Brief Communications

Case Report: Coexistence of SUNCT and Hypnic Headache in the Same Patient

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Background.—Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and hypnic headache (HH) are two exceedingly rare and distinctly classified primary headaches. The hypothalamus seems to be a crucial region involved in the pathophysiology of both conditions, but no cases of SUNCT and HH co-occurrence have been described so far.

Case results.—A 49-year-old woman who has been suffering from SUNCT for years, with alternation of symptomatic periods and remissions, developed a new headache with different clinical features, presenting exclusively during sleep and with a dramatic responsiveness to caffeine, that met the diagnostic criteria for HH.

Conclusions.—The available literature suggests that SUNCT and HH are different conditions but the association in the same patient that we describe supports the concept that they are not mutually exclusive. Further studies are needed to establish if they share a common pathophysiological mechanism.

Key words: short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing, hypnic headache, caffeine, trigeminal, tautonomic

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) belongs to the trigeminal autonomic cephalalgias (TACs). It is characterized by short attacks of orbital, supraorbital, or temporal pain with stabbing character of 1–600 s duration, with frequency of at least one a day.[1] The patients, the majority being males, may experience up to 30 attacks per hour,[2] and the disorder can have an episodic or chronic pattern of occurrence.[3] Hypnic headache (HH) is another uncommon primary headache with recurrent pain attacks developing only during sleep, and causing the patient's awakening. It occurs more than 10 times per month for at least 3 months, and lasts from 15 minutes to 4 hours after waking, without cranial autonomic symptoms or restlessness.[1] We describe the case of a patient who developed these two rare primary headaches.

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CASE DESCRIPTION

A 49-year-old Caucasian otherwise-healthy woman suffered from spontaneous single painful stabs in the left orbital-temporal region lasting few seconds, every 6–7 months, since the age of 23. In the following 17 years, the single stabs became progressively longer in duration, up to 10 seconds, and were accompanied by conjunctival injection and

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tearing ipsilaterally to pain. Frequency also increased to 20 episodes per day for 2–3 consecutive months, with 1-month pain free intervals. She had no personal or family history of migraine, never had side-shift attacks and did not report triggers. Attacks occurred only during the day, and never caused her awakening. Analgesics had a small effect on pain intensity, but did not modify the temporal pattern. She was empirically treated with amitriptyline 40 mg/day and indomethacin 50 mg t.i.d. for three months by her general practitioner, without effect. At the age of 40, suspecting a secondary headache, the patient underwent a 1.5 T brain magnetic resonance imaging (MRI) comprehensive of MR angiography of intracranial vessels and 3D-T2 driven equilibrium radiofrequency reset pulse (DRIVE) sequences for the cranial nerves. These investigations produced normal results and no neurovascular compression was visible. The pituitary gland was within normal limits.

At the age of 42, the duration of attacks and pain profile were unchanged, in particular she did not report nausea, phono, photo, or osmophobia during attacks. Her neurological examination was normal. She was evaluated in our Headache Centre and SUNCT was diagnosed. She was treated with lamotrigine, gradually increasing the dosage up to 100 mg daily. The attacks decreased at the dosage of 50 mg and completely disappeared at maximum dosage. The drug was gradually discontinued after a 3-month pain free period. Headache recurred after 4 months, and lamotrigine was reintroduced, at the same regimen with complete benefit. This treatment was continued for 3 years. Then, she noted that pain recurred at the dosage of 75 mg of lamotrigine per day, so she was forced to continue with the full dose of 100 mg daily.

