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CASE REPORT / OLGU SUNUMU

Two cases of primary stabbing headache

Primer saplanıcı başağrısı: İki olgu sunumu

Murat GUNTEL,[1] Ozge HURDOGAN,[2] Derya ULUDUZ,[2] Taskin DUMAN[1]

Summary

Primary stabbing headache is an excruciating and relatively rare type of headache that typically lasts for only a few seconds. Pain is predominantly felt in the distribution of the first division of the trigeminal nerve and can be experienced as single stabs or as a series of stabs, either per day or every few days. Primary stabbing headache has been well-defined for decades and must be kept in mind during diagnosis. Exclusion of other possible causes is necessary in order to establish diagnosis. Indomethacin has classically been considered the first treatment option, but therapeutic failure occurs in up to 35% of cases. Recent studies have suggested that cyclooxygenase-2 inhibitors, gabapentin, nifedipine, paracetamol, and melatonin are also effective treatments.

Keywords: Primary stabbing headache; primary headache; treatment; indomethacin.

Özet

Primer saplanıcı baş ağrısı; saniyeler süren nadir ve şiddetli bir başağrısıdır. Ağrı özellikle trigeminal sinirin birinci dalı boyunca hissedilir ve günde bir veya birkaç günde bir gelen bir veya birkaç kez tekrarlayan saplanma atakları şeklinde ortaya çıkar. Primer saplanıcı başağrısı yıllardır bilinmekle birlikte tanı için klinisyenin sorgulaması gereklidir. Primer saplanıcı başağrısının tanısını koymada diğer olası nedenlerin dışlanması önemlidir. İndometazin klasik olarak ilk tedavi seçeneğidir ancak olguların %35'e yakını tedaviye direnç gösterir. Son yıllarda cyclooxygenase-2 inhibitörleri, gabapentin, nifedipine, parasetamol, ve melatonin tedavisinin etkinliğini destekleyen çalışmalar bildirilmektedir.

Anahtar sözcükler: Primer saplanıcı başağrısı; primer başağrısı; tedavi; indometazin.

Introduction

Primary stabbing headache (PSH), first defined in 1964 by Lansche as "ophthalmodynia periodica,"[1] was renamed in 1979 by Sjaastad as the "jabs and jolts syndrome"[2] in reference to descriptions given by affected patients. Raskin and Schwarz subsequently defined this pain as "ice pick-like" in patients with migraine.[3] The pain was has also been termed "needle in the eye syndrome" and "sharp short-lived head pain,"[4] although the disorder is now officially defined as PSH in the second edition of the International Classification of Headache Disorders.[5] Patients with PSH usually describe short-lived but severe jabbing stabs of pain accompanied by a shock-like feeling and even jolting head movements and/or vocalizations. Although the exact prevalence of PSH remains unknown, it is estimated to affect 2.0% to 32.5% of the adult population.[6,7] The peak age of onset has been reported to vary from 28 to 47 years.[7,8] Female patients are more susceptible to PHS.[9]

This report describes the clinical characteristics of two female patients with PSH.

Case Report

A 65-year-old woman presented with a 1-month history of sharp, stabbing pain in the right parieto-tem-

[1]Department of Neurology, Mustafa Kemal University Faculty of Medicine, Antakya, Turkey
[2]Department of Neurology, İstanbul University Cerrahpaşa Faculty of Medicine, İstanbul, Turkey

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Correspondence *(İletişim):* Dr. Derya Uluduz. İstanbul Üniversitesi Cerrahpaşa Tıp Fakültesi Nöroloji Anabilim Dalı, 34340, Samatya, Fatih
Tel: +90 - 212 - 414 30 00 **e-mail *(e-posta):*** deryaulu@yahoo.com

poral region and vertex. The attacks were brief, lasting ≤3 s and occurring with an irregular pattern of several times (up to 20 times) per day to once a week. She described the pain as stabs without nausea, vomiting, or autonomic features such as conjunctival redness, eyelid drooping, nasal congestion, runny nose, eyelid edema, facial blushing/sweating, myosis, or ptosis (partial Horner's Syndrome). Her medical history revealed hypertension, for which she was taking the angiotensin-converting enzyme inhibitor perindopril with good control. Her neurological and psychiatric examinations were unremarkable. Blood tests including a complete blood count; sedimentation rate; high-sensitivity C-reactive protein (hs-CRP) level; hepatic, renal, and thyroid function tests; electrolyte levels; and a lipid profile were within normal ranges. Cranial magnetic resonance (MR) imaging and cervical and cranial MR angiography results were normal. An electroencephalogram was interpreted as normal. No cardiac pathology was determined to be present according to the results of cardiac evaluation and echocardiography. We started the patient on indomethacin at 12.5 mg three times per day as the first treatment option for the PSH and increased the dose on alternate days until a response was apparent. The patient responded to an indomethacin dosage of 25 mg three times daily.

