Hemicrania continua with aura

MFP Peres, Hua Chiang Siow & TD Rozen

Department of Neurology, Jefferson Headache Center, Thomas Jefferson University Hospital, Philadelphia, Pennsylvania, USA

Cephalalgia

Peres MFP, Hua Chiang Siow & Rozen TD. Hemicrania continua with aura. Cephalalgia 2002; 22:246–248. London. ISSN 0333-1024

Hemicrania continua is a primary headache disorder that is characterized by a continuous unilateral headache of moderate severity, exacerbations of severe pain and complete responsiveness to indomethacin. We report four patients with a unique variant of hemicrania continua: visual auras that precede or accompany the pain exacerbations. □ *Aura, hemicrania continua, indomethacin*

Mario F.P. Peres, Jefferson Headache Center, Department of Neurology, Thomas Jefferson University Hospital, 111 South Eleventh Street, Gibbon Building, Suite #8130, Philadelphia, Pennsylvania 19107, USA. Tel. +1 215 955 2243, fax +1 215 955 2060, e-mail marioperes@yahoo.com *Received 9 January 2001, accepted 20 November 2001*

Introduction

Hemicrania continua (HC) is characterized by a continuous unilateral headache of moderate severity, exacerbations of severe pain and associated migrainous and autonomic features (tearing, conjunctival injection, ptosis) (1, 2). HC almost invariably has a prompt and enduring response to indomethacin (3).

Even though migrainous features can be part of the HC syndrome, aura has never been reported in HC patients. In 1992, Antonaci (4) reported a patient, with hemicrania continua secondary to a mesenchymal tumour, who described dark spots in his visual fields associated with the headaches, but this was not consistent with a true 'migrainous aura'.

Herein we report four patients with HC and aura-type spells. Case descriptions are summarized in Table 1.

Case 1

A 44-year-old white male with a 25-year history of episodic headaches started to have daily headaches 2 years prior to evaluation. The headaches were strictly right-sided, in a frontal distribution. A baseline, moderate intensity headache was present continuously and exacerbations also occurred. With the exacerbations he experienced unilateral tearing, conjunctival injection, photophobia, phonophobia and nausea. He also complained of seeing light flashes in his left peripheral visual field that lasted 5 min and accompanied the headaches about 5% of the time. The visual symptoms did not precede the pain exacerbations but came on during the increased pain periods. Neurological examination was unremarkable, except for trigger point tenderness in his neck. Vital signs and clinical examination were normal. Computed tomography and magnetic resonance imaging (MRI) were normal. The patient had complete alleviation of his headache and resolution of the auras with indomethacin, 150 mg a day.

The patient has been followed for 8 months and is headache free on 150 mg of indomethacin. He was unable to come off the medication because of headache and aura recurrence with stoppage of indomethacin.

Case 2

A 44-year-old white female had a 20-year history of a continuous left-sided headache, located in the orbital temporal region. Her baseline headache was mild to moderate in intensity. She had exacerbations of more severe pain that occurred every day and lasted 2 h. She had no autonomic symptoms but had nausea, diarrhoea, photophobia and phonophobia were associated with the headache spikes. She also complained of bright white flashes, which she described as sparkles, that lasted 5–10 min. These white flashes usually shortly preceded pain exacerbations but could also accompany them. The visual phenomena were located in her entire visual field (more central than peripheral) and occurred with 10% of her headache exacerbations. She had neck tenderness on examination. Neurological examination and brain MRI were normal. Indomethacin, 75 mg per day, gave complete pain and visual aura

Table 1 Visual symptoms in patients with hemicrania continua

Patient	Side of headache	Side of visual auras	Onset related to exacerbation	Duration (min)	Aura frequency (%)	Shape	Colour
1	Right	Left	Accompanying	5	5	Long, floaters	White flash lights
2	Left	Bilateral	Preceding	5–10	10	Rectangular	Sparkles, white flashes
3	Right	Right	Preceding	5	15–20	Bean	Black spots
4	Right	Right	Preceding	15	20	Round	Bright

relief. The patient has been essentially headache free during 12 months of follow-up; however, the dose of indomethacin had to be increased to 150 mg. She has not been able to decrease the dose to less than 75 mg a day because of headache and aura recurrence.

