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Occipital Neuralgia

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Occipital neuralgia is a form of headache that involves the posterior occiput in the greater or lesser occipital nerve distribution. Pain can be severe and debilitating, with frequent paroxysms. Occipital neuralgia can be difficult to distinguish from other types of headache and, therefore, diagnosis can be challenging. Local anesthetic block of the occipital nerves, either peripherally or more proximally at the C2 and/or C3 nerve root, may aid in diagnosis. Treatment may include medications, minimally invasive percutaneous procedures, and surgical interventions. This issue of *Pain Management Rounds* presents the characteristics of occipital neuralgia and outlines available treatment options.

BACKGROUND

Headache accounts for nearly 20 million outpatient visits per year in the United States and is one of the most common complaints brought to doctors. Nearly 95% of the population will experience a headache at some point in their life. While the parenchyma of the brain is insensate, the scalp, head muscles, periosteum, dura, and blood vessels are all pain-sensitive; thus, there are many possible causes of head and face pain. Occipital neuralgia is a headache syndrome that may be either primary or secondary.

Primary headaches have no clear structural or disease-related cause, (eg, migraine, tension, and cluster headaches). Primary headaches constitute the etiology of >90% of head and facial pain[1] and occipital neuralgia is often confused with other primary headache syndromes, including migraine and cluster headaches.

Secondary headaches have an underlying disease process that may include tumor, trauma, infection, systemic disease, or hemorrhage.

ETIOLOGY

Patients with occipital neuralgia may be divided into those with structural causes and those with idiopathic causes. Structural causes include:

- trauma to the greater and/or lesser occipital nerves
- compression of the greater and/or lesser occipital nerves or C2 and/or C3 nerve roots by degenerative cervical spine changes
- cervical disc disease
- tumors affecting the C2 and C3 nerve roots.

The greater occipital nerve receives sensory fibers from the C2 nerve root and the lesser occipital nerve receives fibers from the C2 and C3 nerve roots. The third occipital nerve (least occipital nerve) stems from the medial sensory branch of the posterior division of the C3 nerve

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root and travels along the greater occipital nerve. It passes through the trapezius and splenius capitus slightly medial to the greater occipital nerve. Clinically, the third occipital nerve may also be involved in causing occipital neuralgia. Cervical spine changes include spondylosis, arthritis of the upper cervical facet joints, and thickening of the ligaments in that area (particularly C1-4 levels).[2] Some cases of presumed occipital neuralgia may in fact be C2 or C3 radiculopathies. Compression of the greater occipital nerve is possible as it travels up the neck, passing through the semispinalis and trapezius muscles. Whiplash or hyperextension injury may lead to this scenario.[3] Other possible causes include localized infections or inflammation, gout, diabetes, and blood vessel inflammation.[4] Although it cannot be quantified, most patients fall in the category of "unknown cause," when no identifiable lesion is found.

CLINICAL FEATURES

Occipital neuralgia symptoms include aching, burning, and throbbing pain that is often unilateral and continuous with intermittent, shocking, shooting pain. The pain usually originates in the suboccipital area and radiates to the posterior and/or lateral scalp. Occasionally, patients report pain behind the eye on the affected side. Pain may also be perceived over the neck, temple, and frontal regions.[5] Pressure over the occipital nerves may amplify the pain, but there is usually no clear trigger. Furthermore, some patients may have a positive Tinel's sign over the occipital nerve. Occasionally, neck movements (eg, extension and rotation) may trigger pain. At times, patients with occipital neuralgia may experience symptoms similar to migraine or even autonomic changes characteristic of cluster headaches. Associated symptoms include posterior scalp paresthesias, photophobia, and dizziness. Many patients with occipital neuralgia report a cycle of pain-spasm-pain.[6]

DIAGNOSIS

Thorough history-taking and a complete physical and neurological examination are necessary in diagnosing headache.[7] A diagnosis is usually made based on the characteristic area of the pain. In addition, finding tender areas that exacerbate the pain aids in diagnosis. It is important to clarify whether the cause of occipital neuralgia is structural or idiopathic. Abnormal findings on neurologic exam usually indicate a structural cause, in which case, computed tomography (CT) or magnetic resonance imaging (MRI) of the head and cervical spine may be indicated. The work-up of occipital neuralgia should include assessment for atlanto-axial joint instability. Patients with a history of rheumatoid arthritis or trauma should receive a thorough spine work-up. Diagnostic occipital nerve blockade also aids in diagnosis.

