Trigeminal Schwannoma simulating orofacial pain: differential diagnosis and treatment. Case report

Schwannoma trigeminal simulando dor orofacial: diagnóstico diferencial e tratamento. Relato de caso

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ABSTRACT

BACKGROUND AND OBJECTIVES: Orofacial pain is a major diagnostic challenge for the most experienced clinicians. Due to the complexity regarding the trigeminal-cervical joint, orofacial pain with the same etiology may present different symptoms, and pain with similar symptomatology may have different causes. The objective of this study was to alert health professionals about the importance of differential diagnosis in the hypothesis of trigeminal neuralgia, where the inclusion of the dentist in the medical-hospital team is of paramount importance in establishing the correct diagnosis.

CASE REPORT: Twenty-nine-year-old female patient complained of electric shock and pulsatile orofacial pain that covered the third division of the fifth cranial nerve on the right side. Magnetic resonance imaging revealed the presence of trigeminal Schwannoma, causing neuralgia due to its neural compressive nature. Two different neurosurgery departments suggested tumor resection. However, after the evaluation by a third neurosurgery department, in which a dentist, specialized in orofacial pain was part of the team, the complete evaluation established the final diagnosis of right lower first molar odontalgia, with irreversible acute pulpitis as the cause of the symptoms and the expansive lesion was only a radiological finding.

CONCLUSION: Interdisciplinary evaluation among physicians and dental surgeons is necessary to obtain the correct diagnosis when considering the hypothesis of trigeminal neuralgia.

Keywords: Facial pain, Neurilemmoma, Referred pain, Trigeminal neuralgia.

RESUMO

JUSTIFICATIVA E OBJETIVOS: As dores orofaciais representam grande desafio diagnóstico ao mais experiente clínico. Devido à complexidade do conjunto trigeminocervical, as dores orofaciais com a mesma etiologia podem apresentar sintomas diferentes, e dores com o mesmo sintoma podem ter etiologias diferentes. O objetivo foi alertar o profissional da saúde sobre a importância do diagnóstico diferencial quando aventada a hipótese diagnóstica de neuralgia trigeminal, e a presença do cirurgião-dentista na equipe médico-hospitalar é de suma importância para o estabelecimento do diagnóstico.

RELATO DO CASO: Paciente do sexo feminino, 29 anos, apresentou dor orofacial em choque elétrico e pulsátil que percorria o trajeto da terceira divisão do V par craniano, do lado direito. A ressonância nuclear magnética evidenciou Schwannoma trigeminal, sendo diagnosticada neuralgia trigeminal secundária à essa lesão expansiva. Foi sugerida ressecção tumoral em dois serviços de neurocirurgia. Contudo, após a avaliação de um terceiro serviço de neurocirurgia, com a participação de cirurgião-dentista especialista em dor orofacial, foi estabelecido o diagnóstico de odontalgia do primeiro molar inferior direito, com pulpite aguda irreversível, sendo essa a causa do quadro sintomático, e a lesão expansiva, apenas um achado radiológico.

CONCLUSÃO: A avaliação interdisciplinar entre médicos e cirurgiões-dentistas é necessária para o diagnóstico correto quando a hipótese diagnóstica for neuralgia trigeminal.

Descritores: Dor facial, Dor referida, Neuralgia do trigêmeo, Neurilemoma.

INTRODUCTION

Orofacial pain (OFP) comprises pain conditions located in the oral cavity and on the face. Other extra-trigeminal anatomical structures can also promote pain referred to this segment, such as the skull and the cervical region. This is due to a rich innervation and broad somatosensory representation of the central nervous system (CNS), which makes the clinical diagnosis a great challenge, even for the most experienced professional[1].

The differential diagnosis of OFP can be related to numerous diseases and/or conditions that affect these regions primarily or secondarily, that is, the site of the pain is not necessarily the primary source of the painful stimulus, on the contrary, it can be a symptom of diseases set in distant regions, such as intracranial and cervical[2].

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OFP can also result from several diseases of inflammatory, infectious, and/or neoplastic nature, which directly affect these structures[2,3].

