Trigeminal Autonomic Cephalalgia and Facial Pain: A Review and Case Presentation

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Trigeminal autonomic cephalalgias (TAC) are primary headache disorders that are characterized by severe unilateral pain along the distribution of the trigeminal nerve with corresponding activation of the autonomic nervous system. The clinical characteristics and presentation of TAC are unique; however, there may be an overlap of these features with other painful conditions affecting the orofacial region, which can be a diagnostic challenge for the clinician. This article reports a case history and discusses the differences between clinical characteristics of TAC and other painful orofacial conditions. Refractory pain conditions and the occurrence of episodic pain attacks with accompanying autonomic symptoms necessitate a thorough evaluation to rule out rare causes of head and face pain. *J Oral Facial Pain Headache 2019;33:e1–e7. doi: 10.11607/ofph.2143*

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Trigeminal autonomic cephalalgias (TAC) are a type of primary headache disorder characterized by severe unilateral pain along the distribution of the trigeminal nerve with corresponding activation of the autonomic nervous system. The autonomic features consist of at least one of the following: conjunctival injection or lacrimation; nasal congestion or rhinorrhea; eyelid edema; facial or forehead sweating; facial or forehead flushing; sensation of fullness in the ear; and miosis or ptosis. Based on the clinical features and responses to therapeutic modalities, TAC are further subdivided into four categories: cluster headache (CH), paroxysmal hemicrania (PH), hemicrania continua (HC), and short-lasting neuralgiform headaches.[1,2]

CH is an episodic headache disorder that presents for 15 minutes to 3 hours once every other day up to eight times per day. This disorder is associated with ipsilateral autonomic activation and primarily affects men. The prevalence of CH is estimated to be between 0.09% and 0.32%[3]; however, it is significantly higher among smokers or former smokers and those who drink alcohol. Similarly, there is significant evidence of a strong genetic background in CH.[1,4]

PH is a type of TAC with clinical features similar to that of CH. However, it is characterized by relatively shorter (2 to 30 minutes) duration of pain episodes and more frequent (at least five attacks per day) episodes.[2,5] Furthermore, PH has an absolute response to indomethacin. The prevalence of PH is estimated to be between 0.021% and 0.07%, is more common in women, and has no evidence for a genetic predisposition. The average age of onset is the fourth decade, and the average duration of illness is nearly 13 years.[1,2,5]

HC is a rare type of persistent headache disorder, with only a few hundred reported cases in the literature. However, recent investigations suggest that it may be more prevalent than previously anticipated.[6] This disorder is more common in women and has no evidence of a genetic background. The headache is strictly unilateral, constant (with or without varying intensity), and associated with autonomic activation. In some instances, it may accompany restlessness, photophobia, and/or phonophobia. Similar to PH, it has an absolute response to indomethacin therapy.[2,5,6]

Short-lasting unilateral neuralgiform headaches are a rare type of episodic headache disorder with a male preponderance. The headache episodes primarily resemble clinical features of CH, PH, and trigeminal neuralgia. The episodes are intense, short lived (1 to 600 seconds), and occur as a single stab, series of stabs, or in a saw-tooth pattern. They take place at least once a day and primarily affect the orbital, supraorbital, and temporal regions.[5,7] The headaches are accompanied by ipsilateral autonomic symptoms; however, based on the type of ipsilateral autonomic feature, they are further subdivided into short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and short-lasting unilateral neuralgiform headache attacks with autonomic symptoms (SUNA).

The clinical characteristics and presentation of TAC are unique. However, there may be an overlap of these features with other painful conditions affecting the orofacial region, which can be a diagnostic challenge for the clinician. The purpose of this article was to present a case that validates the diagnostic challenge associated with TAC. In addition, TAC and other orofacial pain conditions with overlapping clinical features are reviewed.

Clinical Problem

A 74-year-old female patient sought care at the Massachusetts General Hospital (MGH) Oral and Maxillofacial Pain Center with a complaint of pain in the left frontotemporal and infraorbital region extending toward the ipsilateral angle of the mandible and the lateral and posterior cervical region. The pain had begun 18 months prior as an intermittent, throbbing, and dull ache. The pain was severe in intensity; presented up to three times a day every day for 15 to 30 minutes; and was associated with the feeling of constant pressure in the region and occasionally lacrimation from the ipsilateral eye. After 1 week of onset, the patient also started to experience a constant dull ache of mild to moderate intensity that spread across the left frontotemporal and infraorbital region, angle of the mandible, and lateral and posterior cervical region. Episodic bouts of severe throbbing dull pain were sometimes triggered 30 minutes following mastication. The constant pain would worsen after strenuous physical activity and toward the end of the day. Both the episodic and constant pain were alleviated following rest or with ibuprofen 600 mg, aspirin 325 mg, or topical application of peppermint oil. There were no reports of associated facial or neck sensory abnormalities, facial weakness, or the presence of additional autonomic symptoms in the head and neck regions. There were no reports of photophobia, phonophobia, osmophobia, nausea, vomiting, or vertigo.

