



## Case Report

### A Case of Hemicrania Continua Presenting As Temporomandibular Pain and Responding To Topiramate

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## Summary

Hemicrania continua (HC) is a primary headache disorder, characterized by unilateral, unremitting headache of moderate to severe intensity, associated with ipsilateral autonomic signs and an absolute response to indomethacin. We report a case of a 34-year-old woman who presented with strictly left-sided unremitting headache whose initial symptoms suggested a temporomandibular disorder, but the patient was ultimately diagnosed with hemicrania continua. The patient displayed an absolute response to a trial of indomethacin but she had to quit soon due to gastric intolerance and vertigo. Then topiramate was prescribed which she responded completely. This case and the other reports as well suggests that the differential diagnosis of temporomandibular disorders and HC is challenging. HC generally needs to be treated lifelong and indomethacin may not be a good choice because of its side effect profiles while topiramate may be considered as an alternative. The IHS criteria of HC need to be revised to provide a broad based description thus providing new therapeutic options.

**Key words:** Hemicrania continua, temporomandibular pain, topiramate, indomethacin, autonomic sign

### Temporomandibüler Ağrı Olarak Ortaya Çıkan ve Topiramate Yanıt Veren Hemikrania Continua Olgusu

## Özet

Hemikrania continua, tek taraflı, otonom bulguların eşlik ettiği, şiddetlenmelerle giden, devamlı ortaya çıkan ve indometazine yanıt veren bir primer baş ağrısı türüdür. Başının sol yarısında, devamlı olarak ortaya çıkan baş ağrısı yakınması olan 34 yaşındaki kadın vaka önce ilgili tarafta temporomandibüler patoloji tanısı almış, ardından tarafımızdan hemikrania continua tanısı konulmuştur. Hastanın ağrısı indometazine mutlak olarak yanıt vermiştir, ancak hasta, gastrik hassasiyet ve baş dönmesi nedeniyle tedavisini sürdürememiştir. Hastanın ağrısı topiramate ile de geçmiştir. Bu ve buna benzer vakaların ışığında denilebilir ki, zaman zaman hemikrania continua ile temporomandibüler patolojinin ayırıcı tanısında güçlükler yaşanmaktadır. Hemikrania continua çoğunlukla ömür boyu tedavi gerekmektedir. Tedavide indometazin kullanımı, yan etkileri nedeniyle kısıtlıdır. Bu nedenle topiramate tedavide iyi bir alternatif gibi görünmektedir.

**Anahtar Kelimeler:** Hemikrania continua, temporomandibüler ağrı, topiramate, indometazin, otonomik bulgu

## INTRODUCTION

Hemicrania continua (HC) is a primary headache disorder characterized by strictly unilateral, continuous headaches of moderate intensity and absolute response to indomethacin. It was first described by Sjaastad and Spierings in 1984<sup>(9)</sup>. This severe pain is often associated with autonomic disturbances ipsilateral to the side of the pain, such as nasal congestion, rhinorrhea, lacrimation, conjunctival injection, ptosis and miosis. Some patients have typical migraine symptoms, such as photophobia, phonophobia, nausea and throbbing pain. Response to treatment with indomethacin has been considered as a diagnostic criterion<sup>(5)</sup>.

HC presents in continuous or remitting forms. Headaches occur daily and sometimes for years in the continuous form.

We describe a case of HC presenting with ipsilateral temporomandibular pain that was successfully treated with topiramate, which seems to be a good alternative for indomethacin and has fewer side effects. HC has some common features with temporomandibular disorders and should be considered in differential diagnosis.

## CASE PRESENTATION

A 34-year-old woman reported strictly left-sided headache for 6 years. She experienced a remitting pain that presented mainly at the left preauricular and temporomandibular area radiating to the left orbit. Her pain started shortly after three of her wisdom teeth were removed on the same day. Then, she started to experience the pain daily with moderate intensity. The pain was exacerbated in the evening and was accompanied by ipsilateral eyelid and face edema. She denied any other autonomic feature. These headaches had a sharp quality and were sometimes associated with nausea. She experienced exacerbation of the pain during menstruation, windy weather conditions and stressful events.

At first, the pain which had appeared after the surgical dental procedure, was treated by her dentist with an injection which was probably a local anesthetic, in her left temporomandibular joint. This treatment caused temporary relief. She was given several nonsteroidal anti-inflammatory drugs, which were ineffective. As all these treatments failed, she was sent to a psychiatrist.

The patient was admitted to our clinic because her pain appeared to be unremitting. Her neurological examination was normal. She did not experience pain with mastication or jaw movements and did not report a limited range of mandibular motion. Biochemical screening was normal. Cervical, cranial and temporomandibular joint magnetic resonance imaging (MRI) did not reveal any pathology. These findings were not compatible with temporomandibular disorder.

