ORIGINAL ARTICLE

Cluster headache: a challenging headache

Cefaleia em salvas: uma cefaleia desafiante

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ABSTRACT

Introduction: cluster headaches (CS) are a subtype of primary headache disorder characterized by daily pain attacks of 15-180 minutes for weeks to months, usually interspersed with periods of remission. Painful crises are very intense, of periorbital or orbital location, associated with symptoms of autonomic dysfunction. Despite its unique clinical presentation, CS remains under-recognized and underdiagnosed. **Objectives:** To describe and discuss the diagnostic and therapeutic challenges of CS based on clinical cases. **Methods:** this is a series of five cases of CS under clinical monitoring. **Results:** Of the 467 patients treated at the neurological clinic five had a diagnosis of CS, corresponding to 1.07% of the total. The clinical presentation varied little in relation to the description in the literature; it affected predominantly men, episodically. Time elapsed between onset and diagnosis was usually long. **Conclusion:** Even in a tertiary care neurological center, the number of diagnosed patients is small, which reflects on ignorance about CS and contributes to late diagnosis and lack of specific treatment.

Key words: Headache/diagnosis; Headache/therapy; Cluster Headache/diagnosis; Cluster Headache/therapy; Lithium Carbonate.

RESUMO

Introdução: *a cefaleia em salvas (CS) é um subtipo de cefaleia primária caracterizado por crises de dor entre 15 e 180 minutos diárias por semanas a meses, geralmente intercalados por períodos de remissão. As crises álgicas são muito intensas, de localização orbital ou periorbital, associadas à sintomatologia de disfunção autonômica. Apesar de sua singular apresentação clínica, a CS permanece afecção pouco reconhecida e subdiagnosticada.* ***Objetivos:*** *descrever e discutir os desafios diagnósticos e terapêuticos da CS a partir de casos clínicos.* ***Material e método:*** *trata-se de uma série de cinco casos de CS em acompanhamento clínico.* ***Resultados:*** *dos 467 pacientes assistidos em ambulatório neurológico, cinco possuíam diagnóstico de CS, correspondendo a 1,07% do total. A apresentação clínica variou pouco em relação à descrição da literatura, com predomínio em homens e da forma episódica, além de haver grande latência entre seu surgimento e o diagnóstico.* ***Conclusão:*** *mesmo em centro terciário de atendimento neurológico, o número de pacientes diagnosticados é pequeno, colaborando para o desconhecimento a respeito da CS, o que contribui para o seu atraso diagnóstico e tratamento específico.*

Palavras-chave: *Cefaleia/diagnóstico; Cefaleia/terapia; Cefaleia Histamínica/ diagnóstico; Cefaleia Histamínica/terapia; Carbonato de Lítio.*

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INTRODUCTION

The cluster headache (CH) is an unusual form of primary headache, characterized by episodes of very strong pain, usually located in the orbital, periorbital, or

temporal regions, lasting between 15 and 180 minutes, with frequencies ranging from one crisis every other day to up to eight episodes per day. Three daily crises are referred on average, which tend to occur at the same time following a circadian pattern. These headaches are always associated to at least one symptom of autonomic dysfunction that can be: conjunctival hyperemia, watery eyes, nasal congestion, rhinorrhea, facial sweating, miosis, ptosis and/or palpebral edema, always ipsilateral to the pain.[1] The headache crises occur in outbreaks or clusters (hence the English name of "cluster headache") lasting from weeks to months, usually interspersed by periods of remission that can exceed one month.[1]

They affect four men for every woman[2] and the first outbreak usually occurs between 20 and 30 years of age. Factors such as alcohol consumption, climate changes, odors, and bright lights are described as possible triggers of the crisis.[3]

This study aims to show that CH, even with its singular clinical presentation, can pose a challenge for clinicians and a burden for patients who frequently suffer for years before being diagnosed and treated properly. Here we present the main clinical characteristics of patients with CH being followed up in the Headache Clinic (AMBCEF) at the University Hospital (HC) from the Federal University of Minas Gerais (UFMG) and report one case that illustrates specifically the diagnostic and therapeutic challenges associated with CH.

