinical picture, like HC. If an indomethacin test is negative, then a diagnosis other than HC must be found. If an indomethacin test has not been performed, then it is not allowed to even come in the neighbourhood of a diagnosis of HC. In this case, the diagnosis of HC has been made on an unacceptable foundation.

It is impossible to figure out the real prevalence of HC from the present data. In theory, it may be up to ca. 1%. Evidently, lowering the "diagnostic threshold" by taking away one major condition, i.e., the indomethacin demand, will automatically lead to a falsely high prevalence value.

HC was first described by Sjaastad and Spierings [5] in 1984. It has been claimed that HC was actually described prior to this under the term: "Cluster headache variant: Spectrum of a new headache syndrome responsive to indomethacin" [38]. Although it is termed indomethacin-responsive even in the heading, the article demonstrates that only 50% responded in an absolute way. The available space does not allow an analysis and critique of this item. A fairly complete overview has been given elsewhere [21].

CPH: a comment on its diagnosis

The term chronic paroxysmal hemicrania (CPH) signifies a unilateral headache, with multiple, relatively short-lasting attacks of extreme severity. As all (?) the unilateral headaches, it contains two temporal forms, a remitting and a non-remitting form, the latter CPH form apparently dominating in clinical settings. Both are indomethacin-responsive, in an absolute way [39]. In a particular subgroup of CPH, attacks can be precipitated mechanically [40]. A possible case in this category has been reported herein.

Unilateral "neuralgiform pain" associated with jabs

As for the *status* of this group, the following viewpoints may be advanced: Jabs occur in connection with many headaches. The jabs in this context seemed to be of the ordinary type. Jabs as such in Vågå have been dealt with under *ordinary jabs* [11]. *Prolonged jabs* (?), i.e., jabs with a duration up to 120 s, have been dealt with separately [29]. The present group makes up <1% of jabs in Vågå, where jabs were found in 35.2% of the population. It is only this special combination: "neuralgiform" pain/jabs that has been explored here. Does the starting point itself make this an artificial structure? There is little doubt that these premises determine the frame and contents of the group. Among the four ultimate elements: unilateral head pain, jabs, allodynia and the female sex, could one, as well, have started out with another duplex? Could one have started out with jabs and females? Jabs were present in 35.2% in Vågå, and, of these, 21% were females and 14.2% males. Such a design would, therefore, not have led to much of a selection process. If allodynia had been the only starting point, this would have led to an extremely heterogeneous group.

How can one possibly distinguish between this "neuralgiform pain associated with jabs" and HC [4, 24, 34] (which also is known occasionally to be associated with jabs)? Both are unilateral headaches, and both seem to have a female preponderance. Possibly, the following points may aid in distinguishing it from HC:

1. In spite of a protracted total duration, mean 12.5 years (case 5 is an exception) (Table 3), there were no signs of chronicity of the pain. In a series of HC, the continuous stage:remitting stage ratio in the late phase of HC was 16:2 (approximately 9.5 years after onset) [27], as against 0:8 in the present series. This argument has more than just limited weight.
2. Local, autonomic features were not present. In HC, autonomic features are not infrequently present [27].
3. *Pain maximum* was in the temporo-parietal area, as opposed to the fronto-ocular area in HC. This point carries only moderate weight.
4. The combination with allodynia is remarkable.

Any connection with HC must remain illusory, as *indomethacin* – a diagnostic cornerstone for HC – could not be tried. The information about the hair "soreness" was forwarded to us spontaneously. This fact combined with the relative rareness of allodynia in HC (?), may seem to carry considerable weight in the distinction between HC and the present attacks. Three cases of putative HC fulfilled the two first premises: (1) unilateral HA without side-shift, and (2) jabs. They also proved to be females. However, there was no allodynia. They were, therefore, not included.

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