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REVIEW

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Cluster headache and SUNCT: similarities and differences

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Abstract SUNCT is probably a distinct syndrome, although it shares some common features with cluster headache (CH): male sex preponderance, clustering of attacks, unilaterality of headache without sideshift, pain of non-pulsating type with its maximum in the periocular area, ipsilateral autonomic phenomena (e.g. conjunctival injection, lacrimation, rhinorrhea, increased forehead sweating), systemic blood pressure increment with heart rate decrement, blood flow velocity decrement in the middle cerebral artery, and hyperventilation. In spite of these similarities, SUNCT syndrome differs clearly from CH as regards a number of clinical variables, such as duration, intensity, frequency, and nocturnal preponderance of attacks. The two syndromes also differ markedly as regards precipitation of attacks, the usual age at onset, and efficacy of various treatment alternatives. Laboratory investigations have disclosed differences as regards presence or absence of Horner-like picture and possibly also the respiratory sinus arrhythmia pattern. All in all, these differences seem sufficiently ponderous to make it likely that SUNCT syndrome and CH differ essentially. SUNCT seems to be a "neuralgiform" headache, but different from trigeminal neuralgia.

Key words Cluster headache • SUNCT syndrome • Trigeminal neuralgia • Horner's syndrome • Autonomic nervous system dysfunction

Introduction

Our first case of SUNCT was found in 1978 [1], and a total of three cases were later published under the common name "shortlasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing" in 1989 [2]. Currently, SUNCT is defined by the International Association for the Study of Pain (IASP) as "Repetitive paroxysms of unilateral shortlasting pain, usually 15–120 seconds duration, mainly in the ocular and the periocular areas, of a neuralgiform nature and moderate to severe intensity, usually appearing only during daytime and accompanied by ipsilateral marked conjunctival injection, lacrimation, a low to moderate degree of rhinorrhea, and subclinical forehead sweating", and belongs to the group of neuralgias of the head and face [3]. Furthermore, the pain paroxysms accompanied by autonomic phenomena tend to appear in cluster periods, and there is a clear male preponderance in SUNCT [3]. Thus, SUNCT has even been considered to be in the same group as cluster headache (CH) [4, 5].

It was obvious from the beginning [1, 2] that the main differential diagnoses for SUNCT would be trigeminal neuralgia and CH, since SUNCT shared several features with both these headaches. While the differential diagnosis vs. trigeminal neuralgia has already been discussed [6, 7], such comparisons vs. CH have only been performed on a considerably smaller number of patients [2, 8]. Therefore, since we now are aware of >30 SUNCT cases, we would like to present such a comparison with CH.

The diagnosis of CH is based on five main criteria [9]: (1) the male sex, (2) unilaterality of the pain, (3) cluster phenomenon, (4) ipsilateral autonomic phenomena, and (5) excruciating severity of the pain. Thus, only one cardinal point of CH diagnosis, that of excruciating severity, seems to be missing in SUNCT syndrome. Nevertheless, in some CH patients, the headache may be mild [10] and, in some SUNCT patients, the headache may at times be relatively severe [11].

In spite of several features in common, SUNCT differs from CH in a number of ways, including temporal pattern, severity and treatment.

Similarities

The similarities between CH and SUNCT are summarized in Table 1.

Clinical variables

Male sex preponderance is typical for both CH and SUNCT. Analysis of 18 different studies [9] showed that 84% of CH sufferers on average were men. In fact, the percentage of men with SUNCT is so far similar (81%) [7, 12]. The male preponderance by no means implies that typical SUNCT cases cannot be found among women [13].

Strict unilaterality of the pain (i.e. without sideshift of headache) is also typical for both CH and SUNCT. In Manzoni et al.'s study [14], 84% of 180 CH patients experienced only unilateral headaches without sideshift. Until now, only two patients [2, 6] – in the late stage – developed bilateral headaches. In these two exceptional patients, pain was unilateral at the onset and is still preponderant on that side, as are the accompanying autonomic phenomena. Thus, at present, the frequency of strict unilaterality of the pain in SUNCT patients may be estimated as >90%.

