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Case Report

Paroxysmal Hemicrania-Trigeminal Neuralgia (PH-Tic) syndrome with no structural lesion in a Brazilian woman: the first case report

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Abstract

Introduction
In Brazil there is only one case report of a patient diagnosed with Paroxysmal Hemicrania-Trigeminal (PH-Tic) syndrome reported, however it was observed in a patient with Chiari I malformation.
Objective
Here, we describe the first case of primary PH-Tic syndrome in the country.
Method
Case report. CARE guideline was used to guide the structuring of this article. This case report was approved by the ethics committee and has been registered under the protocol number 70705623.7.0000.5440 on "Plataforma Brasil".
Results
A 72-year-old woman with a five-month history of headaches was admitted at our headache outpatient clinic. The pain was sharp, intense, localized in the periorbital and left temporal regions. Blood counts, liver, renal and thyroid function were normal, as well as brain magnetic resonance imaging. Despite using carbamazepine, the patient had pain in only the left side of the face. Indomethacin was added until the dose of 100 mg a day and resulted in improvement of headache frequency.
Conclusion
PH-Tic should be hypothesized in patients with short-lasting headaches associated with facial pain that partially improve with carbamazepine or indomethacin.

Keywords:
Paroxysmal hemicrania
Trigeminal neuralgia
Trigeminal autonomic cephalalgias
Indomethacin
Carbamazepine

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Introduction

Trigeminal autonomic cephalalgias (TACs) and trigeminal neuralgia (TN, "tic douloureux") manifests as unilateral cranial or facial pain.[1] The coexistence in a same patient is possible and pose a challenge for the neurologist. The association between Paroxysmal Hemicrania (PH), a type of TAC, and TN (PH-tic) syndrome was first described by Hannerz in 1993.[2] Treatment focused at each of these conditions may be necessary for a successful outcome.[3]

To the best of our knowledge, in Brazil there is one case report of a patient diagnosed with Chiari malformation that presented with association of PH and TN.[4] Only 12 cases of this syndrome were reported worldwide.[2, 5-12] Here, we report the first of PH-Tic syndrome in a 72-year-old woman with no structural lesion. This case report was approved by the ethics committee and has been registered under the protocol number 70705623.7.0000.5440 on "Plataforma Brasil".

Case report

A 72-year-old woman with a five-month history of headaches was admitted at our headache outpatient clinic. The pain was localized in the periorbital and left temporal regions. It was daily, sharp, intense, sometimes throbbing, and with diffuse ipsilateral irradiation. Episodes usually lasted about 30 minutes and occurred five times a day. The patient reported left hemiface heating during the attacks, however without conjunctival hyperemia, lacrimation, nasal congestion, or rhinorrhea. She sometimes referred to shocking, short-lasting pain triggered by biting food and touching certain points of the face. Previous treatments with duloxetine, amitriptyline, lamotrigine, and carbamazepine (CBZ), initiated by a neurologist in another clinic, provided no relief. Her previous medical history was remarkable for treatment of hypothyroidism and presence of temporomandibular joint dysfunction (TMJD). Physical examinations during pain-free periods were normal except for TMJD signs. Blood counts, liver, renal and thyroid function were normal, as well as brain magnetic resonance imaging (MRI). Still using CBZ, the patient had pain in only the left side of the face. Therapy was changed to indomethacin until the dose of 100 mg a day, with significant improvement of headache frequency, the patient presented a pain-free period of up to five consecutive days. After two months she was admitted due to recurrence of pain. On this occasion, we could examine her during an attack, and the diagnostic hypothesis was done. The attack was triggered by touching some specific parts of the face, and it was characterized by a very intense sharp facial pain that lasted less than 1 minute and progressed to the frontotemporal area, associated with ipsilateral conjunctival hyperemia and eyelid oedema. Reintroduction of CBZ 400 mg a day in combination with indomethacin resulted in complete improvement. Tables 1 and 2 summarize the medications used to treat the patient and the most important events that occurred until the diagnosis of the disease.

Table 1. Medications used for the treatment of the Paroxysmal Hemicrania-Tic syndrome of the patient reported in this article

Start Date	Medication	Posology	Discontinuation Date	Time	Indication	Suspension	Observations
19/11/2019	Carbamazepine	600 mg TID			Suspected trigeminal neuralgia	Not applicable	She experienced significant dizziness during treatment, so it was necessary to reduce the daily dose. There was improvement in pain referred to as sharp and shocking, reduction in headache frequency.
05/08/2020	Lamotrigine	50 mg BID	21/09/2020	1 month and 13 days	Suspected trigeminal neuralgia	Patient reported allergy and during another neurological evaluation,	During the period of use, there were consultations in which the patient reported irregular use.
05/08/2020	Duloxetin	30 mg MID	17/09/2020	1 month	Mixed mood and anxiety disorder.	There was a worsening of the headache after the use of the medication.	
28/01/2021	Amitriptyline	50 mg BID	27/01/2022	12 months	Pain in the context of associated neuropathic pain.	After completion of the etiological investigation of the headache.	

Table 2. Key events from onset of headache to correct diagnosis of Paroxysmal Hemicrania

12/08/2020	Headache in V1 territory in point/shock and burning but reports that he has had pain all over left side with the same pattern. Attending physicians suspected paroxysmal hemicrania.	Indomethacin with a target dose of 25 mg TID.
17/10/2020	She was admitted to an emergency hospital with a headache of the same type. She also complains of sharp jabs of pain. Attacks last 30 minutes, average frequency of five times a day. Local hyperthermia and conjunctival hyperemia, in addition to eyelid edema were frequently observed. Carbamazepine was reintroduced	Indomethacin was maintained and carbamazepine 200 mg BID was introduced.
22/10/2020	She noticed significant pain relief and did not experience headache for five consecutive days.	Medications were maintained.
27/01/2022	She reported that she had two headache days a month, with significantly less intensity in relation to the pain at the beginning of the disease. At this time, she was using indomethacin 50 mg/d and carbamazepine 300 mg/d.	

