

Primary stabbing headache: a new dural sinus stenosis-associated primary headache?

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Abstract Primary stabbing headache (PSH) is a primary syndrome of unknown aetiology, characterised by brief, jabbing stabs predominantly felt in the orbital, temporal and parietal areas, whose frequency may vary from one to many per day, usually responding to indomethacin. PSH frequency in the general population is not well defined, but recent evidence suggests it could be more frequent than previously thought. In clinical series, PSH incidence was 33/100,000 per year, while in a population study 35.2 % prevalence was found. PSH was previously described as isolated or associated to other headache syndromes, most frequently with migraine. There is evidence that an idiopathic intracranial hypertension without papilledema, a condition usually associated to significant stenosis of dural sinuses (93 % sensitivity and specificity), is much more prevalent than believed and may run asymptotically in up to 11 % of otherwise healthy individuals. In migrainous prone people, a sinus stenosis-associated intracranial hypertension without papilledema (ss-IHWOP) comorbidity may represent a powerful risk factor for progression of pain. Besides migraine, significant sinus stenosis has been found over-represented also in chronic tension type headache as well as in exertional, cough, sexual activity-associated headaches (all indomethacin responsive primary headaches) and in altitude headache (an acetazolamide responsive condition). To explore the possible association between venous outflow disturbances and PSH, we retrospectively investigated the co-occurrence of sinus venous stenosis in patients referring to our headache centre since 2004 diagnosed with PSH who

completed the diagnostic protocol. Out of 50 consecutive patients reporting PSH as the main or as accessory complaint, 8 (6 females, 2 males) performed MR venography (MRV). All MRV revealed significant unilateral or bilateral sinus stenosis. Mean age at PSH onset was 35.3 ± 18.9 years (range 11–67 years). Duration of attacks ranged 1–3 s. Median daily frequency of attacks was 4 (range 2–20); median number of days per month with PSH presentation was 14 (range 4–30). Six patients described attacks in temporal or parietal areas, one at the top of the head, and one in the occipital area. Only one patient had isolated PSH; all the others were diagnosed also with migraine without aura. Seven out of eight patients responded to indomethacin 75 mg/die, and one to topiramate 100 mg/die. Interestingly, both drugs share with acetazolamide a CSF pressure lowering effect. Our findings indicate that PSH is associated with central sinus stenosis and suggest that an undiagnosed ss-IHWOP might be involved in PSH pathogenesis.

Keywords Primary stabbing headache · Sinus stenosis-associated intracranial hypertension without papilledema · Intracranial pressure · Indomethacin · Magnetic resonance venography

Introduction

Primary stabbing headache (PSH) is characterised by brief, jabbing stabs predominantly felt in the orbital, temporal and parietal areas, with unilateral or bilateral localization. Attacks are very short, up to 3 s recurring with irregular frequency. PSH is generally thought to be a primary headache of unknown pathogenesis, mostly occurring in women, commonly associated with migraine and usually responsive to indomethacin [1]. PSH prevalence is not well

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defined in the general population, but it could be much more frequent than previously thought. According to Pareja et al. [2], PSH hospital series incidence is 33/100,000 per year, but this value may be underestimated, since not all patients are evaluated in a neurological/ophthalmological department. A large-scale population study on headache epidemiology by Sjaastad et al. in Vaga, Norway [3], shows a PSH prevalence of 35.2 % among an unselected population aged 18–65 years. Jabs generally last up to 3 s and are more frequently felt, in fronto-temporal areas. PSH was previously described either as an isolated headache or associated to other types of primary headache, mostly migraine, but also tension type headache, cluster headache, chronic paroxysmal hemicrania, and hemicrania continua [4]. Raskin et al. [5] compared the incidence and the clinical characteristics of PSH in 100 migraineurs and 100 control subjects. PSH was found in 42 % of migraineurs versus only 3 % of healthy controls ($p < 0.001$). Half of the cases experienced PSH at least monthly.

A number of primary headache subforms have been recently associated with the presence of impaired cerebral venous outflow sustained by significant sinus stenosis at MR Venography (MRV), a highly predictive marker of raised intracranial pressure (ICP) [6]. These include chronic migraine [7], chronic tension type headache [8], cough, exertional and sexual activity-related primary headache [9, 10], and altitude headache [11]. The aim of our study is to evaluate the association of abnormalities in cerebral venous circulation with PSH.

Patient series and method

We retrospectively investigated the co-occurrence of sinus venous stenosis in all patients referring to our headache centre since 2004, diagnosed with PSH, as the main complain. Only patients with available dural sinus imaging by MRV were included in the study.

Results

Out of 50 consecutive PSH subjects, 8 (6 female, 2 males) performed MRV. Mean age at onset of PSH was 35.3 ± 18.9 years (range 11–67 years). Median duration of attacks was 2 s (1–3). Median daily frequency of attacks was 4 (1.50–20); monthly frequency varied largely from 3.5 to 30 days per month (median 14). Attacks were predominantly felt in the temporal or parietal areas in six patients, at vertex in one, and in the occipital area in another one. Only one patient suffered of isolated PSH, all the others were diagnosed also with migraine without aura.

Seven patients responded to indomethacin, one to topiramate 100 mg/die.

MRV resulted in a significant venous outflow disturbances in all patient with one patient showing multiple apparent flow gaps, two patients showed bilateral transverse sinus flow gaps, two patients showed a unilateral transverse sinus flow gap (both at right transverse sinus level), one patient showed aplasia of the right transverse sinus, and two patients showed a significant narrowing of a transverse sinus (both on the left side).

Discussion

The finding of sinus stenosis in PSH patients is an original and suggestive data. It may open new interesting scenario in the definition of the pathophysiological mechanism and treatment of this primary headache. Venous sinus abnormalities were first associated to idiopathic intracranial hypertension (IIH) [6], an infrequent and enigmatic condition [12] that may run without papilledema (IIHWOP) in a significant percentage of patients [7]. According to a recent evidence [13], IIHWOP can be detected in 11 % of individuals without chronic headache or other signs or symptoms of intracranial hypertension. Such asymptomatic and, therefore, largely undiagnosed IIHWOP only occurs in subjects showing sinus stenosis, a condition observed in about 23 % of healthy people [13]. In primary chronic headache clinical series, the prevalence of cerebral venous outflow abnormalities was 48.9 % and almost all patients (91.6 %) showed a continuous or intermittent IIHWOP; conversely, all patients with normal MRV had an ICP within normal limits [14]. Based on the above observations, we recently proposed that a sinus stenosis-associated intracranial hypertension (ss-IIHWOP), although very common among healthy subjects, is a powerful risk factor for progression and refractoriness of pain in primary headache prone individuals [7]. Moreover, sinus venous outflow disturbances have been recently described in primary cough headache, primary exertional headache, and primary headache associated with sexual activity [10] all known indomethacin responsive primary headaches. Moreover altitude headache, an acetazolamide responsive headache [11], has been recently found to correlate with the presence of dural transverse sinus narrowings (uni or bilateral) in subjects complaining of headache within 24 h after the ascending to high quote (5300 mt). Acetazolamide inhibits carbonic anhydrase isoenzymes and is one of the few drugs with a known effect in ICP lowering [15]. Also indomethacin (reported as efficacy in PSH [16]) and topiramate (found efficacy in one of our cases not responding to indomethacin) share the same ICP lowering effect [17, 18]. Based on these considerations, the high prevalence of

sinus stenosis found in our series suggests that also PSH could be related to an overlooked ss-IHWOP.

Conflict of interest I certify that there is no actual or potential conflict of interest in relation to this article

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