



# Episodic Paroxysmal Hemicrania with an Atypical Clinical Presentation: A Case Report and Review of the Literature

## *Atipik Klinik Prezantasyon Gösteren Epizodik Paroksizmal Hemikraniya: Olgu Sunumu ve Literatürün Gözden Geçirilmesi*

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### Summary

Paroxysmal hemicrania (PH) is characterized by frequent, short-lasting, severe, unilateral headaches that occur in association with cranial autonomic features. The hallmarks of this syndrome are relatively short attacks and a complete therapeutic response to indomethacin. Although PH has been classified as strictly unilateral headache in association with cranial autonomic features, a few cases have been reported that suggest bilateral PH. We report a case of PH with bilateral distribution and bilateral autonomic features. PH may actually represent a clinical spectrum including different phenotypes. (Turkish Journal of Neurology 2015; 21:71-3)

**Key Words:** Paroxysmal hemicrania, bilateral distribution, bilateral autonomic features

### Özet

Bu sendromun en belirgin özelliği atakların nispeten kısa süreli olması ve indometazin tedavisine tam olarak yanıt vermesidir. Paroksizmal hemikraniya, net olarak tek taraflı baş ağrısına eşlik eden, kraniyal otonom bulgulu bir baş ağrısı olarak sınıflanmış olsa da, az sayıdaki olguda iki yanlı olabileceği bildirilmiştir. Biz bu sunumda iki yanlı lokalizasyon ve iki yanlı otonom bulguların gözlendiği bir PH olgusu sunacağız. Paroksizmal hemikraniya, aslında farklı fenotipleri içeren bir klinik spektrumu yansıtır olabilir. Paroksizmal hemikraniya (PH), sık, kısa süreli, şiddetli, tek taraflı baş ağrısına kraniyal otonom bulguların eşlik etmesiyle karakterize bir ağrıdır. (Türk Nöroloji Dergisi 2015; 21:71-3)

**Anahtar Kelimeler:** Paroksizmal hemikraniya, iki yanlı lokalizasyon, iki yanlı otonomik bulgular

### Introduction

Paroxysmal hemicrania (PH) is a rare form of primary headache that usually begins in adulthood (1). Along with other trigeminal autonomic cephalalgias (TACs), it is classified in Group 3 in the Third Edition of The International Classification of Headache Disorders (ICHD-III) (2). The disorder has two forms: episodic, which produces attacks lasting from 7 days to 1 year, separated by pain-free periods lasting 1 month or more; and chronic, which produces attacks that occur for more than 1 year without remission or with remissions less than 1 month.

The clinical phenotype of PH includes the following features: 1) headaches are strictly unilateral and are consistently focused in the orbital, supraorbital, or temporal regions (or any combination

of these sites) in the majority of patients; 2) PH attacks invariably occur in association with ipsilateral cranial autonomic features, including conjunctival injection and/or lacrimation, nasal congestion and/or rhinorrhea, eyelid edema, forehead and facial sweating, forehead and facial flushing, sensation of fullness in the ear, and miosis and/or ptosis; 3) most episodes involve more than five attacks per day, and the pain, which is described as excruciating, usually lasts 2-30 minutes; and 4) attacks are always prevented by therapeutic doses of indomethacin (2).

Eventhough PH has often been described as strictly unilateral head pain in association with cranial autonomic features, a few cases exhibiting atypical presentation have been found in the literature. There have been several case reports of patients presenting with short-lasting, frequent, bilateral headaches that respond to indomethacin

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but are without cranial autonomic features (1). We report here an atypical case of episodic paroxysmal hemicrania (EPH) indicating bilateral distribution with bilateral autonomic features.

## Case Report

A 35-year-old woman was referred to the outpatient clinic with a 5-year history of severe, throbbing, and recurrent headaches. She reported bilateral attacks centered over the occipital and parietal areas. The headaches occurred in bouts lasting 2-10 weeks. These attacks occurred irregularly, at a rate of 10-30 per day, and each attack lasted 10-20 minutes. She reported striking reddening of her eyes, tearing, swelling of her eyelids, rhinorrhea, and restlessness during headache episodes. These autonomic symptoms occurred bilaterally and equally.

Her medical history was insignificant and included no family history of headache. Routine evaluations, including systemic physical examination, neurological examination, hematological and biochemical standardized laboratory tests, chest x-ray, and electrocardiograms, were all within normal limits. Magnetic resonance imaging of the brain revealed normal findings, and she responded well to indomethacin (50 mg three times daily).

## Discussion

We report here a case of short-lasting, frequent, bilateral headaches centered over the occipital and parietal regions of the brain with accompanying bilateral autonomic symptoms. The patient showed a complete response to indomethacin, which is an important feature in PH diagnosis. Even though it did not strictly adhere to the traditional definition of PH, including unilateral pain with ipsilateral cranial autonomic features, we consider this case to be a bilateral form of EPH.

