Neuralgia do Trigêmeo Bilateral. Relato de Caso *

Bilateral Trigeminal Neuralgia. Case Report

Caio Marcio Barros de Oliveira, TSA [1], Luis Gustavo Baaklini [2], Adriana Machado Issy [3], Rioko Kimiko Sakata, TSA [4]

RESUMO

Oliveira CMB, Baaklini LG, Issy AM, Sakata RK - Neuralgia do Trigêmeo Bilateral. Relato de Caso.

JUSTIFICATIVA E OBJETIVOS*: A neuralgia do nervo trigêmeo é uma condição intensamente dolorosa, caracterizada por surtos de dor lancinante e súbita, tipo choque, com duração de poucos segundos a dois minutos e geralmente unilateral. Sua incidência anual é de cerca de 4,3 em 100.000 na população geral, tendo manifestação bilateral em apenas 3% desses casos. O objetivo deste artigo foi descrever um caso raro de neuralgia do trigêmeo primário bilateral.*

RELATO DO CASO*: Paciente de 61 anos, maranhense, casada, do lar, com antecedente de hipertensão arterial e há seis anos com queixa de dor intensa em V2-V3 à esquerda, com duração de 5 a 10 segundos, em região lateral do nariz e mandibular, com piora ao falar, mastigar e com diminuição da temperatura. Já havia utilizado clorpromazina (3 mg a cada oito horas) e carbamazepina (200 mg a cada oito horas) durante seis meses sem alívio da dor. Ao exame físico apresentava alodinia térmica e mecânica em regiões de V2-V3. Estava em uso de gabapentina (1.200 mg ao dia) com alívio parcial da dor. Foi então aumentada a gabapentina para 1500 mg ao dia e introduzida amitriptilina 12,5 mg à noite. Evoluiu com dor leve e esporádica com diminuição da intensidade da dor ao longo de 10 meses de tratamento, sendo reduzida progressivamente a gabapentina para 600 mg ao dia e mantida a amitriptilina 12,5 mg ao dia. Após um ano, começou a apresentar dor de característica semelhante em região mandibular à direita, tendo melhorado com aumento de gabapentina para 900 mg ao dia. Não apresentava exames alterados de tomografia ou ressonância magnética de encéfalo.*

CONCLUSÕES*: A carbamazepina é o fármaco de primeira escolha para tratamento de neuralgia trigeminal, porém a gabapentina tem sido cada vez mais utilizada como primeira medida farmacológica ou em casos refratários à terapia convencional.*

Unitermos: DOR, Crônica: neuralgia de trigêmeo; DROGAS, Anticonvulsivante: carbamazepina, gabapentina.

SUMMARY

Oliveira CMB, Baaklini LG, Issy AM, Sakata RK – Bilateral Trigeminal Neuralgia. Case Report.

BACKGROUND AND OBJECTIVES: *Trigeminal neuralgia is an extremely painful condition characterized by recurrent episodes of sudden, lancinating, shock-like pain lasting from a few seconds to two minutes usually unilateral. It has an annual incidence of approximately 4.3 in 100,000 in the general population and only 3% of those cases present bilateral manifestation. The objective of this report was to describe a rare case of bilateral trigeminal neuralgia.*

CASE REPORT: *A 61 years old housewife from Maranhão, Brazil, married, with a history of hypertension, presented with a six-year history of severe pain in the left V2-V3 regions, lasting 5 to 10 seconds, in the lateral aspect of the nose and mandible, worsening by talking, chewing, and with a decrease in temperature. She had been treated with chlorpromazine (3 mg every eight hours) and carbamazepine (200 mg every eight hours) during six months without improvement. On physical exam, the patient presented thermal and mechanical allodynia in the V2-V3 regions. She was using gabapentin (1,200 mg/day) with partial relief of the pain. The dose of gabapentin was increased to 1,500 mg/day and amitriptyline 12.5 mg at night was added to the therapeutic regimen. The patient evolved with mild and sporadical pain and a reduction in pain severity during 10 months; the dose of gabapentin was progressively reduced to 600 mg/day, and amitriptyline was maintained at 12.5 mg/day. After one year, the patient developed similar pain in the region of the right mandible, which improved with an increase in the dose of gabapentin to 900 mg/day. Head CT and MRI did not show any abnormalities.*

CONCLUSIONS: *Carbamazepine is the first choice for the treatment of trigeminal neuralgia; however, the use of gabapentin as the first pharmacological choice or in cases refractory to conventional therapy has been increasing.*

Keywords: DRUGS, Anticonvulsants: carbamazepine, gabapentin; PAIN, Chronic: trigeminal neuralgia.