Pain of a non-pulsating type with its maximum in the periocular area is another common feature of CH and SUNCT [3, 15]. Whether the pain in CH is "deeper" than that in SUNCT is uncertain.

Clustering of attacks, i.e. the occurrence of attacks within a relatively limited time-span, which led to the commonly used appellation cluster headache, is actually not an exclusive feature of this headache, but is also quite typical for other unilateral headaches [16], including SUNCT.

Table 1 Similar between cluster headache and SUNCT syndrome

	Cluster headache	SUNCT syndrome
Male sex	84%	81%
Unilaterality without sideshift	84%	>90%
Pain maximum in the periocular area	+	+
Pain of non-pulsating type	+	+
Clustering of attacks	+	+
Ipsilateral autonomic phenomena during attacks:		
Conjunctival injection	+	++
Lacrimation	+	++
Increased forehead sweating	+	+
Rhinorrhea/nasal stenosis	+	+
IOP/CIP amplitude increments	+	++
Corneal temperature increment	+	+
Systemic blood pressure increment and heart rate decrement during attacks	+	+
Blood flow decrement in MCA during attacks	+	+
Hyperventilation during (and outside?) attacks	+	+
Pathological findings with orbital phlebography	61%[a]	≥80%

[a] See text as for reservations
IOP/CIP, intraocular pressure/corneal indentation pulse

Ipsilateral, localized autonomic phenomena during attacks are obligatory for the diagnosis of CH as well as SUNCT [3, 15]. Already in the first, world-wide accepted description of CH [17], the following ipsilateral phenomena were reported during attack: lacrimation, conjunctival injection, rhinorrhea, nasal stuffiness, facial flushing, dilatation of vessels in the temporal region, and increased facial temperature (+1°–3° C). Later, it has been shown that CH attacks also are associated with ipsilateral increments in forehead sweating [18, 19], intraocular pressure (IOP) [20, 21], corneal indentation pulse (CIP) amplitudes [20, 21], and corneal temperature [20].

Since ipsilateral, autonomic phenomena are obligatory for the diagnosis of SUNCT [3], they have been present in all described cases [1, 2, 5, 11, 13, 22–29]. It seems that ipsilateral lacrimation and conjunctival injection are the most common features accompanying SUNCT attacks, but rhinorrhea/nasal stenosis are also quite common [7]. In addition, laboratory studies have revealed increased forehead sweating [30], as well as relative increment of IOP and CIP amplitudes, and periocular skin/corneal temperature [31] during SUNCT paroxysms, all on the symptomatic side. Thus, virtually all localized, ipsilateral autonomic phenomena accompanying the attacks are common for CH and SUNCT syndromes. There are some differences in the intensity of particular autonomic phenomena between CH and SUNCT, e.g. it seems that attack-related IOP increment is clearly more marked in SUNCT than in CH. They are by no means pronounced and/or regular enough to serve as a basis for differentiation between these two headache types in the single case.

Laboratory investigations

Systemic blood pressure increments with heart rate decrements have been described during spontaneous [32–35] and nitroglycerine-provoked [36, 37] CH attacks. While there is still little information on blood pressure changes, especially from studies applying beat-to-beat technique [34], a relative bradycardia during spontaneous attacks is rather well documented in CH [20, 32–35, 38–42]. Russell and Storstein's detailed study indicated that the heart rate on average increases at the attack onset, decreases during the middle part of the attack, only to increase again while the attack is terminating [42]. In addition, studies utilizing beat-to-beat techniques, on a limited number of CH patients, have revealed increased blood pressure and increased heart rate variability during spontaneous [34] and provoked [37] attacks.

A similar pattern of heart rate and blood pressure changes has been observed during SUNCT attacks [43]. However, in SUNCT, there was a lack of increased variability in heart rate and blood pressure [43] in contradistinction to what was the case in CH. In CH, heart rate variability was partially linked to sudden decrements in heart rate, occurring several times during an attack, described "as though gradually applying a brake" [34]. The same type of change could be observed in SUNCT only once during each (short-lasting) attack [43]. The discrepancy between SUNCT and CH may, thus, partially be due to the different duration of attacks.

