

Facial representation of paroxysmal hemicrania and associated temporomandibular disorder: a case report

Hemicrania paroxística com representação na face e associação com disfunção temporomandibular: relato de caso

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ABSTRACT

Paroxysmal Hemicrania is a trigeminal autonomic cephalalgia described as a severe and strictly unilateral pain, which occurs in paroxysms at orbital, supraorbital and/or temporal region. A 45-year-old woman presented to an orofacial pain specialist reporting severe, burning, throbbing, strictly right-sided headache associated to

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ipsilateral autonomic symptoms and orofacial pain. The pain was perceived on the maxillary region followed by pain spread to the head. Interdisciplinary evaluation, along with absolute responsiveness to indomethacin and normal Brain Magnetic Resonance imaging, led to the diagnosis of primary Episodic paroxysmal hemicrania with facial representation and myofascial pain of masticatory muscles. Dentists should be aware of paroxysmal hemicrania with facial representation and the possibility of temporomandibular disorder coexistence, in order to avoid misdiagnosis and inadequate management. Paroxysmal hemicrania may be first perceived on the face and may be associated with interparoxysmal pain. In these cases, efficient anamnesis and clinical evaluation followed by interdisciplinary approach is needed.

Indexing terms: Facial pain. Paroxysmal hemicrania. Temporomandibular joint disorders.

RESUMO

A Hemicrania Paroxística é uma cefalalgia autonômica trigeminal descrita como uma dor severa e estritamente unilateral, que ocorre em paroxismos na região orbital, supraorbital e/ou temporal. Uma mulher de 45 anos de idade apresentou-se a um especialista em dor orofacial, referindo uma cefaleia intensa, ardente, latejante, estritamente do lado direito, associada a sintomas autonômicos ipsilaterais e dor orofacial. A dor era sentida na região maxilar, seguida de dor que se estendia à cabeça. A avaliação interdisciplinar, aliada à resposta absoluta à indometacina e à normalidade em ressonância magnética cerebral, levou ao diagnóstico de hemicrania paroxística episódica primária com representação facial e dor miofascial dos músculos mastigatórios. Os médicos dentistas devem estar atentos à hemicrania paroxística com representação facial e à possibilidade de coexistência de Disfunção Temporomandibular, de modo a evitar diagnósticos incorretos e um tratamento inadequado. A hemicrania paroxística pode ser percebida inicialmente na face e pode estar associada à dor interparoxística. Nesses casos, é necessária uma anamnese e avaliação clínica eficientes, seguidas de abordagem interdisciplinar.

Termos de indexação: Dor facial. Hemicrania paroxística. Transtornos da articulação temporomandibular.

INTRODUCTION

According to the International Classification of Headache Disorders (ICHD) [1] headache is described as a head-located pain above the orbitomeatal line and/or nuchal ridge. Additionally, facial pain is described as pain below the orbitomeatal line, anterior to the pinnae and above the neck. [1] Although the designation headache indicates a head-located pain, it is also recognized that some headache syndromes may have representations in the face and are usually ipsilateral to headache attacks [2].

In 2020, due to the need of a clear classification of orofacial pains, the first edition of the International Classification of Orofacial Pain (ICOP) [2] was published in a Beta version. Based on that and according to the ICOP's guidelines, patients who present pain exclusively in the facial region, resembling primary headaches, should be diagnosed according to that classification.

Dentists are often the first professionals sought in cases of orofacial pain [3] and the knowledge of conditions that may represent pain in the orofacial regions is imperative to reduce misdiagnosis and inadequate management. Temporomandibular disorder (TMD) is a collective term embracing a number of clinical problems that involve the masticatory muscles, the temporomandibular joint (TMJ) and associated structures [4]. It is the most common chronic pain to affect the face [5] and it is frequently associated with primary headaches [6].

Trigeminal autonomic cephalalgias (TAC) share clinical findings of unilateral headache and cranial parasympathetic autonomic features ipsilateral to the headache [1]. Paroxysmal Hemicrania (PH) is classified as a TAC. It is described as a severe, strictly unilateral pain, which is orbital, supraorbital, temporal, or in any combination of these sites, lasting 2-30 minutes and occurring at least 5 times per day. The attacks are usually associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis, and/or eyelid oedema. The headache is absolutely sensitive to indomethacin [1].