** Recebido da (**Received from**) Disciplina de Anestesiologia, Dor e Terapia Intensiva Cirúrgica da Escola Paulista de Medicina da Universidade Federal de São Paulo (EPM/UNIFESP), São Paulo, SP*

1. Anestesiologista; Mestre em Medicina, Especializando em Dor pela EPM/UNIFESP
2. Anestesiologista; Especializando em Dor pela EPM/UNIFESP
3. Professora Adjunta da Disciplina de Anestesiologia, Dor e Terapia Intensiva Cirúrgica da EPM/UNIFESP
4. Professora Associada da Disciplina de Anestesiologia, Dor e Terapia Intensiva Cirúrgica e Responsável pela Clínica de Dor da EPM/UNIFESP

*Apresentado (**Submitted**) em 30 de outubro de 2008*
*Aceito (**Accepted**) para publicação em 08 de abril de 2009*

*Endereço para correspondência (**Correspondence to**):*
Dr. Caio Marcio Barros de Oliveira
R. Dr. Diogo de Faria, 539/47
Vila Clementino
04037-001 São Paulo, SP
E-mail: caio.oliveira@unifesp.br

Bilateral Trigeminal Neuralgia. Case Report

Caio Marcio Barros de Oliveira, TSA, M.D. [1], Luis Gustavo Baaklini, M.D. [2], Adriana Machado Issy, M.D. [3], Rioko Kimiko Sakata, TSA, M.D. [4]

INTRODUCTION

Trigeminal neuralgia is an uncommon disorder characterized by sudden, recurrent, paroxysmic episodes of lancinating shock-like pain, lasting from a few seconds to two minutes, and usually involving the maxillary branch [1]. It is almost always unilateral, but it is bilateral in rare cases, and the right side is affected more often than the left side (1.5:1), probably due to greater narrowing of the round and oval foramina on that side [2,3]. Pain is typically triggered by speaking, drinking, brushing the teeth, shaving, chewing, light touch, or by a breeze, and it can occur repeatedly during the day [2]. Trigeminal neuralgia has an annual incidence of 4.3 per 100,000 in the general population with discrete predominance of females (3:2). It has a peak around 60 to 70 years, and it is rare before the age of 40 [2,4]. Hypertensive patients have a higher risk of developing trigeminal neuralgia than the general population [5]. Approximately 80 to 90% of the cases classified as idiopathic are caused by compression of the trigeminal nerve (5th cranial nerve) when it leaves the brain stem by an aberrant arterial or venous loop, especially of the superior cerebellar artery [2,5]. Tumor, multiple sclerosis, abnormalities of the base of the skull, or arteriovenous malformation is the cause in 5 to 10% of the patients [1]. Multiple sclerosis is the disease more frequently associated with trigeminal neuralgia affecting 1 to 5% of the patients [2,6]. It has been proposed that trigeminal neuralgia is caused by demyelinization of the nerve, which leads to ephaptic impulse transmission [7]. The diagnosis is eminently clinical, although imaging studies or specialized tests might be necessary in patients with atypical manifestations: younger than 40 years, bilateral symptoms, dizziness or vertigo, hearing loss or changes, pain episodes lasting more than two minutes, pain outside the distribution of the trigeminal nerve, and visual changes [2]. Patients with trigeminal neuralgia have typical episodes and changes in the site, severity, or pain quality should alert for the possibility of alternative diagnoses [5]. The objective of this report was to describe a rare case of primary bilateral trigeminal neuralgia.

