Hemicrania continua and Temporomandibular Disorders Co-Presentation. Case Report

Co-presentación hemicránea continua y trastornos temporomandibulares. Reporte de caso

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ABSTRACT: Hemicrania continua is an uncommon headache disorder that requires complete response to indomethacin for its diagnosis, that becomes on most of the cases chronic pain resilient to treatment. On the other hand, temporomandibular pain is very common on the general population and happens to be undiagnosed most of the time when headache or migraines are happening at the same time. In this case we aim to highlight the importance of treating all concomitant diagnosis for a better long-term prognosis. In this paper, a case of a 51-year-old male with chronic resilient facial pain is presented. The patient described right sided facial pain with six years of evolution that started with no triggers. The pain was described as constant bilateral pressure being worse on the right side, located on the temples, masseteric and preauricular areas; with an intensity of 6 on a scale of 0 to 10. It is accompanied by episodes of paroxysmal pulsating hemifacial pain with autonomic symptoms (rhinorrhea, conjunctival injections and lacrimation). The pain during exacerbations was located on the right periorbital and hemifacial area and with an intensity of 10 on a scale of 0 to 10, lasting a few minutes to more than 2 hours. After the clinical examination and pain history, the patient was given the diagnosis of hemicrania continua and masticatory myofascial pain. He was started on Indomethacin 25mg twice a day and started on temporomandibular therapy (self-care, jaw exercises, motivation and advice for stress management). Three months after the first evaluation the patient reported total resolution of his symptoms. The main goal of this article is to highlight that the presence of temporomandibular disorders is common in patients with headaches, for
which it is necessary to carry out a comprehensive approach for both entities, which include the therapies for treating axis I diagnosis and behavioral management for axis II, this including most of the time a multidisciplinary human centered approach.

KEYWORDS: Temporomandibular joint disorders; Trigeminal autonomic cephalalgias; Masticatory myofascial pain; TMD; Case report.

RESUMEN: La hemicránea continua es un trastorno de cefalea poco común que requiere una respuesta completa a la indometacina para su diagnóstico, lo cual deja como resultado la mayoría de los casos en un dolor crónico resistente al tratamiento. Por otro lado, el dolor temporomandibular es muy común en la población general y no se diagnostica la mayor parte del tiempo cuando el dolor de cabeza o las migrañas ocurren al mismo tiempo. En este caso pretendemos resaltar la importancia de tratar todos los diagnósticos concomitantes para un mejor pronóstico a largo plazo. En este artículo se presenta el caso de un hombre de 51 años con dolor facial crónico resistente. El paciente refirió dolor facial en lado derecho de seis años de evolución que comenzó sin desencadenantes. El dolor se describió como presión constante bilateral, siendo peor en el lado derecho, localizado en temporales, zona maseterina y preauricular; con una intensidad de 6 en una escala de 0 a 10. Se acompaña de episodios de dolor hemifacial pulsátil paroxístico con síntomas autonómicos (rinorrea, inyecciones conjuntivales y lagrimeo). El dolor exacerbado, se localizaba en la zona periorbitaria y hemifacial derecha y con una intensidad de 10 en una escala de 0 a 10, con una duración de unos minutos a más de 2 horas. Tras el examen clínico y la historia del dolor, se le dio al paciente el diagnóstico de hemicránea continua y dolor miofascial masticatorio. El paciente inició tratamiento con Indometacina 25mg dos veces al día y comenzó con terapia temporomandibular (autocuidado, ejercicios de mandíbula, motivación y consejos para el manejo del estrés). Tres meses después de la primera evaluación el paciente reportó resolución total de sus síntomas. El objetivo principal de este artículo es resaltar que la presencia de trastornos temporomandibulares es frecuente en pacientes con cefalea, por lo que es necesario realizar un abordaje integral de ambas entidades, que incluya las terapias para el tratamiento diagnóstico del Eje I y manejo conductual para el Eje II, lo anterior incluye la mayoría de las veces un abordaje multidisciplinario centrado en el ser humano.

PALABRAS CLAVE: Trastornos de la articulación temporomandibular; Cefaleas autonómicas del trigémino; Dolor miofascial masticatorio; TTM; Reporte de caso.
INTRODUCTION

Hemicrania continua is within the variety of headaches called trigeminal autonomic cephalalgias (TAC), which are a group of disorders that share symptoms such as unilateral location in the orbital, supraorbital, and temporal regions (but that can occur in other trigeminal innervation sites), severe throbbing pain exacerbated by movements or physical activities and different temporal/frequency patterns (1). Furthermore, the major pathognomonic features of these conditions are ipsilateral autonomic symptoms: conjunctival injection, periorbital edema, facial redness, lacrimation, rhinorrhea that occur during attacks or exacerbations (2).