Approximately 17 months ago, the patient had visited her primary care physician (PCP) with this complaint. Brain, maxillofacial, and cervical magnetic resonance imaging (MRI) studies were ordered. The brain and maxillofacial MRI studies were noncontributory. However, the cervical MRI study suggested the presence of cervical degenerative joint disease (DJD) along the cervical vertebrae.[4–7] She was diagnosed by her PCP as having cervical myofascial pain disorder and cervicogenic headache associated with cervical DJD. The PCP prescribed her pharmacologic intervention consisting of hydrocodone 5 mg with acetaminophen 325 mg every 4 hours as needed and referred her to a pain anesthesiologist. The pain anesthesiologist prescribed her nortriptyline 10 mg three times a day, 12 sessions of cervical physical therapy, and performed consecutive series of three intramuscular trigger point injections using local anesthetic and corticosteroids. This intervention, pharmacotherapy, and physical therapy did not resolve her symptoms, and she was referred back to the care of her PCP.

She was later referred to an oral and maxillofacial surgeon, who diagnosed her as having masticatory and cervical myalgia pain disorder, prescribed her carisoprodol 350 mg twice a day, and recommended additional physical therapy. She underwent another 12 sessions of masticatory and cervical physical therapy. This helped to resolve the constant dull ache but did not impact the severity, intensity, or frequency of severe episodic pain episodes. She was subsequently referred to a rheumatologist. The rheumatologist performed multiple blood studies and ruled out musculoskeletal, inflammatory, and immunologic/autoimmune etiology and referred her to a neurologist. The neurologist gave her the diagnosis of trigeminal neuralgia and placed the patient on gabapentin 600 mg three times a day and oxcarbazepine 150 mg three times a day. She also underwent another brain MRI study, which was reported as normal. After 2 months of pharmacotherapy, the patient's symptoms did not resolve; however, she developed fatigue, lethargy, and weakness, for which she was hospitalized. During her hospital stay, she was diagnosed with an electrolyte imbalance disorder associated with polypharmacy. All of her neuropathic medications were discontinued, and she was placed on lidocaine 5% cutaneous patch and hydrocodone 5 mg with acetaminophen 325 mg every 4 hours as needed. Subsequently, her PCP referred her to another oral and maxillofacial surgeon. This surgeon ruled out any odontogenic causes of pain and referred her to the Oral and Maxillofacial Pain Clinic at MGH for further evaluation.

Her medical history was significant for hypertension, asthma, gastroesophageal reflux disease, dyslipidemia, depression, and diverticulosis. Her current medications were atenolol 25 mg once a day, mometasone inhaler 220 mcg as needed, albuterol inhaler (1 to 2 puff[s] as needed), omeprazole 20 mg once a day, aripiprazole 5 mg once a day, and mirtazapine 7.5 mg once a day.

On physical examination, there was no gross head and neck asymmetry, discolorations, lesions, or swelling. The mandibular range of motion was within normal limits but associated with pain in the left masseter muscle and temporalis muscle. The opening and closing jaw movements were straight. There was palpable crepitus bilaterally in the temporomandibular joints (TMJ) during vertical and horizontal mandibular movements. The patient reported pain during palpation of the left masseter muscle, the left temporalis muscle, and the posterior mandibular area/styloid fossa. The cervical range of motion was within normal limits but was associated with pain in the left trapezius muscle. The patient reported pain during palpation of the left anterior, middle, and posterior scalene muscles; levator scapulae muscle; semi-spinalis muscle; splenius capitis muscle; and trapezius muscles. There was no cervical lymphadenopathy or thyromegaly. Intraoral examination was noncontributory. The cranial nerves (II–XII) and spinal nerves (C2–T1) were grossly intact. Complex motor skills revealed normal coordination. Deep tendon reflexes at the biceps, triceps, and brachioradialis were normal. Carotid pulsations were full and bilaterally symmetric, and there was no bruit on auscultation.