Decrease of blood flow velocities in middle cerebral arteries (MCA), estimated with transcranial Doppler ultrasound technique, has been reported during CH attacks [44–47]. Bilateral decrease of blood flow velocities in middle cerebral arteries has also been described during SUNCT attacks [48]; the numbers of examined patients and recorded attacks are nevertheless limited.

Hyperventilation during attacks is still another common, though not readily explicable, feature of both CH [9, 49] and SUNCT [2, 50]. In addition, it seems that there is a tendency towards hyperventilation in basal conditions (i.e. outside attacks, both inside and outside bouts of attacks), when both headache types are compared with controls [50–52]. However, virtually no signs of peripheral chemoreceptor hypersensitivity have been demonstrated in either of these two headache types [50–52].

Pathological findings with orbital phlebography were first described in the Tolosa-unt syndrome [53], as far as headache research is concerned. Later, such findings were confirmed in a larger number of Tolosa-Hunt patients [54–56]. The pathological findings included narrowing/calibre variations of the superior ophthalmic vein and partial occlusion of the cavernous sinus. However, similar findings were also described in CH [57, 58] and SUNCT [11, 59]. In fact, when the frequency of pathological findings in series of non-medicated patients with Tolosa-Hunt syndrome [55, 56], episodic CH [58] and SUNCT [59] were compared, the percentages of pathological results amounted to around 70%, 60% and 80%, respectively. Thus, the percentage of abnormal phlebograms seemed to be at least as high in SUNCT as in CH. However, a blinded study in different headache types revealed that such orbital phlebography findings are non-specific, being present not only in CH but also in migraine, tension headache, and cervicogenic headache patients [60]. As a matter of fact, 10 of 12 CH patients had normal phlebograms in the latter study [60]. Interestingly, the same expert evaluated blindly 3 out of 4 phlebograms of SUNCT patients as pathological [59]. Thus, it might have been more appropriate to place the discussion of orbital phlebography in SUNCT and CH among "Differences".

Differences

The differences between CH and SUNCT are summarized in Table 2. It seems that in spite of several similarities, there are many striking differences distinguishing these two headache types.

Table 2 Differences between cluster headache and SUNCT syndrome

	Cluster headache	SUNCT syndrome
Usual age at onset, years	< 40	> 40
Duration of attacks	15–180 min	10–120 s
Severity of attacks (0 – +++++ scale)	+++++	++ – +++
Frequency of attacks, n/day	<1–3[a]	28 (6–77)[b]
Nocturnal preponderance of attacks	++	–
Mechanical precipitation of attacks	–	+
Precipitation of attacks by alcohol	+	– (?)
Therapeutic effect of ergotamine, dihydroergotamine, sumatriptan, prednisone, methysergide, lithium, verapamil	+	–
"Horner-like" picture		
Miosis, ptosis, decreased forehead sweating	+	–
Anisocoria following topical administration of hydroxyamphetamine and phenylephrine	+	–
Forehead sweating asymmetries during heating and pilocarpine tests (outside attacks)	+	–
Respiratory sinus arrhythmia outside attacks	≤ normal	≥ normal

[a] Up to 8 attacks per day

[b] Unweighted mean (range). Up to 30 paroxysms per hour

Clinical variables

Age at onset is typically under 40 years for CH [3, 15]. In the series studied by Manzoni et al. [14], approximately 80% of patients had their first attack in the second to fourth decades (mean age at onset, 28.9 years). In Ekbom's study, the mean age at onset was 27.5 years [61]. The mean of means from several different studies was 31.5 years [9]. This clearly contrasts with SUNCT syndrome, for which onset is during middle to old age [3]. Age at onset for SUNCT is on average 50.7 years [7]. Thus, the difference in mean age at onset between CH and SUNCT is quite clear, i.e. approximately 20 years. Despite the modest number of SUNCT patients, it is improbable that this age difference is simply a chance finding. In fact, if we apply Student's *t* test to compare age at onset for 21 SUNCT patients with 105 CH patients in Ekbom's series [61], the difference is highly significant (50.7±14.8 vs. 27.5±11.3 $p<0.00000001$). Of course, it can be claimed that the prevailing pattern is like this: when CH starts early in life, attacks are of medium duration; if it starts late in life, attacks are more short-lasting. This would anyhow be a weak argument.

