

Expert Opinion

Primary and Secondary Stabbing Headache

Matthew S. Robbins, MD; Randolph W. Evans, MD

Eight out of the 33 cases of primary stabbing headache seen in a general neurology clinic (40% have headache as their chief complaint) in the last 3.5 years are presented. The epidemiology, association with other primary headache disorders, secondary associations, testing, and treatment of primary stabbing headache are reviewed.

Key words: primary stabbing headache, idiopathic stabbing headache, ice pick, jabs and jolts, symptomatic

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Primary stabbing headache is well known to headache medicine specialists. One of the authors (RWE) who sees patients that are teens and older in a general neurology clinic (40% of the patients are seen for headache) has seen 33 patients in the last 3.5 years, only 6 males, ranging in age from 16 to 78. Forty-eight percent of the females were migraineurs. One of the 6 males had migraine, and no other type of headache and one had new daily persistent headache. Other headaches were present in the 27 females: episodic tension type, 3; chronic tension type, 2; and primary thunderclap headache also in one of the migraineurs.

The following are some of the cases.

From the Montefiore Headache Center, Saul R. Korey Department of Neurology, Albert Einstein College of Medicine, Bronx, NY, USA (M.S. Robbins); Department of Neurology, Baylor College of Medicine, Houston, TX, USA (R.W. Evans).

Address all correspondence to M.S. Robbins, Montefiore Headache Center, Saul R. Korey Department of Neurology, Albert Einstein College of Medicine, 1250 Waters Place, Tower 2, 8th floor, Bronx, NY 10461, USA.

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CASE HISTORIES

Case 1. This is a 26-year-old female with a history of migraine with aura and visual aura without headache since age 20. For the last 2 years, she has had a discrete left parietal stabbing pain with an intensity of 3-4/10 lasting seconds that can be multiple times per day for a few days in a row and then not occur for a few weeks. Medical history is negative. Neurological examination was normal. She declined a magnetic resonance imaging (MRI) of the brain.

Case 2. This is a 78-year-old woman with an 8-month history of headaches daily since onset described as a right or left frontal or right or left occipital or top of the head aching with an intensity of 6-10/10 without associated symptoms lasting about 30 seconds 5-10 times per day. She had a past medical history of hypertension and Meniere's disease. Neurological examination was normal. MRI of the brain was negative. Westergren erythrocyte sedimentation rate was 31.

Case 3. This is a 50-year-old female with a history of migraine without aura for 20 years. For the last 9 years, she has sharp or stabbing pains with an intensity of 8-9/10 that can occur up to 27 times per day on a daily basis, daily for a week or not for a month. The pain may be over her left ear, behind the left ear, over

the left or right eye, the back of the head on either side, and the right or left side of the head lasting 1-2 seconds. Medical history was negative. Neurological examination was normal. MRI of the brain 4 years prior was negative.

Case 4. This is a 16-year-old male with a 3-week history of pain occurring 3-4 days per week described as a right parietal and occasionally bifrontal shooting pain with an intensity of 3-8/10 lasting 2 seconds occurring 3 times in a row about 1-4 times in a day without associated symptoms. A history of migraine with and without aura was present, and he had a medical history of anxiety disorder. Neurological examination was normal. An MRI of the brain was normal.

Case 5. This is a 55-year-old female with a 6-month history of occasional right stabbing pain with an intensity of 8/10 lasting seconds that may be right or left parietal, occipital, or frontal. No history of headaches. She also reported a 4-month history consistent with right hemifacial spasm. She had a past medical history of Grave's disease, iritis, and seronegative rheumatoid arthritis on prednisone and methotrexate. Neurological examination was normal. MRI of the brain was negative except for a few non-specific white matter abnormalities.

Case 6. This is a 35-year-old female with an 8-month history of daily headaches described as a stabbing and sharp pain with an intensity of 6/10 lasting a couple of seconds several times a day that occurs in a small discrete area anywhere on the right more often than left side of the head. She had a past medical history of rheumatoid arthritis not on medication, anxiety, and gastroesophageal reflux. Neurological examination was normal. MRI of the brain was normal.