Panoramic radiographs in the open and closed mouth positions were significant for bilateral flattening along the anterior margin of the condylar process and subcondylar sclerosis on the left side. These were considered to be noncontributory to the patient's clinical symptoms.

A tentative diagnosis of paroxysmal hemicrania and masticatory and cervical myalgia pain disorder was made. The patient was placed on indomethacin (extended release) 75 mg twice a day, omeprazole 40 mg once a day, and metaxalone 800 mg twice a day and referred for multimodal physical therapy of the masticatory and cervical regions. Within 24 hours of using indomethacin, the patient experienced relief in the severe episodic frontotemporal and periorbital pain symptoms and subsequently experienced complete relief in the background aching pain after a few sessions of physical therapy and with the use of metaxalone. The patient continues to be on indomethacin (extended release) 75 mg once a day, omeprazole 40 mg once a day, and metaxalone 800 mg once a day, and, at the time of writing this report, reported complete relief in symptoms for a period of 16 months.

Differential Diagnosis

Painful conditions of the orofacial region may have multiple overlapping clinical features. However, these conditions do have unique clinical characteristics through which they can be identified and managed. In order to correctly diagnose the underlying cause, a meticulous analysis of the history (signs and symptoms) and the nature of the orofacial pain condition (pattern) along with a detailed and thorough physical examination of the head, face, and neck regions are essential. This aids in establishing a definitive diagnosis, which holds the key to a successful therapeutic outcome (Table 1).

Seventeen months prior to her referral to the MGH Oral and Maxillofacial Pain Center, the patient presented to a pain anesthesiologist with left side headache, neck, and jaw pain. The differential diagnosis at that time was cervical myofascial pain disorder and cervicogenic headache in the setting of cervical DJD.

Cervical myofascial pain disorder is a regional muscle pain disorder characterized by localized dull aching pain and originating from cervical muscles and surrounding fascia.[1,8] Its clinical presentation consists of mostly unilateral pain, but bilateral pain may also be present. The pain can be continuous or intermittent and may be present for hours. The pain is aggravated with cervical functional and parafunctional movements and is associated with the presence of tightness in the posterior neck region, limited cervical range of motion, and pain on palpation of the cervical region. Furthermore, it may result in referral of pain to the ipsilateral frontotemporal, orbital, retro-orbital, or masticatory regions, vertex, and may result in sensory and motor alterations in the upper extremities.[1,8] In the present case, the patient reported episodic pain lasting 15 to 30 minutes in the frontotemporal and periorbital regions that was associated with a feeling of pressure and ipsilateral lacrimation. Similarly, there was no relief in these episodic pain symptoms following the conventional treatment for cervical myalgia pain disorder.

Cervicogenic headache is typically ipsilateral to the side of the cervicogenic disorder (eg, cervical myofascial pain disorder, cervicalgia, or cervical DJD). This headache disorder is mostly unilateral, but bilateral headaches can also take place. The pain is distributed over the occipital region and tends to radiate anteriorly toward the frontotemporal region. The pain can be continuous or intermittent, nonthrobbing in quality, and aggravated by cervical functional and parafunctional movements. The headache may be accompanied by associated nausea, vomiting, phonophobia, photophobia, and/or periorbital edema.[2,9] The management consists primarily of treatment of the underlying musculoskeletal disorder. Similarly,

Table 1 History and Clinical Features of Common Conditions That May Affect the Orofacial Region

Parameter	Myofascial pain	Ischemic stroke	Giant cell arteritis	Trigeminal neuralgia	Migraine headache disorder
Age of onset (y)	20–40	50–60	50–70	50–60	20–40
Gender ratio (M:F)	1:3	1.3:1	1:2.5	1:2.3	1:3
Clinical features					
Pain location	Masticatory and cervical muscles	Highly variable	Temporal, parietal	Confined to distribution of ophthalmic, maxillary, and mandibular divisions of trigeminal nerve	Frontotemporal, orbital, occipital regions
Laterality	Unilateral or bilateral	Unilateral	Unilateral	Unilateral	Unilateral or bilateral
Pain quality	Dull, pressing, sharp	Burning, stabbing, electric	Throbbing, pressing	Electric shock–like, stabbing, shooting, or sharp	Throbbing
Pain duration	Minutes to days	Varying	Up to 2 h	Less than 2 min	4 to 72 h
Pain frequency	Near daily	Differs	Up to 5 times/d	Varies[a]	Differs[b]
Pain intensity	Mild to severe	Mild to severe	Severe	Severe	Moderate to severe
Additional features	Tightness Pain with jaw or cervical movements	Sensory discrepancy Motor weakness Altered sensorium Visual alteration Speech changes	Scalp allodynia Jaw claudication Polymyalgia rheumatica Malaise Weakness	Innocuous trigger stimuli, such as shaving, smiling, or washing or touching face	Photophobia Phonophobia Osmophobia Nausea Vomiting Worsened by exercise Vertigo