MATERIALS AND METHODS

This was a descriptive and transversal study including five patients with a CH diagnosis, followed up in the AMBCEF-UFMG between January and August 2012. The assistance in this tertiary center is performed by neurologists, psychiatrists, dentists, and nutritionists; the target audience is individuals over 12 years old. Referrals come from doctors in the Emergency Room of the Neurology Service and from other services at the University Hospital from UFMG.

The consultations in the AMBCEF-UFMG are based on a semi-structured approach involving an interview and clinical neurological examination. After each consultation, the team of neurologists discusses the diagnosis, which is established according to criteria followed by the International Classification of Headaches (2004)[1], and the therapeutic strategies to be applied. After this evaluation, the patients are assisted by a multidisciplinary team, which updates the therapeutic guidelines.

This study integrates a research project previously approved by the COEP-UFMG 0500.0.203.000-10.

RESULTS

Until August of 2012, 467 patients were regularly followed up by the AMBCEF-UFMG. Of these, five had their diagnoses established as CH, which represents 1.07% of the total number of patients (Table 1). The male/female ratio was 1.5:1 and the age at the onset of symptoms ranged from 14 to 71 years old. The time interval between the onset of symptoms and establishment of a diagnosis was of nine years on average.

CH evolved in two cases without the period of remission between "clustered" pain (chronic form) and in three cases with the period of remission between "clustered" pain (episodic form). The remission periods ranged from 3 to 8 months and the periods of crisis between 3 weeks and 3 months.

The daily frequency of headache episodes ranged from 1 to 4 with a duration between 30 and 90 minutes. The location of pain predominated on the left side and orbital and supraorbital regions. In one of the patients, the episodes of pain and autonomic symptoms alternated between the sides of the face, however, occurring predominantly on the left side.

All autonomic symptoms were ipsilateral to the location of pain; conjunctival hyperemia and watery eyes were present in all cases. Nasal congestion and palpebral ptosis were also frequent, while rhinorrhea, facial sweating, and miosis were less common. Agitation or psychomotor restlessness was also observed during crises in four patients.

All patients reported using analgesic self-medication without significant impact on the headache; two managed to abort the crisis with oxygen therapy; prophylaxis was achieved in four patients with verapamil and in one with lithium carbonate.

CASE REPORT

This is a male patient resident in Belo Horizonte. At the age of 71 years, he presented an intense algic crisis at the left orbital region associated with conjunctival hyperemia, watery eyes, nasal congestion, and ipsilateral palpebral ptosis. The daily crises occurred at every 2 hours and lasted continuously from 30 to 60 minutes.

Table 1 - Clinical data from patients with diagnosis of cluster headache followed up at the Headache Clinic at the University Hospital from the UFMG

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age (years)	32	36	79	55	72
Gender	M	F	M	F	M
Age at onset of symptoms	14	11	71	50	68
Age at diagnosis	26	24	79	55	69
Time interval †	12	13	8	5	1
Site of pain	Orbital Supraorbital Temporal	Orbital Temporal Frontal	Orbital Supraorbital	Orbital	Retro-orbital
Side involved	L	L	L	Predominantly – L Occasionally – R	L
Crisis duration ‡	90 min	60 min	30 min	60 min	40-60min
Crisis frequency	2x/day	1-6x/day	2-3x/day	3-4x/day	2x/day
Duration of pain period – cluster	2 months/year	1-3 months/year	Continuous	3 weeks/3-6 months	Continuous
Pain intensity	Very intense	Very intense	Very intense	Very intense	Very intense
Autonomic symptoms	Conjunctival hyperemia Watery eyes Nasal congestion Rhinorrhea Facial sweating Miosis Palpebral ptosis	Conjunctival hyperemia Watery eyes Palpebral ptosis	Conjunctival hyperemia Watery eyes Nasal congestion Palpebral ptosis	Conjunctival hyperemia Watery eyes Nasal congestion Facial sweating	Conjunctival hyperemia Watery eyes Palpebral ptosis Rhinorrhea
Neurological symptoms	Photophobia Psychomotor agitation	Psychomotor agitation	Photophobia	Psychomotor agitation Photophobia Phonofobia Osmofobia Local paresthesia	Psychomotor agitation
Other symptoms	No	Night terrors Discomfort Nausea Gastric fullness	No	Discomfort Nausea	Nausea

† Between symptoms onset and diagnosis. ‡ When not treated.