Case 7. This is a 41-year-old female referred by a general neurologist with a history of migraine without aura for 17 years that had become chronic and a 1-month history of a stabbing pain behind the right eye or above the left brow with an intensity of 8/10 lasting a second to 30 seconds occurring about 6 times a day. Neurological examination was normal. The pain was not reproduced with digital pressure over the greater occipital nerves. MRI of the brain showed a small pineal cyst. The next month, her general neu-

rologist diagnosed mild generalized myasthenia gravis and started pyridostigmine.

Case 8. This is a 60-year-old female with an over 10-year history of a sharp stabbing pain with an intensity of 5-6/10 occurring about twice a week occurring in different locations of the right or left side or back of the head for a split second. She had a history of tension-type headaches since childhood and a medical history of hypertension. She had seen an otolaryngologist 5 years earlier for brief vertigo. MRI of the brain showed pansinusitis.

Questions. (1) What is the epidemiology of primary stabbing headache (PSH)? (2) Is having a 4.5 female to male ratio in my practice unusual? (3) Is PSH comorbid with other primary headache types? (4) MRI scans of the brain were negative. Should MRI scans be done and if so, on which patients? (5) Two of the patients have rheumatoid arthritis and one has myasthenia gravis. Is PSH associated with autoimmune diseases? Is this just a chance association or a causal one? Should patients be evaluated for possible autoimmune disease (other than temporal arteritis in those over age 50)? (6) What medications might be effective for treatment, if indicated?

EXPERT COMMENTARY

Primary stabbing headache is an interesting headache disorder because of the wide variety of clinical contexts in which it can present. Though previously labeled with many different terms in the literature including ice-pick headache, jabs and jolts syndrome, and idiopathic stabbing headache, the International Classification of Headaches Disorders, 3rd edition beta (ICHD-3 β) dictates that "primary stabbing headache" is the preferred term.¹ The essential clinical features include paroxysms of transient stabs of pain anywhere in the head that are spontaneous, of short duration, and unassociated with any other symptoms (Table 1). The previous ICHD-2 classification² required the distribution of pain paroxysms in the first division of the trigeminal nerve, but clinical studies^{3,4} were not supportive of this location restriction and led to the removal of this requirement.

Table 1.—International Classification of Headache Disorders, 3rd Edition Beta Diagnostic Criteria for Primary Stabbing Headache

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- A. Head pain occurring spontaneously as a single stab or series of stabs.
 - B. Each stab lasts for up to a few seconds.
 - C. Stabs occur with irregular frequency, from one to many per day.
 - D. No cranial autonomic symptoms.
 - E. Not better accounted for by another ICHD-3 diagnosis.
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ICHD-3 = International Classification of Headache Disorders, 3rd edition.

Although the ICHD-3 β states that the pain may last up to a few seconds, no specific numerical value is given. Many studies have used 0-3 seconds as inclusion criteria. Pareja et al included stabs of longer duration in a series of 38 patients who reported a wide range of durations: 1 second, 69%; 2 seconds, 8%; 3 seconds, 5%; 5 seconds, 8%; 6 seconds, 5%, and 10 seconds, 5%.³ Similarly, in a series of 80 patients, Fuh and colleagues reported the following duration of single stabs: < 3 seconds, 80%; 3-5 seconds, 16%; and 6-60 seconds; 4%.⁴ Some patients may report the attack duration of a grouped volley of pain instead of a single pain paroxysm, which should be distinguished when taking a history. One study reported that most patients have single stabs only,⁵ although a small series reported 38% having single stabs, 30% with a series or volleys of stabs, and 32% with both.⁴

EPIDEMIOLOGY OF PRIMARY STABBING HEADACHE

Epidemiologic studies of primary stabbing headache vary in prevalence estimates. The most rigorous ascertainment of its prevalence was undertaken in the Vaga study of headache epidemiology in Norway, where up to 35% of subjects reported any type of stabbing head pain at any point in their lives.⁶ Other population-based studies have shown a prevalence of 0.2% to 2%.^{7,8} The population-based Vaga study found a female to male ratio of 1.49,⁶ though headache clinic-based studies have revealed higher ratios, from 2 to 6:1,⁹ akin to the series reported here. A recent review⁹ of all adult cases revealed that onset is

typically between ages 23 and 47 years. A childhood onset is not uncommon and is typically between ages 4 and 9 years.