Duration of attacks is one of the most characteristic features distinguishing CH and SUNCT. According to the International Headache Society (IHS) diagnostic criteria, CH attacks last 15–180 min [15]. On the other hand, the usual duration of SUNCT paroxysms is described in the IASP classification as 15–120 s [3]. A prospective study of 77 spontaneous attacks in 22 CH patients [62] revealed that duration of nocturnal attacks ranged from 20–149 min (median, 40 min) and duration of daytime attacks from 8–238 min (median, 37 min). This is far from the range seen in SUNCT attacks. Attack duration was objectively estimated in 348 attacks in 11 SUNCT patients [63]. The mean weighted duration of attacks was 49 s, with a range of 5–250 s. The unweighted mean was 61 s, with a range of 24–125 s. More than 95% of all 348 attacks lasted between 10 and 120 s [63]. Obviously, if statistics were to be applied to these figures, a markedly significant difference would be the result. There is, thus, probably no or close to no overlap as regards this variable.

However, in addition to attacks of usual duration, SUNCT patients may very exceptionally experience pain lasting 30–60 min or even 1–2 hours [26, 27, 29]. Such long-lasting pain was often described as a low-grade, background pain or like a series of short-lasting, but overlapping attacks (when pain did not quite reach the baseline before the next attack started). Occasional patients had 1- to 2-hour episodes of "plateau-like" pain, disclosing otherwise all typical characteristics of SUNCT paroxysms [29]. Such duration of pain would fit with the diagnosis of CH. It should be emphasized, however, that the *usual* pattern in cases exhibiting this exceptional pattern is the short-lasting variety. It is also well known that CH attacks may, under extreme circumstances, last up to 1–2 days [9]. Such temporal patterns fit with "status". In most (or all) types of headaches, a "status" is occasionally seen. It has also been observed in CPH and migraine.

Pain intensity of attacks seems to be much stronger in CH than in SUNCT. In the IASP classification, pain in CH attacks is described as excruciatingly severe, while in SUNCT the range is between moderate and severe [3]. Of course, not all CH attacks are of the same intensity [9]. Thus, some attacks may occasionally be moderate or even mild [62]. Furthermore, there are cases of otherwise typical CH patients who experience only attacks of mild to moderate intensity [10]. However, even in such patients, attacks may eventually become more severe with time and at times even excruciatingly severe [64]. Thus, patients with "mild" CH seem to be exceptional. The expression coined by Horton [65], "suicide headache", well characterizes the degree of pain in CH.

On the other hand, severe headache paroxysms [11] are rather exceptional in SUNCT, where pain mostly is moderate and only rarely severe [12]. While estimation of the pain intensity by a patient is a subjective matter, it seems more objective to observe patients' behavior during attacks. CH patients prefer to assume the erect position during attacks, pace the floor, and even bang their heads against the wall; because of the excruciating pain they are unable to lie down [65]. This is so typical that it was included in the description of the main features of CH by IASP [3]. Generally, SUNCT patients do not seem to be affected by the pain to the same extent. They can sit (or lie) still and continue talking in a rational way during paroxysms [7]. Thus, although there seems to be the occasional overlap between pain intensity in CH and SUNCT [10, 11], these two headache types generally differ – and probably significantly so – as regards pain intensity.

Frequency of attacks usually ranges from <1 to 3 per 24 hours in CH [9]. In Russell's prospective study [62], the frequency of attacks ranged from <1 to 4 per 24 hours, with a mean of 1.68. The IHS criteria are rather liberal, allowing a wide range for the frequency of CH attacks, i.e. from 1 every other day to 8 per day [15]. SUNCT patients may experience up to 25–30 attacks per hour [1, 2], or – at the other extreme – report a frequency of attacks typical for CH. However, even such exceptional patients experience periods of exacerbation with much higher frequency of attacks than in CH [22]. A total of 585 consecutive attacks were included in a study of attack frequency in SUNCT [63]. The mean weighted frequency of attacks was 16 per day (range, 1–86), while the unweighted mean was 28 per day (range, 6–77). Thus, means and upper ranges of attack frequency differ very clearly between CH [62] and SUNCT [63].