He was initially diagnosed in 2003 with trigeminal neuralgia; the patient received an outpatient treatment with carbamazepine (800 mg/day) for three months with no satisfactory response. In 2004, he was submitted to percutaneous rhizotomy by radiofrequency of the V cranial nerve and showed remission of symptoms for two years.

After this period, the pain of strong intensity returned accompanied by the same autonomic symptoms and periodicity without intercritical interval. He had undergone several drug treatments in multiple health services, including the Emergency Room at the HC from the UFMG where, at the end of 2011, eight years after the onset of his symptoms, he was diagnosed with CH.

Since then, he has been followed up in the AMBCEF-UFMG, with an initial use of verapamil (80 mg/TID), showing a significant improvement. The medication was discontinued after an episode of dyspnea, bradycardia, and cardiac branch block on the electrocardiogram. He currently and regularly uses lithium carbonate (300 mg/BID) and is in complete remission of crises, however, the medication causes moderate kinetic tremor in his right hand (Figure 1), which is well tolerated. There was an attempt to reduce this dosage in order to mitigate the tremor; however, the "threat of pain" led the patient himself to request returning to the original dose. The V cranial nerve ablation resulted in permanent paresthesia of the left hemiface.

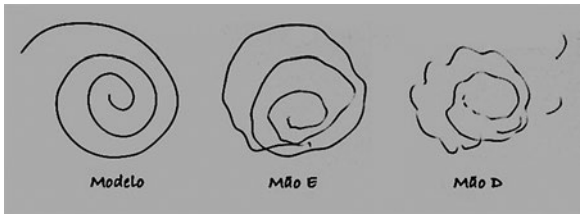

Figure 1 - Kinetic tremor resulting from the use of lithium carbonate.

DISCUSSION

The CH prevalence of 1.07% observed in this study is relatively high compared to its estimated prevalence in the general population as around 0.12%.[2] This may be explained, at least in part, by the group of patients being treated in a specialized service. It should be noted that, even in this context, the number of cases is reduced. The small number of CH cases is observed in other health centers in the country[5], which might result from under-diagnoses because the disease is poorly recognized by doctors.

In this study, the average delay in diagnosis was nine years, and in accordance with the findings of Todd et al.[3], who reports five-year average diagnostic delays in 42% of 1,134 American patients with CH, and 10 years delays or more in 22% of these patients.

The case reported illustrates the trajectory of many patients who, for years, undergo several health centers and, without being properly diagnosed, see themselves subjected to treatments that do not solve their symptomatology.

The drug treatment for CH includes abortive and prophylactic therapies. The most commonly employed abortive therapies include sumatriptans, dihydroergotamine derivatives, and inhalation of high-flow oxygen.[4] With the onset of symptoms and prior to an established diagnosis, self-medication with over-the-counter painkillers and oral anti-inflammatories is common as well as the inadvertent prescription of potent painkillers, usually without satisfactory response and often used in abusive ways. Corticosteroids or dihydroergotamine are used as a transitional prophylactic therapy for short periods of time. The most common prophylactics of continuous usage are verapamil, lithium carbonate, methysergide, sodium divalproex, and, more rarely, melatonin and topiramate. It should be noted that through proper treatment CH is controllable and many are the patients who go into a complete remission of their symptoms.[3,4]

The differential diagnosis includes migraine, dental and facial sinuses diseases, and allergies in addition to the trigeminal neuralgia. Invasive procedures, such as those reported in the case described are performed frequently, such as tooth extraction, surgery of the sinuses, and occipital nerve electrical stimulation.[6] Diagnostic errors probably result from lack of knowledge and similarities with migraines. Indeed, the presence of nausea, vomiting, and visual aura is relatively common and represent symptoms that should guide the CH diagnosis.[7] There are reports that the frequent triggering factors in CH patients are also observed in patients who suffer from migraines - alcohol use, climate changes, odors, and bright lights.[2,6]

CONCLUSION

CH is an uncommon condition that, although presenting clinical characteristic manifestations, is still little known. This lack of knowledge is a determining factor in its diagnosis that when not properly done leads to several unnecessary, ineffective, and often invasive treatments.

There are few large-scale studies on CH and none of them were performed in Brazil. The number of CH patients who wait for years to receive a proper diagnosis and treatment is considerable. Further studies are needed to increase the knowledge about CH, which will allow for adequate diagnosis, treatment, and prophylaxis and less suffering to CH patients.

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