PH is also a rare primary headache with infrequent facial representation [3-6]. Due to its location, it may be confounded [7] or coexist with TMD [8]. Therefore, understanding facial involvements of primary headaches may be useful to better comprehend its pathophysiology and to establish adequate treatment regimens. [3] The aim of this paper is to report a case of PH with facial representation and associated TMD.

CASE REPORT

A 45-year-old brown female presented to an orofacial pain specialist complaining of concomitant and unilateral headache, facial pain, and otalgia. Headache was described as severe (graded as 10 in a Verbal Rating Scale), burning, throbbing pain, and strictly right sided, initiating in masseter muscle, spreading to frontal-orbital, auriculotemporal and maxillary regions (figure 1). Pain attacks were associated with ipsilateral autonomic symptoms (conjunctival injection, ptosis, nasal congestion, lacrimation, facial edema). Photophobia and phonophobia were also present.

The patient reported the onset of pain attacks when she was 23 years old. At that time, and repeatedly over several years, she underwent dental evaluations with a variety of professionals, all of whom diagnosed her with TMD and sleep bruxism. Pain attacks used to occur several times within a period of 20 days, separated by remission periods that usually lasted two to four years. The patient mentioned difficulties



Figure 1. Delimitation of pain site.

to establish the duration of headache attacks, but reported pain episodes used to last approximately 6 hours, characterized by attacks of severe pain intercalated with periods of discomfort. Attacks presented nocturnal preponderance, waking her up from sleep, and were more common when she was stressed. Previous treatments included occlusal appliance wearing during sleep and muscle relaxant intake, which were useful to reduce facial pain. Moreover, a right upper second premolar extraction was performed, since the patient reported odontalgia and believed it to be the pain source.

Intra-oral evaluation evidenced class V non-carious cervical lesion on right first upper premolar and absence of the right second upper pre-molar. Panoramic radiograph evidenced no abnormalities. Clinical evaluation showed allodynia and hyperalgesia on frontal-orbital and auriculotemporal regions and myofascial pain on right temporalis (with pain referral to TMJ) and masseter (with pain referral to right upper first molar and edentulous region of right upper premolar) muscles.

Pain characteristics and associated symptoms met Episodic Paroxysmal Hemicrania (EPH) diagnostic criteria of the ICHD [1] and, to evaluate indomethacin response [1], oral indomethacin (25mg/day) along with a gastro protector were prescribed. Due to positive indomethacin response, EPH was confirmed, and the patient was referred to a neurologist.

Neurological examination was normal, except for dysesthesia in the right-sided first branch of the trigeminal nerve (V1). Brain Magnetic Resonance imaging and Magnetic Resonance Angiography revealed no abnormalities.

After interdisciplinary (dentist and neurologist) evaluation, the patient was diagnosed with primary EPH and ipsilateral myofascial pain of masticatory muscles. Patient also had self-reported awake and sleep bruxism. Neurological treatment was mainly pharmacological, which included daily intake of prednisone 40mg for 7 days and indomethacin 50mg up to twice a day with significant pain relief. After 1 month, the patient was reevaluated by the neurologist and reported only one episode of pain attack during this period of time. TMD therapies included self-management programs (education, thermal therapy, self-massage therapy and oral parafunctional habits monitoring and avoidance) [9], wearing occlusal appliance during sleep, pharmacotherapy (nortriptyline 10mg for 2 months and cyclobenzaprine 5mg for 15 days), and 5 sessions of dry needling over the painful muscle points. Regular dental treatment was also performed (not related to pain management). Historical and current information was organized as a timeline and presented. From the patient's perspective, after several years of unsuccessful treatments, it was really rewarding to meet professionals who were able to achieve the accurate diagnosis and treatment.

DISCUSSION

This case report aimed to alert clinicians to the existence and clinical features of EPH with facial representation, in addition to possible coexistence of TMD, which may delay TACs diagnosis.

It has been suggested that pain and other symptoms of PH may be due to a dysfunction in the hypothalamus, resulting in destabilization of the trigemino-vascular system and central disinhibition of the trigeminal autonomic reflex, possibly through direct hypothalamic-trigeminal connections [10]. PH attacks occur more frequently during the day, but nocturnal attacks have also been described [6].