Nocturnal preponderance of attacks is typical for CH [3]. The "degree" of such preponderance to a large extent depends on the way the data are displayed [9, 14, 62]. There is anyhow a clear difference between CH and SUNCT patients, who only occasionally report nocturnal attacks. The rare nocturnal occurrence of attacks is hardly due to the brevity of SUNCT paroxysms, solitarily. V_1 trigeminal neuralgia paroxysms [7] are generally shorter, and nevertheless they not infrequently lead to nocturnal awakenings. It seems highly likely that the rare nocturnal awakenings partly are due to the relatively mild degree of pain of SUNCT attacks. The most reliable information is obtained from a comparison of prospective studies in CH [62] and SUNCT [63]. Thus, 40% of 77 CH attacks [62] and only 2% of 585 SUNCT attacks [63] occurred between 11 PM and 7 AM. Again, if statistics were to be applied, a markedly significant difference would be found between SUNCT and CH.

Mechanical precipitation of attacks, although not entirely obligatory, is quite typical for SUNCT [1, 2, 11, 13, 22, 24–26, 28]. Eighteen out of 21 patients described a variety of precipitating mechanisms [12]. The precipitation mechanisms [12, 13, 22] mostly concern the trigeminal innervation area (e.g. skin trigger zones, sneezing), but some precipitation mechanisms are related to extratrigeminal areas (e.g. neck movements, walking on a hard surface). No such trigger mechanisms, neither from trigeminal innervation nor extratrigeminal areas, have so far been demonstrated in an incontrovertible manner in CH [9]. This is a *major distinguishing feature* and not a chance finding in our estimation. It probably indicates a fundamental difference in the generation of attacks in the two disorders. Why such a precipitation mechanism is not present in every SUNCT case is another, still unsolved question.

Precipitation of attacks with alcohol is a typical feature of CH [3, 15]. It is difficult to study the influence of alcohol on attacks in SUNCT in the same way as in CH, especially in the SUNCT patient in a chronic stage with multiple paroxysms. However, some authors reported specifically that alcohol did not precipitate attacks in SUNCT patients [11, 13]. At present, therefore, there is no definite evidence that attacks in SUNCT patients, be it in the symptomatic period or in the remission phase, can be precipitated by alcohol intake. Furthermore, it does not seem that alcohol has any influence on severity, duration, or frequency of such attacks.

A therapeutic effect of the drugs effective in CH, used either to treat solitary attacks or prophylactically [4, 9, 66] has not been demonstrated in SUNCT. The tendency to attacks of CH is in most cases abated by prednisone, verapamil, lithium, ergotamine, subcutaneously administered sumatriptan or 100% oxygen; SUNCT syndrome is generally refractory to drug therapy of any kind or anesthetic blockades. From our own data [67] and many other reports [11, 23, 25, 27, 68–74], more than 20 SUNCT patients have received many different kinds of treatments and only solitary cases have been reported with a good response to individual drugs. For practical reasons, not all drugs can be tried in every patient.

Individual SUNCT patients claimed that steroids were of some help. Only 3 patients [11, 67] out of 11 [11, 67, 70, 71, 73, 74] on prednisone had some positive effect [11, 67] or

even a complete relief [70] during the time in which the drug was maintained. One patient on prednisolone combined with carbamazepine had a favorable response [69]. Verapamil did not provide any relief in three patients [67, 69, 70], and 3 others experienced worsening of headache attacks with this drug [67]. Lithium has also been tried in 2 SUNCT patients [67] without positive effect. Parenterally administered ergotamine and intravenously given dihydroergotamine have also been used to treat attacks of CH. Of 9 reported SUNCT patients [11, 25, 67, 70] receiving these drugs, only one had a partial effect [25]. Sumatriptan has been tested in some patients: orally (100–300 mg/day) in 3 patients [67] and in a single subcutaneous dose [11, 70, 74]. One patient with the oral formula had a slight improvement. Another patient receiving sumatriptan subcutaneously [11] had an apparent positive effect, with absence of spontaneous attacks during the 5 h following the administration (attacks could still be triggered). Thus, overall, no drug effective in episodic CH invariably had a lasting, complete effect in SUNCT.

