https://doi.org/10.4082/kjfm.24.0178 • Korean J Fam Med 2025;46:48-51

KJFM
Korean Journal of Family Medicine

eISSN: 2092-6715

■ Case Report

Hemicrania continua with rhinosinusitis: a case report

Dae Hyun Kim[1,*], Yoonseo Lee[2]

[1]Department of Family Practice, Keimyung University School of Medicine, Daegu, Korea
[2]Department of Neuroscience, Wellesley College, Wellesley, MA, USA

Hemicrania continua (HC) is an indomethacin-responsive headache, characterized by unilateral and continuous headaches with cranial autonomic symptoms. Various pathologies, including sinus-related conditions, are associated with HC. Here, we report the case of a 62-year-old man with HC and rhinosinusitis. The patient complained of a unilateral continuous headache with ipsilateral cranial autonomic symptoms, conjunctival injection, and eyelid edema for 20 months. HC was identified as a potential diagnosis based on the symptom criteria, and a response to indomethacin confirmed the diagnosis. Trigeminal autonomic cephalalgia should be considered in patients with unilateral headaches and cranial autonomic symptoms.

Keywords: Headache; Cephalalgia; Sinusitis; Autonomic; Case Reports

Received: July 29, 2024, Revised: October 16, 2024, Accepted: November 4, 2024
*Corresponding Author: **Dae Hyun Kim** https://orcid.org/0000-0002-7313-2384
Tel: +82-53-258-4175, Fax: +82-53-258-4171, E-mail: dhkim@dsmc.or.kr

Introduction

Hemicrania continua (HC) is a primary headache disorder that presents as unilateral continuous headache with ipsilateral cranial autonomic symptoms (CAS) [1]. It is categorized as a trigeminal autonomic cephalalgia (TACs). The diagnostic criteria of HC as per the 3rd edition of the International Classification of Headache Disorders (ICHD-3) states that the headache should be present for more than 3 months with severe exacerbations at moderate or greater intensity [2]. Moreover, the patient should show at least one of the following CAS symptoms: conjunctival injection and/or lacrimation, nasal congestion and/or rhinorrhea, eyelid edema, forehead and facial sweating, miosis and/or ptosis, or a sense of agitation or aggregated pain by movement [2]. Headache from HC consists of continuous background pain, which tends to be dull with pressure-like tension, and exacerbations that vary in intensity, duration, and frequency [3]. The prevalence of HC is estimated to be between 1.3% and 2.3% of all patients presenting with headaches [4], with a more recent meta-analysis demonstrating 1.8% of patients [5]. It is more common in females by a ratio of 1:1.8 [4].

Medina and Diamond [6] were the first researchers to describe the symptoms of HC, which were categorized as background vascular headaches from cluster headache variant. This type of headache was defined as HC by Sjaastad and Spierings [7]. Some important features of HC that distinguish it from other headache disorders are unilateral continuous background pain and patient response to indomethacin. However, background pain is often not considered by physicians and patients, leading to misdiagnosis as other primary headaches, such as migraines [3]. The mean delay of diagnosis for HC is currently 8±7.2 years [3], and the diagnosis is made certain with the patient's improvement with indomethacin, although some patients are unresponsive to the drug.

Case Report

A 62-year-old man visited the outpatient clinic with a history of 1-year unresolved left-sided headache. He had previously visited other hospitals and private clinics where he did not receive a diagnosis or effective medicine for his pain. His first symptoms appeared in September 2022, presenting as restlessness, agitation, CAS of conjunctival injection, left-sided periorbital tingling, nausea, and anorexia. Periorbital pain occurred 5–6 times per day and persisted for 5 minutes each. He rated the pain as 2/10 on the Visual Analog Scale (VAS) and reported no stress factors that could trigger his pain. Dexibuprofen, gabapentin, amitriptyline, mirtazapine, and sumatriptan were administered in several hospitals without significant symptom improvement.

