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Management of cluster headache in the Emergency Department

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Abstract Although cluster headache (CH) is considered one of the most distinctive and painful primary headache disorders in clinical practice because of the brevity of each attack, its management is not always ergonomic or possible in the Emergency Department. In case of a previously competent diagnosis, the Emergency Department's team should send the patient to a headache centre where specialists in the management of CH can handle the pathology in the best way. In our headache centre we treat patients with CH attacks with a hyperbaric chamber, confirming the effectiveness of hyperbaric oxygen in CH patients.

Key words Cluster headache • Management • Emergency Department • Hyperbaric oxygen

Introduction

Cluster headache (CH) is one of the most distinctive and painful primary headache disorders in clinical practice. It is characterised by a series of sudden attacks of severe pain, lasting from 30 min to 2 h, localised invariably on one side and the side affected generally is consistent for every attack and every cluster period.

Most headache disorders seen in emergency departments are of a benign aetiology, with migraine being responsible for 60% of all headache presentations. In contrast, CH accounts for only 2% [1]. Although CH is uncommon in this and other settings, an accurate diagnosis is important because it allows for effective treatment for CH and helps to avoid unnecessary diagnostic and treatment interventions.

Barriers to accurate diagnosis and appropriate treatment include the brevity of the attacks, which may end

before evaluation, the rarity of the disorder and consequent lack of physician familiarity with its presentation, and the lack of specificity of the autonomic features.

Epidemiology

CH is considerably less common than migraine and is predominantly a disease of men. The gender ratio varies between 5.0:1 and 6.7:1, however recent reviews suggest a progressive reduction in the male to female ratio.

Estimations in the literature indicate that lifetime prevalence of CH is between 0.1% and 0.4% and the incidence is between 2.5% and 9.8% cases per 100,000 people each year.

CH usually begins between 20 and 40 years (peak to 20 in episodic), but it may begin at any age [2].

Clinical manifestations

The pain is of terrible intensity and is typically described as boring, tearing and may be likened to a "hot poker in the eye" or as if "the eye as being pushed out". Onset is typically abrupt or preceded by a brief sensation of pressure or mild discomfort in the soon-to-be painful area. The pain intensifies rapidly, peaking in 5–10 min and may stay at maximal intensity for 45–90 min. After the attack the patient is pain-free but exhausted [3].

A notable feature of CH is its short duration; in fact each untreated attack typically lasts for 15–180 min (average 30–120 min), with more than 75% of attacks reported lasting less than 60 min [4]. The location of pain is strictly unilateral and almost always remains on the same side of the head between cluster periods. Rarely, the pain may occur on the opposite side in a subsequent period and even less frequently, attacks may switch to the opposite side within the same cluster period. The pain is typically in the retro-orbital and temporal region on the affected side. The pain may radiate into the forehead, temple, cheek and jaw. In contrast to migraine, patients are restless during the attack.

The attacks commonly occur one to three times daily, although they may be as variable as one every other day to up to eight daily. The daily attacks usually last for 1–2 months (the cluster period). The headaches then remit spontaneously only to recur again in a cluster of daily headaches months to years later [5, 6].

Some patients experience variable sensations antecedent to their attacks of CH. Those occurring several minutes to 1 hour before the attack are referred to as prodromal symptoms. Those occurring several hours or days before the attacks are called premonitory symptoms. These

symptoms may include sensory phenomena such as paraesthesias, burning sensations, sharp pains and stabbing pains; mood changes such as nervousness; alteration in the sleep pattern such as snoring, erratic sleep and insomnia [7, 8].

We can define two different states of CH: episodic CH is characterised by attacks that occur in periods lasting 7 days to 1 year separated by pain-free periods lasting one month [9]; chronic CH is present when attacks occur without remission or with remission lasting less than one month.

Treatment of cluster headache

Because of the rapid onset and short time to peak intensity of the pain of CH, a fast acting symptomatic therapy is imperative [10]. The solutions that give the most effective, rapid and reliable relief for attacks of CH are the following.