Antineuralgic drugs have also been tried in SUNCT [11, 23, 25, 27, 67–71, 73, 74]: carbamazepine, phenytoin, clonazepam, baclofen, sodium valproate, lamotrigine, and gabapentin. Of all these drugs, carbamazepine provided slight or moderate effect in 11 of 21 patients treated [11, 23, 27, 67, 69, 71, 73, 74]. Lamotrigine [68] and gabapentin [70] were tried in one patient each, with a complete response to the drug. Even more, percutaneous left trigeminal ganglion microcompression [73, 74] has been tried in 2 SUNCT patients, with a complete relief in one of them [73].

However, special care must be taken in drawing conclusions concerning drug effect in headache types with paroxysms appearing in clusters or with erratic temporal pattern, as SUNCT. Some of the positive results obtained should be viewed in this light.

Laboratory investigations

"Horner-like" picture on the symptomatic side is a typical, although not obligatory, feature in CH. It may also be present outside attacks, both in and outside a bout [9]. There seems to be a tendency for it to be more clearly "expressed" clinically during a bout, and even more so during the solitary attack. Miosis, ptosis and decreased forehead sweating (outside attacks) may be prominent enough to be seen with the naked eye or be present only at the subclinical level, detectable only with laboratory methods [38, 65, 75–82].

It may be most proper to use the term Horner-like picture concerning the pupil in CH, since the pupillometric pattern generally does not seem to be identical with that of the first-third sympathetic neuron dysfunctions [83, 84]. After topical stimulation with indirectly acting sympathicomimetics (e.g. hydroxyamphetamine, tyramine), in case of first or second sympathetic neuron dysfunctions, the pupil on the symptomatic side dilated more than that on the non-symptomatic side [85]. A similar stimulation in third neuron dysfunction resulted in virtually no dilatation on the symptomatic side [86, 87]. In CH, there is generally an "intermediate" pattern. In contrast to what is the case in third neuron dysfunction, mydriasis usually ensues on the symptomatic side, although not as markedly as on the non-symptomatic side [78–80]. Only a few CH patients exhibit the pupillometric pattern seen in Horner's syndrome due to third neuron dysfunction [87, 88]. Pupillometry is less likely to show differences between Horner's syndrome and CH after administration of directly acting sympathicomimetics (e.g. phenylephrine) than after administration of indirectly acting sympathicomimetics. Both in Horner's syndrome and CH, there seems to be a moderate pupil hypersensitivity to phenylephrine on the symptomatic side [78–80, 85].

As far as forehead sweating is concerned, deficient sweat secretion is observed on the symptomatic side after body heating, whereas sweating is increased after pilocarpine stimulation, both in CH [79, 81, 82] and in Horner's syndrome [85, 86].

As regards SUNCT, no definite Horner-like picture has been reported so far, either with clinical observations or laboratory tests [30, 89]. There was no statistically significant anisocoria in basal conditions prior to pharmacological stimulation with sympathicomimetic agents in SUNCT [89]. As far as topical stimulation of hydroxyamphetamine is concerned, only individual patients may have a slight tendency towards a reduced increase in pupil diameter on the symptomatic side [89]. In fact, the dilatation on the symptomatic side was not significantly different from that in a control population [78]. No supersensitivity reaction to phenylephrine stimulation was found on the symptomatic side, like in Horner's syndrome [84–86] and CH (in the latter it admittedly is modest) [78–80, 84]. The forehead sweating response to heating was fairly symmetrical in SUNCT patients [30]. Generally, there was no trend towards increased sensitivity of the sweat glands on the symptomatic side, using the pilocarpine test [30]. The symmetrical forehead sweating pattern after heating and pilocarpine resembles that of control individuals [81] and is quite different from what is found in Horner's syndrome [85, 86] and many cases of CH [81, 82]. Increased symptomatic side forehead sweating is part of the attack both of SUNCT and CH, but the mechanisms underlying its production appear to differ. Thus, so far, there is no definite evidence that Horner's syndrome or a Horner-like picture, not infrequently seen in CH, is present in SUNCT patients.