The objective of the study was to warn health professionals about the importance of having a dental surgeon, specialist in OFP, in the hospital medical team for multi and interdisciplinary care.

CASE REPORT

A twenty-nine-year-old female patient reported that in October 2018 she had the first episode of facial pain, like an electric shock, with paroxysms, on the path of the third trigeminal division (V3) on the right side, with intensity 8/9 on the visual analog scale (VAS). The neurologist requested a nuclear magnetic resonance imaging (MRI) of the skull base with a focus on the trigeminal ganglion, which showed nodular foci along the distal segment of the cisternal portion of the V cranial nerve on the right, which could cause compression and medial displacement of the cisternal path. After gadolinium contrast, these foci were impregnated, more marked next These foci were impregnated after the gadolinium contrast, markedly next to the trigeminal sensory root, raising the hypothesis of Schwannoma (Figure 1).

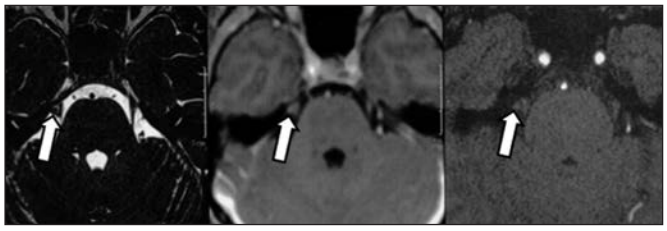

Figure 1. Expansive lesion by trigeminal Schwannoma

The patient was medicated with carbamazepine (400mg) twice a day, nortriptyline (50mg) once a day, and tramadol (100mg) three times a day, with partial pain relief. She was informed about the need to resection the Schwannoma, as it was housed in the trigeminal region and was responsible for the pain. However, the patient did not agree with the surgery. With moderate pain, five, according to VAS, the patient sought another neurosurgery service that corroborated the diagnosis of trigeminal neuralgia, secondary to Schwannoma, with the indication of surgery and pregabalin, 75mg in the morning and 150mg at night. The pulsatile and persistent shock-like pain continued with intensity four, by VAS. In February 2019, she sought another neurosurgery service when a hypoesthesia was observed in the second and third trigeminal divisions (V2 and V3) on the right, in addition to the same painful symptom, five. During hospitalization for venous analgesia, the patient also reported pain with, intensity eight, in the right mandibular hemiarco, which worsened during chewing. The dental surgeon of the team examined the patient, and at the thermal test with ice spray at -50°C to check the pulp vitality of the right lower 1st molar, tooth 46, the patient reported an unbearable painful crisis of intensity 10, which lasted about three minutes, gradually regressing to intensity =5. The indication for endodontics of tooth 46 was made and the periapical radiography showed a deep restoration that bordered the dental pulp (Figure 2).

