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Therapeutic approaches to Occipital Neuralgia – a case report

Abstract

Occipital neuralgia (ON), is a painful condition affecting the posterior head in the distributions of the greater occipital nerve (GON), lesser occipital nerve (LON), third occipital nerve (TON), or a combination of the three. In 90% of cases, ON is a result of GON pathology. This condition results almost always from the compression of one of these nerves at one of several anatomic points. Irritation of the GON and/or LON by chronically contracted muscles and spondylosis of the upper cervical spine is often implicated. Clinically, these patients refer a shooting or stabbing pain in the neck, usually unilateral, that radiates over the cranium, characterized as persistent, paroxysmally aggravating, and of variable distribution. The primary conservative treatment approach for patients with ON aims at reducing muscle tension and improving posture. If non-pharmacological and pharmacological treatments are unsuccessful, minimally invasive interventional procedures should be considered. Anesthetic blocks of the GON and LON can be utilized both for diagnostic purposes and as a therapeutic option.

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Introduction

Occipital neuralgia (ON), first described in 1821 by J. Beruto y Lentijo and M.M. Ramos,[1] is a secondary headache disorder with occipital pain as its key feature.[2] It is a painful condition affecting the posterior head in the distributions of the greater occipital nerve (GON), lesser occipital nerve (LON), third occipital nerve (TON), or a combination of the three.[3]

The GON, also known as Arnold's nerve, is the dorsal branch of the second cervical nerve (C2) which is a mixed nerve, primarily sensory. Along its course, it splits in two branches: the lateral branch, which supplies the splenius capitis muscle, and the medial branch, which anastomizes with the branches of C1 and C3 nerves. The medial branch also innervates the semispinalis and lower oblique muscles, and provides sensory innervation to the skin in the occipital region.[2]

In 90% of cases, ON is a result of GON pathology and 10% of cases are due to LON causes. The involvement of TON is very rare. This condition results almost always from the compression of one of these nerves at one of several anatomic points.[3]

Irritation of the GON and/or LON by chronically contracted muscles and spondylosis of the upper cervical spine is often implicated. Additionally, there are other rare etiologies of ON that include compression from intra or extracranial blood vessels, giant cell arteritis, callus formation after vertebral fractures, schwannomas, and other masses. Its incidence remains uncertain.[4] A study in the Dutch general population reported a relatively low incidence of 3.2 per 100,000, showing a female dominance but not significant, and no seasonal variation was found.[4]

Clinically, these patients refer a shooting or stabbing pain in the neck, usually unilateral, that radiates over the cranium, characterized as persistent, paroxysmally aggravating, and of variable distribution. Due to connections with the VIII, IX, and X cranial nerves and the cervical sympathicus, vision impairment/ocular pain (67%), tinnitus (33%), dizziness (50%), nausea (50%), and congested nose (17%) may occur.(4-6) Upon physical examination, this may present with tenderness to pressure over the course of the GON ou LON and hypo- or dysesthesia in these areas. A positive Tinel's sign can be present as well.[5] When patients lie on a pillow and hyperextend or rotate their neck, pain may occur ("pillow sign").[4]

The diagnosis is confirmed by a local anesthetic block on the suspected nerve, which is, according to International Classification of Headache Disorder (ICHD-II), an essential step to confirm the diagnosis.[3] When a GON block is effective, pain relief usually begins within 20-30 minutes and can last from several hours to several months.[6]

Single diagnostic blocks have a false-positive rate of up to 40%, so performing a second block is advisable. If both blocks yield positive results, the provider can be more confident in the accuracy of the diagnosis. Since compression is believed to be the primary cause of ON, imaging should be considered if there is any suspicion of a lesion or mass at the affected site.[3] An ultrasound is commonly used to evaluate the course of the GON and can be useful to determine if there is a nerve entrapment or if it appears enlarged or swollen. X-ray and computed tomography (CT) can be used to exclude bony sources. Magnetic resonance imaging (MRI) is typically preferred to evaluate soft tissues in the occipital and cervical areas.[7]

Regarding treatment and management, the primary conservative treatment approach for patients with ON aims at reducing muscle tension and improving posture. Recommended treatments include rest, warm or cold compresses, massage, and physical therapy to help alleviate muscle tension.[5] Devices that deliver electrical impulses have been employed in the conservative treatment of painful conditions, since there are case reports that highlight the potential advantages of using transcutaneous electrical nerve stimulation therapy (TENS) for treating ON.[8]

Nonsteroidal anti-inflammatory drugs (NSAIDs), acetaminophen, and muscle relaxants are commonly used. Anticonvulsants, including gabapentin, pregabalin, carbamazepine, and oxcarbazepine, along with selective serotonin reuptake inhibitors (SSRIs) and tricyclic antidepressants (TCAs) like nortriptyline, may help reduce the

severity and frequency of ON over the long term. However, opioids are not recommended due to a lack of supporting evidence.[8]