[a]Mostly related to triggering events.
[b]Episodic migraine headache takes place up to 14 days per month and chronic migraine headaches at least 15 days per month.

nerve blockade of the greater and lesser occipital nerves using local anesthetic may provide intermittent relief in symptoms.[9] In the present case, the patient reported throbbing, dull, aching pain that was associated with the presence of pressure and lacrimation. Likewise, there was no resolution in the episodic headache symptoms following the standard cervical myalgia treatments.

Intermittent pain in the frontotemporal region can be referred from ipsilateral masticatory muscles. Masticatory myofascial pain disorder is characterized by regional, unilateral or bilateral, and dull or aching pain in the masticatory muscles that is affected by jaw movement, function, or parafunction.[1,10,11] The pain is often associated with a subjective feeling of tightness in the masticatory muscles, limited mouth opening, pain on palpation of the masticatory muscles, and referral of pain beyond the anatomical boundary of the masticatory muscles.[10,11] The management may consist of physical therapy, pharmacotherapy, orthotic jaw device therapy, and/or injection therapy.[11] In the present case, the patient did report triggering of episodic pain sometimes following mastication. However, this is unlikely to explain the associated lacrimation and lack of relief after physical therapy and muscle relaxant pharmacotherapy.

Onset of a new headache in individuals above 50 years of age should raise suspicion for a secondary headache disorder, such as headache attributed to ischemic stroke or transient ischemic attack, nontraumatic intracerebral hemorrhage, or intracranial neoplasia.[1,2] The clinical features depend on the type and location of the injury or lesion. The headache is often accompanied by neurologic deficits (sensory changes, motor weakness) or systemic symptoms (fever, malaise, weight loss) that present in an acute setting in patients older than 50 years of age.[2] The headache is typically of a different type than previously experienced by the patient. Brain imaging studies are often indicative of underlying pathology, and treatment depends on the underlying causative factor.[1,2,8] In the present case, brain imaging was normal, and the patient did not report any loss of sensory or motor function or any systemic symptoms in association with the pain.

Similarly, in individuals above 50 years, unilateral frontotemporal pain can be associated with giant cell arteritis (GCA).[12] GCA is a disease in which there is inflammation of cranial arteries resulting in occlusion of blood vessels. Commonly affected arteries include ophthalmic, posterior ciliary, maxillary, superficial temporal, and other branches of the external carotid

	Cluster headache	Paroxysmal hemicrania	Short-lasting neuralgiform headache	Hemicrania continua
	27–31	34–41	40–50	30–40
	4:1	1:1.6	7:1	1:2.8
	Temporal, parietal, periorbital, maxillary regions	Temporal, parietal, periorbital, maxillary regions	Temporal, periorbital	Frontotemporal, parietal, occipital
	Unilateral	Unilateral	Unilateral	Unilateral
	Boring, sharp, stabbing, throbbing	Stabbing, throbbing, boring	Sharp, electric, stabbing	Pressure, dullness, throbbing
	15 to 180 min	2 to 30 min	Up to 10 min	Continuous
	Once every other day up to maximum of 8 attacks/d	At least 5 attacks per day more than half of the time	Single stab, series of stabs; at least one episode per day	Continuous
	Severe	Moderate to severe	Severe	Moderate
	Ptosis or miosis Periorbital edema Facial or forehead sweating or redness Conjunctival injection or tearing Nasal congestion or rhinorrhea Ear fullness Pacing the floor or agitation Phonophobia Photophobia	Ptosis or miosis Periorbital edema Facial or forehead sweating or redness Conjunctival injection or tearing Nasal congestion or rhinorrhea Ear fullness	Ptosis or miosis Periorbital edema Facial or forehead sweating or redness Conjunctival injection or tearing Nasal congestion or rhinorrhea Ear fullness	Ptosis or miosis Periorbital edema Facial or forehead sweating or redness Conjunctival injection or tearing Nasal congestion or rhinorrhea Ear fullness Pacing the floor or agitation Phonophobia Photophobia

system. The involved artery may be enlarged, firm, and tender to palpation, often with decreased pulsations. Patients present with a headache that worsens upon lying flat, malaise, weakness, weight loss, and jaw claudication. Laboratory studies can indicate an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).[8,12] However, these tests are nonspecific, and temporal artery biopsy is required for a definitive diagnosis.[12] In the present case, the patient did not report modification of symptoms with changes in body posture, jaw claudication, or constitutional symptoms of illness, and during the examination, the patient did not report tenderness to palpation of temporal arteries. Likewise, her ESR and CRP were within normal limits.

