

Side-shifting hemicrania continua with aura (migraine with aura with autonomic symptoms responsive to indomethacin?)

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Atypical features of hemicrania continua (HC), including both visual aura and side shifting, have been reported previously. However, auras and variable unilaterality have never been reported together in HC. We report two patients with side-shifting HC with aura. These patients' symptoms are unilateral headaches, visual aura, autonomic features, throbbing pain, nausea and photo/phonophobia. One could speculate that the unilaterality and/or the autonomic symptom modules are indomethacin responsive. The patients can also be classified as chronic migraine with aura, with autonomic symptoms, responsive to indomethacin. Neither migraine subtype nor side-shifting HC with aura is included in the current International Headache Society (IHS) classification, so these patients are not classifiable. Side-shifting HC with aura implies the need to revisit the traditional IHS categorization of headaches into unique diagnostic groups. The modular headache theory may be a tool for the understanding of these rare and complex cases.

□ *Aura, hemicrania continua, migraine, modular headache theory*

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Introduction

Hemicrania continua (HC) is an indomethacin-responsive headache, characterized by a continuous unilateral headache with exacerbations of severe pain and associated migrainous and autonomic features (tearing, nasal congestion, conjunctival injection, ptosis, rhinorrhoea and eyelid oedema) (1). Migrainous features are seen in most patients in the exacerbation period (59% with photophobia and phonophobia, 53% with throbbing pain (2)), thus HC could be misdiagnosed as chronic migraine or chronic tension-type headache with superimposed migraine.

HC was once thought to be a rare headache disorder, but recent reports have shown that HC is more common than previously believed; it is an underrecognized headache syndrome (2).

Atypical features of HC have been reported. Evers (3) has described a patient with familial hemiplegic migraine and HC. Peres et al. (4) first reported four

patients diagnosed with HC with typical visual aura, preceding or accompanying headache exacerbations, supporting the concept that auras are not a migraine-dependent phenomenon. Auras have been shown to occur with cluster headaches (5) and chronic paroxysmal hemicrania (CPH) (6).

One of the essential features of HC is unilateral headache. However, bilateral (7, 8) and side-shifting cases have been reported. Newman (9) reported a patient with a remitting form of HC with strictly unilateral attacks, which alternated sides. Trucco (10) has also reported a side-shifting headache patient.

Ekbom (11) reported side alternation in 10% of episodic cluster patients. Bilateral cases have been reported in the literature in CPH (12) and in CH and a mechanism of failed contralateral suppression has been proposed by Young and Rozen (13).

Bilateral or side-shifting cases of HC may occur. However, auras and variable unilaterality have never been reported together in HC. We report two patients with side-shifting HC with aura never

previously presented, using the modular headache theory (14) to explain diagnosis, classification and clinical implications.

Case 1

A 33-year-old caucasian woman presented with a 12-year history of an episodic headache transformed to a continuous, daily headache for the past 6 months. The headache alternated sides, usually on one side for at least 24 h, up to 7 days, on average 48 h, then turning to the other side, and occurring 70% of days on the left side and 30% on the right side. Pain quality was pulsating with nausea, vomiting, photo- and phonophobia when severe. Ipsilateral autonomic symptoms (tearing, ptosis, conjunctival injection and a sensation of 'sand in the eye') accompanied headache. Visual symptoms lasting 15 min appeared every other day and preceded the pain exacerbation. They occurred in either left- or right-sided headaches. The patient responded to indomethacin 75–125 mg/day, remaining headache free for 10 months of follow-up. She could not stop the medication for more than a week without return of the headache. The patient also responded partially to rofecoxib 50 mg/day and completely to celecoxib 400 mg/day.

Case 2

A 34-year-old caucasian woman with a 18-year history of headaches had developed continuous headaches in the past year. The headache was always unilateral; however, it shifted sides, 50% on the left side, 50% on the right side, usually 72 h on each side. Ipsilateral autonomic symptoms (tearing, ptosis and nasal congestion) followed the pain. Photophobia and phonophobia occurred during exacerbations. Headache was throbbing type. Black spots and flashing lights lasting 20 min appeared every other week. Auras occurred in right- and left-sided headaches and did not predict the switch of sides. The patient responded to indomethacin 150 mg/day. Other treatment options were not tried.

Discussion

We report two patients with similar features: continuous unilateral headaches, side shifting, accompanied by autonomic symptoms, visual aura, photo- and phonophobia, and complete response to indomethacin (Fig. 1). These patients represent a diagnostic and classification challenge. They can be classified as side-shifting HC with aura, a condition that has not been previously reported, or as chronic

migraine with aura, with autonomic symptoms, responsive to indomethacin.

The report implies the need to revisit the traditional International Headache Society (IHS) categorization of headaches into unique diagnostic groups. Neither migraine subtype nor side-shifting HC with aura is included in the current IHS classification, so these patients are not classifiable. Absolute indomethacin responsiveness is considered to be a diagnostic feature of HC and to be characteristic of several other disorders, not including migraine.

The modular headache theory explains how headaches with the features of several different primary headache disorders may occur. It may be a tool for understanding classification and diagnosis of these rare and complex cases. These patients' headaches involve aura modules, unilaterality modules, autonomic symptom modules, the throbbing pain module and the nausea and the photo/phonophobia modules.

Based on the IHS classification, the cases reported here have two atypical features of HC: side shifting and aura. According to the current IHS criteria, side shifting is an exclusionary feature, yet cases which shift sides have been previously described, and then excluded from the classification system. Like cluster headache, it is reasonable to accept that the basic biology of HC could be maintained with side shifting. Other typical features, including continuous headache, autonomic symptoms and absolute indomethacin response and, in case 1, the sensation of 'sand in the eye', are present. On the other hand, the occurrence of autonomic symptoms and complete response to indomethacin are atypical of migraine, or at least not included in the IHS diagnostic criteria for migraine. Aura and unilaterality have long been recognized as typical symptoms in migraine. The above patients are diagnosable in both distinct ways.

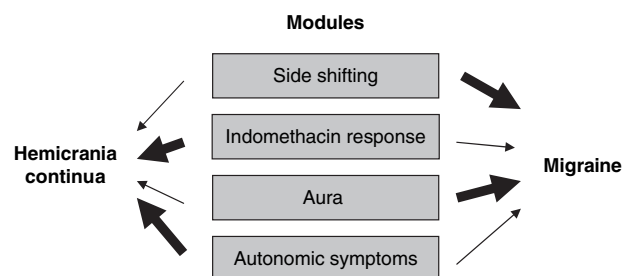


Figure 1 Modules of unilaterality, complete response to indomethacin, visual aura, autonomic symptoms and their possible links with migraine or hemicrania continua diagnosis.

A more useful, descriptive approach, with a broader view than the IHS diagnostic classification system, is needed for the classification of rare headache syndromes. One could speculate that the unilaterality modules and/or autonomic symptom modules are indomethacin responsive. Future studies regarding the indomethacin response rate in migraine with particular features, such as strict unilaterality, autonomic symptoms or both, may clarify our understanding of the underlying biology of indomethacin-responsive headaches and improve migraine management.

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