Case 3

A 44-year-old white female had a 25-year history of unilateral, continuous, right-sided ocular headaches. She had pain exacerbations with accompanying symptoms of right eye ptosis, nausea, vomiting, phonophobia and photophobia. Preceding 20% of the headache exacerbations she reported seeing 10–20 bean-shaped black spots in her right visual field, lasting 5 min. The spots were centrally located for the first few minutes and would then travel to the right peripheral visual field. Clinical and neurological examination were normal. MRI of the brain was normal. The headache and visual symptoms completely responded to indomethacin, 50 mg a day.

In 18 months of follow-up, the patient tried to discontinue the medication several times but the headaches and auras recurred.

Case 4

A 58-year-old white female had a 4-year history of a continuous right-sided headache in her fronto-orbital region. Her baseline headache was mild to moderate in intensity. She had exacerbations of more severe pain that lasted 2 h and occurred every day. She had associated ptosis, photophobia and phonophobia. She also complained of seeing bright, round-shaped flashes with 20% of her pain exacerbations, lasting 15 min, always preceding the pain spikes. Visual symptoms were located in the right upper quadrant of her visual field. No visual symptoms were observed independently to the headaches. Physical examination and brain MRI were normal. Her headaches responded to indomethacin, 75 mg per day, and the auras did not appear while she was on the medication. The patient has been followed for 12 months. Any attempt to discontinue indomethacin resulted in headache and aura recurrence.

Discussion

Auras do not appear to be a migraine-dependent phenomenon. Auras have been shown to occur with cluster headaches and now we report aura with HC. This appears to be the first description of aura with HC.

All of our patients had typical HC with unilateral continuous headaches, pain exacerbation with associated migrainous and autonomic features, and responsiveness to indomethacin. All the cases had typical visual aura descriptions. In two patients (cases 3 and 4) the visual symptoms occurred on the same side as the headache, and in one patient (case 1) the aura occurred in the contralateral visual field. Three patients experienced their auras just before the pain exacerbation while the other one experienced the aura during pain exacerbation. These patients never experienced auras without headaches. Their auras met the International Headache Society criteria for migraine with aura. Indomethacin provided complete relief for both the headaches and the visual symptoms, suggesting that the auras might be pathophysiologically related to the headaches in HC. It is important to note that a past personal history of migraine, or family history of migraine or auras were absent in all the described patients.

Migraine and HC have been linked in the past. Evers (5) reported a patient with familial hemiplegic migraine and HC and many migraine-associated symptoms have been reported during HC exacerbations (2, 6, 7). HC may be a migraine subtype, which is why aura can be associated with it. Auras only occur preceding or during a pain exacerbation period, the time HC patients develop migrainous features. More probable is that auras can be an independent phenomenon that can accompany any form of primary headache disorder.

If aura is really an independent phenomenon, one gene could be responsible for the aura and another gene

for the primary headache. This would help explain why aura-type symptoms have been described with headache disorders other than migraine (5, 8). Genetic studies and functional imaging of these HC with aura patients could help clarify this issue, while further descriptions are necessary to better understand this probable new HC subtype.

References

1 Silberstein SD, Lipton RB. Chronic daily headache. Curr Opin Neurol 2000; 13:277–83.
2 Bordini C, Antonaci F, Stovner LJ, Schrader H, Sjaastad O. 'Hemicrania continua': a clinical review. Headache 1991; 31:20–6.
3 Pareja J, Sjaastad O. Chronic paroxysmal hemicrania and hemicrania continua. Interval between indomethacin administration and response. Headache 1996; 36:20–3.
4 Antonaci F, Sjaastad O. Hemicrania continua: a possible symptomatic case, due to mesenchymal tumor. Funct Neurol 1992; 7:471–4.
5 Evers S, Bahra A, Goadsby PJ. Coincidence of familial hemiplegic migraine and hemicrania continua? A case report. Cephalalgia 1999; 19:533–5.
6 Newman LC, Lipton RB, Solomon S. Hemicrania continua: ten new cases and a review of the literature [see comments]. Neurology 1994; 44:2111–4.
7 Espada F, Escalza I, Morales-Asin F, Navas I, Inignez C, Mauri JA. Hemicrania continua: nine new cases. Cephalalgia 1999; 19:442 (Abstract).
8 Silberstein SD, Niknam R, Rozen TD, Young WB. Cluster headache with aura. Neurology 2000; 54:219–21.