The patient was admitted to the hospital to undergo a magnetic resonance imagining (MRI) scan, diffusion, and magnetic resonance angiography of the brain and carotid gland, and seemed to have sinusitis in the frontal, ethmoid, and sphenoid sinuses. However, sinusitis treatment did not improve the headaches. The pain intensity gradually increased to 3/10 on the VAS and became continuous. He also started to feel discomfort in his throat. In April 2024, he was readmitted for detailed examinations and tests. In addition to previously reported symptoms, the patient experienced fatigue and pain in the right foot. The nerve conduction velocity test and blink reflux study results were normal. A thyroid function test and throat imaging were conducted to address the throat discomfort, but his thyroid results showed no abnormalities. Other routine examinations did not deliver meaningful results, besides showing the progression of degenerative spondylosis. However, blood tests demonstrated abnormally high uric acid levels at 9.8 mg/dL (normal range, 3.4–7 mg/dL). Suspecting that this may have caused the arthralgia, 40 mg/T febuxostat was prescribed without any improvement in periorbital cephalalgia.

The patient underwent computed tomography (CT) of the chest and paranasal sinus (PNS). Chest CT showed mild emphysema in both lungs, a few tiny nodules in the left upper lung that were probably benign, and small reactive lymph nodes in the mediastinum and hilar areas due to smoking history of 40 pack-years.

The PNS CT results in 2024 indicated that the patient had previously undergone bilateral uncinectomies, maxillomandibular advancement, and ethmoidectomies, which he reported having undergone in 2008. The CT showed polypoid soft tissue thickening in both nasal cavities and ethmoid sinuses, with a few areas of mild enhancement (Figure 1). The right frontal sinus demonstrated a soft-tissue lesion with soft-tissue opacification and bubbly features. There was some mild polypoid soft tissue thickening on the left frontal sinus and left maxillary sinus, and scanty soft tissue thickening of the right maxillary and both sphenoid sinuses. The right nasal cavity showed a polypoid lesion with mild contrast enhancement. These results suggest a case of chronic rhinosinusitis in the frontal, ethmoid, and sphenoid sinuses.

The patient met all the ICHD-3 criteria for HC. The patient showed a continuous periorbital left-sided headache that lasted for more than 3 months and CAS of conjunctival injection, eyelid edema, diaphoresis

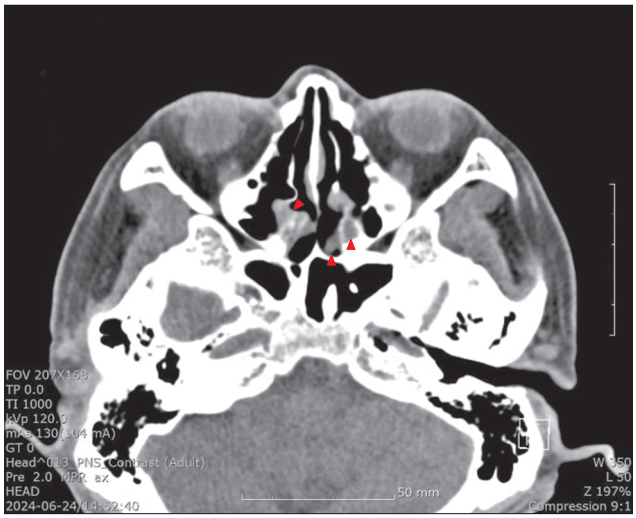

Figure 1. Paranasal sinus computed tomography results. Red arrowheads indicate areas with nasal sinus wall thickening.

https://doi.org/10.4082/kjfm.24.0178

Table 1. Comparison of HC diagnostic criteria with the patient's symptoms

Diagnostic criteria for HC	The patient's symptoms
A. Unilateral headache fulfilling criteria B–D	Continuous left-side periorbital headache
B. Present for >3 months, with exacerbation of moderate or greater intensity	Present for a year
C. Either or both of the following: 1. At least one of the following symptoms or signs, ipsilateral to the headache - Conjunctival injection and/or lacrimation - Nasal congestion and/or rhinorrhea - Eyelid oedema - Forehead and facial sweating - Miosis and/or ptosis 2. Sense of restlessness or aggravation of the pain by movement	Conjunctival injection, eyelid oedema, diaphoresis, and ptosis. Showed restlessness, agitation, and aggravation of the pain by movement
D. Responds absolutely to therapeutic doses of indomethacin	Responded to 75 mg of indomethacin
E. Not better accounted for by another ICHD-3 diagnosis	

HC, hemicrania continua; ICHD-3, the 3rd edition of the International Classification of Headache Disorders.