Some theories about its pathophysiology support that it is given by the activation of the trigeminal-autonomic reflex what triggers on the superior salivary nucleus a mediated ipsilateral autonomic response along with neurogenic sterile inflammation. Due to the severity and intensity of the attacks, these alterations are highly incapacitating, they can appear from youth and remain under diagnosed for several years, which presumes a high burden for the person who suffers it. TACs are considered primary headache syndromes, however, in some cases they appear secondary to alterations such as disorders of the paranasal sinuses or alterations of the pituitary gland (3).

According to the Headache Classification Committee of the International Headache Society (IHS) in its third edition (2018); Hemicrania continua is defined as a persistent, strictly unilateral headache that can be associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis and/or eyelid edema, and/or with restlessness or agitation. In addition, responds completely to indomethacin (2). Within its diagnostic criteria, there are important features to highlight on the pain history: it is a unilateral headache, which in the first instance has lasted for more than three months, with exacerbations of moderate or severe intensity, not necessary aggravated by masticatory function or physical activity.

The difference between these trigeminal autonomic cephalalgias lies in the duration, frequency and response to treatment. These headaches include cluster headache (CH), paroxysmal hemicrania (PH), short-term unilateral neuralgiform headaches with conjunctival injection and lacrimation (SUNCT), short-term unilateral neuralgiform headaches with cranial autonomic features (SUNA) and continuous hemicrania (HC). These headache disorders generate a diagnostic challenge, to the point that sometimes it takes up to 6 years to reach an adequate diagnosis (4,5).

Hemicrania continua is a rare entity, found between 1.3 and 2.3% of all patients seen in headache clinics(1,5,6). Its incidence on the general population is difficult to estimate due to the lack of reported case or cases that go underdiagnosed (7). Hemicrania continua is thought to be the one with the least intensity of all TACs (usually mild to moderate, although it may have exacerbations with severe pain), but unlike the others, it never completely remits. It can be differentiated from migraine, in that, unlike this, the patient tends to become restless and agitated in pain attacks. It is more common in women and roughly, there are two variants: remitting and incessant, the latter being the most common. On the treatment side, leaving indomethacin aside, there few treatment reported on clinical cases with good response greater occipital nerve, blocks, gabapentin, and topiramate (8,9).

On the other hand, temporomandibular disorders (TMDs) are recognized as a significant burden for society affecting 5-12% of the population according to the National Institute of Craniofacial and Dental Research(10). TMDs are one of the most common muscle skeletal problems after low back pain. It also currently better understood
the fact that people suffering from TMDs or any form of chronic pain suffer from biopsychosocial comorbidities such as anxiety, depression, somatization (11,12). Currently, there are many standardized and internationally accepted diagnostic tools for Orofacial pain and TMDs, what makes more feasible for the clinician to approach a case when multiple diagnosis are being presented at the same time. Diagnostic criterion like the International Classification of Orofacial Pain (13) and the Diagnostic Criteria for Temporomandibular Disorders (14) are a must to know to every clinician working in this field.

Painful TMDs have being probed on prospective studies as common on the general population (15,16). Furthermore, its prevalence is even higher on people suffering from headache disorders. This meaning that both entities usually present as comorbidities (17).

In this paper we present a case in where masticatory miofascial pain and TMJ arthralgia were presenting at the same time than hemicrania continua, and it is shown how the management of both conditions at the same time was vital for and adequate prognosis.

CASE REPORT

A 51-year-old married male patient presented to the Orofacial Pain outpatient clinic of the San Juan de Dios Hospital referred for temporomandibular pain, he was under treatment in another facility for the diagnosis of cluster headache with a clinical course of approximately 5-6 years of headache predominantly in the right side of the face (mainly in the temple area) with constant pain on both sides and aggravated by mastication, and no history of headaches before. The patient at the time was unemployed for approximately 4 months due to his condition, but he is a former business administrator. He denied smoking or drug use. His medical history included high blood pressure treated with enalapril 10mg. The patient gave written consent to the publication of his case for academic or research purposes.

