Case Report

Trigeminal neuralgia in a young person: case report and literature review

Neuralgia trigeminal em jovem: Relato de caso e revisão da literatura

Gabriela de França Ribeiro Espindola¹, Marianna Siqueira Cabral Queiroz², Anderson Nunes Teixeira², Miguel de Lemos Neto².

¹ Medical Student, Faculdade de Medicina de Campos (FMC), Campos dos Goytacazes, RJ, Brazil. ² Professor, Faculdade de Medicina de Campos (FMC), Campos dos Goytacazes, RJ, Brazil. Corresponding author: Gabriela de França Ribeiro Espindola Contact: gabrielafribeiroe@gmail.com

ABSTRACT

Trigeminal neuralgia is the most common type of neuropathic pain affecting the head and neck region. It is more prevalent in individuals over 60 years old and uncommon in those under 40. Mediators are released that overload the trigeminal nerve with an excess flow of information, reaching the cortex and being interpreted as pain, causing allodynia and hyperalgesia. The diagnosis of trigeminal neuralgia is predominantly clinical and can be classified as typical or atypical. Imaging exams may be necessary for those under 40 years old or to rule out secondary causes. The first-choice treatment is carbamazepine, which is effective in 70% of cases. This report aims to characterize a rare case of trigeminal neuralgia in a 21-year-old patient and to conduct a literature review on the pathophysiology, diagnosis, and treatment of this condition.

INTRODUCTION

Neuropathic pain is defined as a morbidity that directly affects the somatosensory system, being a chronic condition and therefore difficult to treat and highly incapacitating, causing impairments in various areas such as sleep disturbances, anxiety, depression, and other psychiatric demands. Trigeminal neuralgia is the most common type of neuropathic pain affecting the head and neck region. The trigeminal nerve is the fifth cranial nerve and is a mixed nerve with both motor and sensory roots. The sensory branches form the maxillary and ophthalmic nerves, and the mandib-
ular by somatic afferent fibers, responsible for the somatic sensitivity of most of the head. These fibers conduct external and proprioceptive stimuli, while its motor portion innervates the muscles involved in mastication.

Trigeminal neuralgia is a condition that causes intense suffering and limitations in affected individuals, with a lifetime prevalence estimated at 0.16%-0.3% and an annual incidence of 4-29 per 100,000 person-years. It is more prevalent in women than in men (female to male ratio of 3:2). The incidence increases with age, with an average onset age of 53 to 57 years and a range of 24 to 93 years in adult series. Pediatric cases of trigeminal neuralgia are rare.

Patients report excruciating lancinating pain in the regions innervated by the fifth cranial nerve (trigeminal nerve), worsening during crisis periods, which can occur at any time, even with correct treatment. The sensation is often described as "shocks" in short bursts, which can be triggered by simple and everyday actions such as eating, talking, brushing teeth, or even touching the affected area, with the maxillary and mandibular branches being the most affected. Its highest incidence is in people over 60 years old, being uncommon in those under 40.

The information contained in this report was obtained from medical records, photographic records of diagnostic methods to which the patient was submitted, as well as a descriptive literature review. The case report was approved by the local research ethics committee under number CAAE 36907620.4.0000.5244.

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**CASE REPORT**

A 21-year-old female patient, born in Rio de Janeiro, single, student, and without a history of comorbidities. For five years, she experienced isolated episodes of intense pain (score of 10 on the verbal numeric scale) in the right periorbital region, not radiating, which did not cease with the use of dipyrone alone but only with the association of dipyrone with promethazine hydrochloride and adifenine hydrochloride, which had a sedative effect and made her sleep, alleviating the symptoms. Four years later, these episodes became more frequent, and she began to have equally intense pain crises (score of 10 on the verbal numeric scale) in the right hemiface, with an average duration of 2-3 hours, accompanied by a sensation of electric shocks and paresthesia, increasing in frequency and intensity, and worsening with mastication, low temperatures, and brushing teeth, receiving a clinical diagnosis of trigeminal neuralgia. Neurological physical examination showed no abnormalities. A computed tomography scan was performed with no significant findings. She started taking carbamazepine (100 mg/day) with persistent episodes. The dose was adjusted to 500 mg/day, and new exams were performed: electroencephalogram, which showed an irregular alpha pattern revealing dysrhythmic and diffuse irritative bursts, predominant in frontal areas, suggesting episodes of anxiety although this statement lacks scientific support; as well as angiography and radiography of the paranasal sinuses and temporomandibular joints without alterations, not identifying the origin of the symptomatology. Currently, the patient takes carbamazepine (200 mg/day) and now has only sporadic and less intense crises, resolving after administration of dipyrone (1g).