PRIMARY STABBING HEADACHE IN THE CONTEXT OF OTHER PRIMARY HEADACHE DISORDERS

Primary stabbing headache is known to occur in patients who also have other primary headache disorders as evidenced by the coauthor's series in the first paragraph and other headache clinics.⁴ This association was first described by Raskin and Schwartz⁵ in a comparison of 100 migraineurs to 100 control subjects finding 42% of migraineurs with sharp jabbing pains vs 3% of the controls. Stabs seemed to occur simultaneously with migraine and were thought to potentially be a premonitory symptom of an attack. Primary stabbing headache may also occur in many other headache disorders, including tension-type headache, hemicrania continua, and cluster headache^{10,11} with prevalence reported anywhere from 21% to 84% in these other disorders.⁹

It is not clear if the pathogenesis of primary stabbing headache is different in patients who have other primary headache disorders vs when it occurs in isolation. The prime example of this discrepancy may be its occurrence in patients who have cluster headache. When trigeminal neuralgia occurs in cluster headache,¹¹ it is almost always in the V1 distribution, features short paroxysms of stabbing pain, and is without any associated autonomic symptoms. The bouts occur almost exclusively ipsilateral to the side of pain in the patients' typical cluster attacks. Therefore, "trigeminal neuralgia" occurring in these patients may be indistinguishable from primary stabbing headache. In my experience, this type of primary headache in cluster headache may occur in the setting of only partially effective prophylactic medication or be a harbinger of a looming attack period, and increasing or initiating a medication utilized for cluster prophylaxis such as verapamil may help suppress the stabbing attacks in such patients. In other circumstances, verapamil is not known to be effective for primary stabbing headache, and this may indicate that its

clinical expression in the presence of another primary headache disorder has shared pathophysiology with the index disorder.

SYMPTOMATIC STABBING HEADACHE

Stabbing headache has been described in association with etiologies including encephalitis, neoplasm (cerebral or pituitary), vasculitis (ie, giant cell arteritis),⁹ and in one case I encountered contralateral to an acute thalamic hematoma.¹² A recent study described stabbing headache in association with autoimmune disorders in 14 of 26 patients,¹³ though this association may be limited by sample size, referral bias, and difficulty in ascribing causality. However, these patients had primary stabbing headache according to ICHD-2 criteria with a wide variety of autoimmune disorders that could potentially affect the central and peripheral nervous systems. Two recent studies have demonstrated associations of stabbing headache in the context of a relapse of multiple sclerosis.^{14,15} One group proposed that subpial grey matter and meningeal inflammation may sensitize trigeminal sensory fibers that lead to spontaneous activity of trigeminal branches.¹⁴ It is not clear if a similar mechanism on a smaller scale would be relevant to the occurrence of primary stabbing headache in the general population. Finally, one case series described 8 patients with primary stabbing headache, 7 of whom also had migraine, who on magnetic resonance venography had unilateral or bilateral transverse sinus stenosis.¹⁶ The authors posited that since this finding is also occasionally associated with idiopathic intracranial hypertension without papilledema, a similar pathophysiology could be relevant. However, more prevailing hypotheses relate to the spontaneous firing of nerve branches of trigeminal or upper cervical origin, with involvement of central pain control mechanisms because of the frequent presence of allodynia.^{4,9}