If non-pharmacological and pharmacological treatments are unsuccessful, minimally invasive interventional procedures should be considered. Anesthetic blocks of the GON and LON can be utilized both for diagnostic purposes and as a therapeutic option.[8]

A prospective study involving 44 patients who received unilateral or bilateral GON blocks, or a combination of GON and LON blocks with local anesthetic and corticosteroids, demonstrated a significant reduction in pain scores. The same result has been seen in a retrospective study involving 46 patients who underwent thermal radiofrequency (RF) ablation of the GON and LON. However, RF ablation is not without risks, so pulsed RF that use temperature below the level of nerve destruction, has a reduced risk of deafferentation pain syndrome caused by thermal RF. Cryoneurolysis is an alternative to continuous and pulsed RF treatments since it uses internally cooled RF probes, which act as heat sinks, reducing the risk of tissue charring. This technique creates deeper and larger lesions compared to continuous RF ablation.[9]

Multiple studies have demonstrated that Botulinum toxin A (BoNT-A) infiltration provides pain relief that lasts longer than its muscle relaxant effects.[4] A retrospective study[4] concluded that BoNT-A has been shown to effectively reduce the sharp, shooting pain typically associated with occipital neuralgia, but not the dull or aching pain. Improved quality of life was obtained for several months.[10] Surgical management is considered a last resort when medical and minimally invasive treatment options have failed.[8]

In some cases, diagnostic injections using local anesthetics, with or without steroids, can provide pain relief lasting several months. The advanced treatments previously mentioned may lead to improvements that can last from a few weeks to several years.[3]

Clinical case description

This clinical case concerns a 30-year-old, healthy young woman, who developed a cervical muscle contracture following a gym session. The patient attributes this to improperly performing a series of sit-ups, which caused excessive engagement of the neck muscles instead of the abdominal muscles.

After 24 hours, the patient reported pain in the left cervical region associated with muscle stiffness, radiating to the ipsilateral superior nuchal line. She also mentioned a limitation in neck movements. She started taking anti-inflammatory medication (ibuprofen 600 mg every 12 hours) and a muscle relaxant (cyclobenzaprine 10 mg, one tablet at night) for three days, with partial improvement in the pain.

Subsequently, the patient reported the onset of pain with characteristics different from the initial pain, specifically with neuropathic descriptors (a sensation of electric shock and burning), which prompted her to seek a consultation in Physical Medicine and Rehabilitation. She mentioned that the pain would wake her up at dawn, caused by the pressure of her head on the pillow. Upon physical examination, she exhibited limited cervical rotation, especially to the right, pain upon palpation of the upper trapezius fibers, and at its insertion at the upper nuchal line. She also had painful swellings/nodules upon palpation in the retro auricular and posterior cervical regions. She had a positive Tinel's sign and reported moderate pain (7/10 on the numeric rating scale – NRS).

The DN4 questionnaire was administered, with a score of 5/10. In this context, in addition to the analgesic medication she was already taking, pregabalin 75 mg was prescribed, which she took for 15 days, along with cervical muscle stretching exercises (especially the trapezius), warm compresses and massage. After approximately 2-3 weeks, the pain improved, but she still experienced some when combing her hair (NRS 6/10) and in the early morning upon waking (NRS 4/10). At this point, the DN4 was repeated, with a score of 1/10 (due to the pain when combing). After a month, there was an improvement in resting pain (NRS 0/10) and an almost complete resolution of the condition, since she still continued to experience a sensation of electric shock on the outer lateral edge of the ear when combing her hair, a complaint that gradually subsided. Fortunately, this improvement avoided the need for a more invasive approach, such as an ultrasound-guided infiltrative technique, specifically the great occipital nerve block. Temporary relief is often achieved with standard interventional treatments. She was informed that if there was a recurrence of the clinical condition, she should request another appointment, which did not happen.