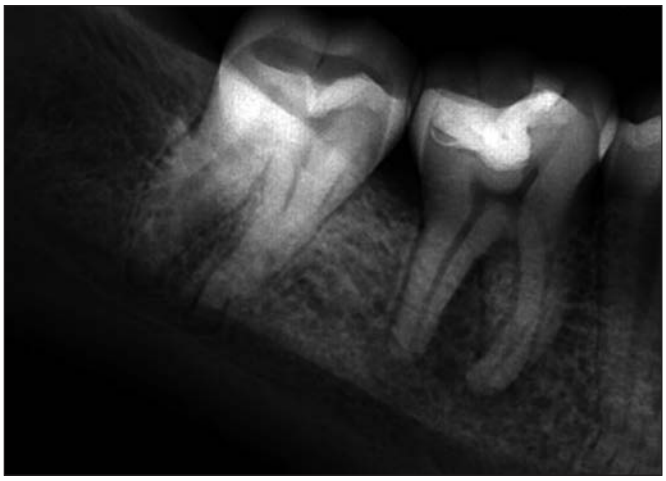

Figure 2. Initial periapical radiography

This tooth had been restored approximately two months before the onset of the painful crises, and the teeth became extremely sensitive to thermal stimuli after the restorative procedure. When performing the endodontics of tooth 46, it was observed signs of vital failure in the dental pulp, and irreversible acute pulpitis (IAP) was diagnosed. In the first session, the root canals were prepared using the Reciproc Blue (VDW) system and intracanal drug insertion with calcium hydroxide paste (Ultracal, Ultradent), after the temporary sealing with stick gutta-percha and glass ionomer. Ibuprofen (600mg) was prescribed at every eight hours, and viminol (70mg), if necessary. At that time, the patient was no longer using other analgesic drugs. After 15 days, a canal filling session was performed using the single cone technique in the mesial canals and lateral condensation in the distal canal using bioceramic endodontic cement BioRoot RCS (Septodont) (Figure 3), and temporary dental restoration with glass ionomer before the final restoration. Currently, the patient is asymptomatic in the follow-up of the Schwannoma evolution.

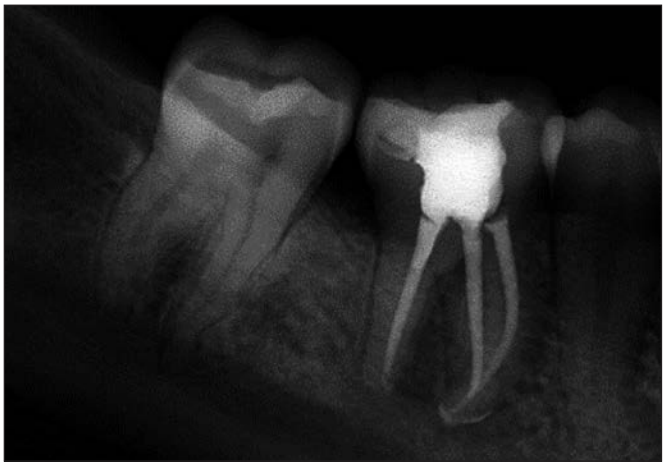

Figure 3. Periapical radiography after the endodontic treatment

DISCUSSION

The trigeminal nerve (TN) is considered the largest pair of cranial nerves, being classified as a mixed nerve because it has sen-

sory and motor roots that receive sensory input from the face. Due to its extensive distribution in the cephalic segment, from central regions to peripheral endings, a range of diseases can affect this complex system and often mimic pain that does not correspond to the reported etiological factor. However, they can present the same symptom[4].

Secondary trigeminal neuralgia can be caused by an underlying disease, such as Schwannoma, being evidenced by MRI that shows the nervous compression and, clinically, sensory alterations with the presence of paroxysms ipsilateral to the radiological finding[5].

Intracranial trigeminal Schwannomas (ITS) are rare neoplasms of the Schwann cells that can occur in any peripheral, cranial, or autonomic nerves. They have slow, insidious growth and are predominantly benign. They affect individuals in adulthood between the 4th and 6th decades of life. They have a slightly higher incidence in women, accounting for approximately 0.07 to 0.33% of all intracranial neoplasms, and 1 to 8% of all intracranial Schwannomas[6-9].

Clinically, the most common symptoms are sensory changes such as hypoesthesia, paresthesia, in addition to OFP, which can involve the trigeminal pathways of the first, second and third division of this cranial nerve, according to the location of the neoplasia[7-10].

The patient presented with a Schwannoma in the trigeminal cisternal region, correlated with pain in the V2 and V3 paths on the right side. However, studies report that neoplasms involving the trigeminal ganglion usually present continuous pain, whereas neoplasms involving their roots are asymptomatic[7,11] which would justify further investigation of the case.

Study[6] evaluated 42 patients diagnosed with ITS who underwent surgical intervention for neoplastic resection and observed at the time of hospital admission that 62% of the individuals had hypoesthesia or paresthesia, whereas only 7% developed facial pain[6]. Similarly, another study[7] conducted with 68 patients with the same diagnosis, found that facial pain was less common compared to facial hypoesthesia.

In the present study, the diagnosis of TN secondary to Schwannoma was based on symptoms and imaging results. However, no exclusion diagnosis was carried out in full. Despite the presence of trigeminal Schwannoma, the proper assessment of the oral cavity should have been performed by a dental surgeon before any neurosurgical procedure, an assessment that was performed only at the third neurosurgery service, when the appropriate investigation procedure was followed (Table 1).