HISTORY OF THE CHIEF COMPLAINT

By the time of the first evaluation the patient described facial pain with six years of evolution and worsened in the last year. The pain started close to the death of his son, after this event, he divorced his wife with whom he shared 27 years. The pain was described as constant bilateral pressure, located on the temples, masseteric and preauricular areas; with an intensity of 6 on a scale of 0 to 10. It is accompanied by episodes of paroxysmal pulsating hemifacial pain with autonomic symptoms (rhinorrhea, conjunctival injections and lacrimation) (Figure 1). The exacerbations are located in the right periorbital and hemifacial area and happened with a severity of 10 on a scale of 0 to 10, lasting a few minutes to more than 2 hours. The patient does not identify aggravating factors but recognizes that lack of sleep and emotional tension could be affecting. He reports that the pain is slightly relieved by the combination of celecoxib 200mg and potassium diclofenac. Previous treatments with 40mg amitriptyline and 800mg of carbamazepine had no impact on the pain and treatment was discontinued due to sides effects surpassing the benefit.
PHYSICAL EXAM

The patient has no facial asymmetry or lymphadenopathy detected during inspection of the head and neck structures. Under the clinical examination using the Diagnostic Criteria for Temporomandibular Disorders (DC / TMD) criterion, a mandibular range of motion greater than 55mm is observed. There were no noises on auscultation of the Temporomandibular Joint (TMJ). Extraoral and intraoral palpation was performed in the masticatory muscles in which part of the main complaint (constant pain) is duplicated in palpation of temporal muscles, masseters and temporomandibular joint on the right side. On dental examination, stable occlusion and moderate wear facets are observed. Evaluation of the cranial nerves presents with no abnormality on examination.

DIAGNOSTIC PROCESS

After a detailed pain history and clinical examination, an initial diagnosis of masticatory myofascial pain and right TMJ arthralgia was determined for the bilateral constant facial pain. In addition, to the documented presence of autonomic symptoms a secondary diagnosis of hemicrania continua is also given, so it was decided to start treatment with indomethacin as part of the confirmatory process.

THERAPEUTIC APPROACH

The patient was indicated therapeutic myofascial therapy base on self-care, jaw/relaxation exercises and advice for stress management (18). He was also started on Indomethacin 25mg twice a day.

FOLLOW-UP

The patient reported a positive reaction to indomethacin three days after treatment. However, first in person follow-up was carried out 4 weeks after the first appointment, this length was decided mainly to evaluate the effectiveness of the temporomandibular exercises/self-care program. That day, the patient reported an overall improvement of 60%. He reported that the pain is less intense and frequent and recognized that emotional stress is his main aggravating factor. It was decided to add gabapentin 300mg in a tapering dose for a total dose of 900mg per day.

At the second follow-up, 2 months after the first appointment, the patient reported an improvement of above 75%. During this interval his father passed away, and although it was difficult for him to accept it, he stated that he was able to manage the pain even with the emotional overload. An
evaluation with a health psychologist was recommended to the patient.

During the third follow-up, 9 weeks later, the patient presented as an emergency consultation describing a recurrent crisis of facial pain. He states that for no apparent reason the intense paroxysmal right hemifacial pain returned as stabbing and electric pain accompanied by rhinorrhea and lacrimation more than 30 episodes per day lasting from a few seconds to thirty minutes (Figure 2). He states that it may have been caused by an increase in his blood pressure, however, it was under control. On this occasion, introral blockade of the sphenopalatine ganglion was performed through the greater palatal canal with 2% lidocaine anesthetic with epinephrine, this procedure gave him immediate pain relief.

The fourth follow-up was carried out 12 weeks after the initial evaluation, he reported 100% improvement and to be free of pain he recognizes that muscle pain and emotional stress as his main aggravating factors. Moreover, currently he has reconciliated with his wife and they are back together.

**DISCUSSION**

This clinical case presented here, shows how a pain history and through examination are crucial for an adequate diagnosis and therefore prognosis (19). The frequency and duration of the facial pain attacks in this case: unilateral pain, lasting from 30 minutes to two hours, accompanied by autonomic features; following the IHSC criteria the diagnosis to hemicrania continua was given. In addition, the almost immediate response to Indomethacin confirmed it. However, the bilateral constant ache reported by the patient was not like any headache diagnosis being in fact of natural common characteristics of temporomandibular pain.

Similar cases have been seen by Porporatti et al. (20) in where bilateral jaw pain was happening concomitantly with facial pain attacks but in this case the duration of the attacks where less that 30 minutes leading to paroxysmal hemicrania as the headache diagnosis (another trigeminal autonomic cephalalgias). Base on this, it is important to remark that although painful temporomandibular disorders are more common on the general population than trigeminal autonomic cephalalgias (15), both share the similarity that can go under-diagnosed during years before getting adequate treatment (3,11).