CASE REPORT

This is a 61 year-old married housewife with a history of hypertension. She had a six-year history of severe (score of 10 in the Verbal Numeric Scale), shock-like and throbbing pain in the left V2-3 region, lasting 5 to 10 seconds that increased by talking, chewing, and with cold weather. She did not sleep

well because of the pain. Physical exam showed thermal and mechanical allodynia in the left V2-3 regions. She had used the following medications: chlorpromazine (3 mg every eight hours) for one year, discontinued eight months ago; carbamazepine (200 mg every eight hours) for six months and discontinued five months ago. She had been using gabapentin (1,200 mg a day) for five months with partial improvement. In the Pain Clinic, the dose of gabapentin was increased for 1,500 mg/day and amitriptyline 12.5 mg/day was added. Imaging exams of the head (CT and MRI) were requested and did not show any abnormalities. The patient presented a good evolution during 10 months of treatment with only sporadic, mild pain when it was cold. The dose of gabapentin was reduced progressively to 600 mg/day and the dose of amitriptyline was maintained at 12.5 mg at night. After one year of follow-up she complained of similar pain in the left mandible (V3) that improved with an increase in the dose of gabapentin to 900 mg/day.

DISCUSSION

In general, trigeminal neuralgia is unilateral affecting the maxillary (35%), mandibular (30%), maxillary and mandibular (20%), ophthalmic and maxillary (10%), and ophthalmic (4%) branches, and all branches (1%) of the trigeminal nerve. It has an incidence of 4.3 per 100,000/year, of which approximately 3% are bilateral [12]. Diagnostic criteria are defined by the IASP (International Association for the Study of Pain) and by the ICHD/IHS (International Classification of Headache Disorders/International Headache Society), and include [8,9]:

1. Paroxysmal episodes lasting from a fraction of a second to two minutes, affecting one or more divisions of the trigeminal nerve;
2. The pain has at least one of the following characteristics:
   a) severe, sudden, superficial, or stabbing;
   b) initiated by trigger-factors or trigger points.
3. Episodes are similar among patients;
4. Patients do not have clinically evident neurologic changes; and
5. It is not attributed to other disorder.

One should be alert for the presence of atypical signs and symptoms that indicate the presence of an underlying disorder like: abnormal neurological exam; abnormal oral, odontological, or ear exam; age below 40 years: bilateral symptoms; dizziness or vertigo; hearing loss or impairment; numbness; pain lasting more than two minutes; pain outside the distribution of the trigeminal nerve; and visual changes. Imaging exams are mandatory [2,4,5]. The physical exam of patients with classical trigeminal neuralgia is predominantly normal. Therefore, sensorial abnormalities in the trigeminal area, loss of the corneal reflex, or weakness in any facial muscle should raise the suspicion of secondary causes of neuralgia [10].

The most common disorders involved in the differential diagnosis include: bursts of headaches, dental pain, giant cell arteritis, glossopharyngeal nerve neuralgia, intracranial tumor, migraine, multiple sclerosis, otitis media, paroxysmal hemicrania, postherpetic neuralgia, sinusitis, SUNCT headache (Short-lasting, Unilateral, Neuralgiform pain with Conjunctival injection and Tearing), temporomandibular joint syndrome, and trigeminal neuralgia [10].

It is very difficult to undertake controlled therapeutical studies in trigeminal neuralgia because the cases are rare and withholding treatment for a patient in the placebo group is unacceptable [1]. Carbamazepine is the drug of choice for the initial treatment of trigeminal neuralgia [11]. It has a number needed to treat (NNT) of 2.5, and its dose varies from 100 to 2,400 mg/day; most cases respond with 200 to 800 mg/day in two divided doses [2].

When the side effects of carbamazepine are severe, it is possible to administer oxcarbamazepine (200 to 2,400 mg/day), a byproduct of carbamazepine that is better tolerated. However, the risk of crossed allergic reaction between carbamazepine and oxcarbamazepine is approximately 25% [12]. In the case of partial pain relief with carbamazepine, a second agent can be added or the drug can be changed [12]. Gabapentin has been used as a first-line agent or in cases of trigeminal neuralgia resistant to the traditional therapy, with complete or almost total remission in 27% of the cases [13,14]. Gabapentin is considered a second-line medication and definitive scientific evidence of its efficacy in the treatment of trigeminal neuralgia does not exist [13]. Its clinically effective dose when used as monotherapy varies from 900 to 1,200 mg/day, but it can be as high as 3,600 mg/day. Gabapentin does not cause direct reduction of ectopic discharges in the trigeminal ganglion, but it interferes with nociceptive transmission in the central nervous system, acting on the $\alpha_2\delta$ subunit of voltage-dependent calcium channels [14,15]. The main collateral effects of this drug include dizziness and somnolence, and confusion, ataxia, and peripheral edema are associated with high doses [14].