Discussion

ON is a painful condition of the occipital nerves (GON, LON, TON).[3] It is more commonly unilateral (85%), and its hallmark is a paroxysmal lancinating pain that patients often describe as stabbing, shooting, electric or "shock-like".[11] It can last from seconds to minutes, often originated from the involvement of the nerves.[3] In the vast majority of cases, ON is a result of GON pathology and less frequently are due to LON causes.[3] There are several etiologies of ON but more frequently, its irritation is by chronically contracted muscles and spondylosis of the upper cervical spine.[4]

Given that the symptoms of ON can be similar to those of other headache disorders, it's important to also take into account other conditions like migraines, cluster headaches or tension-type headaches. Therefore, a careful history and physical exam are key components to accurately identify this condition.[11]

At the time of evaluating the patient, an ultrasound was not available, which, given the clinical history and context, could have been useful in evaluating a possible nerve entrapment caused by a contracted muscle, which is a common cause and probably the cause of this ON. The sensation of an electric shock means that there is a nerve damage, as other neuropathic descriptors. Several conditions share similarities with (ON), such as pain in the back of the neck and head, making it challenging to differentiate them without additional symptoms. Clinicians should be particularly aware of two major categories of disease. The first category includes tumors, infections, and congenital anomalies which must be accurately diagnosed to avoid severe consequences if overlooked. Additionally, ON can be confused with migraine, cluster headaches, tension headaches, or hemicrania continua, so the key point is that ON is neuralgia of the occipital nerve, whereas cervicogenic headache involves nociceptive referred pain from cervical structures.[4]

Even though 90% of cases of ON is a result of GON pathology,[3] in this case the LON is probably affected, once she presented neuropathic pain on the lateral edge of the ear, which corresponds to LON territory.

Conservative measures were prescribed early in the acute phase of the pain, and there was a favorable response to them. In this clinical case, there was a near-complete resolution of the condition, with no impact on daily activities, without the need to proceed to interventional procedures. According to literature, if non-pharmacological and pharmacological treatments do not result, minimally invasive interventional procedures should be considered.[8]

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Anesthetic blocks of the GON and lesser occipital nerve LON are highly effective both for confirming a diagnosis and for providing temporary pain relief. Research has shown that occipital nerve blocks can significantly alleviate ON. In a study by Kuhn et al., ten patients were given an initial injection of 0.5% bupivacaine into the GON, followed by a corticosteroid. All patients reported at least 80% pain relief immediately after the treatment. During follow-up, three patients experienced relief for one week, three for two weeks, one for one month, and two patients had their headaches completely resolved for 2.5 and 4 months, respectively. Loukas determined that the ideal injection site is located 2 cm lateral and 2 cm inferior to the external occipital protuberance.[11]

For patients with ON that doesn't respond to medication or occipital nerve blocks (ONB), neuromodulation techniques such as pulsed radiofrequency (PRF) and subcutaneous nerve stimulation present promising alternative treatment options.[11] PRF is believed to reduce pain by creating a low-intensity electrical field around sensory nerves, which inhibits long-term potentiation in pain-transmitting afferent nerves. In a prospective study conducted by Vanelderen,[5] involving 19 patients with ON treated with PRF, the researchers observed a significant reduction in both the mean Visual Analog Scale (VAS) scores for pain and medication usage, as measured by the Medication Quantification Scale. Just over half of the patients reported substantial pain improvement, as assessed by a Likert scale, at a 6-month follow-up.[11]

Ultrasound-guided percutaneous cryoablation of the GON is a procedure frequently performed by many clinicians. When applied at the right temperature, it temporarily stuns the nerve without causing permanent damage; however, nerve injury can occur at temperatures below -70 degrees Celsius. In a 2018 study by Kastler et al., seven patients underwent cryoneurolysis with positive results, though follow-up was limited to 3 months. According to the editor, treatment benefits typically last between 3 months and 1.5 years, with the average duration being around 6 months.[3]

It's crucial to take into account key elements of the patient's lifestyle and how the condition, along with any suggested treatment, might impact them.[3]

Several studies have demonstrated that BoNT-A provides an analgesic effect that lasts longer than its muscle relaxant properties. Various theories have been proposed to explain this pain-relieving effect. One suggests that BoNT-A inhibits sensory nerve mediators such as substance-P, calcitonin gene-related peptide, and glutamate, which contribute to pain transmission. Additionally, BoNT-A may reduce local neurogenic inflammation directly and inhibit central sensitization indirectly by significantly decreasing the activity of wide dynamic range neurons, which play a role in pain processing.[4]

Conclusion

The patient has improved from the condition without new symptoms or complications, only with conservative measures, thus avoiding the risks of interventional procedures. Clinicians should always keep in mind that destructive procedures carry serious risks. Once an anatomical structure is destroyed, it cannot be easily, if ever, restored. Additionally, any destructive procedure poses the risk of developing painful neuroma or causalgia, which may be more difficult to manage than the initial condition.[4]

It is essential to take into account key aspects of the patient's lifestyle and consider how the condition, as well as any proposed treatment, might impact them.[3] A multidisciplinary approach, incorporating psychosocial support as well as medical and non-medical interventions, is likely necessary to effectively manage this condition.[7]

Acknowledgment

None.

Conflicts of interest

The authors have no conflicts of interest to declare.

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