Four characteristic features are necessary to satisfy the ICHD-III criteria for a PH diagnosis. However, the literature contains five cases of PH that indicate bilateral headaches responding to indomethacin (1,3,4,5), and only one of these cases included cranial autonomic features (4).

Two case reports of short-lasting, frequent, bilateral, indomethacin-responsive headaches without cranial autonomic features have been presented as a bilateral chronic form of PH by two different authors, Pollmann and Mulder and et al. (6,7). Bingel and Weiller reported a case with a temporal headache profile highly characteristic of EPH and a significant response to indomethacin. However, the patient's pain was symmetrically bilaterally localized and, instead of autonomic symptoms, the patient experienced migrainous features such as sensitivity to light and sound and allodynia of the affected skin area. This case, which shows bilateral localized pain without autonomic features, is the first reported atypical episodic form of PH (5). Abu Bakar et al. presented a case with features that were similar to those of previously published cases of headaches that resembled chronic PH in their temporal profile and were responsive to indomethacin but were without unilateral localization and cranial autonomic symptoms (3).

Evans reported a case of bilateral PH that did include autonomic symptoms (4). The patient's headache was always bilateral, either behind both eyes, bitemporal, or behind the nose, resulting in excess tear production in both eyes and clear drainage from both nares. This patient fulfils the criteria for chronic bilateral PH with autonomic symptoms and is the first reported bilateral case of either the episodic or chronic form.

In addition, there are rare case reports of patients with otherwise typical PH, including a good indomethacin response, who exhibit no autonomic features (1,8). Our case includes two

**Table 1. Demographical and clinical characteristics of patients with "bilateral paroxysmal hemicrania"**

	Pöllmann, Pfaffenrath (R, 6)	Mulder, Spierings (R, 7)	Bingel, Weiller (R, 5)	Abu Bakar, Chard, Matharu (R, 3)	Evans (R, 4)	Our case	
Age of onset (years)	65	35	34	66	55	35	
Gender (Female/Male)	Female	Male	Female	Male	Male	Female	
Features of pain	Location	Occiput, temples	Forehead	Forehead, temples	Occiput	Retroorbital, temporal, nasal	Occiput, parietal
	Quality	Piercing	Stabbing	Throbbing	Sharp, throbbing	Throbbing, aching	Throbbing
	Severity	Intense	Severe	Severe	Severe	Severe	Severe
Frequency of attacks (number/day)	2-12	4-20	1-24	12-25	1-10	10-30	
Duration of attacks	Several seconds- 8 minutes	Several seconds-30 minutes	5-10 minutes	10-30 minutes	8-20 minutes	10-20 minutes	
Autonomic features	No	No	No	No	Yes	Yes	
Response to indomethacin (mg/day)	Yes 25	Yes 25-50	Yes 75	Yes 25-150	Yes 75	Yes 150	

\*R, number of reference

atypical features: bilateral localization and bilateral autonomic symptoms (see Table 1). To our knowledge, this is the first case report of bilateral EPH with bilateral autonomic symptoms.

The International Headache Society criteria divide TACs into five categories: cluster headaches, PHs, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), hemicrania continua, and probable TACs (2). TACs are characterized primarily by severe and unilateral headache attacks with ipsilateral cranial autonomic features. Despite these common elements, the TACs differ in attack duration and frequency as well as in their response to indomethacin. SUNCT attacks are brief (1-600 s) and occur at least once per day for over half the time the disorder is active. Cluster headaches are longer in duration (15-180 minute), and their frequency varies between once every other day and eight times per day for over half the time the disorder is active. Hemicrania continua and PHs always respond to indomethacin. Probable TACs do not meet one of the criteria for any of these subtypes and do not fulfil all the criteria for another headache disorder (2).

Episodic paroxysmal hemicrania is a very rare variant of the trigeminal autonomic cephalalgias. Moreover, the pathophysiological mechanisms are completely unknown. Matharu and Goadsby considered these presentation differences, including bilateral localization of headaches and lack of autonomic symptoms, to be cardinal and hypothesized that these headaches may represent a novel syndrome. They proposed that these cases should be classified separately from PH and proposed the term “bilateral paroxysmal cephalalgia” to describe this novel phenotype (1).

Both the Evans case and our case are characterized by short-lasting, frequent, bilateral, indomethacin-responsive headaches similar to those described in other bilateral PH cases. However, Evans' case and ours also included bilateral autonomic symptoms. We propose that PH may actually represent a clinical spectrum including different phenotypes.

Descriptions of cases with different clinical symptoms may help in identifying the phenotypic and pathophysiologic features

of the disease. However, these cases of atypical presentation should be studied separately from PH until their pathophysiological basis is better understood.

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