Occipital neuralgia often is confused with migraines and other headache syndromes (Table 1). In some cases, occipital neuralgia is misdiagnosed as fibrocytis or fibromyalgia, cervical spine arthritis, or cervical disc disease.

TREATMENT OPTIONS

If the cause is structural, then surgical treatment may be indicated. Because the majority of patients have no clear structural cause, their treatment is usually symptomatic. Local nerve blocks, medications, occipital nerve stimulator implantation, surgical decompression, or lesioning of the C2 and/or C3 nerve roots, or even the greater and/or lesser occipital nerves, may be considered. Occipital neuralgia is often difficult to manage because it can easily be mistaken for other headache syndromes.[8] Management of occipital neuralgia follows the usual course, starting with the recommended conservative treatment, conventional therapy, and medications such as non-steroidal anti-inflammatory drugs (NSAIDs), neuropathic medications (seizure medications, tricyclic antidepressants), and possibly opioids.

Conservative treatment

Physical therapy, massage, acupuncture, and heat are other treatments that can be used for the treatment of occipital neuralgia.[9,10]

Medications

Medications that may help relieve pain in occipital neuralgia include gabapentin 300-3600 mg/day, carbamazepine 400-1200 mg/day, phenytoin 300-600 mg/day, valproic acid 500-2000 mg/day, and baclofen 40-120 mg/day. NSAIDs and opioids may also be beneficial.

NERVE BLOCKS

Nerve blocks consisting of steroids and local anesthetics may also be considered for treatment of occipital neuralgia.[11]

Occipital nerve block

Occipital nerve block is indicated for the diagnosis or treatment of occipital neuralgia. The greater occipital nerve is 2.5 to 3 cm lateral to the external occipital protuberance and medial to the occipital artery. The third occipital nerve is medial to the greater occipital nerve and the lesser occipital nerve is about 2.5 cm lateral to the artery.

TABLE 1: Differential diagnosis of common headaches

Names	Clinical features	Epidemiology	Pathophysiology
Migraine headache	Unilateral hemicranial, pulsating, throbbing, with sensitivity to light and sound, and nausea. May have visual aura. Lasts 4-72 hours if untreated	Peak incidence 25-34 years old; 3-4 times more common in women than men. Family history of migraine common.	Neurovascular headache associated with cranial perivascular inflammation via the trigeminal nerve. May be some serotonergic involvement.
Tension headache	Usually bilateral, dull, pressing, squeezing, bandlike quality. May last from 30 minutes to 7 days. Sensitivity to light and sound, but no nausea. May affect frontal, fronto-occipital, occipital, orbital area.	Most common headache. Affects both men and women equally.	Precise mechanisms unknown; likely multifactorial. May be activation of peripheral nociceptors within neck muscles or ligaments.
Cluster headache	Excruciating, painful, drilling, boring quality that is often debilitating. May be so severe that many patients contemplate suicide. Severe, unilateral orbital pain. If untreated, may last from 15 to 180 minutes. At least one autonomic sign on painful side (eg, lacrimation, nasal congestion, rhinorrhea, miosis, eye edema, ptosis, conjunctival injection). May occur from once a day to 8 times a day in cycles from 1 week to every year.	Peak incidence 20-40 years old; 5-6 times more common in men than in women.	Precise mechanism unknown. May be change in hypothalamic, endocrine, brain stem, and central nervous system functioning. May be trigeminovascular involvement like in migraine headache.
Cervicogenic headache	May have similar presentation as occipital neuralgia, cluster, tension, and migraine headaches. Usually caused by neck movement or change in head position. Ipsilateral shoulder, neck, or arm pain that is nonradicular. Usually unilateral, and can involve neck, occiput, temple, or periorbital region. Typically constant or intermittent, but rarely throbbing or lancinating. May have associated nausea and dizziness.	No specific age range. May affect men and women equally.	Various anatomic structures may transmit nociceptive signals. Structures involved include: atlanto-occipital joint, atlanto-axial joint, C2-3 facet joint, C2-3 disc, suboccipital and upper cervical muscles, trapezius, and sternocleidomastoid muscles.
Occipital neuralgia	Constant, burning, aching, shooting, pain in occiput and posterior scalp usually. May be unilateral or bilateral. Usually worse with extension and rotation of neck or pressure over occiput. Retro-orbital pain may occur with severe attacks.	No specific age range. May affect men and women equally.	Usually no known structural cause. Some cases may have structural cause which may include trauma to the greater and/or lesser occipital nerves, compression of the greater and/or lesser occipital nerves or C2 and/or C3 nerve roots by degenerative cervical spine changes, cervical disc disease, and tumors affecting the C2 and C3 nerve roots.