Respiratory sinus arrhythmia (RSA) is a reflex mediated by stimulation of stretch receptors in the lungs during the respiratory cycle. Since the pathways of this reflex are locat-

ed in the vagal nerves, this reflex has been widely used for estimation of vagal function. This reflex has also been assessed in CH, both outside [90, 91] and during a limited number of nitroglycerine-provoked [37] and spontaneous [91] attacks, as well as in SUNCT patients outside and during attacks [92]. There were clear and not readily explainable differences between SUNCT and CH as regards RSA outside attacks: in SUNCT, RSA was significantly higher [92], whereas in CH it was significantly lower than in controls [90, 91]. Hence, RSA was significantly higher in SUNCT than in CH. The most appropriate comparison of SUNCT and CH in this respect is probably to use studies performed in the same laboratory and, furthermore, using the same technique [91, 92]. Thus, 2 of 6 SUNCT patients disclosed RSA values *above* the upper normal limit according to Smith [93]. It is probably of some significance that results above this stipulated upper normal limit [93] were *never* obtained in controls (n=49), or in CH (n=33) or chronic paroxysmal hemicrania (CPH) (n=4) patients [91]. In the solitary case, however, RSA cannot be used as a distinguishing factor.

RSA during attacks is a more complicated matter. RSA was generally decreased during SUNCT attacks [92], while no general pattern emerged in CH [37, 91]. What can be stated at present is that while RSA increment was occasionally seen during the CH attacks [37, 91], no SUNCT patients disclosed increased RSA during attacks [92]. For tenable verdicts to be obtained for attack-related RSA in SUNCT/CH, a study of a larger number of spontaneous attacks should be conducted.

SUNCT vs. chronic paroxysmal hemicrania

A few words are in order concerning CPH, which has been classified by the IHS [15] as a subtype of the CH syndrome. Differential diagnosis of SUNCT vs. CPH is more straightforward, since complete relief from indomethacin is mandatory by definition in the latter headache [3, 9, 15]. This drug has been shown to have no effect on headache in SUNCT [67]. Furthermore, CPH differs from SUNCT as regards attack duration (mean duration of CPH attacks, 13 min; range, 3–46 min [94]), the even distribution of attacks throughout day and night [94], and the sex preponderance [9, 95], to set down only a few of the many distinguishing features.

Conclusions

In spite of many similarities (Table 1), SUNCT differs from CH in many respects (Table 2). These differences are so characteristic that the typical SUNCT patient is rather easily distinguishable from a CH patient. The typical, distinguishing SUNCT characteristics include short duration of attacks, moderate (only occasionally severe) pain intensity, high frequency of attacks, only rare nocturnal attacks, mechanical precipitation of attacks, and – probably – lack of alcohol influence on paroxysms. Presence of these features speaks for SUNCT and against a CH diagnosis.

Unfortunately, not all these typical features are invariably present in every case of SUNCT. There are, however, other features also favor a SUNCT diagnosis, especially if more of them are present. We include in this group of SUNCT characteristics: lack of therapeutic effect of drugs used in CH (especially if several types of drugs have been tried), and age at onset over 40 years. Other features may also be added: increased respiratory sinus arrhythmia outside attacks and absence of a Horner-like picture. These features, however, necessitate laboratory facilities.

SUNCT seems to be a "neuralgiform" disorder [1, 2]. SUNCT has, nevertheless, been demonstrated to be rather clearly distinguishable from tic douloureux [6], especially the V_1 variety [7]. SUNCT accordingly most likely is a distinct, separate syndrome. The present review indicates that SUNCT probably differs essentially from CH.

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