(temporarily), and ptosis (temporarily). The patient also experienced an aggregated headache due to certain movements and demonstrated nausea with restlessness and agitation. Based on these symptoms, we concluded that the patient could be experiencing HC with chronic rhinosinusitis. The diagnosis was confirmed by the patient's response to 25 mg indomethacin three times daily (Table 1).

Discussion

The patient had suffered from continuous left-side periorbital headache for 1 year and reported feeling discomfort in his throat. He reported experiencing four different CAS, increased headache pain due to movement, restlessness, agitation, and nausea. Based on these symptoms, HC was identified as a potential diagnosis, and indomethacin was prescribed, which provided the patient complete relief and confirmed the diagnosis. The patient simultaneously presented with tissue thickening and lesions in the frontal, ethmoid, and sphenoid sinuses, suggestive of chronic rhinosinusitis.

The cause and mechanism of HC are unknown, but HC has been reported to appear secondarily to certain conditions. Secondary HC most commonly occurs due to trauma, dissection, prolactinoma, and lung carcinoma [3]. It can also appear because of sinus pathology [1] such as bacterial infection of the sinus. One study described a case of secondary HC-tic syndrome, a disorder characterized by the overlap of HC and trigeminal neuralgia (TN) associated with fungal sphenoiditis [8]. After symptoms of TN appeared, TN was treated with carbamazepine and was relieved for some period [8]. However, a new form of headache that matched the descriptions of HC developed and was treated with indomethacin, which ameliorated the patient's pain for a week; however, the pain eventually returned [8]. As the patient's CT scan showed the presence of sphenoiditis, he was treated with transnasal sphenoidotomy, which resolved his pain and headache [8]. Similarly, there was a case of secondary HC in which the patient responded to indomethacin due to the reduced intensity of the continuous headache [9]. However, the symptoms of HC persisted, and MRI was performed, which revealed lesions in the sinus region [9]. The patient consecutively underwent endoscopic sinus surgery, antigen testing, intranasal ethmoidectomy, and anti-fungal medicine until they became pain-free [9]. These studies concluded that sphenoid sinusitis caused secondary HC, which completely resolved after curing the sinusitis [9]. This is likely not the case for our patient but is significant for emphasizing the relationship between sinusitis and HC.

Rhinosinusitis refers to the inflammation and swelling of the sinuses caused by infections [10]. As the sinus region is a drainage system that expels harmful or unnecessary particles from the air, an infection in one sinus region can spread to different areas [10]. Headaches caused by rhinosinusitis are often confused with primary headaches because of overlapping symptoms [11] and because headaches from sinusitis are poorly characterized [12]. Among the 100 patients with self-diagnosed sinus headache, 52% had migraines, and only 3% were accurately diagnosed with headache secondary to sinusitis [12]. However, some patients may have comorbid sinusitis and primary headache [11]. One study showed that approximately 71% of their participants had sinusitis or rhinologic disease and approximately 49% of their participants reported meeting the criteria for a primary headache disorder [13]. Based on our patient's previous history, it is likely that he comorbidly acquired primary HC with chronic rhinosinusitis.