Several studies have found coexistence of headache disorders and TMDs, this could be attributed to potential shared pathological mechanisms. Andersson et all found that more that than a 10% of patients presenting headache, presented the diagnosis of miofascial masticatory pain and around 90% of the patients presented headache had a temporomandibular joint pain diagnosis (17).

In addition to the primary headaches, it is important to remark that the classification for headache disorders includes on the secondary
headaches what is known as headache attributed to TMD, in which the presence of a primary diagnosis of myalgia or arthralgia is required, as it is in the case presented. In this patient, a familiar pain will be reproduced in the temporal region, which is modified with mandibular movement, function, or parafunction (2).

The location of familiar headache in the temporal muscle is confirmed by palpation of the temporal muscle and jaw movement as established by the international diagnostic criteria and delimits the difference with tension headache by the fact that the latter is not aggravated or relieved by mandibular function.

In this case, the marked periods of exacerbations lasting from 30 minutes to 3 hours (according to the patient), what made the pivotal point for the diagnosis was the presence of autonomic symptoms in this case conjunctival injection, rhinorrhea, and lacrimation. Furthermore, the question of is this patient suffering of cluster headache along with TMD comes up and the diagnostic decision arrives from the point that exacerbations reported by the patient last more than 30-180 minutes, that is the time frame for cluster headache according to the diagnostic criteria along with the fact that there were no pain free periods before treatment.

Multiple publications have revealed that the management of temporomandibular disorders begins with an adequate diagnosis and patient education about their condition. Currently, it has been shown that painful temporomandibular disorders and the headache associated with these conditions present an effective therapeutic response to conservative medical treatment. Leaving surgical treatment for cases in which there is frank pathology or trauma to the temporomandibular joint. Most TMD patients compromise benefited from simple treatments including physical therapy, behavioral therapy, and oral appliances. However, patients with chronic TMD pain and headaches disorders need a combined therapeutic multidisciplinary approach (21,22).

The case presented here showed that the combination of patient education, counseling and motivation, self-care, stress management and pharmacological treatment with Indomethacin 50mg and gabapentin 900mg per day, gave complete resolution to the patient main complaint. The key component for treatment was approaching the headache diagnosis along with the temporomandibular pain. Understanding both diagnoses gave confidence to the patient and had a positive impact on his motivation and emotional state. It has being reported that patients with chronic Orofacial pain and specially TMD, have higher levels of reported anxiety and catastrophizing (11,23). In addition, this emotional burden creates a cycle of resistance to conservatives therapies being the main reason why biological (Axis I diagnosis) should be assessed and treated at the same time that psychosocial factors (Axis II); comorbidities such as emotional tension and multiple harmful habits that contribute to the worsening of symptoms. In this particular case severe grief, no adequate treated.

Painful temporomandibular disorders are common on the general population, even more on the ones suffering from headache disorders. Validated diagnostic tools are available for TMDs, adequate diagnosis and treatment of different pain entities presenting altogether; is vital for a good prognosis and long-term relief for patients suffering from these conditions.

It is of importance for the clinicians to consider the coexistence of TMDs when treating headache disorders, in particular those who do not response to conventional treatment. Treating both conditions simultaneously could be the key for adequate pain relief for people suffering from this debilitating pain conditions.
Data Sharing statement: Data sharing is not applicable to this article as no new data were created or analyzed in this study.

All the authors declare no conflict of interest related to the publication.

List of Abbreviations (In alphabetic order).

CH: Cluster headache.
DC/TMD: Diagnostic criteria for temporomandibular disorders.
IHS: International Headache Society.
PH: Paroxysmal hemicrania.
SUNA: Short-term unilateral neuralgiform headaches with cranial autonomic features.
HC: Hemicrania continua.
SUNCT: Short-term unilateral neuralgiform headaches with conjunctival injection and lacrimation.
TAC: Trigeminal autonomic cephalalgias.
TMD: Temporomandibular disorder.
TMDs: Temporomandibular disorders.
TMJ: Temporomandibular Joint.

ETHICAL APPROVAL AND CONSENT TO PARTICIPATE

Publication was approved by the bioethical committee and the patient gave written informed consent for publication of the case and images.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

COMPETING INTERESTS

All the authors declare no conflict of interest related to the publication.

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AUTHOR CONTRIBUTION STATEMENT

Contribute to the report and treatment for the patient: A.C.CH. and R.R.G.

DATA AVAILABILITY STATEMENT

All data related with the case is available upon request.

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