**DISCUSSION**

Literature data indicate that injuries in any body structure promote the release of various mediators such as pro-inflammatory cytokines, growth factors, enzymes, and NO (nitric oxide) that increase the excitability of the fiber, exacer-
The most relevant central mechanism seems to be the increased expression of IL-1β in the trigeminal nuclei, also responsible for allodynia, hyperalgesia, and pain chronicity.

The causes behind these changes may include nerve root compression by a tumor, anatomical abnormalities of the skull base that may compress the trigeminal nerve, arteriovenous malformations, or multiple sclerosis, with the latter being the most commonly related to this condition. With the advent of magnetic resonance imaging, the vascular origin is increasingly frequent, and it is postulated that the pulsation of these vessels, usually arterial, contributes to demyelination of the fibers, increasing their afferent stimuli.

There is also familial trigeminal neuralgia, which is extremely rare and can be caused by inherited anatomical changes or mutations in the calcium channel gene, contributing to hyperexcitability. Refractoriness is also a hallmark of trigeminal neuralgia, which can last for months or even years. Its pathophysiology is not well described but may be related to hyperpolarization of sensory neurons.

Currently, the condition can be diagnosed with the patient’s clinical history according to the criteria of the International Headache Society (IHS) and the International Association for the Study of Pain (IASP), being: 1. Character (shooting, electric shock, lancinating, superficial); 2. Intensity (moderate to very intense); 3. Duration (less than 2 minutes, numerous throughout the day); 4. Periodicity (weeks or months without pain); 5. Location (distribution area of the trigeminal nerve); 6. Radiation (within the innervated area); 7. Trigger factors (innocuous stimuli); 8. Relief factors (usually sleep and anticonvulsants); 9. Associated factors (trigger zones, low quality of life, depression).

As the main diagnostic source is the patient’s clinical history, it is important to pay attention to possible indications of differential diagnoses such as previous symptoms indicative of herpes zoster, dental procedures that may have caused trauma to the nerve root, tension headaches, and...
temporomandibular joint disorders, for example\textsuperscript{11}.

Imaging exams are used for complementary investigation and are indicated for young patients (under 40 years old) with atypical symptoms or those who do not respond to treatment, to determine the origin of the condition and to rule out other diagnostic hypotheses. The main complementary exams are: computed tomography, magnetic resonance imaging, with magnetic angioresonance being the gold standard exam to identify vascular compressions in the nerve root\textsuperscript{9}.

Trigeminal neuralgia can be classified as typical when the symptomatology is classic, or atypical; and as primary when the origin of the condition is not elucidated or secondary when the pain is triggered by some basal alteration such as brain tumors, for example\textsuperscript{8}. When classified as atypical, it can be mixed or neuropathic, where there will be persistent discomfort between pain peaks or may be followed by loss of sensitivity and will usually be of secondary origin, although it can also be rarely found in idiopathic cases\textsuperscript{6, 12}. The patient should seek a specialist such as a neurosurgeon or neurologist if they exhibit symptoms characteristic of atypical neuralgia, such as burning pain between crises, loss of sensitivity, or any abnormal neurological signs, lack of effectiveness of treatment with carbamazepine or gabapentin, or if it occurs in individuals under 40 years old.