The greater and third occipital nerves are blocked slightly above the superior nuchal line, just medial to the occipital artery, which is easily palpated. After antiseptic preparation, a 25 gauge $1^1/_2$ inch needle attached to a 5 ml syringe is placed just medial to the artery at the above location. For diagnostic indications, 1 ml of local anesthetic is injected. For treatment, 3-5 ml of local anesthetic combined with steroid is injected. Anesthesia in the region of the greater occipital nerve usually occurs within 10 to 20 minutes. The most serious complication is piercing the occipital artery and bleeding. Compression of the occipital artery is usually effective in avoiding any significant problems.

C2 and/or C3 ganglion block

C2 and/or C3 ganglion block has proven successful in treating some patients. One case report demonstrated that a patient with severe intractable occipital neuralgia

became pain-free for >2 months when given a C2 ganglion block.[12] However, repeat blocks with steroids may have adverse effects. A case report published in 2001 demonstrated that a 39-year-old female who had 6 bilateral greater occipital nerve blocks over a period of 3 months developed signs of Cushing's syndrome. Signs and symptoms were intermittent hypertension, severe muscle weakness, and fluid retention.[13]

BOTULINUM TOXIN

Botulinum Toxin Type A (botox) is an accepted treatment for migraine headache and muscle spasm-related pain with relief up to 4 months.[14] Botox was originally used to treat strabismus and cervical dystonia.[15] One trial demonstrated that botox helped chronic daily headache and appeared to have a cumulative effect with subsequent injections.[16] Treatment with botox is generally well-tolerated; side effects are minimal and include minor discomfort or bleeding at the time of injection.[17] Clinical trials have shown that botox injections for migraine headaches reduced the duration, length, and severity of the headaches, as well as the intake of migraine medications.[18] Botox has been shown to be effective in the treatment of whiplash-associated disorders that often cause occipital neuralgia. It improved the pain and increased the range of motion in these patients. Because of its success in the treatment of muscle spasms and migraines, botulinum toxin may prove to be a reasonable treatment option for occipital neuralgia in the future.

SURGICAL OPTIONS

Occipital neuralgia can occasionally be treated successfully with microvascular nerve decompression. Surgical procedures such as epifacial electric stimulation, dorsal cervical rhizotomy, neurolysis of the greater occipital nerve, and radiofrequency rhizotomy may also be considered. Selective C2 and/or C3 dorsal rhizotomy is another option, although few papers have been published assessing its utility. Dubuisson followed 14 patients over a period of 33 months after partial posterior rhizotomy at C1-3. He found that 10 of 14 patients (71%) had continuing significant relief over that period of time.[19] CT or fluoroscopy-guided percutaneous C2 and/or C3 nerve block is also useful for confirmation of occipital neuralgia and as a preoperative guide for dorsal cervical rhizotomy.[20]

RADIOFREQUENCY THERMOCOAGULATION

Radiofrequency thermocoagulation (RF) is another widely used method to treat occipital neuralgia. It has many advantages, including safety, efficacy, a rapid recovery period, and no permanent scarring. C2 ganglionotomy by RF lesion generator has also been performed and resulted in cases of significant pain relief. Pulsed radiofrequency (PRF) is yet another technique used to treat occipital neuralgia. In a case report, a patient was treated with PRF and, after a 12-month follow-up, was pain-free.[21] Recently, a new surgical treatment was reported consisting of neurolysis of the greater occipital nerve and sectioning of the inferior oblique muscle.[22]

OCCIPITAL NERVE STIMULATOR IMPLANTATION

Surgical implantation of a subcutaneous electrode along the C1-C3 nerve level has been shown to significantly reduce the pain of occipital neuralgia in patients who have failed conservative therapies.[23] In one study of 19 patients, 95% reported improvement in their quality of life and would undergo the procedure again.[24] In another study of 13 patients, 12 reported good-to-excellent pain control at up to 6 years of follow-up.[25] The benefit of this procedure is that it is minimally invasive and there is no permanent destruction of nerves or other vital structures. Another advantage is that patients can first undergo a percutaneous trial of temporary lead placement for several days prior to permanent lead implantation. Depending on the results of the temporary percutaneous trial, patients may or may not undergo the more invasive permanent lead implantation. It has been postulated that a successful temporary percutaneous lead trial, in combination with a successful diagnostic occipital nerve block, may predict a highly effective permanent occipital nerve stimulator implantation.