DIAGNOSTIC WORKUP OF PRIMARY STABBING HEADACHE

There are no clear recommendations from the literature on the necessity of a diagnostic workup for stabbing headache when it occurs in isolation. The ICHD-3^β suggests, “When stabs are strictly localized

to one area, structural changes at this site and in the distribution of the affected cranial nerve must be excluded.” However, no study has examined the correlation of a symptomatic cause with a fixed or unilateral pain location, and many patients without a secondary cause report a static location of stabbing pain.⁴

Recommendations for diagnostic workup of headache with any phenotype, including isolated stabbing headache, should still presumably follow accepted recommendations to rule out secondary causes in the presence of red flags.¹⁷ These red flags include systemic disease or symptoms, neurological examination abnormalities including papilledema, an abrupt onset, onset at an older age, a positional component, or any progression of a previously well-established headache pattern. In a new onset stabbing headache presenting in a patient older than age 50 years, it is reasonable to obtain an erythrocyte sedimentation rate and C-reactive protein as giant-cell arteritis has been associated with this headache phenotype.¹⁸ Unless the review of systems is suggestive for an underlying systemic or autoimmune disorder, widespread screening for underlying autoimmune disease would seem to be low yield.

TREATMENT OF PRIMARY STABBING HEADACHE

Because of the brevity of pain paroxysms, therapy is aimed at prophylactic suppression of the attacks. When primary stabbing headache occurs in the presence of another headache disorder, therapy directed at the index disorder may be appropriate and successful. When primary stabbing headache occurs in isolation, therapy may only be indicated if the attack frequency is marked. Primary stabbing headache is considered an indomethacin responsive headache syndrome,¹⁹ though unlike hemicrania continua and paroxysmal hemicrania, the response is neither universal^{3,4,20} nor part of the diagnostic criteria.¹ Alternative agents reported to be effective in small series of patients include cyclooxygenase type 2 inhibitors, melatonin, gabapentin, amitriptyline, and botulinum toxin A.⁹

REVIEW OF THE CASES

Case 1: The periodic occurrence of stabbing headache in this patient does not seem worrisome for a superimposed secondary cause of all of her headaches. It would be interesting to know if her stabbing headache had contemporaneous worsening or improvement with the course of her migraine, which might inform therapeutic decisions.

Case 2: This patient seems to have primary stabbing headache without any clear underlying secondary cause, though the late age of onset is atypical. If the duration of jabs was actually 30 seconds and she was not reporting a volley of jabs, this case would be similar to some of the patients reported by Fuh et al.⁴

Case 3: For this patient, it would also be useful to know if her stabbing headache had simultaneous worsening with her migraine attacks. If so, I would be inclined to treat with a medication that may confer prophylaxis against both disorders. In some patients, the course of stabbing headache and migraine may diverge, and then each disorder would need individually targeted therapy.

Case 4: A childhood or adolescent age of onset of stabbing headache is not atypical, and I would approach this patient in the same manner as case 1 and case 3.

Case 5: This patient is interesting for two reasons: the first is her coexisting autoimmune disorders that perhaps place her at an elevated risk of stabbing headache, though this is unproven. The second interesting characteristic is that she has hemifacial spasm, which could imply focal demyelination of the right 7th cranial nerve. As small regions of focal demyelination have been implicated in stabbing headache, this mechanism may be plausible in ascribing her stabbing headache to this pathogenesis. It would be interesting to know if stabbing headache in this patient emerged with a declining level of therapeutic immunosuppression for autoimmune disorders.

Case 6: It is difficult to elucidate if this patient has stabbing headache that is somehow associated with her rheumatoid arthritis or is a completely separate entity. It would be interesting to know if her stabbing headache occurred in the context of emerging symptoms referable to her rheumatoid arthritis.

Case 7: This case is also interesting because of the later emergence of an autoimmune disorder and the long duration of some attacks similar to case 2. In the case series by Rampello and colleagues,¹³ myasthenia gravis was not an encountered autoimmune disorder among the 26 patients who had stabbing headache.

Case 8: This patient has primary stabbing headache, and I would not ascribe her MRI finding of sinusitis to her headache syndrome.

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