Trigeminal neuralgia (classical) is a painful neuropathic disorder characterized by brief (< 120 seconds) paroxysms of severe, unilateral, and electric shock–like, sharp, or lancinating pain in the trigeminal nerve distribution.[13] In some cases, there may be a constant dull background ache in the affected area (classical trigeminal neuralgia with concomitant persistent facial pain).[2] The most commonly affected branches of the trigeminal nerve are the maxillary and mandibular divisions. The average age of onset is approximately 60 years, and women are slightly more affected than men. Characteristically, the pain is not associated with any neurologic deficits (sensory changes or motor weakness) or autonomic phenomena. Similarly, the pain may be spontaneous or precipitated by nonpainful tactile stimuli such as washing or gently touching the face, brushing the teeth, or even wind.[13] In the present case, the patient had episodes of severe pain that lasted at least 15 minutes in duration, and the dull, aching pain was spread beyond the distribution of the trigeminal nerve. Similarly, the patient had no improvement in symptoms after using anticonvulsant therapy for 2 months.

Migraine headache disorder may manifest as pain in the craniofacial region.[1,2,4,14] Migraine headache is a recurrent headache disorder associated with severe pain episodes that last between 4 and 72 hours. They are mostly unilateral in distribution, pulsating in quality, and result from aggravation of symptoms with physical activity. They are commonly associated with reports of nausea, vomiting, photophobia, phonophobia, osmophobia, or vertigo. In some instances, a transient neurologic deficit (aura) in the form of visual, sensory, or speech disturbance may precede the headache episode.[1,2] In this case, the patient reported pain attacks of a much shorter duration and had no reports of associated accompanying symptoms of

Table 2 International Classification of Headache Disorders 3 Criteria for Paroxysmal Hemicrania

A. At least 20 attacks fulfilling criteria B–E.
B. Severe unilateral orbital, supraorbital, and/or temporal pain lasting 2 to 30 minutes
C. At least one of the following symptoms or signs ipsilateral to the pain:
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
Miosis and/or ptosis
D. Attacks have a frequency above five per day for more than half of the time
E. Attacks are prevented absolutely by therapeutic doses of indomethacin
F. Not better accounted for by another ICHD-3 diagnosis

Table 3 International Classification of Headache Disorders 3 Criteria for Subdivisions of Paroxysmal Hemicrania

Episodic paroxysmal hemicrania
Attacks fulfilling criteria for paroxysmal hemicrania and occurring in bouts
At least two bouts lasting from 7 days to 1 year (when untreated) and separated by pain-free remission periods of ≥ 1 month
Chronic paroxysmal hemicrania
Attacks fulfilling criteria for paroxysmal hemicrania and criterion below.
Occurring without a remission period or with remissions lasting < 1 month for at least 1 year

nausea, vomiting, photophobia, phonophobia, osmophobia, or vertigo.

In the present case, the presence of short-duration unilateral headaches in the distribution of the trigeminal nerve with corresponding activation of the autonomic nervous system (lacrimation) suggested a TAC as a possible cause for the episodic severe pain. At the initial consultation, the episodic pain was given a tentative diagnosis of chronic paroxysmal hemicrania because of the clinical features and nature of the headache episodes, female gender, and lack of familial history. Similarly, the constant pain was given a diagnosis of masticatory and cervical myalgia pain disorder based on location and duration of pain, modification of pain with functional and parafunctional movements, and replication of pain with palpation of the masticatory and cervical muscles. The patient was empirically placed on a trial of indomethacin 75 mg (extended release) twice a day for the management of chronic paroxysmal hemicrania and omeprazole 40 mg once a day (to counter any gastrointestinal adverse effects associated with the use of indomethacin), and for the management of myalgia, the patient was placed on metaxalone 800 mg twice a day and referred for multimodal manual physical therapy with appropriate adjunctive modalities.