Discussion

In this article we reported the first case in Brazil of primary PH-Tic syndrome, a rare association between paroxysmal hemicrania and trigeminal neuralgia. The age of onset is usually between the fifth and seventh decades of life.[13] Features such as pain of different durations, unilateral involvement of the face and partial response to indomethacin should alert to the overlap between PH and TN.

In Brazil, the coexistence of these disorders[4] as well as the combination of cluster headache and TN14 were already reported. These two articles show female patients in their sixth decade of life who had their symptoms relieved with specific medications for each type of headache - TAC and TN. The woman diagnosed with PH-Tic syndrome had a Chiari I malformation on MRI.[4] Our patient, however, had no structural lesions revealed on neuroimaging, consistent with the other cases reported in the literature.

CPH was improved with initiation of indomethacin and TN with reintroduction of CBZ. In the case reported by Monzillo et al.[4], indomethacin combined with CBZ resulted in decreased pain frequency of both types of pain. Duloxetine was ineffective and, interestingly, these medications were also discontinued in a patient diagnosed with short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT)-TIC syndrome. Lamotrigine was tried in only one patient with a combination of PH, SUNCT, and TN.[12]

PH-Tic is uncommon; however, it should be hypothesized in patients with short-lasting headaches associated with facial pain that partially improve with carbamazepine or indomethacin.

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Conflict of interest
The authors have no conflicts of interest to declare.

Author's contribution
All authors had the same contribution.

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References

1. Olesen J. **International Classification of Headache Disorders**. *The Lancet Neurology* 2018; 17(5): 396-397 Doi:10.1016/s1474-4422(18)30085-1
2. Hannerz J. **Trigeminal Neuralgia With Chronic Paroxysmal Hemicrania: the CPH-tic Syndrome.** *Cephalalgia* 2016; 13(5): 361-364 Doi:10.1046/j.1468-2982.1993.1305361.x
3. Wober C. **Tics in TACs: A Step into an Avalanche? Systematic Literature Review and Conclusions**. *Headache* 2017; 57(10): 1635-1647 Doi:10.1111/head.13099
4. Monzillo P, Nemoto P, Costa A and Rocha AJ. **Paroxysmal Hemicrania-Tic and Chiari I Malformation: An Unusual Association**. *Cephalalgia* 2016; 27(12): 1408-1412 Doi:10.1111/j.1468-2982.2007.01362.x
5. Hannerz J. **The Second Case of Chronic Paroxysmal Hemicrania-Tic Syndrome**. *Cephalalgia* 2016; 18(3): 124-124 Doi:10.1046/j.1468-2982.1998.1803123-4.x
6. Sanahuja J, Vazquez P and Falguera M. **Paroxysmal Hemicrania-Tic Syndrome Responsive to Acetazolamide.** *Cephalalgia* 2016; 25(7): 547-549 Doi:10.1111/j.1468-2982.2005.00882.x
7. Martinez-Salio A, Porta-Etessam J, Perez-Martinez D, Balseiro J and Gutierrez-Rivas E. **Chronic Paroxysmal Hemicrania-Tic Syndrome**. *Headache: The Journal*

of Head and Face Pain 2000; 40(8): 682-685 Doi:10.1046/j.1526-4610.2000.040008682.x

8. Zukerman E, Peres MFP, Kaup AO, Monzillo PH and Costa AR. **Chronic paroxysmal hemicrania-tic syndrome**. *Neurology* 2000; 54(7): 1524-1526 Doi:10.1212/wnl.54.7.1524
9. Caminero AB, Pareja JA and Dobato JL. **Chronic Paroxysmal Hemicrania-Tic Syndrome.** *Cephalalgia* 2016; 18(3): 159-161 Doi:10.1046/j.1468-2982.1998.1803159.x
10. Goadsby PJ and Lipton RB. **Paroxysmal Hemicrania-Tic Syndrome. Headache**: *The Journal of Head and Face Pain* 2001; 41(6): 608-609 Doi:10.1046/j.1526-4610.2001.041006608.x
11. Boes CJ, Matharu MS and Goadsby PJ. T**he Paroxysmal Hemicrania-Tic Syndrome.** *Cephalalgia* 2016; 23(1): 24-28 Doi:10.1046/j.1468-2982.2003.00406.x
12. Leone M, Mea E, Genco S and Bussone G. **Coexistence of TACS and Trigeminal Neuralgia: Pathophysiological Conjectures**. *Headache: The Journal of Head and Face Pain* 2006; 46(10): 1565-1570 Doi:10.1111/j.1526-4610.2006.00537.x
13. Prakash S and Rathore C. **Two Cases of Hemicrania Continua-Trigeminal Neuralgia Syndrome: Expanding the Spectrum of Trigeminal Autonomic Cephalalgia-Tic (TAC-TIC) Syndrome.** *Headache: The Journal of Head and Face Pain* 2017; 57(3): 472-477 Doi:10.1111/head.12965
14. Alves MRPB and Valença MM. **Cluster-tic syndrome: case report and literature review**. *Headache Medicine* 2017; 58-59 Doi:10.48208/HeadacheMed.2017.15