Sumatriptan

Although the mechanism of migraine attacks remains unexplained, it is thought that an important role is played by serotonin receptors, vasodilatation in certain regions and opening of arteriovenous communication in the head. Sumatriptan is an agonist of 5-HT1-like receptors and exerts a selective vasoconstricting effect on the arteries of the head, particularly in the rami of the carotid artery.

Subcutaneous sumatriptan is the most effective self-administered medication for the symptomatic relief of CH [11]. In a placebo-controlled study, 6 mg of sumatriptan delivered subcutaneously was significantly more effective than placebo, with 74% of patients having complete relief by 15 min compared with 26% of patients treated with placebo.

However, sumatriptan is not effective when used before an expected attack in an attempt to prevent an oncoming attack, nor is it useful as a prophylactic agent. The overall efficacy of sumatriptan has been reported to be approximately 8% less in patients with chronic CH than in patients with episodic cluster headache. Patients with chronic CH respond very well but to a somewhat lesser extent and more slowly than patient with episodic CH.

Zolmitriptan

Zolmitriptan is an effective oral agent for the acute treatment of migraine. Recently, a double-blind controlled trial compared the efficacy of 5 and 10 mg oral zolmitriptan

with placebo for the treatment of acute CH attacks [12]. With headache response defined as a two-point reduction on a five-point pain intensity scale at 30 min, response rates following placebo, 5 and 10 mg of zolmitriptan, were 29%, 40% and 47%, respectively. The difference reached statistical significance for 10 mg zolmitriptan compared with placebo. In addition, significantly more patients reported mild or no pain 30 min after treatment with 5 and 10 mg of zolmitriptan than following placebo.

Zolmitriptan is the first orally administered triptan to demonstrate efficacy in the treatment of CH and remains a therapeutic option in patients who cannot tolerate oxygen or subcutaneous sumatriptan or for those in whom an oral medication is desired.

Oxygen

Oxygen inhalation has been the standard of care for the symptomatic relief of CH since it was introduced as an effective therapy by Horton. If delivered at the onset of an attack via a non-rebreathing facial mask at a flow rate of 7 l/min for 15 min, approximately 70% of patients will obtain relief within 15 min. In some patients, oxygen is most effective if taken when the pain is at maximal intensity, whereas in others the attack is delayed for minutes to hours rather than completely aborted [13, 14].

Hyperbaric oxygen

It has been shown that exposure to hyperbaric oxygen is able not only to interrupt a cluster headache but also to prevent the successive attacks occurring for 2 or 3 days after the treatment [15]. The mechanism for this effect has

not been completely clarified but we can already verify the effect of exposure to hyperbaric condition on the content of the immunoreactivity to substance P in the nasal mucosa of CH patients [16, 17]. One of the first experiments was based on 14 patients suffering from an episodic form of CH [18]. The diagnosis was made following the criteria of the International Headache Society [9]. Before admission to the trial, each patient provided a complete history and underwent a physical examination (ears, nose, throat and a radiography of the chest and electrocardiography). The subjects were placed in a multiplace-pluriambient hyperbaric chamber (Galeazzi, La Spezia, Italy). While in the hyperbaric chamber the subjects breathed either 100% oxygen saturation (administered through a facial mask) or environmental air.

Discussion

We have been working on clinical and experimental findings since 1996, in collaboration with the Anaesthetic and Reanimation Department of Umberto I, and in this time we have found several confirmations of our first hypothesis about the implementation of this treatment. Although CH is considered one of the most distinctive and painful primary headache disorders in clinical practice, because of the brevity of each attack the management of CH is possible in the Emergency Department only in the case of a previous competent diagnosis or after partial or complete recovery, which may have occurred by the time of evaluation. In this case, the Emergency Department team may provide the right prophylaxis to the patient or send the subject to a headache centre equipped with a hyperbaric chamber. Today we can certainly affirm the positive effect of hyperbaric oxygen on episodic CH and triptan non-responder chronic CH.

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