A 31-year-old right-handed woman presented with a 2-year history of thunder-like stabbing headaches in the right or left temporal region that radiated to the vertex. She reported daily multiple episodes (<10 attacks) that occurred nearly once per minute, each lasting a fraction of a second. The frequency and duration of the patient's previous headache attacks were three times a year and 10 days, respectively, but their frequency had changed to daily during the last 6 months. During these excruciatingly painful attacks she had to hold her head in both hands and lean her head forward for slight relief. She denied nausea, vomiting, or any cranial autonomic features. Her medical history was remarkable for a 5-year history of migraine attacks without aura. During her migraine attacks, she occasionally experienced moderate stabbing headaches. Her medical and family history was otherwise unremarkable. Neurological and psychiatric examinations revealed no abnormal signs. Laboratory tests including a blood cell count, hepatic and renal function, electrolyte levels, thyroid function tests, sedimentation rate, and hsCRP level were normal. Cranial MR imaging and MR angiography revealed no abnormalities. Indomethacin treatment was started and the patient reported a major improvement in symptoms within days. She experienced a complete clinical recovery with indomethacin at 50 mg three times a day.

Discussion

PSH is characterized by repetitive, short-lived (3–5 s) stabbing pains that are not attributed to another disorder. Together with first-branch trigeminal neuralgia, PSH is the shortest-lasting headache known. Although the pathophysiologic mechanisms underlying PSH remain unknown, irritation of the trigeminal or other nerves has been proposed. Alternatively, intermittent deficits of the central pain control mechanisms that permit spontaneous synchronous discharge of neurons receiving impulses from the area of stabbing pain may also play a role in patients with PSH.[10]

The age of onset of PSH varies from 12 to 70 years. There is a female preponderance. Most patients exhibit a sporadic or irregular pattern. An erratic, unpredictable alternation between symptomatic and asymptomatic periods is a characteristic trait. In the vast majority of patients, paroxysms last from a fraction of a second to a couple of seconds (with 80% ending within 3 s).[7,8] Occasional attacks might last up to 5 to 10 s. Typically, pain can be expected to occur with different frequencies throughout the day. The frequency of attacks varies from annual occurrences to rare cases in which the pain occurs 50 times a day.[4] In very rare cases, knife-like pain persists for days or recurs very often. PSH is a diurnal disorder; most attacks are randomly distributed throughout the day. In rare cases, pain occurs repetitively for days, and paroxysmal occurrences last for a week. Patients occasionally report mild, blunt headaches lasting minutes to hours subsequent to a severe stabbing pain.[9] In our second patient, the pain had lasted 6 months and lacked an association with other types of headaches.

Stabbing pains are more dominant in the first branch of the trigeminal nerve (orbital, temporal, and parietal areas) and might be confined to a single hemisphere of the brain or alternatively move from one side to the other. In cases in which the stabbings

are restricted to a small area, further investigation is necessary to determine the organic etiology. Extratrigeminal paroxysms may also occur, particularly in the auricular, posterior parietal, occipital, and nuchal areas.[11,12]

Short-lived primary headache syndromes may be divided into those exhibiting autonomic activation and those without autonomic activation. Accompanying autonomic features and other symptoms are usually absent in PSH,[13] and in 30% of cases, patients report migraine headaches. If migraine and PSH attacks occur simultaneously, the localization is mainly in the same region as presented in our second patient, suggesting that PSH is an incomplete migraine.[3] Short-lived or sharp stabbing headaches have also been described in patients with temporal arteritis, tension headache, chronic paroxysmal hemicrania, episodic paroxysmal hemicrania, SUNCT, trigeminal neuralgia, and cluster headache, all of which should be considered differential diagnoses.[8,14] The absence of cutaneous triggering factors and the location and duration of the pain can make a diagnosis of trigeminal neuralgia unlikely, and the lack of accompanying autonomic symptoms can help rule out a diagnosis of SUNCT and chronic or episodic paroxysmal hemicrania.

PSH is considered an indomethacin-responsive headache syndrome along with hemicrania continua and chronic paroxysmal hemicraina.[15] Although the underlying mechanisms are not entirely understood, studies have suggested that indomethacin lowers cerebral blood flow and decreases cerebrospinal fluid pressure.[15,16] The response to indomethacin is variable, ranging from complete relief as shown in our cases to none at all in 35% of cases; however, indomethacin remains the first-choice treatment. Few alternatives are available for patients unresponsive to indomethacin treatment or who have a contraindication to indomethacin. Antiepileptic and antidepressant drugs represent two such choices. Some studies have suggested that agents such as cyclooxygenase-2 inhibitors, gabapentin, nifedipine, paracetamol, and melatonin might also be effective treatment options.

Conclusion

Although PSH has been well defined for decades, it must be recalled by clinicians for diagnosis. Exclusion of other possible causes is necessary to establish a diagnosis of PSH. Indomethacin has classically been considered the first treatment option.

Competing Interests

The authors declare that they have no competing interests.

Conflict-of-interest issues regarding the authorship or article: None declared.

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