Similarly to paroxysmal hemicrania, one of the most essential diagnostic criteria for HC is the patient's response to indomethacin, but it is still unknown how indomethacin relieves HC since the cause or mechanism of HC remains unknown. It is likely that the activation of the trigeminovascular system triggers pain via nerve fibers that innervate pain-inducing cranial vessels and the dura mater [14]. As these structures project to the trigeminocervical complex, it subsequently activates and stimulates the inner superior salivatory nucleus (SuS) [14]. SuS is the origin of cells for cranial parasympathetic autonomic vasodilator pathway [14]. From SuS, the parasympathetic reflex is activated through the petrosal nerve, sphenopalatine ganglion, and facial nerve, likely causing HC-associated CAS (conjunctival injection, nasal congestion, and lacrimation) [14]. One study demonstrated that SuS activation in animals exhibited both neuronal trigeminovascular and autonomic symptoms of TACs, and these symptoms were inhibited by indomethacin, which likely acts directly on trigeminovascular neurons and parasympathetic outflow to the cranial vasculature [14].

Therefore, TACs (trigeminal neuralgia, HC, and paroxysmal hemicranias) should be considered in patients with unilateral headache and ipsilateral CAS in primary care.

Article Information

Conflict of interest
No potential conflict of interest relevant to this article was reported.

Funding
None.

Data availability
Not applicable.

ORCID
Dae Hyun Kim: https://orcid.org/0000-0002-7313-2384
YoonSeo Lee: https://orcid.org/0009-0009-0341-5125

Author contribution
Conceptualization: DHK, YL. Data curation: YL. Formal analysis: DHK, YL. Investigation: DHK. Methodology: DHK. Software: YL. Validation: DHK. Visualization: DHK, YL. Project administration: DHK. Writing–original draft: YL. Writing–review & editing: DHK. Final approval of the manuscript: DHK.

References

1. Al-Khazali HM, Christensen RH, Lambru G, Dodick DW, Ashina H. Hemicrania continua: an update. Curr Pain Headache Rep 2023;27:543-50.
2. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia 2018;38:1-211.
3. Prakash S, Rawat KS. Hemicrania continua: an update. Neurol India 2021;69(Supplement):S160-7.
4. Prakash S, Patel P. Hemicrania continua: clinical review, diagnosis and management. J Pain Res 2017;10:1493-509.
5. Al-Khazali HM, Al-Khazali S, Iljazi A, Christensen RH, Ashina S, Lipton RB, et al. Prevalence and clinical features of hemicrania continua in clinic-based studies: a systematic review and meta-analysis. Cephalalgia 2023;43:3331024221131343.
6. Medina JL, Diamond S. Cluster headache variant: spectrum of a new headache syndrome. Arch Neurol 1981;38:705-9.
7. Sjaastad O, Spierings EL. "Hemicrania continua": another headache absolutely responsive to indomethacin. Cephalalgia 1984;4:65-70.
8. Wang C, Zhang X, Yang Y, Wan D. Secondary hemicrania continua-tic syndrome associated with fungal sphenoiditis: a case report. Clin Case Rep 2021;9:e04297.
9. Hwangbo S, Choi B, Choi I, Kim HS, Chu MK, Shin HY, et al. Invasive sphenoid sinus aspergillosis presenting hemicrania continua-like headache. J Korean Neurol Assoc 2018;36:113-5.
10. Sinus infection (Sinusitis or rhinosinusitis) [Internet]. Yale Medicine; 2019 [cited 2024 Nov 20]. Available from: https://www.yalemedicine.org/conditions/sinus-infection-sinusitis-or-rhinosinusitis
11. Ceriani CE, Silberstein SD. Headache and rhinosinusitis: a review. Cephalalgia 2021;41:453-63.
12. Eross E, Dodick D, Eross M. The Sinus, Allergy and Migraine Study (SAMS). Headache 2007;47:213-24.
13. Lal D, Rounds A, Dodick DW. Comprehensive management of patients presenting to the otolaryngologist for sinus pressure, pain, or headache. Laryngoscope 2015;125:303-10.
14. Akerman S, Holland PR, Summ O, Lasalandra MP, Goadsby PJ. A translational in vivo model of trigeminal autonomic cephalalgias: therapeutic characterization. Brain 2012;135(Pt 12):3664-75.