The patient was contacted 5 days after initiation of the indomethacin trial for a follow-up. She reported complete resolution in the episodic pain symptoms within 24 hours of using indomethacin and moderate relief in the more constant, dull, aching symptoms associated with masticatory and cervical myalgia pain disorder. She had started physical therapy and was scheduled for a total of 12 sessions (2 sessions per week). The diagnosis of chronic paroxysmal hemicrania was felt to be confirmed. The patient was advised to continue taking the prescribed medications and to complete the course of physical therapy. She was scheduled for a follow-up 6 weeks later.

On her subsequent follow-up visit, the patient reported complete resolution in pain and associated symptoms. She did not report any pain on examination of the head and neck regions. At this time, an initial taper of the dose of indomethacin and metaxalone was started. At 3 months after her initial consultation, she was taking indomethacin 75 mg once a day, omeprazole 40 mg once a day, and metaxalone 800 mg once a day. Any further taper in the dosage of indomethacin or metaxalone resulted in the onset of episodic and constant pain symptoms. At her last follow-up at the Oral and Maxillofacial Pain Clinic, 16 months after her initial consultation, the patient was completely pain free and doing very well. She will be followed annually at this time. Meanwhile, her PCP will follow up with her every 3 months to perform laboratory studies (renal function test, liver function test, and complete blood count) to ensure that she does not experience any serious adverse effects—such as nephrotoxicity, hepatotoxicity, anemia, or blood dyscrasias—from the regular use of medications.

Discussion

The International Classification of Headache Disorders, third edition (ICHD-3) describes paroxysmal hemicrania as attacks of severe, unilateral pain in the orbital, supraorbital, and/or temporal regions lasting 2 to 30 minutes and occurring more than five times a day for more than half of the time. These attacks should be accompanied by the presence of at least one ipsilateral autonomic sign (lacrimation, conjunctival injection, rhinorrhea, nasal congestion, forehead and/or facial sweating, forehead and/or facial flushing, miosis, ptosis, fullness in the ear, and/or eyelid edema). Similarly, paroxysmal hemicrania has an absolute response to indomethacin therapy (Table 2). It is further divided into episodic and chronic subtypes based on the remission period between headache episodes (Table 3).

Although in the present case the patient did not experience more than five episodes of headache per day, she presented with typical clinical features of PH. Furthermore, she had characteristic absolute resolution of pain with the use of indomethacin. The headache frequency of less than five headache episodes per day is not uncommon in PH and has previously been reported.[15]

The pathophysiology of PH is complex. It has been postulated that altered metabolic activity in the hypothalamic region can subsequently result in activation of the trigeminal brainstem nucleus via the anatomical pathways present between the hypothalamic nuclei and the trigeminal nucleus (trigemino-thalamic pathways).[16] Furthermore, trigeminovascular stimulation can take place as a result of reflex activation of the parasympathetic outflow from the superior salivatory nucleus (SSN). Stimulation of the trigeminovascular system leads to vasodilation of the peripheral blood vessels, which can cause neuropraxic injury to the perivascular sympathetic plexus, resulting in sympathetic dysfunction. This explains the strong autonomic component of these headache attacks.[16,17]

Paroxysmal hemicrania has an absolute response to indomethacin. The mechanism is poorly understood; however, it has been suggested that in addition to cyclooxygenase inhibitory properties, indomethacin results in inhibition of the central nociceptive system and nitric oxide synthesis.[18] The goal of physical therapy and muscle relaxant therapy in masticatory and cervical myalgia pain disorders is to relieve pain and restore function. This patient had previously received appropriate therapy for masticatory and cervical myalgia pain disorder; however, it did not provide her with complete relief of symptoms. This may be due to the ongoing headaches associated with chronic PH. Activation of trigeminovascular afferents results in activation of the trigeminal nucleus and cervical divisions (C1 and C2) through the trigeminocervical complex, which may result in initiation and perpetuation of muscle pain in the masticatory and cervical regions.[1,11] Similarly, ongoing muscle pain may act as an initiating and perpetuating factor for the headaches. Due to this, it was necessary to address the two disorders together to have an optimal outcome.

Conclusions

This case illustrates the importance of a thorough history and clinical examination. In the present case, the clinical symptoms associated with episodic pain in the temporal region could have been mistaken for referred pain associated with masticatory and cervical myofascial pain disorder, neuropathic pain, or odontogenic disease. However, the unilateral presentation, duration and frequency of episodes, and the association of pain with ipsilateral symptoms of lacrimation suggested a diagnosis of paroxysmal hemicrania. Patients with uncommon and refractory signs and symptoms require a thorough investigation for the rare causes of headache and facial pain.

Acknowledgments

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