The treatment for these cases, besides the use of anticonvulsants, includes the administration of antidepressants such as amitriptyline and neuroleptics such as chlorpromazine. Carbamazepine is a first-choice anticonvulsant drug for 70% of trigeminal neuralgia cases. It works by blocking sodium channels, reducing neuronal excitability, which causes significant adverse effects such as drowsiness, dizziness, skin rashes, and tremors\textsuperscript{8}. Oxcarbazepine is indicated if the patient does not respond well to carbamazepine, as it causes fewer adverse effects and poses less risk of drug interactions. It is important to consider these undesirable effects because, in most cases, treatment inefficacy is not due to the medication dosage but rather the bothersome reactions that lead to therapeutic discontinuation\textsuperscript{9}. The management of these drugs must be careful due to their toxicity (especially carbamazepine) and the development of tolerance over time\textsuperscript{22}, which reduces therapeutic efficacy by 50% despite the dose increase\textsuperscript{8}.

Other drugs that can be used as adjuvants in therapy include lamotrigine, which blocks sodium and calcium channels, baclofen, which acts on GABAb receptors, pregabalin, or gabapentin. If drug therapy is not effective, surgical interventions may be an alternative\textsuperscript{3}.

The surgical approach to trigeminal neuralgia has become increasingly early due to the possibility of drug toxicity and tolerance\textsuperscript{3}. One of the techniques used is non-destructive management, which consists of neurovascular decompression, directly treating the cause of the problem while preserving nerve function. Another type of treatment is called destructive management, which includes percutaneous techniques and radiosurgery, which has the advantage of being a non-invasive procedure\textsuperscript{22} but may be associated with recurrences in 50% of cases within five years\textsuperscript{8}.

Some factors presented by the patient support the diagnostic hypothesis of trigeminal neuralgia, such as pain characterization (intense with shock episodes precipitated by specific trigger factors), duration (seconds to a few minutes), periodicity, absence of neurological deficits on examination, and sleep as the main relief factor, which are clinical aspects considered in the diagnosis according to IHS and IASP\textsuperscript{3, 7}.

Despite all these manifestations being common in typical trigeminal neuralgia cases, some factors support the diagnosis of atypical trigeminal neuralgia, such as the paresthesia reported by the patient, which is not a characteristic symptom of the classic manifestation of the disease, and the patient’s age, as this condition is more common in people over 60 years old\textsuperscript{2, 7}.

However, despite clinically fitting into the category of atypical trigeminal neuralgia, whose
ideal treatment would be the combination of an anticonvulsant with antidepressant and neuroleptic drugs, the patient is treated only with the anticonvulsant carbamazepine, showing significant improvement in symptoms with a reduction in intensity and frequency of pain crises.

The pathophysiology of trigeminal neuralgia is diverse and involves a series of inflammatory cascades that generally culminate in allodynia (painful sensation with innocuous stimulation) and hyperalgesia (exaggerated sensitivity to pain). The diagnosis of trigeminal neuralgia is predominantly clinical according to IHS and IASP criteria, and it can be classified as typical when the symptoms fit the criteria. Imaging exams may be requested to rule out secondary causes and in patients under 40 years old. The most commonly used exams are computed tomography and magnetic resonance imaging of the brain, in addition to magnetic angiography, which has high sensitivity for identifying microvascular causes.

The first-choice treatment for this condition is the anticonvulsant drug carbamazepine, which is effective in 70% of cases, and can be associated in specific situations with antidepressants such as amitriptyline and neuroleptics such as chlorpromazine. In cases of intolerance to the adverse effects of carbamazepine, a safer alternative is oxcarbazepine. In case of refractoriness to pharmacological treatment or in situations of very severe adverse effects, surgical approaches are considered.

In summary, this article is relevant for exposing a case of trigeminal neuralgia in a young patient and providing information on the pathophysiology and treatment of this condition, which, although rare, causes many limitations in its sufferers, highlighting the importance of further studies for a better understanding of the subject.

REFERENCES