doi:10.1111/j.1468-2982.2007.01445.x

CLINICAL CORRESPONDENCE

Bilateral paroxysmal hemicrania with autonomic symptoms: the first case report

Randolph W Evans

Baylor College of Medicine, Houston, USA

Randolph Warren Evans, Baylor College of Medicine, 1200 Binz #1370, Houston 77004, USA. Tel. +1 713 528 0725, fax +1 713 528 3628, e-mail rwevans@pol.net

Received 29 April 2007, accepted 4 July 2007

Introduction

Paroxysmal hemicrania (PH) is a rare headache disorder of unknown aetiology with a mean age of onset of 34 years, with a range of 6–81 years. The pain is always unilateral, often severe, with a duration between 2 and 30 min (occasionally 2 h), and a frequency of attacks of 2–30 per 24 h with an even circadian distribution (1). Ipsilateral autonomic symptoms occur during attacks with the following frequencies: lacrimation, 62%; conjunctival injection, 36%; nasal congestion, 42%; and rhinorrhea, 36% (2). Eyelid oedema, ptosis, miosis and facial sweating are less frequently reported. There is a female to male predominance of about 2.36 : 1. Side shift has been rarely reported (3). There are two forms: the episodic, which occurs during a period lasting 7 days to 1 years separated by pain-free periods lasting 1 month or more; and the chronic, which has attacks that occur for more than 1 year without remission or with remissions lasting less than 1 month.

To my knowledge, this is the first case report of bilateral PH with cranial autonomic symptoms.

Case report

This 55-year-old white male was referred by his neurologist for a headache consultation. Almost 4 years previously, the patient developed a new type of headache with a duration of 8 min to 20 min, with an average of about 15 min, occurring any time of day or night. The headache was always bilateral in location, either behind both eyes or bitemporal or behind the nose associated with tearing of both eyes and a little clear drainage from both nares. There was no ptosis, conjunctival injection, congestion of the nares, eyelid oedema, or facial sweating. During the first 3 days of attacks, the pain would be throbbing with an intensity on a visual analogue scale of 4–5/10 and, then, for the remainder of the attack, would become an aching with an intensity of 10/10. The maximum intensity of the pain would occur within about 1 min. During a headache, he would prefer to sit still. The headache would not be precipitated by bending or rotating of the head or alcohol.

The patient had kept a headache diary for the prior 6 months from June through November 2006 including dates of attacks, number per day, and time of day divided into four, 6-h blocks. His recollection was that the pattern had been similar since the onset. He had multiple daily headaches for 11–19 days with pain-free periods of 13–25 days as follows (headache days in bold), 19-**11**-25-**15**-15-**16**-16-**19-**13, and then 26 headache days out of the last 30 (with the 4 pain-free days in the last 2 weeks). The number of attacks per day ranged from 1 to 10, occurring any time of day or night with no predilection for any particular time. The headaches could awaken him from sleep as many as three times nightly. The number of headaches per day for each episode was as follows:

2-3-6-9-10-10-9-10-7-9-8-10-5-2-1;
4-2-4-7-8-6-6-2-2-2-2-3-3-1-2;
2-4-5-7-7-9-8-7-6-4-6-?-2-2-3-5-6-1;
2-3-2-2-3-7-6-3-5-6-3-5-6-4-7-2-3-3-4-1-1-2-2-3-1-2-2-1-2-5.

The first neurologist asked the patient to stop drinking beer for 1 week but there was no improvement in the headaches. He then started the patient on doxepin 10 mg at bedtime for 5 weeks and then 20 mg at bedtime for an additional 7 weeks without benefit. The first neurologist then discontinued doxepin and started indomethacin 25 mg three times daily. The headaches ceased immediately after the first dose.

The patient had previously been evaluated by his primary care physician but did not take any medications for the headache. A CT scan of the brain with and without contrast in 2004 was normal. A Westergren erythrocyte sedimentation rate on 30 November 2006 was 1 mm/h.

There was a prior history of headaches occurring for many years about once every 6 months, lasting 2–3 days, which had not occurred for a few years. The headaches were described as a generalized aching with an intensity of 2–3/10 without associated symptoms, for which he would take acetaminophen or aspirin.

There was a past medical history of a myocardial infarction requiring pacemaker implantation in February 2003, hypertension, and hyperlipidemia. Medications included ramipril, amlodipine and metoprolol. He smoked two packs of cigarettes weekly and drank four beers daily. Family history was negative for migraine or similar headaches. Neurological examination was normal.

The patient was followed-up by telephone twice. Two weeks after the consultation, he had only two headaches within a few hours and had taken 25 mg of indomethacin. On follow-up 5½ months later, he reported approximately two to three similar headaches within 4 or 5 h about once per month (he had stopped keeping a headache diary). He would take a 25-mg of indomethacin after the second or third, and then have one or no more headaches until the next month.

Discussion

PH is defined by the following International Headache Society (IHS) 2nd edition criteria: 20 attacks of headache; severe, unilateral orbital, supraorbital or temporal pain that lasts 2–30 min; headache is accompanied by at least one of the following ipsilateral signs, conjunctival injection and/or lacrimation, nasal congestion and/or rhinorrhea, eyelid oedema, forehead and facial sweating, miosis and/or ptosis; attacks have a frequency above 5 per day for more than half of the time, although periods with lower frequency may occur; attacks are prevented completely by therapeutic doses of indomethacin, and not attributed to another disorder (4).

Sjaastad reported that some patients with PH had pain that crossed the midline slightly (5, 6, 7). There are three case reports (6, 7, 8) of short-lasting, frequent, bilateral, indomethacin-responsive headaches without cranial autonomic features that have been presented as bilateral PH. However, Matharu and Goadsby (9) propose that these cases are not bilateral PH at all but may constitute a new category of primary headache and propose the term, 'bilateral paroxysmal cephalalgia'. Using the IHS 2nd edition criteria for PH and changing unilateral to bilateral, our patient fulfils the criteria for chronic bilateral PH with cranial autonomic symptoms, the first reported bilateral case of either the episodic or chronic form. Interestingly, once started on indomethacin, the pattern of headaches changed and became much less frequent, at least for the first few months of follow-up. However, the significance of this pattern is not certain, as the natural history of PH is not well described (1). Additional case reports will certainly be of interest.

Acknowledgements

I thank Richard A. Sawyers, MD, of Victoria, Texas, USA, for referring the patient. He started the patient on indomethacin.

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