The patient was given a trial of 75 mg indomethacin daily. The patient experienced complete relief of pain, which confirmed the diagnosis of HC. Despite the good response to indomethacin, she had to discontinue the medication because of side effects such as nausea, dizziness and hypotension. She was started on topiramate, which was gradually increased to a daily dose of 150 mg. The patient had a complete response to topiramate as well.

## DISCUSSION

In the differential diagnosis of HC, new daily persistent headache<sup>(3)</sup>, unilateral chronic migraine, chronic daily headache and the other trigeminal autonomic cephalalgias, such as cluster headache, chronic paroxysmal hemicrania and secondary causes of unilateral headache should be considered. These were excluded with patient history, accompanying symptoms and as well as by normal cranial and cervical MRI findings. The headache was strictly unilateral and never paroxysmal contrary to migraine. It had a positive response to indomethacin<sup>(3)</sup>.

Either not experiencing pain with mastication or jaw movements, or absence of limitation in mandibular joint together with normal examination and normal temporomandibular joint MRI and unsuccessful treatment focusing temporomandibular joint, argued against temporomandibular joint disorder.

The patient reported unilateral eyelid and face edema as autonomic features. These autonomic features are not included in the International Headache Society (IHS) criteria of HC; however, eyelid and face edema are included as autonomic features in the IHS cluster headache criteria. The complete response to indomethacin and the exclusion of other unilateral headaches together with typical clinical picture and autonomic features is compatible with HC, and no other diagnosis is likely<sup>(5)</sup>. Even though the mentioned cranial autonomic features are not included in the 2004 IHS criteria, there are reports mentioning a wider range of cranial autonomic features in HC<sup>(2)</sup>. The patient responded to indomethacin therapy but could not continue because of serious side effects, such as dizziness and gastrointestinal intolerance. Indomethacin treatment is not tolerated by many. HC is considered as a life long disorder<sup>(8)</sup>; life long indomethacin may not be a good choice because of its side effect profiles. Therefore, there is need for alternative medicine. Topiramate, which elicited a positive response from our patient, may be a promising alternative treatment for HC. Topiramate efficacy is also observed in other reported patients with HC<sup>(6,1)</sup>.

HC is a new disease entity. The diagnostic criteria are constantly evolving and as new data are reported, diagnostic criteria have been repeatedly modified and revised<sup>(5,7)</sup> to provide a broad based description of the syndrome<sup>(2)</sup>. This was also previously the case for cluster headache. With additional clinical observations, the autonomic criteria for CH were widened to cover more features. The IHS criteria of HC for

autonomic features need to be revised. It should include a wider range of autonomic features<sup>(5)</sup>. As shown by the observed autonomic features of this case and others in the literature, many HC patients do not fulfill the IHS criteria because of limited worldwide clinical experience in HC.

Goadsby and Lipton have proposed a new set of criteria, where indomethacin responsiveness should be a confirmatory trait, instead of a sine qua non diagnostic criterion<sup>(4)</sup>. To our opinion this proposal is appropriate to facilitate efforts for drug trials with less side effects so that new drugs providing new therapeutic options can be established.

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## REFERENCES

1. *Brighina F, Palermo A, Cosentino G, Fierro B. Prophylaxis of hemicrania continua: two new cases effectively treated with topiramate. Headache. 2007 Mar;47(3):441-3.*
2. *Cittadini E, Goadsby PJ. Hemicrania continua: a clinical study of 39 patients with diagnostic implications. Brain 2010 133; 1973-1986)*
3. *Goadsby PJ, Boes C. New daily persistent headache. J Neurol Neurosurg Psychiatry. 2002 Jun;72 Suppl 2:ii6-ii9.*
4. *Goadsby PJ, Lipton RB. The hemicrania continua diagnosis. Cephalalgia 2002 Sep;22(7):563-4; author reply 564-5.*
5. *7. Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders, 2nd edn. Cephalalgia 2004; 24 (Suppl.1): 1-160.*
6. *Matharu MS, Bradbury P, Swash M. Hemicrania continua: side alteration and response to topiramate. Cephalalgia 2006 26(3):341-4*
7. *Pareja JA, Antonaci F, Vincent M. The hemicrania continua diagnosis. Cephalalgia 2001;21:940-6.J.*
8. *Prakash S, Husain M, Sureka DS, Shah NP, Shah ND. Is there need to search for alternatives to indomethacin for hemicrania continua? Case reports and a review. J Neurol Sci. 2009 Feb 15;277(1-2):187-90. Epub 2008 Nov 29.*
9. *Sjaastad O, Spierings EL. 'Hemicrania continua' : another headache absolutely responsive to indomethacin. Cephalalgia 1984; 4: 65-70*