CONCLUSION

Occipital neuralgia is a headache syndrome that requires careful attention to enable proper diagnosis and treatment. Typically, there is no clear structural cause, although appropriate work-up should be considered in order to rule-out pathologic structural causes. The occipital nerve block is a valuable, simple, and safe diagnostic and therapeutic tool that should be considered early in the course of treatment.

If the pain persists despite preliminary therapies, including occipital nerve blockade with local anesthetic and steroid, then botulinum toxin or permanent implantation of a percutaneous occipital nerve stimulator should be considered before destructive C2 and/or C3 root surgical procedures are implemented.

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Abstracts of Interest

Botulinum neurotoxin for the treatment of migraine and other primary headache disorders.

BLUMENFELD AM, DODICK DW, SILBERSTEIN SD, SAN DIEGO, CA

Clinical data and experience to date have demonstrated that BoNT-A is an effective and well-tolerated therapy for the prevention of migraine and other headache disorders. It has a long duration of action that may last over 4 months with no systemic or serious AEs. Several issues remain to be defined, however, including dosing, location, and number of injections; optimal dilution of BoNT-A; specific headache types that respond best to BoNT-A; and long-term efficacy and safety. Data from ongoing well-designed trials that include a larger patient population investigating these issues may confirm a role for BoNT-A as a first-line agent for migraine prevention. Neurotoxin therapy is part of a broader headache management approach. Because the injection techniques for headache are unique and vary depending on the primary headache disorder being treated and the location and pattern of pain referral, the use of BoNT-A for headache is not simply an extension of its use for cosmesis. The use of BoNT-A in the overall management of primary headache disorders should be reserved

for medical practitioners who not only have experience with BoNT-A injections, but possess the expertise in the diagnosis and management of complex headache disorders. Educating patients and addressing headache triggers and optimizing acute treatment improve the outcome of any preventive program.

Dermatol Clin 2004;22(2):167-75.

Peripheral neurostimulation for control of intractable occipital neuralgia

WEINER RL, REED KL, DALLAS, TEXAS

OBJECTIVE: To present a novel approach for treatment of intractable occipital neuralgia using percutaneous peripheral nerve electrostimulation techniques.

METHODS: Thirteen patients underwent 17 implant procedures for medically refractory occipital neuralgia. A subcutaneous electrode placed transversely at the level of C1 across the base of the occipital nerve trunk produced paresthesias and pain relief covering the regions of occipital nerve pain.

RESULTS: With follow-up ranging from 1-1/2 to 6 years, 12 patients continue to report good to excellent response with greater than 50% pain control and requiring little or no additional medications. The13th patient (first in the series) was subsequently explanted following symptom resolution.

CONCLUSIONS: In patients with medically intractable occipital neuralgia, peripheral nerve electrostimulation subcutaneously at the level of C appears to be a reasonable alternative to more invasive surgical procedures following failure of more conservative therapies.

Neuromodulation 1999;2(3):217-221.

Stimulation methods for neuropathic pain control

STOJANOVIC MP, BOSTON, MA

Neurostimulation methods for control of chronic neuropathic pain have recently gained in popularity. The reasons for this are multifactorial. As opposed to nerve ablation, these methods are minimally invasive and reversible. The improvements in hardware design simplified implantation techniques and prolonged equipment longevity. Stimulation trials have become less invasive, allowing patients to test its effects before final implantation. Finally, the scientific evidence has shown good outcomes of neurostimulation methods for chronic neuropathic pain control. Recent research efforts have revealed new potential mechanisms of action of neurostimulation. Whereas its action was widely explained by gate control theory in the past, it seems that neuromodulation acts also by modulation of neurotransmitters in the central nervous system. Three neurostimulation methods are currently used in clinical practice: spinal cord stimulation (SCS), peripheral nerve stimulation (PNS), and deep brain stimulation (DBS). The SCS and PNS are excellent treatment choices for certain forms of neuropathic pain. The new indications for SCS are end-stage peripheral vascular disease and ischemic heart disease, whereas PNS is used for the treatment of occipital neuralgia and chronic pelvic pain. DBS is reserved for carefully selected patients in whom the other treatment modalities have failed. In a minority of patients the "tolerance" to neurostimulation develops after long-term use. Further research is needed to establish better outcome predictors to neurostimulation and possibly improve patient selection criteria.

Curr Pain Headache Rep 2001;5(2):130-7.

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