Surgical treatment can be used when the clinical treatment fails or in patients who do not tolerate the pharmacological treatment [1,2]. Surgical procedures can be percutaneous or opened. The choice of the type of surgical intervention should be based on the preferences of the patient, experience of the surgeon, and potential risks and benefits of each procedure. Percutaneous techniques include balloon compression, radiofrequency rizothomy, and stereotactic radiosurgery with gamma rays. Those are relatively non-invasive procedures, done on an outpatient basis, and lack severe side effects. However, the pain relief they promote is not as long-lasting as that of more invasive procedures (20% recurrence in five years after balloon compression [16]) and cause postoperative sensorial loss in a large proportion of patients [16-22].

Open surgeries include partial rizothomy and microvascular decompression that involve the exploration of the posterior cranial fossa involving the increased risk of severe complications (for example, meningitis and death) [16]. Microvascular decompression is considered the technique that promotes

high long-lasting pain relief with pain control in more than 70% of patients in 10 years [23,24]. Consensus on the best treatment for trigeminal neuralgia, clinical or surgical, does not exist. However, pain relief, recurrence, morbidity, and mortality should be considered [2,16].

REFERENCES

01. Bennetto L, Patel NK, Fuller G - Trigeminal neuralgia and its management. BMJ 2007;334:201-205.
02. Krafft RM - Trigeminal neuralgia. Am Fam Physician 2008;77: 1291-1296.
03. Neto HS, Camilli JA, Marques MJ - Trigeminal neuralgia is caused by maxillary and mandibular nerve entrapment: greater incidence of right-sided facial symptoms is due to the foramen rotundum and foramen ovale being narrower on the right side of the cranium. Med Hypotheses 2005;65:1179-1182.
04. Manzoni GC, Torelli P - Epidemiology of typical and atypical craniofacial neuralgias. Neurol Sci 2005;26(suppl 2):s65-67.
05. Nurmikko TJ, Eldridge PR - Trigeminal neuralgia: pathophysiology, diagnosis and current treatment. Br J Anaesth 2001;87:117-132.
06. Osterberg A, Boivie J, Thuomas KA - Central pain in multiple sclerosis: prevalence and clinical characteristics. Eur J Pain 2005;9:531-542.
07. Love S, Coakham HB - Trigeminal neuralgia: pathology and pathogenesis. Brain 2001;124:2347-2360.
08. Headache Classification Subcommittee of the International Headache Society - The International Classification of Headache Disorders: 2nd edition. Cephalalgia 2004;24(Suppl 1):9-160.
09. Lindena G, Diener HC, Hildebrandt J et al. - Leitlinien zur schmerztherapie: methodische qualitat von leitlinien zur therapie von patienten mit schmerzen. Schmerz 2002;16:194-204.
10. Zakrzewska JM - Diagnosis and differential diagnosis of trigeminal neuralgia. Clin J Pain 2002;18:14-21.
11. Wiffen PJ, McQuay HJ, Moore RA - Carbamazepine for acute and chronic pain. Cochrane Database Syst Rev 2005:CD005451.
12. Canavero S, Bonicalzi V - Drug therapy of trigeminal neuralgia. Expert Rev Neurotherap 2006;6:429-440.
13. Wiffen PJ, McQuay HJ, Edwards JE et al. - Gabapentin for acute and chronic pain. Cochrane Database Syst Rev 2005:CD005452.
14. Cheshire Jr WP - Defining the role for gabapentin in the treatment of trigeminal neuralgia: a retrospective study. J Pain 2002; 3:137-42.
15. Lemos L, Flores S, Oliveira P et al. - Gabapentin supplemented with ropivacain block of trigger points improves pain control and quality of life in trigeminal neuralgia patients when compared with gabapentin alone. Clin J Pain 2008;24:64-75.
16. Ong KS, Keng SB - Evaluation of surgical procedures for trigeminal neuralgia. Anesth Prog 2003;50:181-188.
17. Hall GC, Carroll D, McQuay HJ - Primary care incidence and treatment of four neuropathic pain conditions: a descriptive study, 2002-2005. BMC Fam Pract 2008;9:26.
18. Kalkanis SN, Eskandar EN, Carter BS et al. - Microvascular decompression surgery in the United States, 1996 to 2000: mortality rates, morbidity rates, and the effects of hospital and surgeon volumes. Neurosurgery 2003;52:1251-1262.
19. Lim M, Villavicencio AT, Burneikiene S et al. - CyberKnife radiosurgery for idiopathic trigeminal neuralgia. Neurosurg Focus 2005;18:E9.
20. Massager N, Lorenzoni J, Devriendt D et al. - Radiosurgery for trigeminal neuralgia. Prog Neurol Surg 2007;20:235-243.
21. Massager N, Murata N, Tamura M et al. - Influence of nerve radiation dose in the incidence of trigeminal dysfunction after trigeminal neuralgia radiosurgery. Neurosurgery 2007;60:681-688.
22. Hai J, Li ST, Pan QG - Treatment of atypical trigeminal neuralgia with microvascular decompression. Neurol India 2006;54:53-57.
23. Tomasello F, Alafaci C, Angileri FF et al. - Clinical presentation of trigeminal neuralgia and the rationale of microvascular decompression. Neurol Sci 2008;29(Suppl 1):s191-195.
24. Tronnier VM, Rasche D, Hamer J et al. - Treatment of idiopathic trigeminal neuralgia: comparison of long-term outcome after radiofrequency rhizotomy and microvascular decompression. Neurosurgery 2001;48:1261-1268.