She was pain free for further 3 years; however, at the age of 49, she began suffering from a new occipital, constrictive, moderate, bilateral pain, presenting exclusively during sleep and causing awakening. Pain was occasionally severe and associated with nausea. She had only one attack per night, and the most frequent time of occurrence was between 2 and 4 a.m. The pain lasted about 60 minutes and gradually increased in frequency, becoming nearly daily. Nocturnal blood pressure and glycemia were normal, and sleep disturbances were not reported. Hypnic headache was diagnosed. The patient was treated with a cup of coffee 30 minutes before going to sleep, and she had no headache recurrence in a few days. The treatment was continued on a daily basis for 2 months and then on alternate days for 1 month. No sleep disturbance occurring by drinking coffee at bedtime were reported. Because of moderate intensity, the patient needed no acute therapy. Taking into consideration the change in the headache pattern, a repeat brain MRI was recommendable, but was not performed due to the favorable and rapid treatment response. The neurological examination remained normal.

At 6-month follow-up the patient was pain free.

DISCUSSION

SUNCT and HH are exceedingly rare headaches. They can coexist with other primary headaches, especially migraine.[4,5] A single case of HH in a patient with hemicrania continua, another rare headache belonging to the TACs, was recently described;[4] however, at the best of our knowledge, this is the first description of SUNCT and HH occurring in the same patient.

The pathophysiology of SUNCT and HH is largely unknown. The mechanisms underlying the coexistence of these two rare headaches are intriguing and may involve the trigemino-hypothalamic pathways in both conditions. Hypothalamus regulates endocrine functions, sleep and circadian rhythms, but also autonomic function and pain control. Neuroimaging studies confirmed its role in both SUNCT and HH. Functional MRI (fMRI) studies showed ipsilateral or bilateral hypothalamic activation during SUNCT attacks and a voxel-based morphometric study on HH patients demonstrated a significant gray matter decrease in the posterior hypothalamus.[6,7] In TACs, hypothalamic activation and its influence through descending projections on the trigemino-cervical complex and the superior salivatory nucleus have been invoked to explain attacks occurrence.[8] In HH, previously defined as "alarm clock headache syndrome," the

hypothalamic involvement might explain its strong chronobiological regularity.

Further overlaps between SUNCT and HH involve the clinical pattern, in particular the circadian rhythmicity and cranial autonomic symptoms. SUNCT attacks tend to have an erratic pattern occurring mostly during the day, but a sleep pattern is also described.[9]

HH typically presents without cranial autonomic symptoms and their presence hampers the diagnosis, according to ICHD criteria. This distinction should avoid misdiagnosis with TACs, which may occur at night and cause patient awakening, but noteworthy were some mild manifestations that have been described in about 15% of HH patients in two case series. Whether this could be a clue to hypothalamic activation or simply a nociceptive-induced activation of the trigeminal-autonomic reflex, common to various pain disorders, is not clear, and needs further elucidation.[4] Nevertheless, our patient had typical SUNCT and HH attacks. In particular, SUNCT maintained a diurnal pattern, and HH developed many years after SUNCT onset and treatment, while she was pain free from SUNCT since 2 years, and never presented autonomic features.

Caffeine is the drug of choice in HH and it is also effective when HH develops with other primary headaches.[4] Lamotrigine (LTG) is the most effective prophylaxis in SUNCT and it has been also reported to be effective in two patients with HH at the dose of 50 and 75 mg/day.[10] In our patient, LTG was clearly effective at the dosage of 100 mg/day. When HH developed, the patient was already receiving LTG 100 mg/day. We cannot exclude that LTG could have had some effect on HH, and maybe controlled the emergence of cranial autonomic symptoms; however, we preferred to prescribe a cup of coffee before sleep instead of increasing the dose of LTG, to avoid possible LTG toxic effects.

The current experts' opinion suggests that SUNCT and HH differ in terms of phenomenology and pathophysiology. Even if the hypothalamus is involved in both conditions, some clinical features are at opposite polar extremes, such as the daytime attacks in SUNCT vs the night pattern in HH and the typical presence of autonomic features in SUNCT compared with their usual absence in HH. These contrasts may suggest that different neuroanatomical pathways are involved. However, the association in the same patient that we describe supports the concept that a history of SUNCT does not preclude the diagnosis of HH. Further studies are needed to identify the presence of a hypothetical common patophysiological mechanism.

STATEMENT OF AUTHORSHIP

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