PH is equally prevalent in both males and females, mean age of onset in the 30s; it shows relatively short-lasting duration, strictly unilateral location, ipsilateral cranial autonomic features and absolute responsiveness to indomethacin [1].

Probably due to the rarity of PH, which makes it difficult to conduct well-designed randomized controlled trials, limited therapeutic tools are available [11]. The definitive indomethacin pharmacological response is of great value to differential diagnosis, and it is the most used drug for both acute and prolonged treatment [12]. The mean effective daily dosage of indomethacin ranges from 25 to 300mg [13,14]. Some patients, however, may report adverse effects such as dizziness and gastrointestinal manifestations, thus initiating treatment with a low dose should be considered. In cases where a reduced daily dose of indomethacin is needed, or in cases of its discontinuation, other drugs, such as piroxicam [12], prednisone [12], verapamil [12,13], topiramate [12], amitriptyline [12] and oxygen, may be proposed [6,12].

As previously mentioned, EPH is an orbital, supraorbital and/or temporal pain, and differential diagnosis includes several conditions such as others TACs, paroxysmal hemifacial pain, migraine, trigeminal neuralgia, sinusitis, TMD and dental pain. Paroxysmal hemifacial pain is an orofacial pain that also shares similarities with PH. It is well described in the ICOP as a strictly hemifacial short-lasting pain attack without headache, associated with the same autonomic signs described for TACs, however, with no involvement of the head [2]. Thus, pain location is definitive for PH diagnosis. In the present clinical case, the existence of concomitant headache was mandatory for its diagnosis.

In this way, clinicians should routinely perform detailed anamnesis and physical examination of orofacial pain patients in order to collect specific pain characteristics, leading to early diagnosis and avoiding unnecessary treatments. Previous studies reported PH patients who were submitted to unnecessary interventions such as dental procedures [4], maxillary sinus surgery [7] and TMD treatments [8].

PH patients may exhibit ipsilateral muscle tenderness [3] and, according to Cittadini et al. [6], the involvement of V2, V3, the jaws and the teeth is rare, but it may occur. When analyzing 2,912 patients, Ziegeler and May [3] described only 4 who presented headache with facial involvement in which pain was mainly perceived in V2 and/or V3, but also spread to V1. In the present clinical case, during the attack, the patient first perceived pain on the maxillary region followed by pain spread to the head. She also reported involvement of an ipsilateral upper molar tooth however, clinical examination indicated trigger point located in the right masseter muscle as the pain source. The association between TMD and TACs has not been well established, but the sensitization of the V2 and V3 trigeminal nerve branches has been demonstrated in other primary headaches [14].

PH patients may also present interparoxysmal pain, which is usually milder in intensity [14]. In this case report, the patient described severe pain attacks intercalated by discomfort, although it was not clear if it was a symptom of PH or TMD. Differential diagnosis of hemicrania continua was also considered; however, as it is characterized by continuous and moderate pain [14], this diagnosis was discarded.

In cases where PH is comorbid with other conditions, a headache diary may be useful to provide information regarding pain aspects and to facilitate differential diagnosis [6]. Medication overuse is also common in individuals with primary headaches leading to the high risk of developing medication-overuse headache. Therefore, it should also be considered when filling a headache diary [6].

Although PH is uncommon, future studies regarding its association with facial representation and TMD are suggested in order to elucidate its pathophysiology and improve therapeutic strategies. As a final note, clinicians should be familiarized with both ICOP and ICHD.

CONCLUSIONS

Dentists should be aware of PH with facial representation and of its possible coexistence with TMD, since patients may firstly seek treatment in orofacial pain services. In these cases, interdisciplinary approach is crucial for a well-succeeded treatment.

Collaborators

All authors made individual and substantial contribution to the writing of the article. JAO Buosi, LMSP Fiamengui, PPM Santos and P Braga Neto conducted patient's interdisciplinary evaluation and management. PPM Santos, LMSP Fiamengui and JAO Buosi drafted the article. PCR Conti, HV Chaves and P Braga Neto revised it critically and improved intellectual content. All authors read and approved the manuscript. The authors declare no conflicts of interest.

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