RESUMEN:

Oliveira CMB, Baaklini LG, Issy AM, Sakata RK - Neuralgia del Trigémino Bilateral: Relato de Caso.

JUSTIFICATIVA Y OBJETIVOS*: La neuralgia del nervio trigémino es una condición intensamente dolorosa, caracterizada por brotes de dolor lancinantes y súbitos, del tipo descarga eléctrica, con una duración de pocos segundos a dos minutos y generalmente unilateral. Su incidencia anual es de cerca de 4,3 en 100.000 en la población general, manifestándose bilateralmente en solo un 3% de esos casos. El objetivo de este artículo fue describir un caso raro de neuralgia del trigémino primario bilateral.*

RELATO DEL CASO*: Paciente de 61 años, del estado brasileño de Maranhão, casada, ama de casa, con antecedentes de hipertensión arterial y hace seis años quejándose de dolor intenso en V2-V3 a la izquierda, con una duración de 5 a 10 segundos en la región lateral de la nariz y la mandíbula, con empeoramiento al hablar, masticar y con una reducción de la temperatura. Ya había utilizado clorpromazina (3 mg a cada ocho horas), y carbamazepina (200 mg a cada ocho horas), durante seis meses sin que se le aliviase el dolor. Cuando se le examinó físicamente, presentaba alodinia térmica y mecánica en regiones de V2-V3. Estaba usando gabapentina (1.200 mg al día), con alivio parcial del dolor. Se le aumentó entonces la gabapentina para 1.500 mg al día y se le introdujo la amitriptilina 12,5 mg por la noche. La paciente desarrolló un ligero y esporádico dolor, con una reducción de su intensidad a lo largo de 10 meses de tratamiento, siendo reducida progresivamente la gabapentina para 600 mg al día y mantenida la amitriptilina 12,5 mg al día. Después de un año, empezó a presentar dolor de características similares en la región mandibular a la derecha, y mejoró con el aumento de la gabapentina para 900 mg al día. No presentaba exámenes de tomografía o resonancia magnética de encéfalo alterados.*

CONCLUSIONES*: La carbamazepina es el fármaco de primera elección para el tratamiento de la neuralgia trigeminal, sin embargo la gabapentina ha sido cada vez más utilizada como primera medida farmacológica o en casos refractarios a la terapia convencional.*