The patient's report was decisive in the evolution of her treatment, when she said that the pain on her face were exacerbated when she chewed something hard. Several studies report that extensive carious processes and extensive restorative procedures very close to the pulp cavity have a high chance of causing irreversible acute pulpitis, that is, an irreversible condition of pulp inflammation[13-16].

This clinical condition is associated with severe pain that can be continuous and/or remitting, with no spontaneous resolution, often tricky to locate by the patient[13]. In the initial stage, the patient still manages to identify the affected tooth, which is confirmed by the pulp vitality test with the ice spray at -50°C, which will show a hyperalgesic response greater than 30 seconds. However, in chronic cases, the pain has a diffuse and reflex characteristic to several regions of the ipsilateral skull section, such as teeth, face and head, being exacerbated by heat and the masticatory act, whereas the cold can relieve painful symptoms[12,14,17]. It is sometimes pulsatile, paroxysmal and sharp[14].

Table 1. Patient's history model proposed at the bedside (Adapted)[12]

1- Request and/or ask about the history of previous dental treatments and especially about restorations and/or recent trauma
2- Ask the patient fundamental questions:
a- Where is the pain located?
b- When did the pain start?
c- What is the intensity of the pain?
d- What is the quality of the pain? Examples: burning, stinging, shock, needles, pinprick, etc.
e- What is the duration of the pain?
f- How frequent is the pain?
g- Pain relief factors? Any kind of drug?
h- Pain worsening factors? Examples: cold, hot, chewing.
3- Thorough evaluation of the oral cavity in order to check for the presence of caries, fractured teeth, exposed dentin, defective restorations, soft tissue with swollen and/or hyperemic areas, residual roots, semi-submerged and/or impacted teeth.
4- Perform the vertical percussion test to exclude acute periodontitis.
5- Perform the thermal hyperalgesia test to cold using an ice spray at -50°C or ice stick. It is important to start the thermal test always on the contralateral side to the pain.
6- Perform the thermal hyperalgesia heat test using a heated gutta-percha stick, or even an external heat source, to reproduce the pain.
7- Request, as soon as possible, panoramic, and/or periapical test to identify caries that are not clinically visible, extensive restorations, endodontic and/or periodontal lesions.

The patient reported that she had undergone a recent restorative procedure on element 46, and after surgery, the sensitivity to thermal stimuli was exacerbated. The analysis of the periapical radiography showed close contact of the restorative material with the roof of the pulp chamber. Studies state that bacterial infection is the crucial factor in the pathogenesis of pulp disease, and the infectious process may be due to caries, dental trauma or restorative procedure with pulp exposure and/or contamination[15,18] suggesting that in this clinical case there was pulp exposure and, consequently, its contamination.

The dental pulp is a loose connective tissue inside the pulp chamber and the root canal, containing a vascular-nervous bundle with specific functions of nutrition and sensory input to the dental organ[15]. When suffering some type of injury and/or aggression, it triggers a process called neurogenic inflammation[19] with increased expression of neuropeptides, such as substance P, CGRP, in addition to the release of potent inflammatory media-

tors, such as prostaglandins E2, prostaglandins F2a, interleukins 1 and 6 and TNF-α[15,16]. It also appears to increase neuronal excitability and activate isoforms of nociceptors such as Nav 1.7, 1.8, 1.9, orchestrating an immense cellular immune response[16]. Thus, the clinical conditions arising from this pulp inflammatory process include neuroplasticity, allodynia, hyperalgesia, peripheral sensitization and central sensitization[16], data that help explain the complexity of irreversible acute pulpitis and how easy it can be mistaken for trigeminal neuropathic pain.

CONCLUSION

The patient with OFP needs to be carefully assessed since the pain treatment permeates multifactorial aspects with different etiologies that can be associated with the same symptoms. Therefore, an interdisciplinary evaluation is necessary for the correct diagnosis, since not all paroxysmal pain, whether shock or burning, means the presence of trigeminal neuralgia.

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